The risk of unsuspected surgical emergencies during the neonatal period has decreased over the past several years because of widespread use of ultrasonography for fetal screening. However, surgical emergencies unique to the neonatal period still continue to plague the unsuspecting emergency physician. Most surgically correctable disorders in the neonate will present with vomiting, gastrointestinal bleeding, or respiratory distress. This article will provide the emergency physician with the typical presentation, diagnostic work-up, and treatment options for surgically correctable problems in the neonate.


Neonatal Surgical Emergencies

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The scope of pediatric surgery is broad and encompasses a wide variety of diseases. Many surgical disease processes are age related. There are several common surgical emergencies that physicians caring for neonates will encounter in the emergency department. The most common neonatal surgical emergencies of the gastrointestinal tract are outlined in Table 1. This article will review common neonatal surgical emergencies that present with vomiting, bleeding, or respiratory distress.

Neonatal Surgical Disorders Associated with Vomiting

Surgical disorders associated with vomiting include intestinal obstruction, malrotation and midgut volvulus, Hirschsprung's disease (HD), meconium ileus (MI), and abdominal wall defects. Other common emergencies, such as incarcerated inguinal hernia, hypertrophic pyloric stenosis, and trauma, have been discussed elsewhere in this issue.

Intestinal Obstruction—Atresias

Intestinal atresias involving the duodenum, jejunum, ileum, and colon together are the most common causes of intestinal obstruction in the newly born, affecting between 1 in 2,000 and 1 in 5,000 live births.1 There is equal representation of the sexes. Duodenal obstruction is postulated to occur as a result of incomplete recanalization of the duodenal lumen during the tenth week of gestation. Jejunoileal atresia is believed to be the result of late mesenteric vascular accidents in utero.

Bilious vomiting is the most common presenting symptom in a newborn with intestinal obstruction. Although abdominal distention is typical of distal atresias, the abdomen may be flat or even scaphoid in duodenal lesions. Abdominal distention is most prominent in ileal atresia, whereas polyhydramnios and jaundice
on the type of intestinal atresia, different surgical strategies are indicated. In general, bowel reconstruction is achieved by an end-to-end (or end-to-side) anastomosis.2

Survival rates exceed 90%. Late complications from repair of duodenal atresia occur in approximately 12% to 15% of patients and include megaloduenum, intestinal motility disorders, duodenogastric reflux, gastritis, peptic ulcer disease, gastroesophageal reflux, and choledochal cysts.

Malrotation and Midgut Volvulus

The incidence of malrotation is 1 in 500 live births.3 Twenty-five percent to 40% of patients with symptomatic malrotation present within the first week of life, 50% present within the first month, 75% present before 1 year of age, and there is a 2:1 male predominance in cases presenting in the neonatal period. Malrotation of the midgut occurs when

<table>
<thead>
<tr>
<th>TABLE 1. Common Neonatal Surgical Emergencies of the Gastrointestinal Tract</th>
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<tr>
<td>Intestinal obstruction</td>
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<tr>
<td>Duodenum, jejunum, ileum, colon</td>
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<tr>
<td>Atresia, web, stenosis</td>
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<td>Malrotation and midgut volvulus</td>
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<td>Duplications, Meckel’s diverticulum, mesenteric cysts</td>
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<td>Incarcerated inguinal hernia</td>
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<td>Hypertrophic pyloric stenosis</td>
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<td>Trauma (consider nonaccidental trauma)</td>
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</tbody>
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are more frequently associated with jejunal atresia. Approximately one third of newly born infants with duodenal atresia have associated Down’s syndrome.

Other subtle signs of intestinal obstruction include respiratory difficulty, excessive salivation, presence of an abdominal mass, jaundice (within 24 hours of birth), and lethargy.2

Abdominal plain films show a characteristic double bubble sign, showing air bubbles in the stomach and a dilated proximal duodenum in duodenal atresia (Fig 1). In jejunoileal atresia, abdominal films show air fluid levels proximal to the lesion. The more distal the obstruction, the greater the number of air fluid levels and distended loops of bowel. A contrast enema will show a small unused “microcolon” in most infants with jejunoileal atresia.

