
INTRODUCTION

In his 1953 classic textbook titled *The Surgery of Infancy and Childhood*, Dr. Robert E. Gross summarized the essential challenge of pediatric surgery: "Those who daily operate upon adults, even with the greatest of skill, are sometimes appalled—or certainly are not at their best—when called upon to operate upon and care for a tiny patient. Something more than diminutive instruments or scaled-down operative manipulations are necessary to do the job in a suitable manner." To this day, surgical residents often approach the pediatric surgical patient with a mix of fear and anxiety. Nonetheless, they generally complete their pediatric surgical experience with a clear sense of the enormous ability of children to tolerate large operations, and with a true appreciation for the precision required in their care, both in the operating room and during the perioperative period. The specialty has evolved considerably in its ability to care for the smallest of patients with surgical disorders, so that in utero surgery is now an option in certain circumstances. Similarly, our understanding of the pathophysiology of the diseases that pediatric surgeons face has increased greatly to the point where our focus has shifted from an understanding of anatomy and physiology to an appreciation of the molecular or cellular pathways that regulate tissue growth and differentiation. There are few specialties in all of medicine that provide the opportunity to intervene in such a positive manner in such a wide array of diseases, and to receive the most heartfelt appreciation possible—that of a parent whose child's life has forever been improved.

GENERAL CONSIDERATIONS

Fluid and Electrolyte Balance

In managing the pediatric surgical patient, an understanding of fluid and electrolyte balance is critical, as the margin between dehydration and fluid overload is small. Several surgical diagnoses, such as gastroschisis or short-gut syndrome, are characterized by a predisposition to fluid loss. Others require judicious restoration of intravascular volume in order to prevent cardiac failure, as in patients with congenital diaphragmatic hernia and associated pulmonary hypertension. It is important to realize that the infant's physiologic day is approximately 8 hours in duration. A careful assessment of the individual patient's fluid balance tally—showing fluid intake and output for the previous 8 hours—will prevent dehydration or fluid overload. Clinical signs of dehydration include tachycardia and reduced urine output as well as a depressed fontanelle, lethargy, and poor feeding. Fluid overload is often manifested by the onset of new oxygen requirements, respiratory distress, tachypnea, and tachycardia.

The infant is born with a surplus of body water, which is normally excreted by the end of the first week of life. At birth, fluid requirements are 65 mL/kg (750 mL/m²) and increase to 100 mL/kg (1000 mL/m²) by the end of the first week. Daily maintenance fluids for most children can be estimated using the formula: 100 mL/kg for the first 10 kg plus 50 mL/kg for 11 to 20 kg plus 25 mL/kg for each additional kilogram of body weight thereafter. Because intravenous fluid orders are written as milliliters per hour, this can be...
conveniently converted to 4 mL/kg per hour up to 10 kg by adding 2 mL/kg per hour for 11 to 20 kg, and 1 mL/kg per hour for each additional kilogram of body weight thereafter. For example, a 26-kg child has an estimated maintenance fluid requirement of $(10 \times 4) + (10 \times 2) + (6 \times 1) = 66$ mL/h in the absence of massive fluid losses or shock. Fluid for maintenance is generally provided as 5% dextrose in one quarter normal saline. For short-term intravenous therapy, the administration of 5 mEq/kg per day of sodium and 2 mEq/kg per day of potassium will satisfy the daily need. Fluid and electrolyte losses secondary to protracted vomiting or diarrhea are corrected by modifying this formula according to the measured losses. In infants the normal serum osmolarity is between 280 and 290 mmol/L. Newborns have the ability to concentrate their urine well by the fifth day of life; thus urine concentration as well as output must be considered when ordering intravenous fluids postoperatively. If the child has a significant ongoing fluid loss (e.g., from a nasogastric tube), it is best to properly replace that loss with IV fluids at least every 4 hours. A typical replacement formula is $\text{D}_5 \frac{1}{2} \text{ normal saline +20 mEq KCl/L}$. Whatever the formula used to calculate fluid replacement for the infant or small child, the optimal strategy is to analyze serum electrolytes and fluid losses and to replace the appropriate constituents precisely.

**Acid-Base Equilibrium**

Acute metabolic acidosis usually implies inadequate tissue perfusion, and is a serious disorder in children. Potentially life-threatening causes that are specific for the pediatric population must be sought, including intestinal ischemia from necrotizing enterocolitis (in the neonate), midgut volvulus, or incarcerated hernia. Other causes include chronic bicarbonate loss from the gastrointestinal tract or an acid accumulation as in chronic renal failure. Respiratory acidosis implies hypoventilation, the cause of which should be apparent. Treatment of acute metabolic acidosis should be aimed at restoring tissue perfusion by addressing the underlying abnormality first. For severe metabolic acidemia where the serum pH is less than 7.25, sodium bicarbonate should be administered using the following guideline: base deficit $\times$ weight in kilograms $\times$ 0.5 (in newborns). The last factor in the equation should be 0.4 for smaller children, and 0.3 for older children. The dose should be diluted to a concentration of 0.5 mEq/mL because full-strength sodium bicarbonate is hyperosmolar. One half the corrective dose is given, and the serum pH is measured again. During cardiopulmonary resuscitation (CPR), one half the corrective dose can be given as an intravenous bolus and the other half given slowly intravenously.

Respiratory alkalosis is usually caused by hyperventilation, which is readily correctable. Metabolic alkalosis most commonly implies gastric acid loss, as in the child with pyloric stenosis or overaggressive diuretic therapy. In the child with gastric fluid loss, IV fluids of 5% dextrose, 0.5% normal saline, and 20 mEq KCl/L usually correct the alkalosis.

**Blood Volume and Blood Replacement**

Criteria for blood transfusion in infants and children remain poorly defined. The decision to transfuse a critically-ill pediatric patient may depend on a number of clinical features that include the patient's age, primary diagnosis, the presence of ongoing bleeding, coagulopathy, hypoxia, hemodynamic compromise, lactic acidosis, and cyanotic heart
disease, as well as overall severity of illness. A recent survey of transfusion practices among pediatric intensivists showed that the baseline hemoglobin levels that would prompt them to recommend a red blood cell (RBC) transfusion ranged from 7 to 13 g/dL. Patients with cyanotic heart disease are often transfused to higher hemoglobin values, although the threshold for transfusion in this population remains to be defined. To decrease the need for transfusion, other strategies have been considered. Studies in both critically-ill adults and neonates have shown that administration of erythropoietin decreases RBC transfusion requirements. In general terms, there is a trend toward an avoidance of the use of RBC products whenever possible, as current studies suggest that lower hemoglobin concentrations are well tolerated by many groups of patients, and that administration of RBCs may have unintended negative consequences.

A useful guideline for estimation of blood volume for the infant is 85 mL/kg of body weight. When packed red blood cells (PRBC) are utilized, the transfusion requirement is calculated as 10 mL/kg, which roughly is equivalent to a 500-mL transfusion for a 70-kg adult. At the authors' institution, the following formula is used to determine the volume of blood in mL: \((\text{target hematocrit} - \text{current hematocrit}) \times \text{weight(kg)} \times 80/65 = \text{blood volume}\)

In the child, coagulation deficiencies may rapidly assume clinical significance after extensive blood transfusion. It is advisable to have fresh frozen plasma and platelets available if more than 30 mL/kg have been transfused. Plasma is given in a dose of 10 to 20 mL/kg, and platelets are given in a dose of 1 unit/5 kg. Each unit of platelets consists of 40 to 60 mL of fluid, and platelets can be spun down to a platelet "button" for infants who require restricted fluid administration. Following transfusion of PRBC to neonates, with tenuous fluid balance, a single dose of a diuretic (such as furosemide 1 mg/kg) may help to facilitate excretion of the extra fluid load.

**Hyperalimentation and Nutrition**

The nutritional requirements of the surgical neonate must be met in order for the child to grow and to heal surgical wounds. If inadequate protein and carbohydrate calories are given, the child may not only fail to recover from surgery, but may also exhibit growth failure and impaired development of the central nervous system. Neonates that are particularly predisposed to protein-calorie malnutrition include those with gastroschisis, intestinal atresia, or intestinal insufficiency from other causes such as necrotizing enterocolitis. The protein and caloric requirements for the surgical neonate are shown in Table 38-1.

Nutrition can be provided via either the enteral or parenteral routes. Whenever possible, the enteral route is preferred, because it not only promotes the growth and function of the gastrointestinal system, but also ensures that the infant learns how to feed. There are various enteral feeding preparations available, which are outlined in Table 38-2. The choice of formula is based upon the clinical state of the individual child. Pediatric surgeons are occasionally faced with situations in which oral feeding is not possible. This problem can be seen in the extremely premature infant who has not yet developed the feeding skills, or in the infant with concomitant craniofacial abnormalities that impair
sucking. In these instances, enteral feeds can be administered using either a nasojejunal or a gastrostomy tube.

When the gastrointestinal tract cannot be used because of mechanical, ischemic, inflammatory, or functional disorders, parenteral alimentation must be given. When an extended period of parenteral nutrition is required, central venous catheters are placed. Peripheral intravenous alimentation can be given, utilizing less concentrated but greater volumes of solutions. To prevent the development of trace metal deficiencies, supplemental copper, zinc, and iron are provided to patients receiving long-term total parenteral nutritional (TPN) support.

A major complication of long-term TPN is the development of liver failure. This is characterized by cholestatic liver disease that eventually progresses to end-stage hepatic fibrosis. To prevent this major complication, concomitant enteral feedings must be instituted, and the gastrointestinal tract should be used as soon as possible. In instances in which proximal stomas are in place, continuity of the gastrointestinal tract should be restored as soon as possible. Where intestinal insufficiency is associated with dilation of the small intestine, tapering or intestinal lengthening procedures may be beneficial. Other strategies to minimize the development of TPN-related liver disease include avoidance of infection by meticulous catheter care, aggressive treatment of any infection, and early cycling of parenteral nutrition to include a period during the day when parenteral nutrition is not given.

**Venous Access**

Obtaining reliable vascular access in an infant or child is a major responsibility of the pediatric surgeon. The goal should always be to place the catheter in the least invasive, least risky, and least painful manner, and in a location that is most accessible and facilitates use of the catheter without complications for as long as needed. In infants, the general approach of these authors is to place a central venous catheter using a cutdown approach, either in the antecubital fossa, external jugular vein, facial vein, or proximal saphenous vein. If the internal jugular vein is used, placing a purse-string suture at the venotomy is recommended, if possible, to prevent venous occlusion. In infants over 2 kg and in older children, percutaneous access of the subclavian, internal jugular, or femoral veins is possible in most cases, and central access is achieved using the Seldinger technique. The catheters are tunneled to an exit site separate from the venotomy site. Regardless of whether the catheter is placed by a cutdown approach or percutaneously, a chest x-ray to confirm central location of the catheter tip and to exclude the presence of a pneumothorax or hemothorax is mandatory. When discussing the placement of central venous catheters with parents, it is important to note that the complication rate for central venous lines in children is high. The incidence of catheter-related sepsis or infection approaches 10% in many series. Superior or inferior vena caval occlusion is a significant risk, particularly in the smallest premature patients.

**Thermoregulation**
Careful regulation of the ambient environment of infants and children is crucial, as these patients are extremely thermolabile. Premature infants are particularly susceptible to changes in environmental temperature. Because they are unable to shiver and lack stores of fat, their potential for thermogenesis is impaired. This is compounded by the administration of anesthetic and paralyzing agents. Since these patients lack adaptive mechanisms to cope with the environment, the environment must be regulated. Attention to heat conservation during transport of the infant to and from the operating room is essential. Transport units incorporating heating units are necessary for premature infants. In the operating room, the infant is kept warm by the use of overhead heating lamps, a heating blanket, warming of inspired gases, and coverage of the extremities and head with occlusive materials. During abdominal surgery, extreme care is taken to avoid wet and cold drapes. All fluids used to irrigate the chest or abdomen must be warmed to body temperature. Constant monitoring of the child's temperature is critical in a lengthy procedure, and the surgeon should continuously communicate with the anesthesiologist regarding the temperature of the patient. The development of hypothermia in infants and children can result in cardiac arrhythmias or coagulopathy. These potentially life-threatening complications can be avoided by careful attention to thermoregulation.

**Pain Control**

Despite previously held beliefs to the contrary, it has now been definitively established that neonates experience pain. Therefore, any procedure that is performed on a neonate must be accompanied by the provision of adequate analgesia. There is a range of pain management options that can improve the child's well being, as well as the parents' sense of comfort. The use of a pacifier, which may be dipped in sucrose, has been shown to decrease crying time and neonatal pain scores after minor procedures. For situations in which more pain is expected, intravenous narcotic agents should be used. Morphine and fentanyl have an acceptable safety margin and can be administered judiciously to neonates and children. A recent randomized trial of neonates on ventilators showed that the use of a morphine infusion decreased the incidence of intraventricular hemorrhage by 50%. Additional analgesic modalities include the use of topical anesthetic ointment (e.g., EMLA [eutectic mixture of local anesthetics] cream), and the use of regional anesthesia, such as caudal blocks for hernias, or epidural infusion for thoracic surgery. In the postoperative period, patient-controlled analgesia (PCA) is another excellent method of pain control. By ensuring that the pediatric surgical patient has adequate analgesia, the surgeon ensures that the patient receives the most humane and thorough treatment, and provides important reassurance to all other members of the health care team and to the family that pain control is a high priority.

**NECK MASSES**

**Introduction**

The management of neck masses in children is determined by their location and the length of time that they have been present. Neck lesions are found either in the midline or lateral compartments. Midline masses include thyroglossal duct remnants, thyroid masses, thymic cysts, or dermoid cysts. Lateral lesions include branchial cleft remnants,
cystic hygromas, vascular malformations, salivary gland tumors, torticollis, and lipoblastoma (a rare benign mesenchymal tumor of embryonal fat occurring in infants and young children). Enlarged lymph nodes and rare malignancies such as rhabdomyosarcoma can occur either in the midline or laterally.

**Lymphadenopathy**

The most common cause of a neck mass in a child is an enlarged lymph node, which typically can be found laterally or in the midline. The patient is usually referred to the pediatric surgeon for evaluation after the mass has been present for several weeks. A detailed history and physical examination often helps determine the likely etiology of the lymph node, and the need for excisional biopsy. Enlarged tender lymph nodes are usually the result of a bacterial infection (*Staphylococcus* or *Streptococcus*). Treatment of the primary cause (e.g., otitis media or pharyngitis) with antibiotics often is all that is necessary. However, when the involved nodes become fluctuant, incision and drainage are indicated. More chronic forms of lymphadenitis, including infections with tuberculosis, atypical mycobacteria, as well as cat-scratch fever, are determined based on serologic findings and excisional biopsy. The lymphadenopathy associated with infectious mononucleosis can be diagnosed based on serology. When the neck nodes are firm and fixed, and others also are present in the axillae or groin, or the history suggests the presence of a hematologic malignancy, excisional biopsy is indicated. In these cases, a chest radiograph must be obtained to evaluate whether a mediastinal mass also is present. The presence of a large mediastinal mass should be identified preoperatively, as this may cause airway compression when muscle relaxants are administered. Under these circumstances, tissue should be obtained under local anesthesia. The tissue is sent to pathology fresh for evaluation.

**Thyroglossal Duct Remnants**

Pathology and Clinical Manifestations

The thyroid gland buds off the foregut diverticulum at the base of the tongue in the region of the future foramen cecum at 3 weeks of embryonic life. As the fetal neck develops, the thyroid tissue becomes more anterior and caudad until it rests in its normal position. The "descent" of the thyroid is intimately connected with the development of the hyoid bone. Residual thyroid tissue left behind in the migration may persist and subsequently present in the midline of the neck as a thyroglossal duct cyst. The mass is most commonly appreciated in the 2- to 4-year-old child when the baby fat disappears and irregularities in the neck become more readily apparent. Usually the cyst is encountered in the midline at or below the level of the hyoid bone, and moves up and down with swallowing or with protrusion of the tongue. Occasionally it presents as an intrathyroidal mass. Most thyroglossal duct cysts are asymptomatic. If the duct retains its connection with the pharynx, infection may occur, and the resulting abscess will necessitate incision and drainage, occasionally resulting in a salivary fistula. Submental lymphadenopathy and midline dermoid cysts can be confused with a thyroglossal duct cyst. Rarely, midline ectopic thyroid tissue masquerades as a thyroglossal duct cyst, and may represent the patient's only thyroid tissue. Therefore, if there is any question regarding the diagnosis or
if the thyroid gland cannot be palpated in its normal anatomic position, it is advisable to obtain a nuclear scan to confirm the presence of a normal thyroid gland. Although rarely the case in children, in adults the thyroglossal duct may contain thyroid tissue that can undergo malignant degeneration. The presence of malignancy in a thyroglossal cyst should be suspected when the cyst grows rapidly, or when the ultrasound demonstrates a complex anechoic pattern or the presence of calcification.

Treatment

If the cyst presents with an abscess, treatment should consist of drainage and antibiotics. Following resolution of the inflammation, resection of the cyst in continuity with the central portion of the hyoid bone and the tract connecting to the pharynx, in addition to ligation at the foramen cecum (the Sistrunk operation) is curative. Lesser operations result in unacceptably high recurrence rates, and recurrence is more frequent following infection. According to a recent review, factors predictive of recurrence included more than two infections prior to surgery, age under 2 years, and inadequate initial operation.

Branchial Cleft Anomalies

Paired branchial clefts and arches develop early in the fourth gestational week. The first cleft and the first, second, third, and fourth pouches give rise to adult organs. The embryologic communication between the pharynx and the external surface may persist as a fistula. A fistula is seen most commonly with the second branchial cleft, which normally disappears, and extends from the anterior border of the sternocleidomastoid muscle superiorly, inward through the bifurcation of the carotid artery, and enters the posterolateral pharynx just below the tonsillar fossa. The branchial cleft remnants may contain small pieces of cartilage and cysts, but internal fistulas are rare. A second branchial cleft sinus is suspected when clear fluid is noted draining from the external opening of the tract at the anterior border of the lower third of the sternocleidomastoid muscle. Rarely, branchial cleft anomalies occur in association with biliary atresia and congenital cardiac anomalies, an association that is referred to as Goldenhar's complex.

Treatment

The treatment is surgical, and complete removal of the cyst and tract is necessary for cure. Dissection of the sinus tract is facilitated by passing a fine lacrimal duct probe through the external opening into the tract and utilizing it as a guide for dissection. Injection of a small amount of methylene blue dye into the tract also may be useful. A series of two or sometimes three small transverse incisions in a "stepladder" fashion is preferred to a long oblique incision in the neck, which is cosmetically undesirable. Branchial cleft cysts can present as abscesses. In these cases, initial treatment includes incision and drainage with a course of antibiotics to cover *Staphylococcus* and *Streptococcus* species, followed by excision of the cyst after the infection resolves.

Cystic Hygroma

Etiology and Pathology
Cystic hygroma (lymphangioma) occurs as a result of sequestration or obstruction of developing lymph vessels in approximately 1 in 12,000 births. Although the lesion can occur anywhere, the most common sites are in the posterior triangle of the neck, axilla, groin, and mediastinum. The cysts are lined by endothelium and filled with lymph. Occasionally unilocular cysts occur, but more often there are multiple cysts infiltrating the surrounding structures and distorting the local anatomy. A particularly troublesome variant of cystic hygroma is that which involves the tongue, floor of the mouth, and structures deep in the neck. Adjacent connective tissue may show extensive lymphocytic infiltration. The mass may be apparent at birth or may appear and enlarge rapidly in the early weeks or months of life as lymph accumulates; most present by age 2 years (Fig. 38-1A). Extension of the lesion into the axilla or mediastinum occurs about 10% of the time and can be demonstrated preoperatively by chest x-ray, ultrasound (US), or computed tomographic (CT) scan. Cystic hygromas occasionally contain nests of vascular tissue. These poorly supported vessels may bleed and produce rapid enlargement and discoloration of the hygroma. Infection within the cysts, usually caused by *Streptococcus* or *Staphylococcus*, may occur. In the neck this can cause rapid enlargement, which may result in airway compromise. Rarely, it may be necessary to carry out percutaneous aspiration of a cyst to relieve respiratory distress.

The diagnosis of cystic hygroma by prenatal US before 30 weeks' gestation has detected a "hidden mortality," as well as a high incidence of associated anomalies, including abnormal karyotypes and hydrops fetalis. Occasionally, very large lesions can cause obstruction of the fetal airway. Such obstruction can result in the development of polyhydramnios by impairing the ability of the fetus to swallow amniotic fluid. In these circumstances, the airway is usually markedly distorted, which can result in immediate airway obstruction unless the airway is secured at the time of delivery. Orotracheal intubation or urgent emergency tracheostomy while the infant remains attached to the placenta, the *ex utero intrapartum* technique (EXIT) procedure, may be necessary to secure the airway.

**Treatment**

Surgical excision is the treatment of choice for cystic hygromas. Total removal may not be possible because of the extent of the hygroma and its proximity to, and intimate relationship with, adjacent nerves, muscles, and blood vessels (see Fig. 38-1B). Radical ablative surgery is not indicated for this lesion. Conservative excision and unroofing of remaining cysts is advised, with repeated partial excision of residual hygroma if necessary, preserving all adjacent crucial structures. Postoperative wound drainage is important and is best accomplished by closed-suction technique. Fluid may accumulate beneath the surgically-created flaps in the area from which the hygroma was excised, requiring multiple needle aspirations. Injection of sclerosing agents (OK-432 or bleomycin) with favorable results has been reported. OK-432 is composed of a lyophilized mixture of group A *Streptococcus pyogenes*, and has been used in neonates without systemic toxicity. The use of these agents has not been widely adopted.

**FIG. 38-1.** A. Left cervical cystic hygroma in a 2-day-old baby. B. Intraoperative photograph showing a vessel loop around the spinal accessory nerve.
**Torticollis**

The presence of a lateral neck mass in infancy in association with rotation of the head toward the opposite side of the mass indicates the presence of congenital torticollis. This lesion results from fibrosis of the sternocleidomastoid muscle. The mass may be palpated in the affected muscle in approximately two-thirds of cases. Histologically, the lesion is characterized by the deposition of collagen and fibroblasts around atrophied muscle cells. In the majority of cases, physical therapy is of benefit. Rarely surgical transection of the affected muscle can be curative, if needed.

**RESPIRATORY SYSTEM**

**Congenital Diaphragmatic Hernia**

Pathology

During formation of the diaphragm, the pleural and coelomic cavities remain in continuity by means of the pleuroperitoneal canal. The posterolateral communication is the last to be closed by the developing diaphragm. Failure of diaphragmatic development leaves a posterolateral defect known as a Bochdalek hernia. This anomaly is encountered more commonly on the left (80 to 90%). Incomplete development of the posterior diaphragm allows the abdominal viscera to fill the chest cavity. The abdominal cavity is small and underdeveloped and remains scaphoid after birth. Both lungs are hypoplastic, with decreased bronchial and pulmonary artery branching. Lung weight, lung volume, and deoxyribonucleic acid (DNA) content also are decreased, but these findings are more striking on the ipsilateral side. In many instances, evidence suggests that a paucity of surfactant is present, which compounds the degree of respiratory insufficiency.

Amniocentesis with karyotyping may show chromosomal defects, especially trisomy 18 and 21. Associated anomalies, once thought to be uncommon, are identified in 40% of these infants, and most commonly involve the heart, brain, genitourinary system, craniofacial structures, or limbs.

Prenatal ultrasonography is successful in making the diagnosis of congenital diaphragmatic hernia (CDH) as early as 15 weeks' gestation. Ultrasound findings include herniated abdominal viscera, abnormal anatomy of the upper abdomen, and mediastinal shift away from the herniated viscera (Fig. 38-2). Accurate prenatal prediction of outcome for fetuses who have CDH is difficult. A useful index of severity for patients with left CDH is the lung-to-head ratio (LHR), which is the product of the length and the width of the right lung at the level of the cardiac atria divided by the head circumference (all measurements in millimeters). An LHR value of less than 1.0 is associated with a poor prognosis, whereas an LHR greater than 1.4 predicts a more favorable outcome.