A nasogastric or orogastric tube should be passed to decompress the stomach and duodenum. Intravenous fluids are given to maintain adequate urine output and normoglycemia. Surgery is not urgent. The infant must be evaluated thoroughly for associated malformations, especially cardiac anomalies. Duodenal atresia has been associated with abnormal development of the pancreas (annular pancreas) and Down’s syndrome (30% to 50%). There is also an association with malrotation, congenital heart disease, and esophageal atresia.

For duodenal malformations, most surgeons will perform a diamond-shaped duodenoduodenostomy using a standard side-to-side or proximal transverse-to-distal longitudinal anastomosis. Depending

Figure 1. Double bubble sign in duodenal atresia.
Evaluation, resuscitation, and preoperative preparation proceed simultaneously in patients with suspected malrotation with volvulus. In the emergency department, volume resuscitation, gastric decompression, and broad-spectrum antibiotics are administered. The operative procedure for the correction of malrotation, called the Ladd procedure, includes derotation of the volvulus, division of mesenteric bands, separation of the duodenojejunal mesentery from the cecocolic mesentery, and appendectomy. An appendectomy is performed because the colon is positioned on the left side of the abdomen during a Ladd procedure. Subsequent development of appendicitis could be confusing. A gastrostomy is rarely indicated.

With viable intestine, postoperative care is straightforward, and prognosis is excellent. The lifetime risk of adhesive small bowel obstruction is 1% to 10%.3 Volvulus should not recur. With extensive ischemic bowel and intestinal necrosis, the bowel is untwisted and reduced into the abdominal cavity. Twelve to 24 hours later, a “second look” procedure may be performed to assess bowel viability. This allows the surgeon to resect the necrotic bowel and to create an enterostomy at the distal end of the normal bowel. Bowel reconstruction is performed in a later operation. These patients may develop short gut syndrome and may become dependent on total parenteral nutrition (TPN).

The most serious complication of malrotation is midgut volvulus with necrosis of the entire midgut. Mortality in infants with malrotation ranges from 2.5% to 24% and is influenced by the presence of necrotic bowel at laparotomy, the presence of associated anomalies, and younger patient age.4

Billious vomiting in an otherwise healthy infant must be considered a surgical emergency and should be considered to be malrotation with midgut volvulus until proven otherwise. An emergency UGI must be performed as the patient is being evaluated, not on an elective basis.

Hirschsprung’s Disease

HD is a common cause of intestinal obstruction in newborns. It results from absence of ganglion cells in the myenteric plexus of the intestine. Incidence varies from 1 in 4,000 to 1 in 7,000 live births with a male to female ratio of 4:1.5 It is usually an isolated disorder; however, Down’s syndrome is associated with 4% to 12% of the cases.

HD results from the migratory arrest of vagally derived neuroblasts, which normally reach the rectum by the twelfth week of gestation. Any length of the intestine may be involved; however, 80% to 90%

the normal rotational process and fixation of the intestine fail to occur during the seventh to twelfth week of gestation.

Malrotation typically presents in the first month of life with bilious vomiting and sudden onset of abdominal pain (crying). In older infants and children, symptoms may be vague and may include chronic, intermittent vomiting and cramping abdominal pain, failure to thrive, constipation, bloody diarrhea, and hematemesis. Physical examination may exhibit a normal abdominal examination in 50% of patients. One third of patients present with abdominal distension without tenderness. Bowel distension, abdominal pain, and evidence of peritonitis occurs as intestinal ischemia progresses to necrosis. As ischemia progresses to infarction, fever, peritonitis, abdominal distension, profound dehydration and vascular collapse worsen.