Following delivery, the diagnosis of CDH is made by chest x-ray (CXR) (Fig. 38-3). The differential diagnosis includes congenital cystic adenomatoid malformation, in which the intrathoracic loops of bowel may be confused with multiple lung cysts. The vast majority of infants with CDH develop immediate respiratory distress, which is due to the
combined effects of three factors. First, the air-filled bowel in the chest compresses the mobile mediastinum, which shifts to the opposite side of the chest, compromising air exchange in the contralateral lung. Second, pulmonary hypertension develops. This phenomenon results in persistent fetal circulation, with resultant decreased pulmonary perfusion and impaired gas exchange. Finally, the lung on the affected side is often markedly hypoplastic, such that it is essentially nonfunctional. Varying degrees of pulmonary hypoplasia on the opposite side may compound these effects. As a result, neonates with CDH are extremely sick, and the overall mortality in most series is approximately 60 to 70%.

Treatment

Many infants are symptomatic at birth due to hypoxia, hypercarbia, and metabolic acidosis. Prompt cardiorespiratory stabilization is mandatory. It is interesting that the first 24 to 48 hours after birth are often characterized by a period of relative stability, with high PaO₂ (partial pressure of arterial oxygen) levels and relatively good perfusion. This has been termed the "honeymoon period," and is often followed by progressive cardiorespiratory deterioration in the majority of patients. In the past, correction of the hernia was felt to be a surgical emergency, and these patients underwent surgery shortly after birth. It is now accepted that the presence of persistent pulmonary hypertension that results in right-to-left shunting across the open foramen ovale or the ductus arteriosus, and the degree of pulmonary hypoplasia, are the leading causes of cardiorespiratory insufficiency. Therefore, current management is directed toward preventing or reversing the pulmonary hypertension, and minimizing barotrauma while optimizing oxygen delivery. To achieve this goal, infants are placed on mechanical ventilation using relatively low or "gentle" settings that prevent overinflation of the noninvolved lung. PaO₂ (partial arterial pressure of carbon dioxide) levels in the range of 50 to 60 mm Hg or higher are acceptable, as long as the pH remains 7.25 or greater. If these objectives cannot be achieved using conventional ventilation, high-frequency oscillatory ventilation (HFOV) may be employed to avoid the injurious effects of conventional tidal volume ventilation. Echocardiography is used to assess the degree of pulmonary hypertension and to identify the presence of a coexisting cardiac anomaly. To minimize the degree of pulmonary hypertension, inhaled nitric oxide may be used. In certain patients, this agent significantly improves pulmonary perfusion, as manifested by improved oxygenation. Nitric oxide is administered into the ventilation circuit, and is used in concentrations up to 40 parts per million. Correction of acidosis using bicarbonate solution may minimize the degree of pulmonary hypertension. As the degree of pulmonary hypertension becomes hemodynamically significant, right-sided heart failure develops and systemic perfusion is impaired. Administration of excess intravenous fluid will compound the degree of cardiac failure, and lead to marked peripheral edema. Inotropic support using epinephrine is therefore useful in optimizing cardiac contractility and maintaining mean arterial pressure.

Infants with CDH who remain severely hypoxic despite maximal ventilatory care may be candidates for treatment of their respiratory failure by extracorporeal membrane oxygenation (ECMO). Venovenous or venoarterial bypass is used. Venovenous bypass is established with a single cannula through the internal jugular vein, with blood removed
from and infused into the right atrium by separate ports. Venoarterial bypass is used preferentially by some centers because it provides the cardiac support that is often needed. The right atrium is cannulated by means of the internal jugular vein and the aortic arch through the right common carotid artery. As much of the cardiac output is shunted through the membrane oxygenator as needed to provide oxygenated blood to the infant and remove carbon dioxide. The infant is maintained on bypass until the pulmonary hypertension is reversed and lung function, as measured by compliance, is improved. This is usually seen within 7 to 10 days, but in some infants it may take up to 3 weeks to occur. The use of ECMO is associated with significant risk and because patients require systemic anticoagulation, bleeding complications are the most significant. They may occur intracranially or at the site of cannula insertion, and can be life threatening. Systemic sepsis is a significant problem, and may necessitate decannulation. Criteria for placing infants on ECMO include the presence of normal cardiac anatomy by echocardiography, the absence of fatal chromosome anomalies, and the expectation that the infant would die without ECMO. Traditionally, a threshold of weight greater than 2.5 kg and gestational age greater than 34 weeks has been used to select patients for ECMO, although success has been achieved at weights as low as 1.8 kg. It is important to emphasize that although ECMO may salvage a population of neonates with refractory pulmonary hypertension, the use of this technique remains controversial. A strategy that does not involve the use of ECMO, but instead emphasizes the use of permissive hypercapnia and the avoidance of barotrauma, may provide equal overall outcome in patients with CDH. This likely reflects the fact that mortality is related to the degree of pulmonary hypoplasia and the presence of congenital anomalies, neither of which are correctable by ECMO.

The timing of diaphragm repair is controversial. In patients that are not placed on ECMO, most surgeons perform repair once the hemodynamic status has been optimized. In neonates that are on bypass, some surgeons perform early repair on bypass; others wait until the infant’s lungs are fully recovered, repair the diaphragm, and discontinue bypass within hours of surgery. Still others repair the diaphragm only after the infant is off bypass. Operative repair of the diaphragmatic hernia is best accomplished by an abdominal approach. Through a subcostal incision, the abdominal viscera are withdrawn from the chest, exposing the defect in the diaphragm. Care must be taken when reducing the spleen and liver, as bleeding from these structures can be fatal. The anterior margin is often apparent, while the posterior muscular rim is attenuated. If the infant is heparinized on bypass, minimal dissection of the muscular margins is performed. Electrocautery is used liberally to minimize postoperative bleeding. Most infants who require ECMO support prior to hernia repair have large defects, often lacking the medial and posterior margins. Prior to the availability of ECMO therapy, most of these infants died. About three fourths of infants repaired on bypass require prosthetic material to patch the defect, suturing it to the diaphragmatic remnant or around ribs or costal cartilages for large defects. If there is adequate muscle for closure, a single layer of nonabsorbable horizontal mattress suture is used to close the defect. Just before the repair is complete, a chest tube may be positioned in the thoracic cavity. We tend to reserve the use of chest tubes for patients who are repaired on ECMO, as these patients are at risk for developing a hemothorax, which can significantly impair ventilation. Anatomic closure of the abdominal wall may be impossible after reduction of the viscera. Occasionally a
prosthetic patch of GoreTex or Surgisis may be sutured to the fascia and facilitate closure. The patch can be removed at a later time and the ventral hernia can be closed at that time or subsequently.

If the diaphragm has been repaired on ECMO, weaning and decannulation are accomplished as soon as possible. All infants are ventilated postoperatively to maintain preductal arterial oxygenation of 80 to 100 mm Hg. Very slow weaning from the ventilator is necessary to avoid recurrent pulmonary hypertension. Oscillation ventilation may be switched to conventional ventilation as part of the process of weaning.

**FIG. 38-2.** Prenatal ultrasound of a fetus with a congenital diaphragmatic hernia. Arrows point to the location of the diaphragm. Arrowhead points to the stomach, which is in the thoracic cavity.

**FIG. 38-3.** Chest x-ray showing a left congenital diaphragmatic hernia.

### Congenital Lobar Emphysema

Congenital lobar emphysema (CLE) is a condition manifested during the first few months of life as a progressive hyperexpansion of one or more lobes of the lung. It can be life threatening in the newborn period, but in the older infant it causes less respiratory distress. Air entering during inspiration is trapped in the lobe; on expiration, the lobe cannot deflate and progressively overexpands, causing atelectasis of the adjacent lobe or lobes. This hyperexpansion eventually shifts the mediastinum to the opposite side and compromises the other lung. CLE usually occurs in the upper lobes of the lung (left greater than right), followed next in frequency by the right middle lobe, but it also can occur in the lower lobes. It is caused by intrinsic bronchial obstruction from poor bronchial cartilage development or extrinsic compression. Approximately 14% of children with this condition have cardiac defects, with an enlarged left atrium or a major vessel causing compression of the ipsilateral bronchus.

Symptoms range from mild respiratory distress to full-fledged respiratory failure, with tachypnea, dyspnea, cough, and late cyanosis. These symptoms may be stable or they may progress rapidly or result in recurrent pneumonia. Occasionally, infants with CLE present with failure to thrive, which likely reflects the increased work associated with the overexpanded lung. Diagnosis is made by chest x-ray, which shows a hyperlucent affected lobe with adjacent lobar compression and atelectasis with varying degrees of shift of the mediastinum to the opposite side and compression of the contralateral lung (Fig. 38-4). If definitive diagnosis is unclear by chest x-ray, CT scan may be helpful. Unless foreign body or mucus plugging is suspected as a cause of hyperinflation, bronchoscopy is not advisable because it can produce more air trapping and cause life-threatening respiratory distress in a stable infant. Treatment is resection of the affected lobe. Unless symptoms necessitate earlier surgery, resection can usually be performed after the infant is several months of age. The prognosis is excellent.

**FIG. 38-4.** Congenital lobar emphysema of the left upper lobe in a 2-week-old boy. Mediastinal shift is present.
**Congenital Cystic Adenomatoid Malformation**

This malformation consists of cystic proliferation of the terminal airway, producing cysts lined by mucus-producing respiratory epithelium, and elastic tissue in the cyst walls without cartilage formation. There may be a single cyst with a wall of connective tissue containing smooth muscle. Cysts may be large and multiple (type I), smaller and more numerous (type II), or they may resemble fetal lung without macroscopic cysts (type III). Most congenital cystic adenomatoid malformation (CCAM) occurs in the left lower lobe. However, this lesion can occur in any lobe and may occur in both lungs simultaneously. In the left lower lobe, type I may be confused at birth with a congenital diaphragmatic hernia. Clinical symptoms may range from none at all to severe respiratory failure at birth. The cyst(s), whether single or multiple, can produce air trapping and may be confused with congenital lobar emphysema pneumatoceles or even pulmonary sequestrations. They also can be involved with repeated infections and produce fever and cough in older infants and children. The diagnosis often can be made by CXR. In certain cases US or CT scan may be definitive (Fig. 38-5). Prenatal US may suggest the diagnosis. In the newborn period, US may also be useful, especially to distinguish between CCAM and congenital diaphragmatic hernia. Resection is curative and may need to be performed urgently in the infant with severe respiratory distress. Lobectomy is usually required (Fig. 38-6). Prognosis is excellent.

**FIG. 38-5.** CT scan of the chest showing a congenital cystic adenomatoid malformation of the left lower lobe.

**FIG. 38-6.** Intraoperative photograph showing the left lower lobe congenital cystic adenomatoid malformation seen in Fig. 38-5.

**Pulmonary Sequestration**

Pulmonary sequestration is uncommon and consists of a mass of lung tissue, usually in the left lower chest, occurring without the usual connections to the pulmonary artery or tracheobronchial tree, yet with a systemic blood supply from the aorta. There are two kinds of sequestration. Extralobar sequestration is usually a small area of nonaerated lung separated from the main lung mass, with a systemic blood supply, that is located immediately above the left diaphragm. It is commonly found in cases of congenital diaphragmatic hernia. Intralobar sequestration more commonly occurs within the parenchyma of the left lower lobe, but can occur on the right. There is no major connection to the tracheobronchial tree, but a secondary connection may be established, perhaps through infection or via adjacent intrapulmonary shunts. The blood supply is systemic from the aorta, is often multiple vessels, and frequently originates below the diaphragm (Fig. 38-7). Venous drainage of both types can be systemic or pulmonary. The cause of sequestration is unknown, but most probably involves an abnormal budding of the developing lung that picks up a systemic blood supply and never becomes connected with the bronchus or pulmonary vessels. Extralobar sequestration is asymptomatic and is usually discovered incidentally on CXR. If the diagnosis can be confirmed (e.g., by CT scan), resection is not necessary. Diagnosis of intralobar sequestration, on the other hand,
is usually made after repeated infections manifested by cough, fever, and consolidation in the posterior basal segment of the left lower lobe. Increasingly the diagnosis is being made in the early months of life by US, and color Doppler often can be helpful in delineating the systemic arterial supply. Removal of the entire left lower lobe is usually necessary since the diagnosis often is made late after multiple infections. Occasionally the sequestered part of the lung can be removed segmentally. Prognosis is excellent.

**FIG. 38-7.** Arteriogram showing large systemic artery supply to intralobar sequestration of the left lower lobe.

**Bronchogenic Cyst**

Bronchogenic cysts can occur anywhere along the respiratory tract from the neck to the lung parenchyma. They can present at any age. Histologically, they are hamartomatous, and usually consist of a single cyst lined with respiratory epithelium containing cartilage and smooth muscle. They are probably embryonic rests of foregut origin that have been pinched off from the main portion of the developing tracheobronchial tree, and are closely associated in causation with other foregut duplication cysts arising from the esophagus. Bronchogenic cysts may be seen on prenatal US, but are discovered most often incidentally on postnatal CXR. Although they may be completely asymptomatic, bronchogenic cysts may produce symptoms, depending on their anatomic location. In the paratracheal region of the neck they can produce airway compression and respiratory distress. In the lung parenchyma, they may become infected and present with fever and cough. In addition they may cause obstruction of the bronchial lumen with distal atelectasis and infection. They may also cause mediastinal compression. Rarely, rupture of the cyst can occur. CXR usually shows a dense mass, and CT scan or magnetic resonance imaging (MRI) delineates the precise anatomic location of the lesion. Treatment consists of resection of the cyst, which may need to be undertaken in emergency circumstances for airway or cardiac compression. Resection can be performed either as an open procedure or using a thoracoscopic approach.

**Bronchiectasis**

Bronchiectasis is an abnormal and irreversible dilatation of the bronchi and bronchioles associated with chronic suppurative disease of the airways. These children usually will have an underlying congenital pulmonary anomaly, cystic fibrosis, or immunologic deficiency. Bronchiectasis also can result from chronic infection secondary to a neglected bronchial foreign body. The symptoms include a chronic cough, often productive of purulent secretions, recurrent pulmonary infection, and hemoptysis. The diagnosis is suggested by a CXR that shows increased bronchovascular markings in the affected lobe. Chest CT delineates bronchiectasis with excellent resolution. The preferred treatment for bronchiectasis is medical, consisting of antibiotics, postural drainage, and bronchodilator therapy, since many children with the disease show signs of airflow obstruction and bronchial hyperresponsiveness. Lobectomy or segmental resection is indicated for localized disease that has not responded appropriately to medical therapy. In severe cases, lung transplantation may be required to replace the terminally-damaged, septic lung.
Foreign Bodies

The inherent curiosity of children, and their innate propensity to place new objects into their mouths to fully explore them, places them at great risk for aspiration. Aspirated objects can be found either in the airway or in the esophagus, and in both cases the results can be life threatening.

Airway Ingestion

Aspiration of foreign bodies most commonly occurs in toddlers. Peanuts are the most common object that is aspirated, although other materials (e.g., popcorn) may also be involved. A solid foreign body often will cause air trapping, with hyperlucency of the affected lobe or lung seen especially on expiration. Oil from a peanut is irritating and may cause pneumonia. Delay in diagnosis can lead to atelectasis and infection. The most common anatomic location for a foreign body is the right main stem bronchus or the right lower lobe. The child usually will cough or choke while eating, but may then become asymptomatic. Total respiratory obstruction with tracheal foreign body may occur; however, respiratory distress is usually mild if present. A unilateral wheeze is often heard on auscultation. This wheeze often leads to an inappropriate diagnosis of asthma, and may delay the correct diagnosis for some time. CXR will show a radiopaque foreign body, but in the case of nuts, seeds, or plastic toy parts, the only clue may be nondeflation of the affected lobe on an expiratory film or fluoroscopy. Bronchoscopy confirms the diagnosis and allows removal of the foreign body. It can be a simple procedure or it may be extremely difficult, especially with a smooth foreign body that cannot be grasped easily, or one that has been retained for some time. The rigid bronchoscope should be used in all cases, and utilization of the optical forceps facilitates grasping the inhaled object. Epinephrine may be injected into the mucosa when the object has been present for a long period of time, which minimizes bleeding. Bronchiectasis may be seen as an extremely late phenomenon after repeated infections of the poorly aerated lung, and may require partial or total resection of the affected lobe.

Esophagus Ingestion

The most common foreign body found in the esophagus is a coin, followed by small toy parts. Toddlers are most commonly affected. The coin is retained in the esophagus at one of three locations: the cricopharyngeus, the area of the aortic arch, or the gastroesophageal junction; all areas of normal anatomic narrowing. Symptoms are variable depending on the anatomic position of the foreign body and the degree of obstruction. There is often a relatively asymptomatic period after ingestion. The initial symptoms are gastrointestinal and include dysphagia, drooling, and vomiting. The longer the foreign body remains in the esophagus, the greater the incidence of respiratory symptoms, which include cough, stridor, and wheezing. These findings may be interpreted as signs of upper respiratory infections. Objects that are present for a long period of time—particularly in children who have underlying neurologic impairment—may manifest as chronic dysphagia. The CXR is diagnostic in the case of a coin. A contrast swallow may be required for nonradiopaque foreign bodies. Coins lodged within the upper esophagus for less than 24 hours may be removed using Magill forceps. For all
other situations, the treatment is by esophagoscopy, rigid or flexible, and removal of the foreign body. In the case of sharp foreign bodies such as open safety pins, extreme care is required on extraction to avoid injury to the esophagus. Rarely, esophagotomy is required for removal, particularly of sharp objects. Diligent follow-up is required after removal of foreign bodies, especially batteries, which can cause strictures, and sharp objects, which can injure the underlying esophagus.

**ESOPHAGUS**

**Esophageal Atresia and Tracheoesophageal Fistula**

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are among the most gratifying pediatric surgical conditions to treat. In the distant past, nearly all infants born with EA or TEF died. In 1939 Ladd and Leven achieved the first successful repair by ligating the fistula, placing a gastrostomy, and reconstructing the esophagus at a later time. Subsequently, Dr. Cameron Haight in Ann Arbor, Michigan, performed the first successful primary anastomosis for esophageal atresia, which remains the current approach for treatment of this condition. Despite the facts that there are several common varieties of this anomaly, and the underlying cause remains obscure, a careful approach consisting of meticulous perioperative care and attention to the technical detail of the operation can result in an excellent prognosis in most cases.

**Anatomic Varieties**

The five major varieties of EA and TEF are shown in Fig. 38-8. The most commonly seen variety is EA with distal TEF (type C), which occurs in approximately 75-85% of the cases in most series. The next most frequent is pure EA (type A), occurring in 8 to 10% of patients, followed by TEF without EA (type E). This occurs in 5-8% of cases, and also is referred to as an H-type fistula, based on the anatomic similarity to that letter (Fig. 38-9). EA with fistula between both proximal and distal ends of the esophagus and trachea (type D) is seen in approximately 1-2% of cases, and type B, EA with TEF between proximal segments of esophagus and trachea, is seen in approximately 1% of all cases.

**Etiology and Pathologic Presentation**

The esophagus and trachea share a common embryologic origin. They typically divide into separate tubes by approximately the thirty-sixth day of gestation. Failure of this occurrence can result in the spectrum of anomalies seen in Fig. 38-8. Recent studies have shed light on some of the molecular mechanisms underlying this condition. Mice deficient in the Sonic-hedgehog signaling pathway develop a phenotype that includes esophageal atresia-tracheoesophageal fistula (EA-TEF), suggesting a role for this molecule in the pathogenesis of the anomaly in humans. In support of this theory, Sonic-hedgehog transcripts were absent in human esophageal samples obtained from infants with TEF. Similarly, tissue obtained from the fistula tract was found to express thyroid transcription factor one (TTF-1) and fibroblast growth factor (FGF-10), suggesting that the fistula is of respiratory origin. Although a genetic basis for EA-TEF has not been
definitively established, reports indicate that this anomaly may occur in several
generations of the same family. Twin studies also demonstrate the presence of esophageal
atresia in sets of dizygotic twins.

Other congenital anomalies frequently occur in association with EA-TEF. These defects
are known by the acronyms VATER or VACTERL syndrome, which refers to vertebral
(missing vertebra) and anorectal (imperforate anus) anomalies, cardiac defects (severe
congenital cardiac disease), tracheoesophageal fistula, renal anomalies (renal agenesis
and renal anomalies), and radial limb hyperplasia. In nearly 20% of infants born with
esophageal atresia, some variant of congenital heart disease is present.

Clinical Presentation

The anatomic variant of infants with EA-TEF predicts the clinical presentation. When the
esophagus ends either as a blind pouch or as a fistula into the trachea (as in types A, B, C,
or D), infants present with excessive drooling, followed by choking or coughing
immediately after feeding. As a result, aspiration occurs through the fistula tract. As the
neonate coughs and cries, air is transmitted through the fistula into the stomach, resulting
in abdominal distention. As the abdomen distends, it becomes increasingly more difficult
for the infant to breathe. This leads to further atelectasis, which compounds the
pulmonary dysfunction. In patients with type C and D varieties, the regurgitated gastric
juice passes through the fistula, where it collects in the trachea and lungs and leads to a
chemical pneumonitis, which further exacerbates the pulmonary status. In many
instances, the diagnosis is actually made by the nursing staff, who attempt to feed the
baby and notice the accumulation of oral secretions.

The diagnosis of esophageal atresia is confirmed by the inability to pass an orogastric
tube into the stomach (Fig. 38-10). The dilated upper pouch may occasionally be seen on
a plain chest radiograph. If a soft feeding tube is used, the tube will coil in the upper
pouch, which provides further diagnostic certainty. An important alternative diagnosis
that must be considered when an orogastric tube does not enter the stomach is that of an
esophageal perforation. This problem can occur in infants after traumatic insertion of a
nasogastric or orogastric tube. In this instance, the perforation classically occurs at the
level of the piriform sinus, and a false passage is created which prevents the tube from
entering the stomach. Whenever there is any diagnostic uncertainty, a contrast study will
confirm the diagnosis of EA and occasionally document the TEF. The presence of a TEF
can be demonstrated clinically by finding air in the gastrointestinal tract. This can be
proven at the bedside by percussion of the abdomen, and confirmed by obtaining a plain
abdominal radiograph. Occasionally, a diagnosis of EA-TEF can be suspected prenatally
on ultrasound evaluation. Typical features include failure to visualize the stomach and the
presence of polyhydramnios. These findings reflect the absence of efficient swallowing
by the fetus.

In a child with esophageal atresia, it is important to identify whether coexisting anomalies
are present. These include cardiac defects in 38%, skeletal defects in 19%, neurologic
defects in 15%, renal defects in 15%, anorectal defects in 8%, and other abnormalities in
13%. Examination of the heart and great vessels with echocardiography is important to
exclude cardiac defects, as these are often the most important predictors of survival in these infants. The echocardiogram also demonstrates whether the aortic arch is left sided or right sided, which may influence the approach to surgical repair. Vertebral anomalies are assessed by plain radiography, and a spinal ultrasound is obtained if any are detected. A patent anus should be confirmed clinically. The kidneys in a newborn may be assessed clinically by palpation. An ultrasound of the abdomen will demonstrate the presence of renal anomalies, which should be suspected in the child who fails to make urine. The presence of extremity anomalies is suspected when there are missing digits, and confirmed by plain radiographs of the hands, feet, forearms, and legs. Rib anomalies may also be present. These may include the presence of a thirteenth rib.

**Initial Management**

The initial treatment of infants with esophageal atresia-tracheoesophageal fistula includes attention to the respiratory status, decompression of the upper pouch, and appropriate timing of surgery. Because the major determinant of poor survival is the presence of other severe anomalies, a search for other defects including congenital cardiac disease is undertaken in a timely fashion. The initial strategy after the diagnosis is confirmed is to place the neonate in an infant warmer with the head elevated at least 30 degrees. A sump catheter is placed in the upper pouch on continuous suction. Both of these strategies are designed to minimize the degree of aspiration from the esophageal pouch. When saliva accumulates in the upper pouch and is aspirated into the lungs, coughing, bronchospasm, and desaturation episodes can occur, which may be minimized by ensuring the patency of the sump catheter. Intravenous antibiotic therapy is initiated, and warmed electrolyte solution is administered. Where possible, the right upper extremity is avoided as a site to start an intravenous line, as this location may interfere with positioning of the patient during the surgical repair.