In the newborn, volvulus can rapidly result in significant bowel compromise with abdominal distention, bloody stools, and eventually hypovolemic shock and peritonitis. An upper gastrointestinal (UGI) contrast study is the most reliable method to diagnose malrotation. The malrotated duodenum is often “coiled” to the right of the midline giving a corkscrew appearance (Fig 2). A cutoff appearance of the contrast (“beak”) suggests obstruction from volvulus.

![Figure 2. Malrotation with midgut volvulus.](image-url)
of HD involves only the rectosigmoid colon. There are no “skip lesions” in the intestine, as HD is a failure of normal neuroblasts to complete their migration from the proximal GI tract to the anus. A mutation in the (RET)-proto-oncogene appears to be a major factor in the development of the disease. Also, molecules in the endothelin signaling pathway and the SOX10 transcriptional activator have been implicated.

The typical presentation of a neonate with HD includes failure to pass meconium in the first 24 hours of life, constipation and abdominal distention, bilious emesis, and refusal to feed. In some cases, failure to thrive may be the only initial sign.

Barium enema (80% accuracy) shows a transitional zone (funnel-shaped dilatation of bowel) at the junction for the aganglionic and ganglionic intestine (Fig 3). Rectal suction biopsy (95% accuracy) shows absence of ganglion cells in the myenteric plexus, increased staining of cholinesterase, and the presence of hypertrophied nerve bundles. There are 2 surgical approaches: performance of a colostomy in the newborn period with a definitive pull-through operation at a later date and a primary pull through operation without a colostomy. Currently, there are 3 pull-through procedures being used: Swenson procedure, Duhamel procedure, and Soave’s operation.

A very serious complication of HD is Hirschsprung’s-associated enterocolitis (HAEC). It can occur before diagnosis and treatment of HD or after the definitive pull-through procedure and may be lethal if not aggressively treated. The mean incidence of HAEC is 25% (range, 17% to 50%). There has been a clear decline in the incidence of HAEC over the past 40 years, most likely because of improved and prompt diagnosis of HD. Classic manifestations include abdominal distention, fever, and foul-smelling stool. Other symptoms include explosive diarrhea, vomiting, fever, lethargy, rectal bleeding, and shock. Physical examination reveals marked abdominal distension, which is hyperresonant to percussion. Rectal examination results in explosive discharge of gas and stool.

Abdominal radiographs may show the presence of a distended loop of bowel along the left flank with an abrupt termination in the pelvis. This is known as the intestinal “cut-off” sign. Because of the risk of perforation, a contrast enema is not recommended in clinical HAEC.

The infant is at risk for HAEC before the diagnosis of HD has been made and after the definitive pull-through surgery. Risk factors include delay in diagnosis of HD, increased length of the aganglionic segment, and Down’s syndrome. Once HAEC develops, the infant is at increased risk for future development of enterocolitis.

Treatment includes bowel rest, rectal washouts, and intravenous fluids and antibiotics. Mild cases can be treated with oral metronidazole. An emergency diverting stoma may be required in patients who fail to improve with medical management. This stoma must be performed in normally innervated bowel, as confirmed by frozen section evaluation. Mortality rates of HAEC range up to 33% and tend to be higher in children with other abnormalities, such as Down’s syndrome.

**Meconium Ileus**

MI occurs in 15% of newborns with cystic fibrosis and in 1 per 5,000 to 10,000 live births. It is characterized by impaction of thick meconium in the distal ileum leading to a functional bowel obstruction. Most (90% to 95%) of these infants have cystic fibrosis and thus have deficits of pancreatic enzymes in the intestine. Complicated MI results when a volvulus or perforation of the bowel occurs leading to meconium peritonitis. This may occur prenatally.

MI is usually identified immediately after birth. After a few hours of life, marked abdominal distension and bilious vomiting occurs. Physical exami-
nation shows thickened bowel loops that are often visible and palpable through the abdominal wall. Rectal examination is difficult because of the small caliber of the rectum.