The timing of repair is influenced by the stability of the patient. Definitive repair of the EA-TEF is rarely a surgical emergency. If the child is hemodynamically stable and is oxygenating well, definitive repair may be performed within 1 to 2 days after birth. This allows for a careful determination of the presence of coexisting anomalies and for selection of an experienced anesthetic team.

**Management in the Preterm Infant**

The ventilated, premature neonate with EA-TEF and associated hyaline membrane disease represents a patient who may develop severe pulmonary disease. TEF can worsen the fragile pulmonary status as a result of recurrent aspiration through the fistula, and of increased abdominal distention, which impairs lung expansion. Moreover, the elevated airway pressure that is required to ventilate these patients can worsen the clinical course by forcing air through the fistula into the stomach, thereby exacerbating the degree of abdominal distention and compromising lung expansion. In this situation, the first priority is to minimize the degree of positive pressure needed to adequately ventilate the child. This can be accomplished using HFOV. If the gastric distention becomes severe, a gastrostomy tube should be placed. This procedure can be performed at the bedside under local anesthetic, if necessary. The dilated, air-filled stomach can easily be accessed
through an incision in the left upper quadrant of the abdomen. Once the gastrostomy tube is placed, and the abdominal pressure is relieved, the pulmonary status can paradoxically worsen. This is because the ventilated gas may pass preferentially through the fistula, which is the path of least resistance, and bypass the lungs thereby worsening the hypoxemia. To correct this problem, the gastrostomy tube may be placed under water seal, elevated, or intermittently clamped. If these maneuvers are to no avail, ligation of the fistula may be required. This procedure can be performed in the neonatal intensive care unit if the infant is too unstable to be transported to the operating room. These interventions allow for the infant's underlying hyaline membrane disease to improve, for the pulmonary secretions to clear, and for the infant to reach a period of stability so that definitive repair can be performed.

Primary Surgical Correction

In a stable infant, definitive repair is achieved through performance of a primary esophagoesophagostomy. The infant is brought to the operating room, intubated, and placed in the lateral decubitus position with the right side up in preparation for a right posterolateral thoracotomy. If a right-sided arch was determined previously by echocardiography, consideration is given to performing the repair through the left chest, although most surgeons believe that the repair can be performed safely from the right side as well. Bronchoscopy may be performed to exclude the presence of additional, upper pouch fistulae in cases of esophageal atresia (i.e., differentiation of types B, C, and D), and identification of a laryngeotracheoesophageal cleft.

The operative technique for primary repair is as follows (Fig. 38-11). A retropleural approach is generally used, as this technique prevents widespread contamination of the thorax if a postoperative anastomotic leak occurs. The sequence of steps includes:

1. Mobilization of the pleura to expose the structures in the posterior mediastinum.
2. Division of the fistula and closure of the tracheal opening.
3. Mobilization of the upper esophagus sufficiently to permit an anastomosis without tension, and to determine whether a fistula is present between the upper esophagus and the trachea. Forward pressure by the anesthesia staff on the sump drain in the pouch can greatly facilitate dissection at this stage of the operation. Care must be taken when dissecting posteriorly to avoid violation of either the lumen of the trachea or esophagus.
4. Mobilization of the distal esophagus. This needs to be performed judiciously to avoid devascularization, since the blood supply to the distal esophagus is segmental from the aorta. Most of the esophageal length is obtained from mobilizing the upper pouch, since the blood supply travels via the submucosa from above.
5. Performing a primary esophagoesophageal anastomosis. Most surgeons perform this procedure in a single layer using 5-0 sutures. If there is excess tension, the muscle of the upper pouch can be circumferentially incised without compromising blood supply to increase its length. Many surgeons place a transanastomotic feeding tube in order to institute feeds in the early postoperative period.
6. A retropleural drain is placed, and the incision is closed in layers.

Postoperative Course
The postoperative management strategy of patients with EA-TEF is influenced to a great degree by the preference of the individual surgeon and the institutional culture. Many surgeons prefer not to leave the infants intubated postoperatively, to avoid the effects of positive pressure on the site of tracheal closure. However, it may not be possible in babies with preoperative lung disease either from prematurity or pneumonia, or when there is any cord edema. When a transanastomotic tube is placed, feeds are begun slowly in the postoperative period. Some surgeons institute parenteral nutrition for several days, using a central line. The retropleural drain is assessed daily for the presence of saliva, indicating an anastomotic leak. Many surgeons obtain a contrast swallow 1 week after repair to assess the caliber of the anastomosis and to determine whether a leak is present. If there is no leak, feedings are started.

Complications of Surgery

Anastomotic leakage occurs in 10 to 15% of patients, and may be seen either in the immediate postoperative period, or after several days. Early leakage is manifested by a new pleural effusion, pneumothorax, and sepsis, and requires immediate exploration. In these circumstances, the anastomosis may be completely disrupted, possibly due to excessive tension. Revision of the anastomosis may be possible. If not, cervical esophagostomy and gastrostomy placement is required, with a staged procedure to reestablish esophageal continuity. Anastomotic leakage that is detected after several days usually heals without intervention, particularly if a retropleural approach is used. Under these circumstances, broad-spectrum antibiotics, pulmonary toilet, and optimization of nutrition are important. After approximately a week or so, a repeat esophagram should be performed, at which time the leakage may have resolved.

Strictures are not infrequent (10 to 20%), particularly if a leak has occurred. A stricture may become apparent at any time, from the early postoperative period to months or years later. It may present as choking, gagging, or failure to thrive, but often becomes clinically apparent with the transition to eating solid food. A contrast swallow or esophagoscopy is confirmatory, and simple dilatation is usually corrective. Occasionally, repeated dilatations are required. These may be performed in a retrograde fashion, during which a silk suture is placed into the oropharynx and delivered from the esophagus through a gastrostomy tube. Tucker dilators are then tied to the suture and passed in a retrograde fashion from the gastrostomy tube and delivered out of the oropharynx. Increasing sizes are used, and the silk is replaced at the end of the procedure, where it is taped to the side of the face at one end, and to the gastrostomy tube at the other. "Recurrent" tracheoesophageal fistula may represent a missed upper pouch fistula or a true recurrence. This may occur after an anastomotic disruption, during which the recurrent fistula may heal spontaneously. Otherwise, reoperation may be required. Recently, the use of fibrin glue has been successful in treating recurrent fistulas, although long-term follow-up is lacking.

Gastroesophageal reflux commonly occurs after repair of EA-TEF, potentially due to alterations in esophageal motility and the anatomy of the gastroesophageal junction. The clinical manifestations of such reflux are similar to those seen in other infants with primary gastroesophageal reflux disease (GERD). A loose antireflux procedure, such as a
Nissen fundoplication, is used to prevent further reflux, but the child may have feeding problems after antireflux surgery as a result of the innate dysmotility of the distal esophagus. The fundoplication may be safely performed laparoscopically in experienced hands, although care should be taken to ensure that the wrap is not excessively tight.

Special Circumstances

Patients with type E TEFs (also called H-type) most commonly present beyond the newborn period. Presenting symptoms include recurrent chest infections, bronchospasm, and failure to thrive. The diagnosis is suspected using barium esophagography, and confirmed by endoscopic visualization of the fistula. Surgical correction is generally possible through a cervical approach, and requires mobilization and division of the fistula. Outcome usually is excellent.

Patients with duodenal atresia and EA-TEF may require urgent treatment due to the presence of a closed obstruction of the stomach and proximal duodenum. In stable patients, treatment consists of repair of the esophageal anomaly and correction of the duodenal atresia if the infant is stable during surgery. If not, a staged approach should be utilized, consisting of ligation of the fistula and placement of a gastrostomy tube. Definitive repair can then be performed at a later time.

Primary esophageal atresia (type A) represents a challenging problem, particularly if the upper and lower ends are too far apart for an anastomosis to be created. Under these circumstances, treatment strategies include placement of a gastrostomy tube and performing serial bougienage to increase the length of the upper pouch. Occasionally, when the two ends cannot be brought safely together, esophageal replacement is required, using either a gastric pull-up or colon interposition (see below).

Outcome

Various classification systems have been utilized to predict survival in patients with EA-TEF and to stratify treatment. A system devised by Waterston in 1962 was used to stratify neonates based on birth weight, the presence of pneumonia, and the identification of other congenital anomalies. In response to advances in neonatal care, the surgeons from the Montreal Children's Hospital proposed a new classification system in 1993. In the Montreal experience only two characteristics independently affected survival: preoperative ventilator dependence and associated major anomalies. Pulmonary disease as defined by ventilator dependence, appeared to be more accurate than pneumonia. When the two systems were recently compared, the Montreal system more accurately identified children at highest risk. Spitz and colleagues recently analyzed risk factors in infants with EA-TEF who died. Two criteria were found to be important predictors of outcome: birth weight less than 1500 g and the presence of major congenital cardiac disease. A new classification for predicting outcome in esophageal atresia was therefore proposed as follows: group I: birth weight ≥1500 g, without major cardiac disease, survival 97% (283 of 293); group II: birth weight <1500 g, or major cardiac disease, survival 59% (41 of 70); and group III: birth weight <1500 g, and major cardiac disease, survival 22% (2 of 9).
In general, surgical correction of EA-TEF leads to a satisfactory outcome with nearly normal esophageal function in most patients. Overall survival rates of greater than 90% have been achieved in patients classified as stable, in all the various staging systems. Unstable infants have an increased mortality (40 to 60% survival) because of potentially fatal associated cardiac and chromosomal anomalies or prematurity. However, the use of a staged procedure also has increased survival, even in these high-risk infants.

**FIG. 38-8.** The five varieties of esophageal atresia and tracheoesophageal fistula. A. Isolated esophageal atresia. B. Esophageal atresia with tracheoesophageal fistula between proximal segments of esophagus and trachea. C. Esophageal atresia with tracheoesophageal fistula between distal esophagus and trachea. D. Esophageal atresia with fistula between both proximal and distal ends of esophagus and trachea. E. Tracheoesophageal fistula without esophageal atresia (H-type fistula).

**FIG. 38-9.** Barium esophagram showing H-type tracheoesophageal fistula (arrow).

**FIG. 38-10.** Type C esophageal atresia with tracheoesophageal fistula. Note the catheter that is coiled in the upper pouch, and the presence of gas below the diaphragm, which confirms the presence of the tracheoesophageal fistula.


**Corrosive Injury of the Esophagus**

Injury to the esophagus after ingestion of corrosive substances most commonly occurs in toddlers. Both strong alkalies and strong acids produce injury by liquefaction or coagulation necrosis, and since all corrosive agents are extremely hygroscopic, the caustic substance will cling to the esophageal epithelium. Subsequent strictures occur at the anatomically narrowed areas of the esophagus, cricopharyngeus, midesophagus, and gastroesophageal junction. A child who has swallowed an injurious substance may be symptom free, but usually will be drooling and unable to swallow saliva. The injury may be restricted to the oropharynx and esophagus, or may extend to include the stomach. There is no effective immediate antidote. Diagnosis is by careful physical examination of the mouth, and endoscopy with a flexible or rigid esophagoscope. It is important to endoscope only to the first level of the burn in order to avoid perforation. Early barium swallow may delineate the extent of the mucosal injury. It is important to realize that the esophagus may be burned without evidence of injury to the mouth. Although previously used routinely, steroids have not been shown to alter stricture development or modify the extent of injury. Therefore they are no longer part of the management of caustic injuries. Antibiotics are administered during the acute period.

The extent of injury is graded endoscopically as mild, moderate, or severe (grade I, II, or III). Circumferential esophageal injuries with necrosis have an extremely high likelihood of stricture formation. These patients should undergo placement of a gastrostomy tube.
once clinically stable. A string should be inserted through the esophagus, either immediately or during repeat esophagoscopy several weeks later. When established strictures are present (usually after 3 to 4 weeks), dilatation is performed. Retrograde dilatations are safest, using graduated dilators brought through the gastrostomy and advanced into the esophagus via the transesophageal string. For less severe injuries, dilatation may be attempted in antegrade fashion by either graded bougies or balloons. Management of esophageal perforation during dilatation should include antibiotics, irrigation, and closed drainage of the thoracic cavity to prevent systemic sepsis. When recognition is delayed or if the patient is systemically ill, esophageal diversion may be required, with staged reconstruction at a later time.

Although the native esophagus can be preserved in most cases, severe stricture formation that does not respond to dilation is best managed by esophageal replacement. The most commonly used options for esophageal substitution are the colon (right colon or transverse/left colon) and the stomach (gastric tubes or gastric pull-up). Pedicled or free grafts of the jejunum are less commonly used. The right colon is based on a pedicle of the middle colic artery, and the left colon on a pedicle of the middle colic or left colic artery. Gastric tubes are fashioned from the greater curvature of the stomach, based on the pedicle of the left gastroepiploic artery. When the entire stomach is used, as in gastric pull-up, the blood supply is provided by the right gastric artery. The neoesophagus may transverse: (1) subinternally, (2) through a transthoracic route, or (3) through the posterior mediastinum to reach the neck. A feeding jejunostomy is placed at the time of surgery, and tube feedings are instituted once the postoperative ileus has resolved. In a recent review of patients treated by gastric pull-up, long-term outcome was good. Complications included esophagogastric anastomotic leak (n = 15; 36%), which uniformly resolved without intervention, and stricture formation (n = 20; 49%), which responded to a course of dilation. Long-term follow-up has shown that all methods of esophageal substitution can support normal growth and development, and the children enjoy reasonably normal eating habits. Because of the potential for late complications such as ulceration and stricture, follow-up into adulthood is mandatory, but complications appear to diminish with time.

Gastroesophageal Reflux

Gastroesophageal reflux (GER) occurs to some degree in all children, and refers to the passage of gastric contents into the esophagus. By contrast, GERD describes the situation in which reflux is symptomatic. Typical symptoms include failure to thrive, bleeding, stricture formation, reactive airway disease, aspiration pneumonia, or apnea. Failure to thrive and pulmonary problems are particularly common in infants with GERD, whereas strictures and esophagitis are more common in older children and adolescents. GERD is particularly problematic in neurologically-impaired children.

Because all infants experience occasional episodes of GER to some degree, care must be taken before a child is labeled as having pathologic reflux. A history of repeated episodes of vomiting that interferes with growth and development, or the presence of apparent life-threatening events, are required before the diagnosis of GERD can be made. In older children, esophageal bleeding, stricture formation, severe heartburn, or the development
of Barrett's esophagus unequivocally connote pathologic reflux or GERD. In neurologically-impaired children, vomiting due to GER must be distinguished from chronic retching.

The work-up of patients suspected of having GERD includes documentation of the episodes of reflux and evaluation of the anatomy. A barium swallow should be performed as an initial test. This will determine whether there is obstruction of the stomach or duodenum (due to duodenal webs or pyloric stenosis), and will determine whether malrotation is present. The frequency and severity of reflux should be assessed using a 24-hour pH probe study. Although this test is poorly tolerated, it provides the most accurate determination that GERD is present. Esophageal endoscopy with biopsies may identify the presence of esophagitis, and is useful to determine the length of intra-abdominal esophagus and the presence of Barrett's esophagus. Some surgeons obtain a radioisotope "milk scan" to evaluate gastric emptying, although there is little evidence to show that this test changes management when a diagnosis of GERD has been confirmed using the above modalities.

Most patients with GERD are treated initially by conservative means. In the infant, propping up the baby and thickening the formula with rice cereal are generally recommended. Some authors prefer a prone head-up position. In the infant unresponsive to position and formula changes and the older child with severe GERD, medical therapy is based on gastric acid reduction with an H2-blocking agent and/or a proton pump inhibitor. Medical therapy is successful in most neurologically normal infants and younger children, many of whom will outgrow their need for medications. In certain patients, however, medical treatment does not provide symptomatic relief, and surgery is therefore indicated. The least invasive surgical option includes the placement of a nasojejunal or gastrojejunal feeding tube. Because the stomach is bypassed, food contents do not enter the esophagus, and symptoms are often improved. However, as a long-term remedy, this therapy is associated with several problems. The tubes often become dislodged, acid reflux still occurs, and bolus feeding is generally not possible. Fundoplication provides definitive treatment for GER and is highly effective in most circumstances. The fundus may be wrapped around the distal esophagus either 360° (i.e., Nissen), or to lesser degrees (i.e., Thal). At present, the standard approach in most children is to perform these procedures laparoscopically whenever possible. In children with feeding difficulties and in infants under 1 year of age, a gastrostomy tube should be placed at the time of surgery. Early postoperative complications include pneumonia and atelectasis, often due to inadequate pulmonary toilet and pain control, which leads to abdominal splinting. Late postoperative complications include wrap breakdown with recurrent reflux, which may require repeat fundoplication, and dysphagia due to a wrap that is too tight, which generally responds to dilation. These complications are more common in children with neurologic impairment. The keys to successful surgical management of patients with GERD include careful patient selection and meticulous operative technique.

GASTROINTESTINAL TRACT

Hypertrophic Pyloric Stenosis
Clinical Manifestations

Timely diagnosis and treatment of infants with hypertrophic pyloric stenosis (HPS) is extremely gratifying. It is one of the few instances in surgery in which a relatively simple operation can have such a dramatic long-term effect. HPS occurs in approximately 1 in 300 live births, and classically presents in a first-born male between 3 and 6 weeks of age. However, children outside of this age range also are commonly seen, and the disease is by no means restricted to either males or first-born children. The cause of HPS has not been determined. Studies have shown that HPS is found in several generations of the same family, suggesting a familial link. Administration of erythromycin in early infancy has been linked to the subsequent development of HPS, although the cause is unclear.

Infants with HPS present with nonbilious vomiting that becomes increasingly projectile over the course of several days to weeks. Eventually, the infant develops an almost complete gastric outlet obstruction, and is no longer able to tolerate even clear liquids. Despite the recurrent emesis, the child normally has a voracious appetite, leading to a cycle of feeding and vomiting that invariably results in severe dehydration if left untreated. Jaundice may occur in association with HPS, although the reason for this is unclear. Particularly perceptive caregivers will mention that their infant is passing less flatus, which provides a further clue that gastric outlet obstruction is complete.

Infants with HPS develop a hypochloremic, hypokalemic metabolic alkalosis. The urine pH level is high initially, but eventually drops because hydrogen ions are preferentially exchanged for sodium ions in the distal tubule of the kidney as the hypochloremia becomes severe. The diagnosis of pyloric stenosis usually can be made on physical examination by palpation of the typical "olive" in the right upper quadrant, and the presence of visible gastric waves on the abdomen. When the olive cannot be palpated, ultrasound can diagnose the condition accurately in 95% of patients. Criteria for ultrasound diagnosis include a channel length of over 16 mm and pyloric thickness over 4 mm.

Treatment

Pyloric stenosis is never a surgical emergency, although dehydration and electrolyte abnormalities may present a medical emergency. Fluid resuscitation with correction of electrolyte abnormalities and metabolic alkalosis is essential before induction of general anesthesia for surgery. For most infants, fluid containing 5% dextrose and 0.45% saline with added potassium of 2 to 4 mEq/kg given at a rate of approximately 150 to 175 mL/kg for 24 hours will correct the underlying deficit. It is important to ensure that the child has an adequate urine output (>1 mL/kg per hour) as further evidence that rehydration has occurred. After resuscitation, a Fredet-Ramstedt pyloromyotomy is performed (Fig. 38-12). It may be performed using an open or laparoscopic approach. The open pyloromyotomy is performed through either an umbilical or a right upper quadrant transverse abdominal incision. The former route is cosmetically more appealing, although the transverse incision provides easier access to the antrum and pylorus. In recent years, the laparoscopic approach has gained great popularity. Whether done through an open or laparoscopic approach, the operation involves splitting the pyloric
muscle until the submucosa is seen bulging upward. The incision begins at the pyloric vein of Mayo and extends onto the gastric antrum; it typically measures between 1 and 2 cm in length. Postoperatively, intravenous fluids are continued for several hours, after which Pedialyte followed by formula or breast milk are offered, and are gradually increased to 60 mL every 3 hours. Most infants can be discharged home within 24 to 48 hours following surgery. Recently, several authors have shown that ad lib feeds are safely tolerated by the neonate and result in a shorter hospital stay.

The complications of pyloromyotomy include perforation of the mucosa (1 to 3%), bleeding, wound infection, and recurrent symptoms due to inadequate myotomy. When perforation occurs, the mucosa is repaired with a stitch that is placed to tack the mucosa down and reapproximate the serosa in the region of the tear. A nasogastric tube is left in place for 24 hours. The outcome is generally good.

**FIG. 38-12.** Fredet-Ramstedt pyloromyotomy. A. Pylorus delivered into wound and seromuscular layer incised. B. Seromuscular layer separated down to the submucosal base to permit herniation of mucosa through the pyloric incision. C. Cross section demonstrating hypertrophied pylorus, depth of incision, and spreading of muscle to permit mucosa to be herniated through the incision.

**Intestinal and Rectal Disorders in the Newborn**

The cardinal symptom of intestinal obstruction in the newborn is bilious emesis. Prompt recognition and treatment of neonatal intestinal obstruction can truly be life saving. Intestinal obstruction can be thought of as either proximal or distal to the ligament of Treitz. Proximal obstruction presents as bilious vomiting, with minimal abdominal distention. In cases of complete obstruction, there may be a paucity of gas, and no distal air will be seen on the supine and upright films of the abdomen. In this case, the diagnosis of malrotation and midgut volvulus must be excluded. Distal obstruction presents with abdominal distention and bilious emesis. The physical examination will determine whether the anus is patent. Calcifications on the abdominal plain film may indicate meconium peritonitis; pneumatosi and/or free abdominal air indicates necrotizing enterocolitis, with or without intestinal perforation. A contrast enema will show whether there is a microcolon indicative of jejunoileal atresia or meconium ileus. If a microcolon is not present, the diagnoses of Hirschsprung’s disease, small left colon syndrome, or meconium plug syndrome should be considered. In all cases of intestinal obstruction, it is vital to obtain abdominal films in the supine and upright (lateral decubitus) views. This is the only way to assess the presence of air-fluid levels or free air, and to characterize the obstruction as proximal or distal. Moreover, it is important to realize that in the absence of contrast, it is difficult to determine whether a loop of dilated bowel is part of the small or large intestine, as the neonatal bowel lacks typical features, such as haustra or plicae circulares, that characterize these loops in older children or adults. For this reason, care must be taken to ensure that a complete prenatal history is obtained, to perform a thorough physical examination, and to judiciously determine the need for further contrast studies versus immediate abdominal exploration.

**Duodenal Obstruction**
Whenever the diagnosis of duodenal obstruction is entertained, malrotation and midgut volvulus must be excluded. This topic is covered in further detail below. Other causes of duodenal obstruction include duodenal atresia, duodenal web, stenosis, annular pancreas, or duodenal duplication cyst. Duodenal obstruction is easily diagnosed on prenatal ultrasound, which demonstrates the fluid-filled stomach and proximal duodenum as two discrete cystic structures in the upper abdomen. Associated polyhydramnios is common and presents in the third trimester. In 85% of infants with duodenal obstruction, the entry of the bile duct is proximal to the level of obstruction, such that vomiting is bilious. Abdominal distention is typically not present because of the proximal level of obstruction. In those infants with obstruction proximal to the bile duct entry, the vomiting is nonbilious. The classic finding on abdominal radiography is the "double bubble" sign, which represents the dilated stomach and duodenum (Fig. 38-13). In association with the appropriate clinical picture, this finding is sufficient to confirm the diagnosis of duodenal obstruction. However, if there is any uncertainty, particularly when a partial obstruction is suspected, a contrast upper gastrointestinal series is diagnostic.

Treatment. An orogastric tube is inserted to decompress the stomach and duodenum and the infant is given intravenous fluids to maintain adequate urine output. If the infant appears ill, or if abdominal tenderness is present, a diagnosis of malrotation and midgut volvulus should be considered, and surgery should not be delayed. Typically, the abdomen is soft and the infant is stable. Under these circumstances, the infant should be evaluated thoroughly for other associated anomalies. Approximately one-third of newborns with duodenal atresia have associated Down syndrome (trisomy 21). Patients then should be evaluated for associated cardiac anomalies. Once the work-up is complete and the infant is stable, the patient is taken to the operating room and the abdomen is entered through a transverse right upper quadrant supraumbilical incision under general endotracheal anesthesia. Associated anomalies should be sought at the time of the operation. These include malrotation, anterior portal vein, a second distal web, and biliary atresia. The surgical treatment of choice for duodenal obstruction due to duodenal stenosis or atresia or annular pancreas is a duodenoduodenostomy. This procedure can be most easily performed using a proximal transverse-to-distal longitudinal (diamond-shaped) anastomosis. In cases in which the duodenum is extremely dilated, the lumen may be tapered using a linear stapler with a large Foley catheter (≥24F) in the duodenal lumen. It is important to emphasize that an annular pancreas is never divided. Treatment of duodenal web includes vertical duodenotomy, excision of the web, oversewing of the mucosa, and closing the duodenotomy horizontally. Gastrostomy tubes are not placed routinely. Recently reported survival rates exceed 90%. Late complications from repair of duodenal atresia occur in approximately 12 to 15% of patients, and include megaduodenum, intestinal motility disorders, and gastroesophageal reflux.

Intestinal Atresia

Obstruction due to intestinal atresia can occur at any point along the intestinal tract. Most cases are believed to be caused by in utero mesenteric vascular accidents leading to segmental loss of the intestinal lumen. The incidence of intestinal atresia has been estimated to be between 1 in 2000 to 1 in 5000 live births, with equal representation of the sexes. Infants with jejunal or ileal atresia present with bilious vomiting and
progressive abdominal distention. The more distal the obstruction, the more distended the abdomen becomes, and the greater the number of obstructed loops on upright abdominal films (Fig. 38-14).

In cases in which the diagnosis of complete intestinal obstruction is ascertained by the clinical picture and the presence of staggered air-fluid levels on plain abdominal films, the child can be brought to the operating room after appropriate resuscitation. In these circumstances, there is little extra information to be gained by performing a barium enema. By contrast, when there is diagnostic uncertainty, or when distal intestinal obstruction is apparent, a barium enema is useful to establish whether a microcolon is present, and to diagnose the presence of meconium plugs, small left colon syndrome, Hirschsprung's disease, or meconium ileus. Judicious use of barium enema is therefore required in order to safely manage neonatal intestinal obstruction, based on an understanding of the expected level of obstruction.

Surgical correction of the small intestinal atresia should be performed urgently. At laparotomy, one of several types of atresia will be encountered. In type I there is a mucosal atresia with intact muscularis. In type 2 the atretic ends are connected by a fibrous band. In type 3A the two ends of the atresia are separated by a V-shaped defect in the mesentery. Type 3B is an "apple-peel" deformity or "Christmas tree" deformity, in which the bowel distal to the atresia receives its blood supply in a retrograde fashion from the ileocolic or right colic artery (Fig. 38-15). In type 4 atresia, there are multiple atresias with a "string of sausage" or "string of beads" appearance. Disparity in lumen size between the proximal distended bowel and the small diameter of collapsed bowel distal to the atresia has lead to a number of innovative techniques of anastomosis. However, under most circumstances, an anastomosis can be performed using the end-to-back technique, in which the distal, compressed loop is "fish-mouthed" along its antimesenteric border. The proximal distended loop can be tapered as described above. Because the distended proximal bowel rarely has normal motility, the extremely dilated portion should be resected prior to performing the anastomosis.

Occasionally the infant with intestinal atresia will develop ischemia or necrosis of the proximal segment secondary to volvulus of the dilated, bulbous, blind-ending proximal bowel. Under these conditions, an end ileostomy and mucus fistula should be created, and the anastomosis should be deferred to another time, after the infant stabilizes.

Malrotation and Midgut Volvulus

**Embryology.** During the sixth week of fetal development, the midgut grows too rapidly to be accommodated in the abdominal cavity, and it therefore prolapses into the umbilical cord. Between the tenth and twelfth week, the midgut returns to the abdominal cavity, undergoing a 270 degree counterclockwise rotation around the superior mesenteric artery. Because the duodenum also rotates caudal to the artery, it acquires a C-loop which traces this path. The cecum rotates cephalad to the artery, which determines the location of the transverse and ascending colon. Subsequently, the duodenum becomes fixed retroperitoneally in its third portion and at the ligament of Treitz, while the cecum becomes fixed to the lateral abdominal wall by peritoneal bands. The takeoff of the
branches of the superior mesenteric artery elongates and becomes fixed along a line extending from its emergence from the aorta to the cecum in the right lower quadrant. If rotation is incomplete, the cecum remains in the epigastrium, but the bands fixing the duodenum to the retroperitoneum and cecum continue to form. This results in (Ladd's) bands extending from the cecum to the lateral abdominal wall and crossing the duodenum, which creates the potential for obstruction. The mesenteric takeoff remains confined to the epigastrium, resulting in a narrow pedicle suspending all the branches of the superior mesenteric artery and the entire midgut. A volvulus may therefore occur around the mesentery. This twist not only obstructs the proximal jejunum, but also cuts off the blood supply to the midgut. Intestinal obstruction and complete infarction of the midgut occur unless the problem is promptly corrected surgically.

**Presentation and Management.** Midgut volvulus can occur at any age, though it is seen most often in the first few weeks of life. Bilious vomiting is usually the first sign of volvulus, and all infants with bilious vomiting must be evaluated rapidly to ensure that they do not have intestinal malrotation with volvulus. The child with irritability and bilious emesis should raise particular suspicions for this diagnosis. If left untreated, vascular compromise of the midgut initially causes bloody stools, but eventually results in circulatory collapse. Additional clues to the presence of advanced ischemia of the intestine include erythema and edema of the abdominal wall, which progresses to shock and death. It must be reemphasized that the index of suspicion for this condition must be high, since abdominal signs are minimal in the early stages. Abdominal films show a paucity of gas throughout the intestine with a few scattered air-fluid levels (Fig. 38-16). When these findings are present, the patient should undergo immediate fluid resuscitation to ensure adequate perfusion and urine output, followed by prompt exploratory laparotomy.

Often the patient will not appear ill, and the plain films may suggest partial duodenal obstruction. Under these conditions, the patient may have malrotation without volvulus. This is best diagnosed by an upper GI series that shows incomplete rotation with the duodenojejunal junction displaced to the right. The duodenum may have a corkscrew shape indicating volvulus, or complete duodenal obstruction, with the small bowel loops entirely in the right side of the abdomen. Barium enema may show a displaced cecum, but this sign is unreliable, especially in the small infant in whom the cecum is normally in a somewhat higher position than in the older child.

When volvulus is suspected, early surgical intervention is mandatory if the ischemic process is to be avoided or reversed. Volvulus occurs clockwise, and it is therefore untwisted counterclockwise. This can be remembered using the memory aid "turn back the hands of time." Subsequently, Ladd's procedure is performed. This operation does not correct the malrotation, but does broaden the narrow mesenteric pedicle to prevent volvulus from recurring. This procedure is performed as follows (Fig. 38-17): The bands between the cecum and the abdominal wall and between the duodenum and terminal ileum are divided sharply to splay out the superior mesenteric artery and its branches. This maneuver brings the straightened duodenum into the right lower quadrant and the cecum into the left lower quadrant. The appendix is removed to avoid diagnostic errors in later life. No attempt is made to suture the cecum or duodenum in place. With advanced
ischemia, reduction of the volvulus without the Ladd's procedure is accomplished, and a second-look procedure 24 to 36 hours later will often show some vascular recovery. A transparent plastic silo may be placed to facilitate constant evaluation of the intestine, and to plan for the timing of reexploration. Frankly necrotic bowel can then be resected conservatively. With early diagnosis and correction, the prognosis is excellent. However, diagnostic delay can lead to mortality or to short gut syndrome, requiring intestinal transplantation.

A subset of patients with malrotation will demonstrate chronic obstructive symptoms. These symptoms may result from Ladd's bands across the duodenum, or occasionally from intermittent volvulus. Symptoms include intermittent abdominal pain and intermittent vomiting that may occasionally be bilius. Infants with malrotation may demonstrate failure to thrive, and they may be diagnosed initially as having gastroesophageal reflux disease. Surgical correction using Ladd's procedure as described above can prevent volvulus from occurring and improve symptoms in many instances.

Meconium Ileus

**Pathogenesis and Clinical Presentation.** Infants with cystic fibrosis have characteristic pancreatic enzyme deficiencies and abnormal chloride secretion in the intestine that result in the production of viscous, water-poor meconium. Meconium ileus occurs when this thick, highly viscous meconium becomes impacted in the ileum and leads to high-grade intestinal obstruction. Meconium ileus can be either uncomplicated, in which case there is no intestinal perforation, or complicated, in which case prenatal perforation of the intestine has occurred or vascular compromise of the distended ileum develops. Antenatal ultrasound may reveal the presence of intra-abdominal or scrotal calcifications, or distended bowel loops. These infants present shortly after birth with progressive abdominal distention and failure to pass meconium with intermittent bilius emesis. Abdominal radiographs show dilated loops of intestine. Because the enteric contents are so viscous, air-fluid levels do not form, even when obstruction is complete. Small bubbles of gas become entrapped in the inspissated meconium in the distal ileum, where they produce a characteristic "ground glass" appearance on radiograph.

The diagnosis of meconium ileus is confirmed by a contrast enema, which typically demonstrates a microcolon. In patients with uncomplicated meconium ileus, the terminal ileum is filled with pellets of meconium. In patients with complicated meconium ileus, intraperitoneal calcifications form, producing an eggshell pattern on plain abdominal x-ray.

**Management.** The treatment strategy depends on whether the patient has complicated or uncomplicated meconium ileus. Patients with uncomplicated meconium ileus can be treated nonoperatively. Dilute water-soluble contrast is advanced through the colon into the dilated portion of the ileum under fluoroscopic control. Since these contrast agents act partially by absorbing fluid from the bowel wall into the intestinal lumen, maintaining adequate hydration of the infant during this maneuver is extremely important. The enema may be repeated at 12-hour intervals over several days until all the meconium is evacuated. Failure to reflux the contrast into the dilated portion of the ileum signifies the
presence of an associated atresia or complicated meconium ileus, and thus warrants exploratory laparotomy. If surgical intervention is required because of failure of contrast enemas to relieve obstruction, operative irrigation with a dilute contrast agent, the mucolytic N-acetylcysteine, or saline through a purse-string suture may be successful. Alternatively, resection of the distended terminal ileum is performed and the meconium pellets are flushed from the distal small bowel. At this point, ileostomy and mucous fistula may be created from the proximal and distal ends, respectively. Alternatively, a Bishop-Koop anastomosis may be fashioned, or an end-to-end anastomosis may be performed (Fig. 38-18).

Necrotizing Enterocolitis

Clinical Features. Necrotizing enterocolitis (NEC) is the most common and lethal gastrointestinal disorder affecting the intestine of the stressed, preterm neonate. Over 25,000 cases of NEC are reported annually, and the overall mortality ranges between 10 and 50%. Advances in neonatal care such as surfactant therapy, as well as improved methods of mechanical ventilation, have resulted in increasing numbers of low-birth-weight infants surviving neonatal hyaline membrane disease. An increasing proportion of survivors of neonatal respiratory distress syndrome will therefore be at risk for developing NEC. Consequently, it is estimated that NEC soon will surpass respiratory distress syndrome as the principal cause of death in the preterm infant.

Multiple risk factors have been associated with the development of NEC. These include prematurity, initiation of enteral feeding, bacterial infection, intestinal ischemia resulting from birth asphyxia, umbilical artery cannulation, persistence of a patent ductus arteriosus, cyanotic heart disease, and maternal cocaine abuse. Nonetheless, the mechanisms by which these complex interacting etiologies lead to the development of the disease remain undefined. The only consistent epidemiologic precursors for NEC are prematurity and enteral alimentation, representing the commonly encountered clinical situation of a stressed infant who is fed enteraly. Of note, there is some debate regarding the importance of enteral alimentation in the pathogenesis of NEC. A prospective randomized study showed no increase in the incidence of NEC despite an aggressive feeding strategy, and up to 10% of infants with NEC have never received any form of enteral nutrition.

The indigenous intestinal microbial flora have been postulated to play a central role in the pathogenesis of NEC. Bacterial colonization may be a prerequisite for the development of this disease, as oral prophylaxis with vancomycin or gentamicin reduced the incidence of NEC. The importance of bacteria in the pathogenesis of NEC is further supported by the finding that NEC occurs in episodic waves that can be abrogated by infection control measures, and the fact that NEC usually develops at least 10 days postnatally, when the GI tract is colonized by coliforms. Common bacterial isolates from the blood, peritoneal fluid, and stool of infants with advanced NEC include *Escherichia coli*, *Enterobacter*, *Klebsiella*, and occasionally, coagulase-negative *Staphylococcus* species.

NEC may involve single or multiple segments of the intestine, most commonly the terminal ileum, followed by the colon. The gross findings in NEC include bowel
distention with patchy areas of thinning, pneumatosisis, gangrene, or frank perforation. The microscopic features include the appearance of a "bland infarct" characterized by full-thickness necrosis.

**Pathogenesis.** The exact mechanisms that lead to the development of NEC remain incompletely understood. However, current thinking suggests that in the setting of an episode of perinatal stress, such as respiratory distress syndrome, the premature infant suffers a period of intestinal hypoperfusion. This is followed by a period of reperfusion, and the combination of ischemia and reperfusion lead to mucosal injury. The damaged intestinal mucosa can then be readily breached by indigenous microorganisms that translocate across it. The translocated bacteria then initiate an inflammatory cascade that involves the release of various proinflammatory mediators, which in turn may be responsible for further epithelial injury and the systemic manifestations of NEC. It is postulated that maintenance of the gut barrier is essential for the protection of the host against NEC, and that impairment of the mechanisms that normally repair the damaged mucosal barrier may facilitate propagation of the mucosal injury, and thus NEC.

**Clinical Manifestations.** Infants with NEC present with a spectrum of disease. In general, the infants are premature and may have sustained one or more episodes of stress, such as birth asphyxia, or they may have congenital cardiac disease. The clinical picture of NEC has been characterized by Bell and colleagues as progressing from a period of mild illness to that of severe, life-threatening sepsis. Although not all infants progress through the various "Bell stages," this classification scheme provides a useful format to describe the clinical picture associated with the development of NEC. In the earliest stage (Bell stage I), infants present with formula intolerance. This is manifested by vomiting or by finding a large residual volume from a previous feeding in the stomach at the time of the next feeding. Following appropriate treatment, which consists of bowel rest and intravenous antibiotics, many of these infants will not progress to more advanced stages of NEC. These infants are colloquially described as suffering from "NEC scare," and represent a population of neonates that are at risk of developing more severe NEC if a more prolonged period of stress supervenes.

Infants with Bell stage II have established NEC that is not immediately life threatening. Clinical findings include abdominal distention and tenderness, bilious nasogastric aspirate, and bloody stools, which indicate the development of intestinal ileus and mucosal ischemia. Abdominal examination may reveal a palpable mass indicating the presence of an inflamed loop of bowel, diffuse abdominal tenderness, cellulitis, and edema of the anterior abdominal wall. The infant may appear systemically ill, with decreased urine output, hypotension, tachycardia, and noncardiac pulmonary edema. Hematologic evaluation reveals either leukocytosis or leukopenia, an increase in the number of bands, and thrombocytopenia. An increase in the blood urea nitrogen and plasma creatinine levels may be found, which signify the development of renal dysfunction. The diagnosis of NEC may be confirmed by abdominal radiography. The pathognomonic radiographic finding in NEC is pneumatososis intestinalis, which represents invasion of the ischemic mucosa by gas-producing microbes (Fig. 38-19). Other findings include the presence of ileus or portal venous gas. The latter is a transient finding that indicates the presence of severe NEC with intestinal necrosis. A fixed loop of bowel may
be seen on serial abdominal radiographs, which suggests the possibility that a diseased loop of bowel, potentially with a localized perforation, is present. Although these infants are at risk of progressing to more severe disease, with timely and appropriate treatment, they often recover.

Infants with Bell stage III have the most advanced form of NEC. Abdominal radiographs often demonstrate the presence of pneumoperitoneum, indicating that intestinal perforation has occurred. These patients may develop a fulminant course with progressive peritonitis, acidosis, sepsis, disseminated intravascular coagulation, and death.

**Treatment.** In all infants suspected of having NEC, feedings are discontinued, a nasogastric tube is placed, and broad-spectrum parenteral antibiotics are given. The infant is resuscitated, and inotropes are administered to maintain perfusion as needed. Intubation and mechanical ventilation may be required to maintain oxygenation. Total parenteral nutrition is started. Subsequent treatment may be influenced by the particular stage of NEC that is present. Patients with Bell stage I are closely monitored, and generally remain NPO (nil per OS; nothing by mouth) and on intravenous antibiotics for 7 to 10 days prior to reinitiating enteral nutrition. After this time, providing the infant fully recovers, feedings may be reinitiated.

Patients with Bell stage II disease merit close observation. Serial physical examinations are performed to search for the development of diffuse peritonitis, a fixed mass, progressive abdominal wall cellulitis, or systemic sepsis. If infants fail to improve after several days of treatment, consideration should be given to exploratory laparotomy. Paracentesis may be performed, and if the Gram's stain demonstrates multiple organisms and leukocytes, perforation of the bowel should be suspected, and patients should be treated as Bell stage III patients.

In the most severe form of NEC (Bell stage III), patients have definite intestinal perforation, or have not responded to nonoperative therapy. Two schools of thought direct further management. One group favors exploratory laparotomy. At laparotomy, frankly gangrenous or perforated bowel is resected, and the intestinal ends are brought out as stomas. When there is massive intestinal involvement, marginally viable bowel is retained and a second-look procedure is carried out after the infant stabilizes (24 to 48 hours). Patients with extensive necrosis at the second-look may be managed by placing a proximal diverting stoma, resecting bowel that is definitely not viable, and leaving questionably viable bowel behind distal to the diverted segment. When the intestine is viable except for a localized perforation without diffuse peritonitis, and if the infant's clinical condition permits, intestinal anastomosis may be performed either proximal or distal to the divided segment. In cases in which the diseased, perforated segment cannot be safely resected, drainage catheters may be left in the region of the diseased bowel, and the infant is allowed to stabilize.

An alternative approach to the management of infants with perforated NEC involves drainage of the peritoneal cavity. This may be performed under local anesthesia at the bedside, and can be an effective means of stabilizing the desperately-ill infant by relieving increased intra-abdominal pressure and allowing ventilation. When successful,
this method also allows for drainage of perforated bowel by establishing a controlled fistula. Approximately one-third of infants treated with drainage alone survive without requiring additional operations. Infants that do not respond to peritoneal drainage alone after 48 to 72 hours should undergo laparotomy. This procedure allows for the resection of frankly necrotic bowel and diversion of the fecal stream. As well, laparotomy may allow for more effective drainage.

**Outcome.** Survival in patients with NEC is dependent on the stage of disease, the extent of prematurity, and the presence of associated comorbidities. Survival by stage has recently been shown to be approximately 85%, 65%, and 35% percent for stages I, II, and III, respectively. Strictures develop in 20% of medically or surgically treated patients, and a contrast enema is mandatory before reestablishing intestinal continuity. If all other factors are favorable, the ileostomy is closed when the child weighs between 2 and 2.5 kg. At the time of stoma closure, the entire intestine should be examined to search for areas of NEC. Patients that developed massive intestinal necrosis are at risk of developing short-gut syndrome, particularly when the total length of the viable intestinal segment is below 40 cm. These patients require TPN to provide adequate calories for growth and development, and may develop TPN-related cholestasis and hepatic fibrosis. In a significant number of these patients, transplantation of the liver and small bowel may be required.

Intussusception

Intussusception is the leading cause of intestinal obstruction in the young child. It refers to the condition whereby a segment of intestine becomes drawn into the lumen of the more proximal bowel. The process usually begins in the region of the terminal ileum, and extends distally into the ascending, transverse, or descending colon. Rarely, an intussusception may prolapse through the rectum.

The cause of intussusception is not clear, although current thinking suggests that hypertrophy of the Peyer's patches in the terminal ileum from an antecedent viral infection acts as the starting point. Peristaltic action of the intestine then causes the bowel distal to this point to invaginate into itself. Idiopathic intussusception occurs in children between the ages of approximately 6 and 24 months. Beyond this age group, one should consider the possibility that a pathologic starting point may be present. These include polyps, malignant tumors such as lymphoma, enteric duplication cysts, or Meckel's diverticulum. Such intussusceptions are rarely reduced by air or contrast enema, and thus the starting point is identified when operative reduction of the intussusception is performed.

**Clinical Manifestations.** Since intussusception is frequently preceded by a gastrointestinal viral illness, the onset may not be easily determined. Typically, the infant develops paroxysms of crampy abdominal pain and intermittent vomiting. Between attacks, the infant may act normally, but as symptoms progress, increasing lethargy develops. Bloody mucus ("currant-jelly" stool) may be passed per rectum. Ultimately, if reduction is not accomplished, gangrene of the intussusceptum occurs, and perforation may ensue. On physical examination, an elongated mass is detected in the right upper
quadrant or epigastrium, with an absence of bowel in the right lower quadrant (Dance's sign). The mass may be seen on plain abdominal x-ray, but is more easily demonstrated on air or contrast enema.

**Treatment.** Patients with intussusception should be assessed for the presence of peritonitis and for the severity of systemic illness. Following resuscitation and administration of intravenous antibiotics, the child is assessed for suitability to proceed with radiographic versus surgical reduction. In the absence of peritonitis, the child should undergo radiographic reduction. If peritonitis is present, or if the child appears systemically ill, urgent laparotomy is indicated.

In the stable patient, the air enema is both diagnostic and often curative. It constitutes the preferred method of diagnosis and nonoperative treatment of intussusception. Air is introduced with a manometer, and the pressure that is administered is carefully monitored. Under most instances, this should not exceed 120 mm Hg. Successful reduction is marked by free reflux of air into multiple loops of small bowel, and symptomatic improvement as the infant suddenly becomes pain free. Unless both of these signs are observed, it cannot be assumed that the intussusception is reduced. If reduction is unsuccessful, and the infant remains stable, the infant should be brought back to the radiology suite for a repeat attempt at reduction after a few hours. This strategy has improved the success rate of nonoperative reduction in many centers. In addition, hydrostatic reduction with barium may be useful if pneumatic reduction is unsuccessful. The overall success rate of radiographic reduction varies based on the experience of the center, and is typically between 60 and 90%.

If nonoperative reduction is successful, the infant may be given oral fluids after a period of observation. Failure to reduce the intussusception mandates surgery. Two approaches are used. In an open procedure, exploration is carried out through a right lower quadrant incision, delivering the intussuscepted mass into the wound. Reduction usually can be accomplished by gentle distal pressure, where the intussusceptum is gently milked out of the intussuscipiens (Fig. 38-20). Care should be taken not to pull the bowel out, as this can cause damage to the bowel wall. The blood supply to the appendix often is compromised, and appendectomy is performed. If the bowel is frankly gangrenous, resection and primary anastomosis is performed. In experienced hands, laparoscopic reduction may be performed, even in very young infants. This is performed using a 5-mm laparoscope placed in the umbilicus, and two additional 5-mm ports in the left and right lower quadrants. The bowel is inspected, and if it appears to be viable, reduction is performed by milking the bowel or using gentle traction, although this approach is normally discouraged during manual reduction. Atraumatic bowel graspers allow the bowel to be handled without injuring it.

Intravenous fluids are continued until the postoperative ileus subsides. Patients are started on clear liquids and their diet is advanced as tolerated. Of note, recurrent intussusception occurs in 5 to 10% of patients, independent of whether the bowel is reduced radiographically or surgically. Patients present with recurrent symptoms in the immediate postoperative period. Treatment involves repeat air enema, which is successful in most cases. In patients who experience three or more episodes of intussusception, the presence
of a pathologic starting point should be suspected and carefully evaluated using contrast studies. After the third episode of intussusception, many pediatric surgeons will perform an exploratory laparotomy to reduce the bowel and to resect a pathologic starting point if identified.