Plain abdominal films show distended loops of intestine with thickened bowel walls. Meconium mixed with swallowed air will give a characteristic “ground-glass” appearance without air fluid levels. Calcification, free air, and very large air fluid levels suggest bowel perforation, which requires urgent surgery. A contrast enema demarcates a microcolon (Fig 4). Reflux of contrast into the ileum shows the plugs, which are located in the distal small intestine. The small bowel is of narrower caliber distal to the meconium plug and is dilated proximal to the plug.

Uncomplicated MI may initially be treated with a water-soluble contrast enema and intravenous fluids. The hypertonicity of the radiopaque agent draws fluid into the bowel and facilitates passage and expulsion of meconium. When repeated contrast enemas are unsuccessful, laparotomy is indicated to evacuate the obstructing meconium manually or by enterotomy irrigation.

Abdominal Wall Defects

Abdominal wall defects, such as omphalocele and gastroschisis, are challenging surgical emergencies at the time of birth. Complications after repair are more likely to be encountered by the emergency physician. Table 2 summarizes these abdominal wall defects and their associated complications.

Neonatal Surgical Disorders Associated with GI Bleeding

Infants with upper gastrointestinal (GI) bleeding may present with coffee-ground emesis, hematemesis, melena, or hematochezia. Fortunately, upper GI bleeding in infants is usually benign and self-limiting and rarely requires surgical intervention.

Lower GI bleeding may be more serious. Infants may present with bright red blood from the rectum, melena, or hematochezia. Bleeding can result from Meckel’s diverticulum, polyps (although rare in infants), lymphonodular hyperplasia, anorectal fissures, or intestinal duplications. Necrotizing enterocolitis must be suspected in premature and stressed full-term infants. Other causes, previously discussed, are malrotation, intussusception (rare in the newborn) and HD.

Necrotizing Enterocolitis

Necrotizing enterocolitis (NEC) is primarily a disease of premature, low-birth-weight infants and usually presents in the immediate newborn period. It is the most common gastrointestinal emergency seen in the neonatal intensive care unit, occurring in 3% to 5% of all admitted infants. For the emergency physician, it is important to remember that NEC can also develop in full-term infants. Infants at high risk are those with a history of congenital heart disease, perinatal asphyxia, hypoglycemia, polycythemia, respiratory distress, maternal cocaine use, or maternal pre-eclampsia.

The clinical presentation of a full-term neonate with NEC is similar to the preterm infant. Subtle
signs may include feeding intolerance, abdominal distension, jaundice, and a change in stooling pattern. More ominous signs and symptoms include abdominal tenderness, bilious emesis, grossly bloody stools, lethargy, temperature instability, apneic episodes, or respiratory distress.

The pathologic hallmarks of NEC are coagulation necrosis, inflammation, and hemorrhage in the involved segment of intestine. The origin of NEC is multifactorial. Risk factors include bacterial colonization, intestinal ischemia, hypoxia, and formula feeding. All of these stimulate proinflammatory mediators that lead to bowel necrosis. Abdominal radiograph findings include ileus, persistent loops, pneumatosis intestinalis (the hallmark of NEC), portal venous gas, gasless abdomen, and pneumoperitoneum (Fig 5). The treatment for uncomplicated NEC (no stricture or perforation) is medical. Current treatment recommendations are to place an orogastric tube to low intermittent suction, administer broad-spectrum antibiotics, and to give nothing by mouth for 10 to 14 days. Nutritional support is provided by TPN. This nonoperative management is successful in 75% of patients.14

Surgical intervention is indicated if there is evidence of perforation or intestinal necrosis. Relative indications for surgery include clinical deterioration, refractory acidosis, oliguria, hypotension, thrombocytopenia, ventilatory failure, portal venous gas, fixed dilated loop of bowel, or erythema of the abdominal wall. Surgical interventions include peritoneal drainage, laparotomy with resection and proximal end ostomy, or laparotomy with resection and primary anastomosis.15 Survival rates range from 50% to 70% depending on level of prematurity, severity of disease, and institution. Long-term complications include intestinal strictures and adhesions (10% to 35%) which may result in bowel obstruction, short bowel syndrome (malabsorption syndrome), bacterial overgrowth and life-threatening sepsis, electrolyte and water loss from the ileostomy, or cholestasis secondary to prolonged TPN administration.15