Appendicitis

**Presentation.** Correct diagnosis of appendicitis in children can be one of the most humbling and challenging tasks facing the pediatric surgeon. The classic presentation is known to all students and practitioners of surgery: generalized abdominal pain that localizes to the right lower quadrant, followed by nausea, vomiting, fever, and localized peritoneal irritation in the region of McBurney's point. When children present in this manner, there should be little diagnostic delay. The child should be made NPO, administered intravenous fluids and broad-spectrum antibiotics, and brought to the operating room for an appendectomy. However, children often do not present in this manner. The coexistence of viral syndromes, and the inability of young children to describe the location and quality of their pain, often results in diagnostic delay. As a result, children with appendicitis often present with perforation, particularly those who are under 5 years of age. Perforation increases the length of hospital stay, and makes the overall course of the illness significantly more complex.

**Diagnosis of Appendicitis in Children.** Controversy exists regarding the role of radiographic studies in the diagnosis of acute appendicitis. Because children have less periappendiceal fat than adults, CT scanning is less reliable in making the diagnosis. In addition, radiation exposure resulting from the CT scan may potentially have long-term adverse effects. Likewise, ultrasonography is neither sufficiently sensitive nor specific to accurately make the diagnosis of appendicitis, although it is useful for excluding ovarian causes of abdominal pain. Therefore the diagnosis of appendicitis remains largely clinical, and each clinician should develop their own threshold to operate or to observe the patient. A reasonable practice guideline is as follows: when the diagnosis is clinically apparent, appendectomy should obviously be performed with minimal delay. Localized right lower quadrant tenderness associated with low-grade fever and leukocytosis in boys should prompt surgical exploration. In girls, ovarian or uterine pathology must also be considered. When there is diagnostic uncertainty, the child may be observed, rehydrated, and reassessed. In girls of menstruating age, an ultrasound may be obtained to exclude ovarian pathology (i.e., cysts, torsion, or tumor). If all studies are negative yet the pain persists, and the abdominal findings remain equivocal, diagnostic laparoscopy may be employed to determine the etiology of the abdominal pain. The appendix should be removed even if it appears to be normal, unless another pathologic cause of the abdominal pain is definitively identified and the appendectomy would substantially increase morbidity.

**Management of the Child with Perforated Appendicitis.** The signs and symptoms of perforated appendicitis can closely mimic those of gastroenteritis, and include abdominal pain, vomiting, and diarrhea. Alternatively, the child may present with symptoms of intestinal obstruction. An abdominal mass may be present in the lower abdomen. When the symptoms have been present for more than 4 or 5 days and an abscess is suspected, it
is reasonable to obtain a CT study of the abdomen and pelvis with intravenous, oral, and rectal contrast in order to visualize the appendix and the presence of an associated abscess, phlegmon, or fecalith (Fig. 38-21).

An individualized approach is necessary for the child who presents with perforated appendicitis. When there is evidence of generalized peritonitis, intestinal obstruction, or systemic toxicity, the child should undergo appendectomy. This should be delayed only for as long as is required to ensure adequate fluid resuscitation and administration of broad-spectrum antibiotics. The operation can be performed through a laparotomy or through a laparoscopic approach. One distinct advantage of the laparoscopic approach is that it provides excellent visualization of the pelvis and all four quadrants of the abdomen. At the time of surgery, adhesions are gently lysed, abscess cavities are drained, and the appendix is removed. Drains are seldom used, and the skin incisions can be closed primarily. If a fecalith is identified outside the appendix on CT scan, every effort should be made to retrieve it and to remove it along with the appendix, if at all possible. Often, the child in whom symptoms have been present for more than 4 or 5 days will present with an abscess cavity without evidence of generalized peritonitis. Under these circumstances, it is appropriate to perform image-guided percutaneous drainage of the abscess, followed by broad-spectrum antibiotic therapy. The inflammation will generally subside within several days, and the appendix can be safely removed on an outpatient basis 6 to 8 weeks later. If the child's symptoms do not improve, or if the abscess is not amenable to percutaneous drainage, then laparoscopic or open appendectomy and abscess drainage is required. Patients who present with a phlegmon in the region of a perforated appendix may be managed in a similar manner. In general, children who are younger than 4 or 5 years of age do not respond as well to an initially nonoperative approach, because their bodies do not localize or isolate the inflammatory process. Thus these patients are more likely to require early surgical intervention. Patients who have had symptoms of appendicitis for no more than 4 days should probably undergo early appendectomy, since the inflammatory response is not as excessive during the initial period and the procedure can be performed safely.

Intestinal Duplications

Duplications represent mucosa-lined structures that are in continuity with the gastrointestinal tract. Although they can occur at any level in the gastrointestinal tract, these inguinal anomalies are found most commonly in the ileum within the leaves of the mesentery. Duplications may be long and tubular, but usually are cystic masses. In all cases, they share a common wall with the intestine. Symptoms associated with enteric duplication cysts include recurrent abdominal pain, emesis from intestinal obstruction, or hematochezia. Such bleeding typically results from ulceration in the duplication, or in the adjacent intestine if the duplication contains ectopic gastric mucosa. On examination, a palpable mass is often identified. Children may also develop intestinal obstruction. Torsion may produce gangrene and perforation.

The ability to make a preoperative diagnosis of enteric duplication cyst usually depends on the presentation. CT, ultrasonography, and technetium pertechnetate scanning can be helpful. Occasionally, a duplication is seen on small bowel follow-through or barium
enema. In the case of short duplications, resection of the cyst and adjacent intestine with end-to-end anastomosis can be performed in a straightforward fashion. If resection of long duplications would compromise intestinal length, multiple enterotomies and mucosal stripping in the duplicated segment will allow the walls to collapse and become adherent. An alternative method is to divide the common wall using the Endo-GIA stapler, forming a common lumen. Patients with duplications who undergo complete excision without compromise of the length of remaining intestine have an excellent prognosis.

Meckel's Diverticulum

A Meckel's diverticulum is a remnant of a portion of the embryonic omphalomesenteric (vitelline) duct. It is located on the antimesenteric border of the ileum, usually within 60 cm of the ileocecal valve (Fig. 38-22). It may be found incidentally at surgery or may present with inflammation, masquerading as appendicitis. Perforation of a Meckel's diverticulum may occur if the outpouching becomes impacted with food, leading to distention and necrosis. Occasionally, bands of tissue extend from the Meckel's diverticulum to the anterior abdominal wall, and these may represent starting points around which internal hernias may develop. This is an important cause of intestinal obstruction in the older child who has a scarless abdomen. Similarly to duplications, ectopic gastric mucosa may produce ileal ulcerations that bleed and lead to the passage of maroon-colored stools. Pancreatic mucosa may also be present. Diagnosis may be made by technetium pertechnetate scans when the patient presents with bleeding. Treatment is surgical. If the base is narrow and there is no mass present in the lumen of the diverticulum, a wedge resection of the diverticulum with transverse closure of the ileum can be performed. A linear stapler is especially useful in this circumstance. When a mass of ectopic tissue is palpable, if the base is wide, or when there is inflammation, it is preferable to perform a resection of the involved bowel and end-to-end ileoileostomy.

Mesenteric Cysts

Mesenteric cysts are similar to duplications in their location within the mesentery. However, they do not contain any mucosa or muscular wall. Chylous cysts may result from congenital lymphatic obstruction. Mesenteric cysts can cause intestinal obstruction or may present as an abdominal mass. The diagnosis may be made by abdominal ultrasound or CT. Treatment involves surgical excision. This may require resection of the adjacent intestine, particularly for extensive, multicystic lesions. In cases in which complete excision is not possible due to the close proximity to vital structures, partial excision or marsupialization should be performed.

Hirschsprung's Disease

Pathogenesis. In his classic textbook titled *Pediatric Surgery*, Dr. Orvar Swenson—who is eponymously associated with one of the classic surgical treatments for Hirschsprung's disease—described this condition as follows: "... congenital megacolon is caused by a malformation in the pelvic parasympathetic system which results in the absence of ganglion cells in Auerbach's plexus of a segment of distal colon. Not only is there an
absence of ganglion cells, but the nerve fibers are large and excessive in number, indicating that the anomaly may be more extensive than the absence of ganglion cells."

This description of Hirschsprung's disease is as accurate today as it was nearly 50 years ago, and summarizes the essential pathologic features of this disease: absence of ganglion cells in Auerbach's plexus and hypertrophy of associated nerve trunks. The cause of Hirschsprung's disease remains incompletely understood, although current thinking suggests that the disease results from a defect in the migration of neural crest cells, which are the embryonic precursors of the intestinal ganglion cell. Under normal conditions, the neural crest cells migrate into the intestine from cephalad to caudal. The process is completed by the twelfth week of gestation, but the migration from midtransverse colon to anus takes 4 weeks. During this latter period, the fetus is most vulnerable to defects in migration of neural crest cells. This may explain why most cases of aganglionosis involve the rectum and rectosigmoid. The length of the aganglionic segment of bowel is therefore determined by the most distal region that the migrating neural crest cells reach. In rare instances, total colonic aganglionosis may occur.

Recent studies have shed light on the molecular basis for Hirschsprung's disease. Patients with Hirschsprung's disease have an increased frequency of mutations in several genes, including \textit{GDNF}, its receptor \textit{Ret}, or its coreceptor \textit{Gfra-1}. Moreover, mutations in these genes also lead to aganglionic megacolon in mice, which provides the opportunity to study the function of the encoded proteins. Initial investigations indicate that \textit{GDNF} promotes the survival, proliferation, and migration of mixed populations of neural crest cells in culture. Other studies have revealed that \textit{GDNF} is expressed in the gut in advance of migrating neural crest cells, and is chemoattractive for neural crest cells in culture. These findings raise the possibility that mutations in the \textit{GDNF} or \textit{Ret} genes could lead to impaired neural crest migration in utero, and the development of Hirschsprung's disease.

\textbf{Clinical Presentation.} The incidence of sporadic Hirschsprung's disease is 1 in 5000 live births. There are reports of increased frequency of Hirschsprung's disease in multiple generations of the same family. Occasionally, such families have mutations in the genes described above, including the \textit{Ret} gene. Because the aganglionic colon does not permit normal peristalsis to occur, the presentation of children with Hirschsprung's disease is characterized by a functional distal intestinal obstruction. In the newborn period, the most common symptoms are abdominal distention, failure to pass meconium, and bilious emesis. Any infant who does not pass meconium beyond 48 hours of life must be investigated for the presence of Hirschsprung's disease. Occasionally, infants present with a dramatic complication of Hirschsprung's disease called enterocolitis. This pattern of presentation is characterized by abdominal distention and tenderness, and is associated with manifestations of systemic toxicity that include fever, failure to thrive, and lethargy. Infants are often dehydrated, and demonstrate a leukocytosis or increase in circulating band forms on hematologic evaluation. On rectal examination, forceful propulsion of foul-smelling liquid feces is typically observed, and represents the accumulation of stool under pressure in an obstructed distal colon. Treatment includes rehydration, systemic antibiotics, nasogastric decompression, and rectal irrigations while the diagnosis of Hirschsprung's disease is being confirmed. In children that do not respond to nonoperative management, a decompressive stoma is required. It is important to ensure
that this stoma is placed in ganglion-containing bowel, which must be confirmed by frozen section at the time of stoma creation.

In approximately 20% of cases, the diagnosis of Hirschsprung's disease is made beyond the newborn period. These children have severe constipation, which has usually been treated with laxatives and enemas. Abdominal distention and failure to thrive may also be present at diagnosis.

**Diagnosis.** The definitive diagnosis of Hirschsprung's disease is made by rectal biopsy. Samples of mucosa and submucosa are obtained at 1 cm, 2 cm, and 3 cm from the dentate line. This can be performed at the bedside in the neonatal period without anesthesia, as samples are taken in bowel that does not have somatic innervation, and is thus not painful to the child. In older children, the procedure should be performed using intravenous sedation. The histopathology of Hirschsprung's disease is the absence of ganglion cells in the myenteric plexuses, increased acetylcholinesterase-positive nerve fibers, and the presence of hypertrophied nerve bundles.

It is important to obtain a barium enema in children in whom the diagnosis of Hirschsprung's disease is suspected. This test may demonstrate the location of the transition zone between the dilated ganglionic colon and the distal constricted aganglionic rectal segment. The authors' practice is to obtain this test before instituting rectal irrigations, so the difference in size between the proximal and distal bowel is preserved. Although the barium enema can only suggest, but cannot reliably establish the diagnosis of Hirschsprung's disease, it is useful in excluding other causes of distal intestinal obstruction. These include small left colon syndrome (as occurs in infants of diabetic mothers), colonic atresia, meconium plug syndrome, or the unused colon observed in infants after the administration of magnesium or tocolytic agents. The barium enema in total colonic aganglionosis may show a markedly shortened colon. Some surgeons have found the use of rectal manometry helpful, particularly in older children, although it is relatively inaccurate.

**Treatment.** The diagnosis of Hirschsprung's disease requires surgery in all cases. The classic surgical approach consisted of a multiple stage procedure. This included a colostomy in the newborn period, followed by a definitive pull-through operation after the child weighed over 10 kg. There are three viable options for the definitive pull-through procedure that are currently used. Although individual surgeons may advocate one procedure over another, studies have demonstrated that the outcome after each type of operation is similar. For each of these procedures, the principles of treatment include confirming the location in the bowel where the transition zone between ganglionic and aganglionic bowel exists, resecting the aganglionic segment of bowel, and performing an anastomosis of ganglionated bowel to either the anus or a cuff of rectal mucosa (Fig. 38-23).

Recently it has been shown that a primary pull-through procedure can be performed safely, even in the newborn period. This approach follows the same treatment principles as a staged procedure, and saves the patient from an additional surgical procedure. Many surgeons perform the intra-abdominal dissection using the laparoscope. This approach is
especially useful in the newborn period, as this provides excellent visualization of the pelvis. In children with significant colonic distention, it is important to allow for a period of decompression using a rectal tube if a single-stage pull-through is to be performed. In older children with an extremely distended, hypertrophied colon, it may be prudent to perform a colostomy to allow the bowel to decompress prior to performing a pull-through procedure. However, it should be emphasized that there is no upper age limit for performing a primary pull-through.

Of the three pull-through procedures performed for Hirschsprung's disease, the first is the original Swenson procedure. In this operation, the aganglionic rectum is dissected in the pelvis and removed down to the anus. The ganglionic colon is then anastomosed to the anus via a perineal approach. In the Duhamel procedure, dissection outside the rectum is confined to the retrorectal space, and the ganglionic colon is anastomosed posteriorly just above the anus. The anterior wall of the ganglionic colon and the posterior wall of the aganglionic rectum are anastomosed using a stapler. Although both of these procedures are extremely effective, they are limited by the possibility of damage to the parasympathetic nerves that are adjacent to the rectum. To circumvent this potential problem, Soave's procedure involves dissection entirely within the rectum. The rectal mucosa is stripped from the muscular sleeve, and the ganglionic colon is brought through this sleeve and anastomosed to the anus. This operation may be performed completely from below. In all cases, it is critical to determine the level at which ganglionated bowel exists. Most surgeons believe that the anastomosis should be performed at least 5 cm from the point at which ganglion cells begin. This avoids performing a pull-through in the transition zone, which is associated with a high incidence of complications due to inadequate emptying of the pull-through segment. Up to one-third of patients who undergo a transition zone pull-through will require a reoperation.

The main complications of all procedures include postoperative enterocolitis, constipation, and anastomotic stricture. As mentioned, long-term results with the three procedures are comparable and generally excellent in experienced hands. These three procedures also can be adapted for total colonic aganglionosis, in which case the ileum is used for the pull-through segment.

Anorectal Malformations

**Anatomic Description.** Anorectal malformations describe a spectrum of congenital anomalies that include imperforate anus and persistent cloaca. Anorectal malformations occur in approximately 1 in 5000 live births and affect males and females almost equally. The embryologic basis includes failure of descent of the urorectal septum. The level to which this septum descends determines the type of anomaly that is present, which subsequently influences the surgical approach.

In patients with imperforate anus, the rectum fails to descend through the external sphincter complex. Instead, the rectal pouch ends blindly in the pelvis, above or below the levator ani muscle. In most cases, the blind rectal pouch communicates more distally with the genitourinary system or with the perineum through a fistulous tract. Traditionally, anatomic description of imperforate anus has been characterized as either
"high" or "low," depending on whether the rectum ends above the levator ani muscle complex or partially descends through this muscle (Fig. 38-24). Based upon this classification system, in male patients with high imperforate anus, the rectum usually ends as a fistula into the membranous urethra. In females, high imperforate anus often occurs in the context of a persistent cloaca. In both males and females, low lesions are associated with a fistula to the perineum. In males, the fistula connects with the median raphe of the scrotum or penis. In females, the fistula may end within the vestibule of the vagina, which is located immediately outside the hymen, or at the perineum.

Because this classification system is somewhat arbitrary, Pena proposed a classification system that specifically and unambiguously describes the location of the fistulous opening. In males the fistula may communicate with: (1) the perineum (cutaneous perineal fistula); (2) the lowest portion of the posterior urethra (rectourethral bulbary fistula); (3) the upper portion of the posterior urethra (rectourethral prostatic fistula); or (4) the bladder neck (rectovesical fistula). In females, the urethra may open onto the perineum between the female genitalia and the center of the sphincter (cutaneous perineal fistula), or into the vestibule of the vagina (vestibular fistula) (Fig. 38-25). In both sexes, the rectum may end in a completely blind fashion (imperforate anus without fistula). In rare cases, patients may have a normal anal canal, yet there may be total atresia or severe stenosis of the rectum.

The most frequent defect in males is imperforate anus with rectourethral fistula, followed by rectoperineal fistula, then rectovesical fistula or recto-bladder neck. In females, the most frequent defect is the rectovestibular defect, followed by the cutaneous perineal fistula. The third most common defect in females is the persistent cloaca. This lesion represents a wide spectrum of malformations in which the rectum, vagina, and urinary tract meet and fuse into a single common channel. On physical examination, a single perineal orifice is observed, and is located at the place where the urethra normally opens. Typically, the external genitalia are hypoplastic.

**Associated Malformations.** Approximately 60% of patients have an associated malformation. The most common is a urinary tract defect, which occurs in approximately 50% of patients. Skeletal defects also are seen, and the sacrum is most commonly involved. Spinal cord anomalies, especially a tethered cord, are common, particularly in children with high lesions. Gastrointestinal anomalies occur, most commonly esophageal atresia. Cardiac anomalies may be noted, and occasionally patients present with a constellation of defects as part of the VACTERL syndrome (vertebral [missing vertebra] and anorectal anomalies, cardiac defects [severe congenital cardiac disease], tracheoesophageal fistula, renal anomalies [renal agenesis and renal anomalies], and radial limb hyperplasia).

**Management of Patients with Imperforate Anus.** Patients with imperforate anus are usually stable, and the diagnosis is readily apparent. Despite the obstruction, the abdomen is initially not distended, and there is rarely any urgency to intervene. The principles of management center around diagnosing the type of defect that is present (high versus low), and evaluating the presence of associated anomalies. It may take up to 24 hours before the presence of a fistula on the skin is noted, and thus it is important to observe the
neonate for some time before definitive surgery is undertaken. All patients should therefore have an orogastric tube placed and be monitored for the appearance of meconium in or around the perineum, or in the urine. Investigation for associated defects should include an ultrasound of the abdomen to assess for the presence of urinary tract anomalies. Other tests should include an echocardiogram and spinal radiographs. An ultrasound of the spine should be performed to look for the presence of a tethered cord. To further classify the location of the fistula as either high or low, a lateral abdominal radiograph can be obtained with a radiopaque marker on the perineum. By placing the infant in the inverted position, the distance between the most distal extent of air in the rectum and the perineal surface can be measured. However, this study is imprecise.

The surgical management of infants with imperforate anus is determined by the anatomic defect. In general, when a low lesion is present, only a perineal operation is required, without a colostomy. Infants with a high lesion require a colostomy in the newborn period, followed by a pull-through procedure at approximately 2 months of age. When a persistent cloaca is present, the urinary tract needs to be carefully evaluated at the time of colostomy formation to ensure that normal emptying can occur, and to determine whether the bladder needs to be drained by means of a vesicostomy. If there is any doubt about the type of lesion, it is safer to perform a colostomy rather than jeopardize the infant's long-term chances for continence by performing an injudicious perineal operation.

The type of pull-through procedure favored by most pediatric surgeons today is the posterior sagittal anorectoplasty (PSARP) procedure, as described by Pena and DeVries. This involves dividing the levator ani and external sphincter complex in the midline posteriorly, and bringing down the rectum after sufficient length is achieved. The muscles are then reconstructed and sutured to the rectum. The outcome of 1192 patients who had undergone this procedure was recently reviewed by Pena and Hong. Seventy-five percent of patients were found to have voluntary bowel movements, and nearly 40% were considered totally continent. As a rule, patients with high lesions demonstrate an increased incidence of incontinence, whereas those with low lesions are more likely to be constipated.

**FIG. 38-13.** Abdominal x-ray showing "double bubble" sign in a newborn infant with duodenal atresia. The two "bubbles" are numbered.

**FIG. 38-14.** Intestinal obstruction in the newborn, showing several loops of distended bowel with air-fluid levels. This child has jejunal atresia.

**FIG. 38-15.** Operative photograph of newborn with "Christmas tree" type of ileal atresia.

**FIG. 38-16.** Abdominal x-ray of a 10-day-old infant with bilious emesis. Note the dilated proximal bowel and the paucity of distal bowel gas, characteristic of a volvulus.

**FIG. 38-17.** Ladd's procedure for malrotation. A. Lysis of cecal and duodenal bands. B. Broadening the mesentery. C. Appendectomy.

**FIG. 38-18.** Techniques of intestinal anastomosis for infants with small bowel obstruction. A. The end-to-back distal limb has been incised, creating a "fishmouth"
to enlarge the lumen. \textit{B}. Bishop-Koop anastomosis. The proximal distended limb is joined to the side of the distal small bowel, which is vented by a "chimney" to the abdominal wall. \textit{C}. Tapering. A portion of the antimesenteric wall of the proximal bowel is excised, with longitudinal closure to minimize disparity in the limbs. \textit{D}. A Mikulicz double-barreled enterostomy is constructed by suturing the two limbs together, and then exteriorizing the double stoma. The common wall can be crushed with a special clamp to create a large stoma. The stoma can be closed in an extraperitoneal manner.

\textbf{FIG. 38-19}. Abdominal radiograph of infant with necrotizing enterocolitis. Arrows point to area of pneumatosis intestinalis.

\textbf{FIG. 38-20}. Open reduction of intussusception, showing how the bowel is milked backward to relieve the obstruction.

\textbf{FIG. 38-21}. CT scan of the abdomen showing the presence of a ruptured appendix with pelvic fluid and a fecalith (arrow).

\textbf{FIG. 38-22}. Operative photograph showing the presence of a Meckel's diverticulum (arrow).

\textbf{FIG. 38-23}. The three operations for surgical correction of Hirschsprung's disease. \textit{A}. The Duhamel procedure leaves the rectum in place and brings ganglionic bowel into the retrorectal space. \textit{B}. The Swenson procedure is a resection with end-to-end anastomosis performed by exteriorizing bowel ends through the anus. \textit{C}. The Soave operation is performed by endorectal dissection and removal of mucosa from the aganglionic distal segment and bringing the ganglionic bowel down to the anus within the seromuscular tunnel.

\textbf{FIG. 38-24}. Low imperforate anus in a male. Note the well developed buttocks. The perineal fistula was found at the midline raphe.

\textbf{FIG. 38-25}. Imperforate anus in a female. A catheter has been placed into the fistula, which is in the vestibule of the vagina.

\section*{JAUNDICE}

\textbf{Approach to the Jaundiced Infant}

Jaundice is present during the first week of life in 60\% of term infants and 80\% of preterm infants. There is usually accumulation of unconjugated bilirubin, but there may also be deposition of direct bilirubin. During fetal life, the placenta is the principal route of elimination of unconjugated bilirubin. In the newborn infant, bilirubin is conjugated through the activity of \textit{glucuronosyltransferase}. In the conjugated form, bilirubin is water soluble, which results in its excretion into the biliary system, then into the gastrointestinal tract. Newborns have a relatively high level of circulating hemoglobin, and relative immaturity of the conjugating machinery. This results in a transient accumulation of bilirubin in the tissues, which is manifested as jaundice. Physiologic jaundice is evident by the second or third day of life, and usually resolves within approximately 5 to 7 days. By definition, jaundice that persists beyond 2 weeks is considered pathologic.
Pathologic jaundice may be due to biliary obstruction, increased hemoglobin load, or to liver dysfunction. The work-up of the jaundiced infant therefore should include a search for the following possibilities: (1) obstructive disorders, including biliary atresia, choledochal cyst, and inspissated bile syndrome; (2) hematologic disorders, including ABO incompatibility, Rh incompatibility, and spherocytosis; (3) metabolic disorders, including \( \alpha_1 \)-antitrypsin deficiency, galactosemia, or pyruvate kinase deficiency; and (4) congenital infection, including syphilis and rubella.

**Biliary Atresia**

**Pathogenesis**

The most important surgical cause of jaundice in the newborn period is biliary atresia. The incidence of this disease is approximately 1 in 20,000. This disease is characterized by an obliterative process of the extrahepatic bile ducts, and is associated with hepatic fibrosis. The etiology is unknown. In the classic textbook *Abdominal Surgery of Infancy and Childhood*, Ladd and Gross described the cause of biliary atresia as an "... arrest of development during the solid stage of bile duct formation." More recent evidence suggests an acquired basis for this disease, and studies in both animals and humans have implicated a role for the immune system and systemic viral infections in its pathogenesis.

**Clinical Presentation**

Jaundice, a constant finding, is usually present at birth or shortly thereafter, but may go undetected or may be regarded as physiologic until the child is 2 or 3 weeks old. The infant demonstrates acholic, grey-appearing stools, secondary to obstructed bile flow. Infants with biliary atresia also manifest progressive failure to thrive, and if untreated, progress to develop stigmata of liver failure and portal hypertension, particularly splenomegaly and esophageal varices. The obliterative process involves the common duct, cystic duct, one or both hepatic ducts, and the gallbladder, in a variety of combinations. Approximately 25% of patients have coincidental malformations often associated with polysplenia, and may include intestinal malrotation, preduodenal portal vein, and intrahepatic vena cava.

**Diagnosis**

Generally, a combination of investigations is required in order to ascertain the diagnosis of biliary atresia, as no single test is sufficiently sensitive or specific. In many centers the nuclear medicine scan using technetium-99m \( ^{99m}\text{Tc} \) iminodiacetate (DISIDA), performed after pretreatment of the patient with phenobarbital, has proven to be an accurate and reliable study. If radionuclide appears in the intestine, extrahepatic bile duct patency is ensured and the diagnosis of biliary atresia is excluded. If radiopharmaceutical is normally concentrated by the liver but not excreted despite treatment with phenobarbital, and the metabolic screen, particularly \( \alpha_1 \)-antitrypsin determination, is normal, the presumptive diagnosis is biliary atresia. An ultrasound may be performed to assess for the presence of other causes of biliary tract obstruction, including choledochal cyst. The presence of a gallbladder is also evaluated, although it is important to
emphasize that the presence of a gallbladder does not exclude the diagnosis of biliary atresia. In approximately 10% of patients, the distal biliary tract is patent and a gallbladder may be visualized, even though the proximal ducts are atretic. It is worth noting that the intrahepatic bile ducts are never dilated in the patient with biliary atresia. A percutaneous liver biopsy may at times differentiate biliary atresia from neonatal hepatitis. When these tests point to a diagnosis of biliary atresia, surgical exploration is warranted. At surgery a cholangiogram is performed, using the gallbladder as a conduit. This demonstrates the anatomy of the biliary tree, determines whether extrahepatic bile duct atresia is present, and evaluates whether there is distal bile flow into the duodenum. The cholangiogram may demonstrate hypoplasia of the extrahepatic biliary system. This condition is associated with hepatic parenchymal disorders that cause severe intrahepatic cholestasis, including alpha1-antitrypsin deficiency and arteriohepatic dysplasia (Alagille's syndrome).

The presentation of biliary atresia closely mimics that of inspissated bile syndrome. This term is applied to patients with normal biliary tracts who have persistent obstructive jaundice. Increased viscosity of bile and obstruction of the canaliculi are implicated as causes. The condition has been seen in infants receiving parenteral nutrition, but it is also encountered in conditions associated with hemolysis, or in cystic fibrosis. In some instances, no etiologic factors can be defined. Neonatal hepatitis may present in a fashion similar to biliary atresia. This disease is characterized by persistent jaundice due to acquired biliary inflammation without obliteration of the bile ducts. There may be a viral etiology, and the disease is usually self-limited.

Treatment

If the intraoperative cholangiogram confirms the presence of biliary atresia, then surgical correction should be immediately undertaken. The most effective surgical treatment for biliary atresia is the portoenterostomy, as described by Kasai. The purpose of this procedure is to promote bile flow into the intestine. The procedure is based on Kasai's observation that the fibrous tissue at the porta hepatis invests microscopically patent biliary ductules, that in turn communicate with the intrahepatic ductal system (Fig. 38-26). Transecting this fibrous tissue, which is invariably encountered cephalad to the bifurcating portal vein, opens these channels and establishes bile flow into a surgically constructed intestinal conduit, usually a Roux-en-Y limb of jejunum (Fig. 38-27). Some authors believe that an intussuscepted antireflux valve is useful in preventing retrograde bile reflux, although the data suggest that it does not impact outcome. A liver biopsy is performed at the time of surgery to determine the degree of hepatic fibrosis that is present. The likelihood of surgical success is increased if the procedure is accomplished before the infant attains the age of 8 weeks. Although the outlook is less favorable for patients after the twelfth week, it is reasonable to proceed with surgery even beyond this point, as the alternative is certain liver failure. It is noteworthy that a significant number of patients do have favorable outcomes when operated on at this time.

Bile drainage is anticipated when the operation is carried out early; however, bile flow does not necessarily imply cure. Approximately one-third of patients remain symptom free after portoenterostomy, the remainder require liver transplantation due to progressive
liver failure. Independent risk factors that predict failure of the procedure include bridging liver fibrosis at the time of surgery and postoperative cholangitic episodes. A recent review of the data of the Japanese Biliary Atresia Registry (JBAR), which includes the results of 1381 patients, showed that the 10-year survival rate without transplantation was 53%; with transplantation the rate was 66.7%. A common postoperative complication is cholangitis. There is no effective strategy to completely eliminate this complication, and the effectiveness of long-term prophylactic antibiotics has not been fully resolved.

**FIG. 38-26.** Operative photograph showing the Kasai portoenterostomy. Arrows denote site of the anastomosis. Note the engorged liver.

**FIG. 38-27.** Schematic illustration of the Kasai portoenterostomy for biliary atresia. An isolated limb of jejunum is brought to the porta hepatis and anastomosed to the transected ducts at the liver plate.

### Choledochal Cyst

**Classification**

The term *choledochal cyst* refers to a spectrum of congenital biliary tract disorders that were previously grouped under the name "idiopathic dilation of the common bile duct." After the classification system proposed by Alonso-Lej, five types of choledochal cyst were described. A type I cyst is characterized by fusiform dilatation of the bile duct. This is the most common type and is found in 80 to 90% of cases. A type II choledochal cyst appears as an isolated diverticulum protruding from the wall of the common bile duct. The cyst may be joined to the common bile duct by a narrow stalk. Type III choledochal cysts arise from the intraduodenal portion of the common bile duct and are also known as choledochoceles. Type IVA cysts consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts. Type IVB choledochal cysts are multiple dilatations involving only the extrahepatic bile ducts. Type V (Caroli's disease) consists of multiple dilatations limited to the intrahepatic bile ducts.

Choledochal cysts are most appropriately considered the predominant feature in a constellation of pathologic abnormalities that can occur within the pancreatobiliary system. Frequently associated with choledochal cyst is an anomalous junction of the pancreatic and common bile ducts. The etiology of choledochal cyst is controversial. Babbit proposed an abnormal pancreatic and biliary duct junction, with the formation of a common channel into which pancreatic enzymes are secreted. This process results in weakening of the bile duct wall by gradual enzymatic destruction, leading to dilatation, inflammation, and finally cyst formation. Not all patients with choledochal cysts demonstrate an anatomic common channel, which raises questions regarding the accuracy of this model.

**Clinical Presentation**

Choledochal cysts are more common in females than in males (4:1). Typically these present in children beyond the toddler age group. The classic symptom triad consists of
abdominal pain, mass, and jaundice. However, this complex is actually encountered in fewer than half of the patients. The more usual presentation is that of episodic abdominal pain, often recurring over the course of months or years, and generally is associated with only minimal jaundice that may escape detection. If the condition is not recognized, the patient develops cholangitis, which may lead to the development of cirrhosis and portal hypertension. Choledochal cysts can present in the newborn period, when the symptoms are similar to those of biliary atresia. Often neonates will have an abdominal mass at presentation.

Diagnosis

A choledochal cyst is frequently diagnosed in the fetus at a screening prenatal ultrasound. In the older child or adolescent, abdominal ultrasonography may reveal a cystic structure arising from the biliary tree. CT scan will confirm the diagnosis. These studies will demonstrate the dimensions of the cyst and define its relationship to the vascular structures in the porta hepatis, as well as the intrahepatic ductal configuration. Endoscopic retrograde cholangiopancreatography (ERCP) is reserved for patients in whom confusion remains after evaluation by less-invasive imaging modalities. Magnetic resonance cholangiopancreatography may provide a more detailed depiction of the anatomy of the cyst, and its relationship to the bifurcation of the hepatic ducts and the pancreas.

The cyst wall is composed of fibrous tissue and is devoid of mucosal lining. As a result, the treatment of choledochal cysts is surgical excision followed by biliary-enteric reconstruction. There is no role for internal drainage by cystenterostomy, which leaves the cyst wall intact and leads to the inevitable development of cholangitis. Rarely, choledochal cyst can lead to the development of a biliary tract malignancy. This provides a further rationale for complete cyst excision.

Resection of the cyst requires circumferential dissection. The posterior plane between the cyst and portal vein must be carefully dissected to accomplish removal. The pancreatic duct, which may enter the distal cyst, is vulnerable to injury during distal cyst excision, but this can be avoided by avoiding entry into the pancreatic parenchyma. In cases in which the pericystic inflammation is extremely dense, it may be unsafe to attempt complete cyst removal. In this instance, it is reasonable to dissect within the posterior wall of the cyst, which allows the inner lining of the back wall to be dissected free from the outer layer that directly overlies the portal vascular structures. The lateral and anterior cyst, as well as the internal aspect of the back wall, is removed, yet the outer posterior wall remains behind. Cyst excision is accomplished, and the proximal bile duct is anastomosed to the intestinal tract.

The prognosis for children who have undergone complete excision of a choledochal cyst is excellent. Complications include anastomotic stricture, cholangitis, and intrahepatic stone formation. These complications may develop a long time after surgery has been completed.

DEFORMITIES OF THE ABDOMINAL WALL
Embryology

The abdominal wall is formed by four separate embryologic folds—cephalic, caudal, and right and left lateral folds—each of which is composed of somatic and splanchnic layers. Each of the folds develops toward the anterior center portion of the celomic cavity, joining to form a large umbilical ring that surrounds the two umbilical arteries, the vein, and the yolk sac or omphalomesenteric duct. These structures are covered by an outer layer of amnion, and the entire unit composes the umbilical cord. Between the fifth and tenth weeks of fetal development, the intestinal tract undergoes rapid growth outside the abdominal cavity within the proximal portion of the umbilical cord. As development is completed, the intestine gradually returns to the abdominal cavity. Contraction of the umbilical ring completes the process of abdominal wall formation.

Failure of the cephalic fold to close results in sternal defects such as congenital absence of the sternum. Failure of the caudal fold to close results in exstrophy of the bladder, and in more extreme cases, exstrophy of the cloaca. Interruption of central migration of the lateral folds results in omphalocele. Gastroschisis, originally thought to be a variant of omphalocele, probably results from a fetal accident in the form of intrauterine rupture of a hernia of the umbilical cord.

Umbilical Hernia

Failure of the umbilical ring to close results in a central defect in the linea alba. The resulting umbilical hernia is covered by normal umbilical skin and subcutaneous tissue, but the fascial defect allows protrusion of abdominal contents. Hernias less than 1 cm in size at the time of birth usually will close spontaneously by 4 years of age. Sometimes the hernia is large enough that the protrusion is disfiguring and disturbing to both the child and the family. In such circumstances early repair may be advisable (Fig. 38-28).

Umbilical hernias are generally asymptomatic protrusions of the abdominal wall. They are generally noted by parents or physicians on physical examination, and present for a surgical opinion out of concerns for possible incarceration. Although incarceration rarely is seen in an umbilical hernia, it can happen. Children present with abdominal pain, bilious emesis, and a tender, hard mass protruding from the umbilicus. This constellation of symptoms mandates immediate exploration and repair of the hernia. More commonly, the child is asymptomatic and treatment is governed by the size of the defect, the age of the patient, and the concern that the child and family have regarding the cosmetic appearance of the abdomen. When the defect is small and spontaneous closure is likely, most surgeons will delay surgical correction until 4 or 5 years of age. If closure does not occur by this time, it is reasonable to repair the hernia. If a younger child has an extremely large hernia, or if the family or child is bothered by the cosmetic appearance, then repair is indicated.

Repair of uncomplicated umbilical hernia is performed under general anesthesia as an outpatient procedure. A small curving incision that fits into the skin crease of the umbilicus is made, and the sac is dissected free from the overlying skin. The fascial
defect is repaired with permanent or long-lasting absorbable interrupted sutures that are placed in a transverse plane. The skin is closed using subcuticular sutures.

**FIG. 38-28.** Umbilical hernia in a 1-year-old female.

**Patent Urachus**

During the development of the celomic cavity, there is free communication between the urinary bladder and the abdominal wall through the urachus, which exits adjacent to the omphalomesenteric duct. Persistence of this tract results in a communication between the bladder and the umbilicus. The first sign of a patent urachus is moisture or urine flow from the umbilicus. Recurrent urinary tract infection can result. The urachus may be partially obliterated, with a remnant remaining beneath the umbilicus in the extraperitoneal position as an isolated cyst that may be identified by ultrasound. Such a cyst usually presents as an inflammatory mass inferior to the umbilicus. Initial treatment is drainage of the infected cyst, followed by cyst excision as a separate procedure once the inflammation has resolved.

In the child with a persistently draining umbilicus, a diagnosis of patent urachus should be considered. The differential diagnosis includes an umbilical granuloma, which generally responds to local application of silver nitrate. The diagnosis of patent urachus is confirmed by umbilical exploration. The urachal tract is excised and the bladder is closed. A patent vitelline duct also may present with umbilical drainage. In this circumstance, there is a communication with the small intestine, often at the site of a Meckel's diverticulum (Fig. 38-29). Treatment includes umbilical exploration with resection of the involved bowel.

**FIG. 38-29.** Patent vitelline duct. Note the communication between the umbilicus and the small bowel at the site of a Meckel's diverticulum.

**Omphalocele**

Clinical Presentation

*Omphalocele* refers to a congenital defect of the abdominal wall in which the bowel and solid viscera are covered by peritoneum and amniotic membrane (Fig. 38-30). The umbilical cord inserts into the sac. The abdominal wall defect can measure 4 cm or more in diameter. The incidence is approximately 1 in 5000 live births, and it occurs in association with special syndromes such as extrophy of the cloaca (vesicointestinal fissure), the Beckwith-Wiedemann constellation of anomalies (macroglossia, macrosomia, hypoglycemia, visceromegaly, and omphalocele), and Cantrell’s pentalogy (lower thoracic wall malformations [cleft sternum], ectopia cordis, epigastric omphalocele, anterior midline diaphragmatic hernia, and cardiac anomalies). The size of the defect may be very small, or large enough that it contains most of the abdominal viscera. There is a 60 to 70% incidence of associated anomalies, especially cardiac (20 to 40% of cases) and chromosomal abnormalities. Chromosomal anomalies are more
common in children with smaller defects. Omphalocele is associated with prematurity (10 to 50% of cases) and intrauterine growth restriction (20% of cases).

**Treatment**

Immediate treatment of an infant with omphalocele consists of attending to the vital signs and maintaining the body temperature. The omphalocele should be covered with saline-soaked gauze and the trunk should be wrapped circumferentially. No pressure should be placed on the omphalocele sac in an effort to reduce its contents, as this maneuver may increase the risk of rupture of the sac, or may interfere with abdominal venous return. Prophylactic antibiotics should be administered in case of rupture. Whenever possible, a primary repair of the omphalocele should be undertaken. This involves resection of the omphalocele membrane and closure of the fascia. A layer of prosthetic material may be required to achieve closure.

Occasionally, an infant will have a giant omphalocele (defect greater than 7 cm in diameter) that cannot be closed primarily because there is simply no room to reduce the viscera into the abdominal cavity (see Fig. 38-30). Other infants may have associated congenital anomalies that complicate surgical repair. Under these circumstances, a nonoperative approach can be used. The omphalocele sac can be treated with desiccating substances such as povidone-iodine. It typically takes 2 to 3 months before reepithelialization occurs. In the past, mercury compounds were used, but have been discontinued because of associated systemic toxicity.

**FIG. 38-30.** Giant omphalocele in a newborn male.

**Gastroschisis**

**Clinical Presentation**

Gastroschisis represents a congenital defect characterized by a defect in the anterior abdominal wall through which the intestinal contents freely protrude. Unlike the omphalocele, there is no overlying sac and the size of the defect is much smaller (<4 cm). The abdominal wall defect is located at the junction of the umbilicus and normal skin, and is almost always to the right of the umbilicus (Fig. 38-31). The umbilicus becomes partly detached, allowing free communication with the abdominal cavity. The appearance of the bowel provides some information with respect to the in utero timing of the defect. The intestine may be normal in appearance, suggesting that the rupture occurred relatively late during the pregnancy. More commonly, however, the intestine is thick, edematous, discolored, and covered with exudate, implying a more long-standing process.

Unlike infants born with omphalocele, associated anomalies seen with gastroschisis consist mostly of intestinal atresia. This defect can readily be diagnosed on prenatal ultrasound (Fig. 38-32). There is no advantage to performing a cesarean section over a vaginal delivery. Even though the thickness of the peel on the surface of the bowel
indicates that a shorter gestational time would be less injurious, there is no benefit to early versus late delivery.

Treatment

All infants born with gastroschisis require urgent surgical treatment. In many instances, the intestine can be returned to the abdominal cavity, and a primary surgical closure of the abdominal wall is performed. Techniques that facilitate primary closure include mechanical stretching of the abdominal wall, thorough orogastric suctioning with foregut decompression, rectal irrigation, and evacuation of all meconium. Care must be taken to prevent increased abdominal pressure during the reduction, which would lead to compression of the inferior vena cava, respiratory dysfunction, and result in abdominal compartment syndrome. To avoid this complication, it is helpful to monitor the bladder or airway pressure during reduction. In infants whose intestine has become thickened and edematous, it may be impossible to reduce the bowel into the peritoneal cavity in the immediate postnatal period. Under such circumstances, a plastic spring-loaded silo can be placed onto the bowel and secured beneath the fascia. The silo covers the bowel and allows for graduated reduction on a daily basis as the edema in the bowel wall decreases (Fig. 38-33). Surgical closure can usually be accomplished within approximately 1 week. A prosthetic piece of material (e.g., GoreTex or Surgisis) may be required to bring the edges of the fascia together. If an atresia is noted at the time of closure, it is prudent to reduce the bowel at the first operation, then to return after several weeks once the edema has resolved to correct the atresia. Intestinal function does not typically return for several weeks in patients with gastroschisis. This is especially true if the bowel is thickened and edematous. As a result, these patients will require central line placement and institution of total parenteral nutrition in order to grow.

**FIG. 38-31.** Gastroschisis in a newborn. Note the location of the umbilical cord, and the edematous, thickened bowel.

**FIG. 38-32.** Prenatal ultrasound of a fetus at 30 weeks' gestation with a gastroschisis. Arrows point to the bowel outside within the amniotic fluid.

**FIG. 38-33.** Use of a silo with a gastroschisis to allow the bowel wall edema to resolve and to facilitate closure of the abdominal wall.

**Prune-Belly Syndrome**

Clinical Presentation

Prune-belly syndrome refers to a disorder that is characterized by a constellation of symptoms including extremely lax lower abdominal musculature, dilated urinary tract including the bladder, and bilateral undescended testes (Fig. 38-34). The term *prune-belly syndrome* appropriately describes the wrinkled appearance of the anterior abdominal wall that characterizes these patients. Prune-belly syndrome is also known as Eagle-Barrett syndrome and the triad syndrome, because of the three major manifestations. The incidence is significantly higher in males. Patients manifest a variety of comorbidities. The most significant is that of pulmonary hypoplasia, which can lead to death in the most
severe cases. Skeletal abnormalities include dislocation or dysplasia of the hip and pectus excavatum.

The major genitourinary manifestation in prune-belly syndrome is ureteral dilation. The ureters are typically long and tortuous, and become more dilated distally. Ureteric obstruction is rarely present, and the dilation is thought to be caused by decreased smooth muscle and increased collagen in the ureters. Approximately 80% of individuals will have some degree of vesicoureteral reflux, which can predispose to urinary tract infection. Despite the marked dilatation of the urinary tract, most children with prune-belly syndrome have adequate renal parenchyma for growth and development. Factors associated with the development of long-term renal failure include the presence of abnormal kidneys on ultrasound or renal scan and persistent pyelonephritis.

Treatment

Despite the ureteric dilation, there is currently no role for ureteric surgery unless an area of obstruction develops. The testes are invariably intra-abdominal and bilateral orchiopexy can be performed in conjunction with abdominal wall reconstruction at 6 to 12 months of age. Despite orchiopexy, fertility in a boy with prune-belly syndrome is unlikely, as spermatogenesis over time is insufficient. Deficiencies in the production of prostatic fluid and a predisposition toward retrograde ejaculation contribute to infertility. Abdominal wall repair is accomplished through an abdominoplasty, which typically requires a transverse incision in the lower abdomen extending into the flanks.

FIG. 38-34. Eagle-Barrett (prune-belly) syndrome. Notice the lax, flaccid abdomen.

Inguinal Hernia

An understanding of the management of pediatric inguinal hernias is a central component of modern pediatric surgical practice. Inguinal hernia repair represents one of the most common operations performed in children. The presence of an inguinal hernia in a child is an indication for surgical repair. Surgery to repair an inguinal hernia is termed a herniorrhaphy, because it involves closing off the patent processus vaginalis. This is in contrast with the hernioplasty that is performed in adults, which requires reconstruction of the inguinal floor.

Embryology

In order to understand how to diagnose and treat inguinal hernias in children, it is critical to understand the embryologic origin. It is useful to describe these events to the parents, who are often under the misconception that the hernia was somehow caused by their inability to console their crying child, or the child's high activity level. Inguinal hernia results from a failure of closure of the processus vaginalis, a finger-like projection of the peritoneum that accompanies the testicle as it descends into the scrotum. Closure of the processus vaginalis normally occurs a few months prior to birth. This explains the high incidence of inguinal hernias in premature infants. When the processus vaginalis remains
completely patent, a communication persists between the peritoneal cavity and the groin, resulting in a hernia. Partial closure can result in entrapped fluid, which results in the presence of a hydrocele. A communicating hydrocele refers to a hydrocele that is in communication with the peritoneal cavity, and can therefore be thought of as a hernia. Using the classification system that is typically applied to adult hernias, all congenital hernias in children are by definition indirect inguinal hernias. Children also present with direct inguinal and femoral hernias, although these are much less common.

Clinical Manifestations

Inguinal hernias occur more commonly in males than females (10:1), and are more common on the right side than the left. Infants are at high risk for incarceration of an inguinal hernia because of the narrow inguinal ring. Patients most commonly present with a groin bulge that is noticed by the parents as they change the diaper (Fig. 38-35). Older children may notice the bulge themselves. On examination, the cord on the affected side will be thicker, and pressure on the lower abdomen usually will display the hernia on the affected side. The presence of an incarcerated hernia is manifested by a firm bulge that does not spontaneously resolve, and may be associated with fussiness and irritability in the child. The infant that has a strangulated inguinal hernia will manifest an edematous, tender bulge in the groin, occasionally with overlying skin changes. The child will eventually develop intestinal obstruction, peritonitis, and systemic toxicity.