### Intestinal Duplications

Duplications are rare congenital anomalies that develop along the intestinal tract; occurring anywhere from the mouth to the anus, but most commonly in the ileum (50%). They may be cystic or tubular and possess 3 major characteristics:

1. Each is contiguous and adherent to some part of the alimentary tract.

### Table 2: Comparison of Gastrochisis and Omphalocele

<table>
<thead>
<tr>
<th>Epidemiology</th>
<th>Gastrochisis: 1 in 20,000 births</th>
<th>Omphalocele: 1 in 3,000 to 1 in 10,000 births</th>
</tr>
</thead>
<tbody>
<tr>
<td>Embryology</td>
<td>The anomaly probably results from a rupture at the base of the umbilical cord in an area weakened by the involution of the right umbilical vein.</td>
<td>Failure of development or migration of the folds that form at the umbilical ring results in an anterior wall defect. The umbilical cord inserts into the amniotic sac.</td>
</tr>
<tr>
<td>Clinical</td>
<td>Characterized by an intact umbilical cord with loops of intestine herniated through a small defect to the right side of the cord. There is no sac covering the intestinal defect. Bowel is matted and edematous in appearance.</td>
<td>Characterized by intestinal loops covered by amniotic membrane and peritoneum. Giant omphaloceles may contain liver as well as loops of intestine. Infants have a high incidence of associated anomalies—chromosomal, cardiac, genitourinary and/or craniofacial anomalies.</td>
</tr>
<tr>
<td>Location</td>
<td>Right of intact umbilicus</td>
<td>Proximal part of umbilical cord</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>Antenatal ultrasound</td>
<td>Antenatal ultrasound</td>
</tr>
<tr>
<td>Treatment</td>
<td>Primary repair versus staged repair with silastic silo</td>
<td>Small: One-stage surgical repair</td>
</tr>
<tr>
<td>Other anomalies</td>
<td>Rare; intestinal atresias</td>
<td>Large: Gradual reduction with silastic silo</td>
</tr>
<tr>
<td>Complications</td>
<td>Malrotation, midgut volvulus, hypoperistalsis, intestinal atresias/stenoses, undescended testicles</td>
<td>Malrotation, midgut volvulus</td>
</tr>
</tbody>
</table>

1. Each is contiguous and adherent to some part of the alimentary tract.
2. Each has a smooth muscle coat.
3. Each is lined with mucosa or epithelium similar to that of the stomach, small intestine, or colon.17

Abdominal pain and melena are the most common presenting symptoms. An abdominal mass may be palpated in up to half of infants. Heterotropic gastric mucosa may be present (33%) and may result in ulceration, bleeding, and perforation. Other symptoms, such as vomiting, hematemesis, abdominal distension, melena, abdominal pain, and peritonitis, may occur. Although ultrasonography or barium enema studies may suggest the diagnosis, laparotomy is usually the only definitive method. Sixty percent are diagnosed by 6 months and 80% within the first 2 years of life.17

Small duplications are simply resected followed by a primary anastomosis. Resection of long tubular duplications may not be possible without impairing the vasculature of the normal intestinal tract. Several complicated procedures are advocated; one of which involves mucosal stripping of the duplication.18

**Juvenile Polyps or Retention Polyps**

Polyps are benign hemartomas typically found proximal to the transverse colon. They are uncommon in the newborn period. Bleeding occurs with sloughing of polyps and is painless and rarely massive.

**Anorectal Fissures**

Anorectal bleeding, perhaps the most common cause of lower GI bleeding, is characterized by bright red blood on the outside of the stool and is typically from posterior-midline anal fissures. Fissures are treated with stool softeners and warm sitz baths. Excision is rarely needed.