Usually an incarcerated hernia can be reduced. Occasionally this may require light sedation. Gentle pressure is applied on the sac from below in the direction of the internal inguinal ring. Following reduction of the incarcerated hernia, the child may be admitted for observation, and herniorrhaphy is performed within the next 24 hours to prevent recurrent incarceration. Alternatively, the child may be scheduled for surgery at the next available time slot. If the hernia cannot be reduced, or if evidence of strangulation is present, emergency surgery is necessary. This may require a laparotomy and bowel resection.

When the diagnosis of inguinal hernia is made in an otherwise normal child, operative repair should be planned. Spontaneous resolution does not occur and therefore a nonoperative approach can never be justified. An inguinal hernia in a female frequently contains an ovary rather than intestine. Although the gonad usually can be reduced into the abdomen by gentle pressure, it often prolapses in and out until surgical repair is carried out. In some patients, the ovary and fallopian tube constitute one wall of the hernial sac (sliding hernia), and in these patients the ovary can be reduced effectively only at the time of operation. If the ovary is irreducible, prompt hernia repair is indicated to prevent ovarian torsion or strangulation.

When a hydrocele is diagnosed in infancy and there is no evidence of a hernia, observation is proper therapy until the child is older than 12 months. If the hydrocele has not disappeared by 12 months, invariably there is a patent processus vaginalis, and operative hydrocelectomy with excision of the processus vaginalis is indicated. When the first signs of a hydrocele are seen after 12 months of age, the patient should undergo elective hydrocelectomy, which in a child is always performed through a groin incision.
Aspiration of hydroceles is discouraged, since almost all without a patent processus vaginalis will resorb spontaneously, and those with a communication to the peritoneum will recur and require operative repair eventually.

Surgical Repair

The repair of a pediatric inguinal hernia can be extremely challenging, particularly in the premature child with incarceration. A small incision is made in a skin crease in the groin directly over the internal inguinal ring. Scarpa's fascia is seen and divided. The external oblique muscle is dissected free from overlying tissue, and the location of the external ring is confirmed. The external oblique aponeurosis is then opened along the direction of the external ring. The undersurface of the external oblique is then cleared from surrounding tissue. The cremasteric fibers are separated from the cord structures and hernia sac, and then are elevated into the wound. Care is taken not to grasp the vas deferens. The hernia sac then is dissected up to the internal ring and doubly suture ligated. The distal part of the hernia sac is opened widely to drain any hydrocele fluid. When the hernia is very large and the patient very small, tightening of the internal inguinal ring or even formal repair of the inguinal floor may be necessary, although the vast majority of children do not require any treatment beyond high ligation of the hernia sac.

Controversy exists regarding the role for exploration of an asymptomatic opposite side in a child with an inguinal hernia. Several reports indicate that frequency of a patent processus vaginalis on the side opposite the obvious hernia is approximately 30%, although this figure decreases with increasing age of the child. Management options include never exploring the opposite side or to explore only under certain conditions, such as in premature infants or in patients in whom incarceration is present. The opposite side may be explored laparoscopically. To do so, a blunt 4-mm trocar is placed into the hernia sac of the affected side. The abdominal cavity is insufflated, and the 4-mm 70 degree camera is placed through the trocar such that the opposite side is visualized. The status of the processus vaginalis on the opposite side can be visualized. However, the presence of a patent processus vaginalis by laparoscopy does not always imply the presence of a hernia.

Several authors have now reported a completely laparoscopic approach in the management of inguinal hernias in children. This technique requires insufflation through the umbilicus, and the placement of an extraperitoneal suture to ligate the hernia sac. Proponents of this procedure emphasize the fact that no groin incision is used and there is a decreased chance of injuring cord structures. The long-term results of this technique remain to be established.

Inguinal hernias in children recur in less than 1% of patients, and recurrences usually result from missed hernia sacs at the first procedure, a direct hernia, or a missed femoral hernia. All children should have local anesthetic administered either by caudal injection or by direct injection into the wound. Spinal anesthesia in preterm infants decreases the risk of postoperative apnea when compared with general anesthesia.
FIG. 38-35. Right inguinal hernia in a 4-month-old male. The arrows point to the bulge in the right groin.

GENITALIA

Undescended Testis

Embryology

The term undescended testicle (cryptorchidism) refers to the interruption of the normal descent of the testis into the scrotum. The testicle may reside in the retroperitoneum, in the internal inguinal ring, in the inguinal canal, or even at the external ring. The testicle begins as a thickening on the urogenital ridge in the fifth to sixth week of embryologic life. In the seventh and eighth months the testicle descends along the inguinal canal into the upper scrotum, and with its progress the processus vaginalis is formed and pulled along with the migrating testicle. At birth, approximately 95% of infants have the testicle normally positioned in the scrotum.

A distinction should be made between the undescended testicle and the ectopic testicle. By definition, an ectopic testis is one that has passed through the external ring in the normal pathway, and then has come to rest in an abnormal location overlying the rectus abdominis or external oblique muscle, the soft tissue of the medial thigh, or behind the scrotum in the perineum. A congenitally absent testicle results from failure of normal development or an intrauterine accident leading to loss of blood supply to the developing testicle.

Clinical Presentation

The incidence of undescended testes is approximately 30% in preterm infants, and 1 to 3% at term. For diagnosis, the child should be examined in the supine position, where visual inspection may reveal a hypoplastic or poorly rugated scrotum. A unilateral undescended testicle usually can be palpated in the inguinal canal or in the upper scrotum. Occasionally, the testicle will be difficult or impossible to palpate, indicating either an abdominal testicle or congenital absence of the gonad. If the testicle is not palpable in the supine position, the child should be examined with his legs crossed while seated. This maneuver diminishes the cremasteric reflex and facilitates identification of the location of the testicle.

It is now established that cryptorchid testes demonstrate an increased predisposition to malignant degeneration. In addition, fertility is decreased when the testicle is not in the scrotum. For these reasons, surgical placement of the testicle in the scrotum (orchidopexy) is indicated. It should be emphasized that this procedure does improve the fertility potential, although fertility is never normal. Similarly, the testicle is still at risk of malignant change, although its location in the scrotum facilitates potentially earlier detection of a testicular malignancy. Other reasons to consider orchidopexy include the risk of trauma to the testicle located at the pubic tubercle, an increased incidence of torsion, and the psychologic impact of an empty scrotum in a developing male. The
reason for malignant degeneration is not established, but the evidence points to an inherent abnormality of the testicle that predisposes it to incomplete descent and malignancy, rather than malignancy as a result of an abnormal environment.

Treatment

Males with bilateral undescended testicles are often infertile. When the testicle is not within the scrotum, it is subjected to a higher temperature, resulting in decreased spermatogenesis. Mengel and coworkers studied 515 undescended testicles by histology and demonstrated a decreasing presence of spermatogonia after 2 years of age. Consequently it is now recommended that the undescended testicle be surgically repositioned by 2 years of age. Despite orchidopexy, the incidence of infertility is approximately two times higher in men with unilateral orchidopexy compared to men with normal testicular descent.

The use of chorionic gonadotropin occasionally may be effective in patients with bilateral undescended testes, suggesting that these patients are more apt to have a hormone deficiency than children with unilateral undescended testicle. If there is no testicular descent after a month of endocrine therapy, operative correction should be undertaken. A child with unilateral cryptorchidism should have surgical correction of the problem. The operation is typically performed through a combined groin and scrotal incision. The cord vessels are fully mobilized, and the testicle is placed in a dartos pouch within the scrotum. An inguinal hernia often accompanies a cryptorchid testis. This should be repaired at the time of orchidopexy.

Patients with a nonpalpable testicle present a challenge in management. The current approach involves laparoscopy to identify the location of the testicle. If the spermatic cord is found to traverse the internal ring, or the testis is found at the ring and can be delivered into the scrotum, a groin incision is made and an orchidopexy is performed. If an abdominal testis is identified that is too far to reach the scrotum, a two-staged Fowler-Stephens approach is used. In the first stage, the testicular vessels are clipped laparoscopically. The orchidopexy then is performed through the groin approximately 6 months later, after which time collateral flow supplies the testicle. It is preferable to preserve the testicular vessels whenever possible. When the testicle is within 1 or 2 cm from the ring, its blood supply may be preserved by mobilizing the testicular vessels up to the renal hilum, then releasing the peritoneal attachments. This often provides sufficient length to allow an orchidopexy to be performed through the groin.

Some patients who have an absent testis are greatly bothered by this anatomic deficiency. Prostheses of all sizes are now available, and can be simply inserted into the scrotum, achieving normal appearance and a normal structure for palpation. Any patient who has an undescended testicle corrected surgically should be examined yearly by the surgeon until his mid-teenage years. At that time, the individual should be thoroughly informed about the possibility of malignant degeneration, and be instructed in self-examination, which should be carried out at least twice a year for life.

Vaginal Anomalies
Surgical diseases of the vagina in children are either congenital or acquired. Congenital anomalies include a spectrum of diseases that include simple defects (imperforate hymen) to more complex forms of vaginal atresia, including distal, proximal, and most severe, complete. These defects are produced by abnormal development of mullerian ducts and/or urogenital sinus. The diagnosis is made most often by physical examination. Secretions into the obstructed vagina produce hydrocolpos, which may present as a large, painful abdominal mass. The anatomy may be defined using ultrasound. Pelvic MRI provides the most thorough and accurate assessment of the pelvic structure. Treatment is dependent on the extent of the defect. For an imperforate hymen, division of the hymen is curative. More complex forms of vaginal atresia require mobilization of the vaginal remnants and creating an anastomosis at the perineum. Laparoscopy can be extremely useful in mobilizing the vagina, in draining hydrocolpos, and in evaluating the internal genitalia. Complete vaginal atresia requires the construction of skin flaps or the creation of a neovagina using a segment of colon.

The most common acquired disorder of the vagina is the straddle injury. This often occurs as young girls fall on blunt objects that cause a direct injury to the perineum. Typical manifestations include vaginal bleeding and inability to void. Unless the injury is extremely superficial, patients should be examined in the operating room, where the lighting is optimal and sedation can be administered. Vaginal lacerations are repaired using absorbable sutures, and the proximity to the urethra should be carefully assessed. Prior to discharge, it is important that patients be voiding spontaneously. In all cases of vaginal trauma, it is essential that the patient be assessed for the presence of sexual abuse.

**Ovarian Cysts and Tumors**

Pathologic Classification

Ovarian cysts and tumors may be classified as non-neoplastic or neoplastic. Non-neoplastic lesions include cysts (e.g., simple, follicular, inclusion, paraovarian, or corpus luteum), endometriosis, and inflammatory lesions. Neoplastic lesions are classified based on the three primordia that contribute to the ovary: mesenchymal components of the urogenital ridge, germinal epithelium overlying the urogenital ridge, and germ cells migrating from the yolk sac. The most common variety is germ cell tumors. Germ cell tumors are classified based on the degree of differentiation and the cellular components involved. The least differentiated tumors are the dysgerminomas, which share features similar to the seminoma in males. Although these are malignant tumors, they are extremely sensitive to radiation and chemotherapy. The most common lesions are the teratomas, which may be mature, immature, or malignant. The degree of differentiation of the neural elements of the tumor determines the degree of immaturity. The sex cord stromal tumors arise from the mesenchymal components of the urogenital ridge. These include the granulosa-thecal cell tumors and the Sertoli-Leydig cell tumors. These tumors often produce hormones that result in precocious puberty or hirsutism, respectively. Although rare, epithelial tumors do occur in children. These include serous and mucinous cystadenomas.

Clinical Presentation
Children with ovarian lesions usually present with abdominal pain. Other signs and symptoms include a palpable abdominal mass, evidence of urinary obstruction, symptoms of bowel obstruction, and endocrine imbalance. The surgical approach depends on the appearance of the mass at operation (i.e., whether it is benign-appearing or is suspicious for malignancy). In the case of a simple ovarian cyst, surgery depends on the size of the cyst and the degree of symptoms it causes. In general, large cysts (over 4 to 5 cm) should be resected, as they are unlikely to resolve, may be at risk of torsion, and may mask an underlying malignancy. Resection may be performed laparoscopically, and ovarian tissue should be spared in all cases.

Surgical Management

For ovarian lesions that appear malignant, it is important to obtain tumor markers including alpha-fetoprotein (teratomas), lactate dehydrogenase (dysgerminoma), beta human chorionic gonadotropin (choriocarcinoma), and CA-125 (epithelial tumors). Although the diagnostic sensitivity of these markers is not always reliable, they provide material for postoperative follow-up and indicate the response to therapy. When a malignancy is suspected, the patient should undergo a formal cancer operation. This procedure is performed through either a midline incision or a Pfannenstiel approach. Ascites and peritoneal washings should be collected for cytologic study. The liver and diaphragm are inspected carefully for metastatic disease. An omentectomy is performed if there is any evidence of tumor present. Pelvic and para-aortic lymph nodes are biopsied and the primary tumor is resected completely. Finally, the contralateral ovary is carefully inspected, and if a lesion is seen, it should be biopsied. Dysgerminomas and epithelial tumors may be bilateral in up to 15% of cases. It is occasionally possible to preserve the ipsilateral fallopian tube. More radical procedures are not indicated.

Ovarian Cysts in the Newborn

An increasing number of ovarian cysts are being detected by prenatal ultrasonography. In the past, surgical excision was recommended for all cysts greater than 5 cm in diameter because of the perceived risk of ovarian torsion. More recently, it has become apparent from serial US examinations that many of these lesions will resolve spontaneously. Therefore asymptomatic simple cysts may be observed, and surgery can be performed only when they fail to decrease in size or become symptomatic. Typically, resolution occurs by approximately 6 months of age. A laparoscopic approach may be utilized. By contrast, complex cysts of any size require surgical intervention at presentation.

Ambiguous Genitalia

Embryology

Normal sexual differentiation occurs in the sixth fetal week. In every fetus, wolffian (male) and mullerian (female) ducts are present until the onset of sexual differentiation. Normal sexual differentiation is directed by the sex-determining region of the Y chromosome (SRY). This is located on the distal end of the short arm of the Y chromosome. SRY provides a genetic switch that initiates gonadal differentiation in the
mammalian urogenital ridge. Secretion of mullerian inhibiting substance (MIS) by the Sertoli cells of the seminiferous tubules results in regression of the mullerian duct, the anlagen of the uterus, fallopian tubes, and the upper vagina. The result of MIS secretion therefore is a phenotypic male. In the absence of SRY in the Y chromosome, MIS is not produced, and the mullerian duct derivatives are preserved. Thus the female phenotype prevails.

In order for the male phenotype to develop, the embryo must have a Y chromosome, the SRY must be normal without point mutations or deletions, testosterone and MIS must be produced by the differentiated gonad, and the tissues must respond to these hormones. Any disruption of the orderly steps in sexual differentiation may be reflected clinically as variants of the intersex syndromes.

These may be classified as (1) true hermaphroditism (with ovarian and testicular gonadal tissue), (2) male pseudohermaphroditism (testicles only), (3) female pseudohermaphroditism (ovarian tissue only), and (4) mixed gonadal dysgenesis (usually underdeveloped or imperfectly formed gonads).

True Hermaphroditism

This represents the rarest form of ambiguous genitalia. Patients have both normal male and female gonads, with an ovary on one side and a testis on the other. Occasionally, an ovotestis is present on one or both sides. The majority of these patients have a 46, XX karyotype. Both the testis and the testicular portion of the ovotestis should be removed.

Male Pseudohermaphroditism

This condition occurs in infants with an XY karyotype, but deficient masculinization of the external genitalia. Bilateral testes are present, but the duct structures differentiate partly as phenotypic females. The causes include inadequate testosterone production due to biosynthetic error, inability to convert testosterone to dihydrotestosterone due to 5-alpha-reductase deficiency, or deficiencies in androgen receptors. The latter disorder is termed testicular feminization syndrome. Occasionally, the diagnosis in these children is made during routine inguinal herniorrhaphy in a phenotypic female at which time testes are found. The testes should be resected due to the risk of malignant degeneration, although this should be performed only after a full discussion with the family has occurred.

Female Pseudohermaphroditism

These children commonly have congenital adrenal hyperplasia. They have a 46, XX karyotype, but have been exposed to excessive androgens in utero. Common enzyme deficiencies include 21-hydroxylase, 11-hydroxylase, and 3-beta hydroxysteroid dehydrogenase. These deficiencies result in overproduction of intermediary steroid hormones, which result in masculinization of the external genitalia of the XX fetus. These patients are unable to synthesize cortisol. In 90% of cases, deficiency of 21-hydroxylase causes adrenocorticotropic hormone (ACTH) to stimulate the secretion of excessive quantities of adrenal androgen, which masculinizes the developing female.
These infants are prone to salt loss, and require cortisol replacement. Those with mineralocorticoid deficiency also require fludrocortisone replacement.

**FIG. 38-36.** Ambiguous genitalia manifest as enlarged clitoris and labioscrotal folds in a baby with the adrenogenital syndrome.

**Mixed Gonadal Dysgenesis**

This syndrome is associated with dysgenetic gonads and retained mullerian structures. The typical karyotype is mosaic, usually 45XO,46XY. A high incidence of malignant tumors occur in the dysgenetic gonads, most commonly gonadoblastoma. Therefore they should be removed.

**Management**

In the differential diagnosis of patients with intersex anomalies, the following diagnostic steps are necessary: (1) evaluation of the genetic background and family history; (2) assessment of the anatomic structures by physical examination, ultrasound and/or MRI; (3) chromosome studies; (4) determination of biochemical factors in serum and urine to evaluate the presence of an enzyme defect; and (5) laparoscopy for gonadal biopsy and further evaluation of the anatomy. Treatment should include correction of electrolyte and volume losses in cases of congenital adrenal hyperplasia, and replacement of hormone deficiency. Surgical assignment of gender is controversial, and must take into account considerations of the anatomy as well as the psychosocial effects involved. In most instances, female gender is assigned. There are those who believe that anatomy and endocrine studies may not be sufficiently reliable to accurately assign a particular gender, and that pre- and postnatal hormones have a significant impact on gender identity. In general terms, surgical reconstruction should be performed after a full work-up, and with the involvement of pediatric endocrinologists, pediatric plastic surgeons, and ethicists with expertise in gender issues and the family. This approach will serve to reduce the anxiety associated with these disorders, and will help to ensure the normal physical and emotional development of these patients.

**PEDIATRIC MALIGNANCY**

**Introduction**

Cancer is the second leading cause of death in children after trauma, and accounts for approximately 11% of all pediatric deaths in the United States. Several features distinguish pediatric from adult cancers, including the presence of tumors that are predominantly seen in children, such as neuroblastoma and germ cell tumors, and the favorable response to chemotherapy observed for many pediatric solid malignancies, even in the presence of metastases.

**Wilms' Tumor**

**Clinical Presentation**
Wilms' tumor is the most common primary malignant tumor of the kidney in children. There are approximately 500 new cases annually in the United States, and most are diagnosed between 1 and 5 years with the peak incidence at age 3. Advances in the care of patients with Wilms' tumor have resulted in an overall cure rate of roughly 90%, even in the presence of metastatic spread. The tumor usually develops in otherwise healthy children, as an asymptomatic mass in the flank or upper abdomen. Frequently, the mass is discovered by a parent while bathing or dressing the child. Other symptoms include hypertension, hematuria, obstipation, or weight loss. Occasionally the mass is discovered following blunt abdominal trauma.

Genetics

Wilms' tumor can arise from both germline and somatic mutations, and can occur in the presence or absence of a family history. Nearly 97% of Wilms' tumors are sporadic in that they occur in the absence of a heritable or congenital cause or risk factor. When a heritable risk factor is identified, the affected children often present at an earlier age, and are frequently bilateral. Most of these tumors are associated with germline mutations. It is well established that there is a genetic predisposition to Wilms' tumor in the WAGR syndrome, which consists of Wilms' tumour, aniridia, genitourinary abnormalities, and mental retardation. In addition, there is an increased incidence of Wilms' tumor in certain overgrowth conditions, particularly Beckwith-Wiedemann syndrome and hemihypertrophy. The WAGR syndrome has been shown to result from the deletion of one copy each of the Wilms' tumor gene, \( WT1 \), and the adjacent aniridia gene, \( PAX6 \), on chromosome 11p13. Beckwith-Wiedemann syndrome is an overgrowth syndrome that is characterized by visceromegaly, macroglossia, and hyperinsulinemic hypoglycemia. It arises from mutations at the 11p15.5 locus. There is evidence to suggest that analysis of the methylation status of several genes in the 11p15 locus could predict the individual risk to the development of Wilms' tumor. Importantly, most patients with Wilms' tumor do not have mutations at these genetic loci.

Surgical Treatment

Before operation, all patients suspected of Wilms' tumor should undergo abdominal and chest CT scanning. These studies characterize the mass, identify the presence of metastases, and provide information on the opposite kidney (Fig. 38-37). CT scanning also indicates the presence of nephrogenic rests, which are precursor lesions to Wilms' tumor. An abdominal ultrasound should be performed to detect the presence of renal vein or vena caval extension.

The management of patients with Wilms' tumor has been carefully evaluated within the context of large studies involving thousands of patients. These studies have been coordinated by the National Wilms' Tumor Study Group (NWTSG) in North America and the International Society of Paediatric Oncology (SIOP), mainly involving European countries. Significant differences in the approach to patients that present with Wilms' tumor have been highlighted by these studies. NWTSG supports a strategy of surgery followed by chemotherapy in most instances, whereas the SIOP approach is to shrink the tumor using preoperative chemotherapy. There are instances in which preoperative
chemotherapy is supported by both groups, including the presence of bilateral involvement or inferior vena cava involvement that extends above the hepatic veins, and involvement of a solitary kidney by Wilms' tumor. The NWTSG proponents argue that preoperative therapy in other instances results in a loss of important staging information, and therefore places patients at higher risk for recurrence; alternatively it may lead to overly aggressive treatment in some cases. However, the overall survival rates are no different between the NWTSG and SIOP approaches.

The goals of surgery include complete removal of the tumor. It is crucial to avoid tumor rupture or injury to contiguous organs. A sampling of regional lymph nodes should be included, and all suspicious nodes should be sampled. Typically a transverse abdominal incision is made, and a transperitoneal approach is used. The opposite side is carefully inspected to ensure that there is no disease present. A radical nephroureterectomy is then performed with control of the renal pedicle as an initial step. If there is spread above the hepatic veins, an intrathoracic approach may be required. If bilateral disease is encountered, both lesions are biopsied, and chemotherapy is administered, followed by a nephron-sparing procedure.

Chemotherapy

Following nephroureterectomy for Wilms' tumor, the need for chemotherapy and/or radiation therapy is determined by the histology of the tumor and the clinical stage of the patient (Table 38-3). Essentially, patients who have disease confined to one kidney that is totally removed surgically receive a short course of chemotherapy, and can expect a 97% 4-year survival rate, with tumor relapse rare after that time. Patients with more advanced disease or with unfavorable histology receive more intensive chemotherapy and radiation. Even in stage IV, cure rates of 80% are achieved. The survival rates are worse in the small percentage of patients considered to have unfavorable histology. The major chemotherapeutic agents are dactinomycin and vincristine, with the addition of doxorubicin for more advanced stages.

FIG. 38-37. Wilms' tumor of the right kidney (arrow) in a 3-year-old girl.

Neuroblastoma

Clinical Presentation

Neuroblastoma is the third most common pediatric malignancy, and accounts for approximately 10% of all childhood cancers. The vast majority of patients have advanced disease at the time of presentation, and unlike Wilms' tumor, the overall survival is less than 30%. Over 80% of cases present before the age of 4 years, and the peak incidence is at 2 years of age. Neuroblastomas arise from the neural crest cells and show different levels of differentiation. The tumor originates most frequently in the adrenal glands, posterior mediastinum, neck, or pelvis, but can arise in any sympathetic ganglion.
Two-thirds of these tumors are first noted as an asymptomatic abdominal mass. The tumor may cross the midline, and a majority of patients will already show signs of metastatic disease. Occasionally, children may present with pain from the tumor mass or to bone pain from metastases. Proptosis and periorbital ecchymosis may occur due to the presence of retrobulbar metastasis. Because they originate in paraspinal ganglia, neuroblastomas may invade through neural foramina and compress the spinal cord, causing muscle weakness or sensory changes. Rarely, children may have severe watery diarrhea due to the secretion of vasoactive intestinal polypeptide by the tumor, or with paraneoplastic neurologic findings including cerebellar ataxia or opsoclonus/myoclonus.

Diagnostic Evaluation

Since these tumors derive from the sympathetic nervous system, catecholamines and their metabolites will be produced at increased levels. These include elevated levels of serum catecholamines (dopamine and norepinephrine) or urine catecholamine metabolites (vanillylmandelic acid [VMA] or homovanillic acid [HVA]). Measurement of VMA and HVA in serum and urine aids in the diagnosis and in monitoring adequacy of future treatment and recurrence. The minimum criterion for a diagnosis of neuroblastoma is based on one of the following findings: (1) an unequivocal pathologic diagnosis made from tumor tissue by light microscopy (with or without immunohistology, electron microscopy, or increased levels of serum catecholamines or urinary catecholamine metabolites); and (2) the combination of bone marrow aspirate or biopsy containing unequivocal tumor cells and increased levels of serum catecholamines or urinary catecholamine metabolites as described above.

The patient should be evaluated by abdominal CT scan, which usually shows displacement and occasionally obstruction of the ureter of an intact kidney (Fig. 38-38). Prior to the institution of therapy, a complete staging work-up should be performed. This includes radiograph of the chest, bone marrow biopsy, and radionuclide scans to search for metastases. Any abnormalities on chest x-ray should be followed up with CT of the chest.

Prognostic Indicators

A number of biologic variables have been studied in children with neuroblastoma. An open biopsy is required in order to provide tissue for this analysis. Hyperdiploid tumor DNA is associated with a favorable prognosis, and N-myc amplification is associated with a poor prognosis regardless of patient age. The Shimada classification describes tumors as having either favorable or unfavorable histology based on the degree of differentiation, the mitosis-karyorrhexis index, and the presence or absence of schwannian stroma. In general, children of any age with localized neuroblastoma and infants younger than 1 year of age with advanced disease and favorable disease characteristics have a high likelihood of disease-free survival. By contrast, older children with advanced-stage disease have a significantly decreased chance for cure despite intensive therapy. For example, aggressive multiagent chemotherapy has resulted in a 2-year survival rate of approximately 20% in older children with stage IV disease.
Neuroblastoma in the adolescent has a worse long-term prognosis, regardless of stage or site, and in many cases, a more prolonged course.

Surgery

The goal of surgery is complete resection. However, this is often not possible due to the extensive locoregional spread of the tumor at the time of presentation. Under these circumstances, a biopsy is performed and preoperative chemotherapy is provided based on the stage of the tumor. After neoadjuvant treatment has been administered, surgical resection is performed. The principal goal of surgery is to obtain at least a 95% resection, without compromising major structures. Abdominal tumors are approached through a transverse incision. Thoracic tumors may be approached through a posterolateral thoracotomy or through a thorascopic approach. These may have an intraspinal component.

Neuroblastoma in Infants

Spontaneous regression of neuroblastoma has been well described in infants, especially in those with stage 4S disease. Regression generally occurs only in tumors with a near triploid number of chromosomes that also lack N-myc amplification and loss of chromosome 1p. Recent studies indicate that infants with asymptomatic, small, low-stage neuroblastoma detected by screening may have tumors that spontaneously regress. These patients may be observed safely without surgical intervention or tissue diagnosis.

FIG. 38-38. Abdominal neuroblastoma arising from the right retroperitoneum (arrow).

Rhabdomyosarcoma

Rhabdomyosarcoma is a primitive soft tissue tumor that arises from mesenchymal tissues. The most common sites of origin include the head and neck (36%), extremities (19%), genitourinary tract (21%), and trunk (9%), although the tumor can arise virtually anywhere. The clinical presentation of the tumor depends on the site of origin. The diagnosis is confirmed with incisional or excisional biopsy after evaluation by MRI, CT scans of the affected area and the chest, and bone marrow biopsy. The tumor grows locally into surrounding structures and metastasizes widely to lung, regional lymph nodes, liver, brain, and bone marrow. The staging system for rhabdomyosarcoma is based on the tumor-node-metastasis (TNM) system, as established by the Soft Tissue Sarcoma Committee of the Children's Oncology Group (Table 38-4). Surgery is an important component of the staging strategy, and involves biopsy of the lesion and evaluation of lymphatics. Primary resection should be undertaken when complete excision can be performed without causing disability. If this is not possible, the lesion is biopsied and intensive chemotherapy is administered. It is important to plan the biopsy so that it does not interfere with the subsequent resection. After the tumor has decreased in size, resection of gross residual disease should be performed. Radiation therapy is effective in achieving local control when microscopic or gross residual disease exists following initial treatment. Patients with completely resected tumors of embryonal histology do well
without radiation therapy, but radiation therapy benefits patients with stage 1 tumors with alveolar or undifferentiated histology.

Prognosis

The prognosis for rhabdomyosarcoma is related to the site of origin, resectability, presence of metastases, number of metastatic sites, and histopathology. Primary sites with more favorable prognoses include the orbit and nonparameningeal head and neck, paratestis and vagina (nonbladder, nonprostate genitourinary), and the biliary tract. Patients with tumors under 5 cm in size have improved survival compared to children with larger tumors, while children with metastatic disease at diagnosis have the poorest prognosis. Tumor histology influences prognosis, and the embryonal variant is favorable, while the alveolar subtype has an unfavorable prognosis.

Teratoma

Teratomas are tumors composed of tissue from all three embryonic germ layers. They may be benign or malignant, may arise in any part of the body, and are usually found in midline structures. Thoracic teratomas usually present as an anterior mediastinal mass. Ovarian teratomas present as an abdominal mass, often with symptoms of torsion, bleeding, or rupture. Retroperitoneal teratomas may present as a flank or abdominal mass.

Mature teratomas usually contain well-differentiated tissues and are benign, while immature teratomas contain varying degrees of immature neuroepithelium or blastemal tissues. Immature teratomas can be graded from 1 to 3, based on the amount of immature neuroglial tissue present. Tumors of higher grade are more likely to have foci of yolk sac tumor. Malignant germ cell tumors usually contain frankly neoplastic tissues of germ cell origin (i.e., yolk sac carcinoma, embryonal carcinoma, germinoma, or choriocarcinoma). Yolk sac carcinomas produce alpha-fetoprotein, while choriocarcinomas produce beta human chorionic gonadotropin (bHCG), resulting in elevation of these substances in the serum, which can serve as tumor markers. In addition, germinomas also can produce elevation of serum bHCG, but not to the levels associated with choriocarcinoma.

Sacrococcygeal Teratoma

Sacrococcygeal teratoma usually presents as a large mass extending from the sacrum in the newborn period. Diagnosis may be established by prenatal ultrasound. In fetuses with evidence of hydrops and a large sacrococcygeal teratoma, prognosis is poor; thus prenatal intervention has been advocated in such patients. The mass may be as small as a few centimeters in diameter or as massive as the size of the infant (Fig. 38-39). The tumor has been classified based on the location and degree of intrapelvic extension. Lesions with growth predominantly into the presacral space often present later in childhood. The differential diagnosis consists of neural tumors, lipoma, and myelomeningoceles.

Most tumors are identified at birth and are benign. Malignant yolk sac tumor histology occurs in a minority of these tumors. Complete resection of the tumor as early as possible is essential. The rectum and genital structures are often distorted by the tumor, but
usually can be preserved in the course of resection. Perioperative complications of hypothermia and hemorrhage can occur with massive tumors and may prove lethal. This is of particular concern in small preterm infants with large tumors. The cure rate is excellent if the tumor is excised completely. The majority of patients who develop recurrent disease are salvageable with subsequent platinum-based chemotherapy.

**FIG. 38-39.** Sacrococcygeal teratoma in a 2-day-old boy.

**Liver Tumors**

More than two-thirds of all liver tumors in children are malignant. There are two major histologic subgroups: hepatoblastoma and hepatocellular carcinoma. The age of onset of liver cancer in children is related to the histology of the tumor. Hepatoblastoma is the most common malignancy of the liver in children, with most of these tumors diagnosed before 4 years of age. Hepatocellular carcinoma is the next most common, with a peak age incidence between 10 and 15 years. Malignant mesenchymomas and sarcomas are much less common, but constitute the remainder of the malignancies. The finding of a liver mass does not necessarily imply that a malignancy is present. Nearly 50% of all masses are benign, and hemangiomas are the most common lesions.

Most children with a liver tumor present with an abdominal mass that is usually painless, which the parents note while changing the child's clothes or while bathing the child. The patients are rarely jaundiced, but may complain of anorexia and weight loss. Most liver function tests are normal. Alpha-fetoprotein levels are elevated in 90% of children with hepatoblastomas, but are increased much less commonly in other liver malignancies. Radiographic evaluation of these children should include an abdominal CT scan to identify the lesion and to determine the degree of local invasiveness. For malignant-appearing lesions, a biopsy should be performed unless the lesion can be completely resected easily. Hepatoblastoma is most often unifocal, while hepatocellular carcinoma is often extensively invasive or multicentric. If a hepatoblastoma is completely removed, the majority of patients survive, but only a minority of patients have lesions amenable to complete resection at diagnosis.

A staging system based on postsurgical extent of tumor and surgical resectability is shown in Table 38-5. The overall survival rate for children with hepatoblastoma is 70%, but is only 25% for hepatocellular carcinoma. Children diagnosed with stage I disease and II hepatoblastoma have a cure rate of greater than 90%, compared to 60% for stage III and approximately 20% for stage IV. In children diagnosed with hepatocellular carcinoma, those with stage I have a good outcome, whereas stages III and IV are usually fatal. The fibrolamellar variant of hepatocellular carcinoma may have a better prognosis.

**Surgery**

The abdominal CT scan usually will determine the resectability of the lesion, although occasionally this can only be determined at the time of exploration. Complete surgical resection of the tumor is the primary goal and is essential for cure. For tumors that are
unresectable, preoperative chemotherapy should be administered to reduce the size of the tumor and improve the possibility for complete removal. Chemotherapy is more successful for hepatoblastoma than for hepatocellular carcinoma. Areas of locally invasive disease, such as the diaphragm, should be resected at the time of surgery. For unresectable tumors, liver transplantation has recently been used with some success. The fibrolamellar variant of hepatocellular carcinoma may have a better outcome with liver transplant than other hepatocellular carcinomas.

FIG. 38-40. CT scan of the abdomen showing a hepatocellular carcinoma in a 12-year-old boy.

TRAUMA IN CHILDREN

Introduction

Injury is the leading cause of death among children older than 1 year. In fact, trauma accounts for almost half of all pediatric deaths, more than cancer, congenital anomalies, pneumonia, heart disease, homicide, and meningitis combined. Death from unintentional injury accounts for 65% of all injury deaths in children younger than 19 years. From 1972 to 1992, motor vehicle collisions were the leading cause of death in people aged 1 to 19 years, followed by homicide or suicide (predominantly with firearms) and drowning. Each year, approximately 20,000 children and teenagers die as a result of injury in the United States. For every child who dies from an injury, it is calculated that 40 others are hospitalized and 1120 are treated in emergency departments. An estimated 50,000 children acquire permanent disabilities each year, most of which are the result of head injuries. Thus the problem of pediatric trauma continues to be one of the major threats to the health and well-being of children.

Specific considerations apply to trauma in children that influence management and outcome. These relate to the mechanisms of injury, the anatomic variations in children compared to adults, and the physiologic responses.

Mechanisms of Injury

Most pediatric trauma is blunt. Penetrating injuries are seen in the setting of gun violence, falls onto sharp objects, or penetration by glass after falling through windows. Age and gender significantly influence the patterns of injury. Male children younger than 18 years are exposed to contact sports and drive motor vehicles. As a result, they have a different pattern of injury than younger children, characterized by higher injury severity scores. In the infant and toddler age group, falls are a common cause of severe injury. Injuries in the home are extremely common. These include falls, near-drownings, caustic ingestion, and nonaccidental injuries.

Initial Management

The goals of managing the pediatric trauma patient are similar to those of adults, and follow Advanced Trauma Life Support guidelines as established by the American
College of Surgeons. Airway control is the first priority. In a child, respiratory arrest can proceed quickly to cardiac arrest. It is important to be aware of the anatomic differences between the airway of the child and the adult. The child has a shorter neck, smaller and anterior larynx, floppy epiglottis, short trachea, and large tongue. The child’s fifth digit can provide an estimate of the size of the correct endotracheal tube. Alternatively, the formula (age in years + 16)/4 may be used. It is important to use uncuffed endotracheal tubes in children younger than 8 years in order to minimize tracheal trauma. After evaluation of the airway, breathing is assessed. It is important to consider that gastric distention from aerophagia can severely compromise respirations. A nasogastric tube should therefore be placed early in the resuscitation. Pneumothorax or hemothorax should be treated promptly. When evaluating the circulation, it is important to recognize that tachycardia is usually the earliest measurable response to hypovolemia. Other signs of impending hypovolemic shock in children include changes in mentation, delayed capillary refill, skin pallor, and hypothermia. Intravenous access should be rapidly obtained once the patient arrives in the trauma bay. The first approach should be to use the antecubital fossae. If this is not possible, a cutdown into the saphenous vein at the groin can be performed quickly and safely. Intraosseous cannulation can provide temporary access in infants until intravenous access is established. Percutaneous neck lines should generally be avoided. Blood is drawn for cross-match and evaluation of liver enzymes, lipase, amylase, and hematologic profile, after the intravenous lines are placed.

In patients who show signs of volume depletion, a 20-mL/kg bolus of saline or lactated Ringer's solution should be promptly given. If the patient does not respond to three boluses, blood should be transfused (10 mL/kg). The source of bleeding should be established. Common sites include the chest, abdomen, pelvis, extremity fractures, or large scalp wounds. These should be carefully sought. Care is taken to avoid hypothermia by infusing warmed fluids and by using external warming devices.

**Evaluation of Injury**

All patients should receive an x-ray of the cervical spine, chest, and abdomen with pelvis. All extremities that are suspicious for fracture should also be evaluated by x-ray. Screening blood work that includes aspartate aminotransferase and alanine aminotransferase, and amylase/lipase is useful for the evaluation of liver and pancreatic injuries. Significant elevation in these tests requires further evaluation by CT scanning. The child with significant abdominal tenderness and a mechanism of injury that could cause intra-abdominal injury should undergo abdominal CT scanning using intravenous and oral contrast in all cases. There is a limited role for diagnostic peritoneal lavage (DPL) in children as a screening test. However, it can be useful in the child that is brought emergently to the operating room for management of significant intracranial hemorrhage. At the time of craniotomy, a DPL can be performed concurrently to identify abdominal bleeding. Although abdominal ultrasound is extremely useful in the evaluation of adult abdominal trauma, it has not been widely accepted in the management of pediatric injury. In part this relates to the widespread use of nonoperative treatment for most solid-organ injuries, which would result in a positive abdominal ultrasound scan.

**Injuries to the Central Nervous System**
The central nervous system (CNS) is the most commonly injured organ system, and CNS injury is the leading cause of death among injured children. In the toddler age group, nonaccidental trauma is the most common cause of serious head injury. Findings suggestive of abuse include the presence of retinal hemorrhage on funduscopic evaluation, intracranial hemorrhage without evidence of external trauma (indicative of a shaking injury), and fractures at different stages of healing on skeletal survey. In older children, CNS injury occurs most commonly after falls and bicycle and motor vehicle collisions. The initial head CT scan can often underestimate the extent of injury in children. Criteria for head CT scan include any loss of consciousness or amnesia to the trauma, or inability to assess the CNS status as in the intubated patient. Patients with mild, isolated head injury (Glasgow Coma Scale [GCS] score 14 to 15) and negative CT scans can be discharged if their neurologic status is normal after 6 hours of observation. Young children and those in whom there is multisystem involvement should be admitted to the hospital for a period of overnight observation. Any change in the neurologic status warrants neurosurgical evaluation and repeat CT scanning. In patients with severe head injury (GCS score 8 or less), urgent neurosurgical consultation is required. These patients are evaluated for intracranial pressure monitoring, and for the need to undergo craniotomy.

Thoracic Injuries

The pediatric thorax is pliable due to incomplete calcification of the ribs and cartilages. As a result, blunt chest injury commonly results in pulmonary contusion, although rib fractures are rare. Diagnosis is made by chest radiograph, and may be associated with severe hypoxia requiring mechanical ventilation. Pulmonary contusion usually resolves with careful ventilator management and judicious volume resuscitation. Children who have sustained massive blunt thoracic injury may develop traumatic asphyxia. This is characterized by cervical and facial petechial hemorrhages or cyanosis associated with vascular engorgement and subconjunctival hemorrhage. Management includes ventilation and treatment of coexisting CNS or abdominal injuries. Penetrating thoracic injuries may result in damage to the lung, or to major disruption of the bronchi or great vessels.

Abdominal Injuries

In children, the small rib cage and minimal muscular coverage of the abdomen can result in significant injury after seemingly minor trauma. The liver and spleen in particular are relatively unprotected, and are often injured after direct abdominal trauma. Duodenal injuries are usually the result of blunt trauma, which may arise from child abuse or injury from a bicycle handlebar. Duodenal hematomas usually resolve without surgery. Small intestinal injury usually occurs in the jejunum in the area of fixation by the ligament of Treitz. These injuries are usually caused by rapid deceleration while the child is restrained by a lap belt. There may be a hematoma on the anterior abdominal wall caused by a lap belt, the so-called "seat belt sign" (Fig. 38-41A). This should alert the caregiver to the possibility of an underlying small bowel injury (Fig. 38-41B), as well as to a potential lumbar spine injury (Chance fracture).
The spleen is injured relatively commonly after blunt abdominal trauma in children. The extent of injury to the spleen is graded (Table 38-6), and the management is governed by the injury grade. Current treatment involves a nonoperative approach in most cases, even for grade IV injuries, providing the patient is hemodynamically stable. This approach avoids surgery in most cases. All patients should be placed in a monitored unit, and type-specific blood should be available for transfusion. When nonoperative management is successful, as it is in most cases, an extended period of bedrest is prescribed. This optimizes the chance for healing, and minimizes the likelihood of reinjury. A typical guideline is to keep the children on extremely restricted activity for 2 weeks longer than the grade of spleen injury (i.e., a child with a grade IV spleen injury receives 6 weeks of restricted activity). In children that have an ongoing fluid requirement, or when a blood transfusion is required, exploration should not be delayed. At surgery the spleen can often be salvaged. If a splenectomy is performed, prophylactic antibiotics and immunizations should be administered to protect against overwhelming postsplenectomy sepsis. The liver also is commonly injured after blunt abdominal trauma. A grading system is used to characterize hepatic injuries (Table 38-7), and nonoperative management is usually successful (Fig. 38-42). Recent data have shown that associated injuries are more significant predictors of outcome in children with liver injuries than the actual injury grade. Criteria for surgery are similar to those for splenic injury and primarily involve hemodynamic instability. The intraoperative considerations in the management of massive hepatic injury are similar in children and adults. Renal contusions may occur after significant blunt abdominal trauma. Nonoperative management is usually successful, unless patients are unstable due to active renal bleeding. It is important to confirm the presence of a normal contralateral kidney at the time of surgery.

FIG. 38-41. Abdominal CT scan of patient that sustained a lap belt injury. A. Bruising is noted across the abdomen from the lap belt. B. At laparotomy, a perforation of the small bowel was identified.

FIG. 38-42. Abdominal CT scan of a child demonstrating a grade III liver laceration (arrows).

FETAL INTERVENTION

Introduction

One of the most exciting developments in the field of pediatric surgery has been the emergence of fetal surgery. The performance of a fetal intervention is justified when a defect is present that would cause devastating consequences to the infant if left uncorrected. For the vast majority of congenital anomalies, postnatal surgery is the preferred modality. However, in specific circumstances, fetal surgery may offer the best possibility for a successful outcome. The decision to perform a fetal intervention requires careful patient selection, as well as a multidisciplinary center that is dedicated to the surgical care of the fetus and the mother. Patient selection is dependent in part on highly accurate prenatal imaging, which includes ultrasound and MRI. At the present time, fetal surgery is performed at a few centers in North America, although this number is increasing.
Significant risks may be associated with the performance of a fetal surgical procedure, to both the mother and the fetus. From the maternal viewpoint, open fetal surgery may lead to uterine bleeding due to the uterine relaxation required during the procedure. The long-term effects on subsequent pregnancies remain to be established. For the fetus, in utero surgery carries the risk of premature labor and amniotic fluid leakage. As a result, these procedures are performed only when the expected benefit of fetal intervention outweighs the risk to the fetus of standard postnatal care.

**Surgery for Lower Urinary Tract Obstruction**

Lower urinary tract obstruction refers to a group of diseases characterized by obstruction of the distal urinary system. Common causes include the presence of posterior urethral valves and urethral atresia, as well as other anomalies of the urethra and bladder. The pathologic effects of lower urinary tract obstruction lie in the resultant massive bladder distention that occurs, which can lead to reflux hydronephrosis. This may result in oligohydramnios, and cause limb contractures, facial anomalies (Potter facies), and pulmonary hypoplasia. Carefully selected patients with lower urinary tract obstruction may benefit from vesicoamniotic shunting. By relieving the obstruction and improving renal function, fetal growth and lung development may be preserved.

**Fetal Surgery for Congenital Diaphragmatic Hernia**

Given the high mortality associated with the most severe cases of CDH, tremendous efforts have been undertaken to determine whether fetal intervention could improve the outcome of this disease. In 1990, Harrison and colleagues reported the first open fetal repair for CDH. The high morbidity of the open technique led to the development of fetal tracheal occlusion as a therapeutic approach. This was based on the observation that tracheal occlusion could lead to increased lung growth and reduction of the intrathoracic viscera in animal models. Tracheal occlusion can be achieved in utero by placement of clips that are removed at the time of delivery. Despite initial enthusiasm for this approach, a recent randomized trial that compared fetal tracheal occlusion with standard postnatal care for left-sided CDH showed no improvement in survival for patients treated with tracheal occlusion.

**Fetal Surgery for Myelomeningocele**

*Myelomeningocele* refers to a spectrum of anomalies in which portions of the spinal cord are uncovered by the spinal column. This leaves the neural tissue exposed to the injurious effects of the amniotic fluid, as well as to trauma from contact with the uterine wall. Nerve damage ensues, resulting in varying degrees of lower extremity paralysis, as well as bowel and bladder dysfunction. Initial observations indicated that the extent of injury progressed throughout the pregnancy, which provided the rationale for fetal intervention. The current in utero approach for the fetus with myelomeningocele has focused on obtaining coverage of the exposed spinal cord. Initial results have shown a decrease in the development of obstructive hydrocephalus requiring ventriculoperitoneal shunting. A National Institutes of Health-sponsored trial is currently underway, in which patients are randomized to receive either in utero coverage of the spinal cord or standard postnatal
care. The effects of the treatment approach on neurologic function can then be determined.

**The Ex-Utero Intrapartum Treatment Procedure**

The EXIT procedure is utilized in circumstances in which an airway obstruction is predicted at the time of delivery, due to the presence of a large neck mass such as a cystic hygroma or teratoma (*Fig. 38-43*), or congenital tracheal stenosis. The success of the procedure is dependent on the maintenance of uteroplacental perfusion for a sufficient duration to secure the airway. To achieve this, deep uterine relaxation is obtained during a cesarean section under general anesthesia. Uterine perfusion with warmed saline also promotes relaxation and blood flow to the placenta. On average, between 20 and 30 minutes of placental perfusion can be achieved. The fetal airway is secured either by placement of an orotracheal tube, or performance of a tracheostomy. Once the airway is secured, the cord is cut, and a definitive procedure may be performed to relieve the obstruction postnatally.

*FIG. 38-43.* The EXIT (ex-utero intrapartum treatment) procedure in a baby at 34 weeks' gestation with a large cervical teratoma. Intubation is performed while the fetus is on placental support.

**REFERENCES**