**Neonatal Surgical Emergencies That Present With Respiratory Distress**

**Congenital Diaphragmatic Hernia**

Congenital diaphragmatic hernia (CDH) is a condition in which a defect in the diaphragm allows abdominal viscera to herniate into the thorax. The incidence of CDH is estimated to be 1 per 2,000 to 5,000 births. Approximately 30% of fetuses that have CDH die before birth, usually from chromosomal or lethal nonpulmonary malformations. Eighty-five percent of defects are left-sided, 13% are right-sided, and 2% are bilateral.19 Most studies have found an equal representation of genders. Up to 50% of newborn infants with CDH will have other anomalies, such as cardiac, genitourinary, gastrointestinal, and chromosomal defects.

Although infants with CDH can have multiple anomalies, they all typically have some component of pulmonary hypoplasia and pulmonary artery hypertension. It is hypothesized that lung development proceeds normally until 9 to 10 weeks of development. This is when the midgut returns to the abdomen from the umbilical cord. If the pleuroperitoneal folds fail to close or muscularize at 8 weeks, the intestine is able to pass into the thorax. Intestine in the thorax causes mediastinal shift, compression of the thoracic contents, and impairment of subsequent pulmonary growth resulting in

Figure 5. Pneumatosis intestinalis and portal venous gas in NEC.
pulmonary hypoplasia, not only in the involved thorax, but also the contralateral lung as well.\textsuperscript{20}

CDH is usually diagnosed prenatally by routine obstetric ultrasound screening. Postnatal diagnosis of CDH is usually made within the first 24 hours of life. Classically, affected infants are born with a scaphoid abdomen and develop progressive respiratory distress as swallowed air causes intestinal distension and worsening lung compression. Mediastinal compression may occur causing decreased venous return, poor perfusion, and hypotension. Although most patients who have CDH present within the first day of life, 10% to 20% present later with recurrent respiratory distress, chronic pulmonary infection, or acute GI symptoms caused by volvulus or intestinal obstruction. These late-presenting infants typically do very well, as they clearly have sufficient pulmonary function to survive.\textsuperscript{21}

Infants who present to the emergency room with CDH are typically not as ill as those diagnosed prenatally or at birth. Infants presenting within the first day of life are gravely ill and require endotracheal intubation, nasogastric tube decompression, intravenous fluids, and inotropic support. They should be cared for in a facility capable of providing advanced pulmonary support, including high-frequency ventilation, nitric oxide, and extracorporeal membrane oxygenation (ECMO). Infants who present to the emergency room outside of the newborn period are managed expectantly. Management includes supplemental oxygen, support of respirations (ABCs), nasogastric tube decompression of abdominal contents, and intravenous fluids. Definitive surgery includes decompression of the lungs by reduction of the abdominal viscera and primary closure of the diaphragmatic defect. If the diaphragm is inadequate, reconstruction can be done with use of nearby musculature or prosthetic material (Gortex, W.L. Gore & Associates, Flagstaff, AZ).\textsuperscript{21}

**Congenital Lung Malformations**

Congenital cystic adenomatoid malformations (CCAM), pulmonary sequestrations, congenital lobar emphysema (CLE) and bronchogenic cysts are rare congenital lung malformations that can present with respiratory distress.

Most neonates with CGAM will present with respiratory distress, tachypnea, retractions, and cyanosis at delivery. Other infants who do not experience respiratory distress at birth present with infectious complications (recurrent pneumonias) or incidentally when a chest radiograph is obtained for a work-up. A chest radiograph may show multiple air-filled, thin-walled cysts of various sizes (Fig 6). A computed tomographic (CT) scan of the chest may differentiate among CCAM, CLE, and bronchogenic cysts. Prenatal treatment of CCAM includes fetal surgery, percutaneous shunt placement, and cyst aspiration. CCAMs that present in infancy are usually removed by anatomic lobectomy and occasionally by partial lobectomy. Removal is recommended because of the association between CGAM and rhabdomyosarcoma and other malignant tumors.\textsuperscript{22}

**Congenital Lobar Emphysema**

CLE is a disease characterized by massive distention of a pulmonary lobe. It is primarily caused by a ball-valve bronchial obstruction that allows inflation during inhalation but obstructs the bronchus during exhalation. Bronchial obstruction can be caused by cartilaginous dysplasia, compression by abnormal vessels associated with congenital cardiovascular disease, bronchial stenosis, and redundant bronchial mucosal flaps.\textsuperscript{22} Symptoms may occur at birth, in the first week, or within the first month of life. Infants usually are normal at birth and then develop symptoms of respiratory distress over the first few days or weeks of life. Symptoms include tachypnea, cyanosis, retractions, and, on physical examination, there may be prolonged breath sounds on expiration. Infants with largeCLEs may present with decreased breath sounds and a prominent hemithorax on the involved side, and their trachea may be shifted to the contra-

![Image](image-url)
Pulmonary Sequestration

Pulmonary sequestration is a lung tissue mass that receives its blood supply from an anomalous systemic artery and does not communicate with the bronchial tree by a normal bronchus. Pulmonary sequestrations can be either extralobar or intralobar. An extralobar sequestration is a mass of abnormal lung tissue contained in a pleural envelope separate from that of the normal lung and is usually in the posterior lower chest. In an intralobar sequestration, the arterial supply comes from a systemic artery, usually the aorta or a primary branch of the aorta. In neonates and infants younger than 1 year of age, pulmonary sequestrations are usually diagnosed as an incidental finding. Patients with extralobar sequestrations are diagnosed at an earlier age because of the higher incidence of associated anomalies. Infants present with frequent lower respiratory infections, reactive airway disease, and, occasionally, hemoptysis.

A chest radiograph may show a soft tissue opacity in the posterior basal segment of the lung. CT imaging and ultrasonography are also helpful in defining the abnormal lung tissue. Treatment of pulmonary sequestration is resection; extralobar tissue can be treated with resection alone, and intralobar lesions will usually require lobectomy.

Bronchogenic Cyst

Bronchogenic cysts are lesions that arise from abnormal budding of the trachobronchial tree during airway development. They are located either in the mediastinum (70%) or in the lung parenchyma (30%). Infants with bronchogenic cysts present with respiratory distress, cough, wheezing, stridor, dyspnea, cyanotic spells, and pneumonia. Chest radiographs show segmental or lobar cysts that may contain air, fluid, or an air/fluid level. CT scan confirms the diagnosis. Treatment of symptomatic lesions is partial or total lobectomy. Morbidity and mortality are minimal.

Esophageal Atresia and Tracheoesophageal Fistula

Esophageal atresia (EA) is estimated to affect 1 in 3,570 to 1 in 4,500 live births. Most commonly, patients will also have an abnormal communication between the trachea and lower esophageal segment called a distal tracheoesophageal fistula (TEF). It occurs in about 85% of patients. Less commonly, there may be atresia alone with no fistula (6%), a proximal TEF and no distal fistula (2%), both a proximal and distal fistula (7%), or no esophageal atresia with a congenital TEF (H-type fistula).

A history of maternal polyhydramnios and prematurity is common. Infants present with excessive drooling, excessive mucus or saliva with or without respiratory distress, or with recurrent pulmonary infections. Symptoms, such as choking and coughing caused by aspiration of feeds or excessive tracheal secretions, recurrent respiratory infections, or abdominal distension caused by aerophagia, typically appear at birth. In a large retrospective study of 227 infants, 64% of infants with EA had associated congenital anomalies. Congenital heart disease was noted in 86 (38%), musculoskeletal anomalies in 44 (19%), neurologic defects in 34 (15%), and renal defects in 35 (15%), and anorectal defects in 18 (8%).

In a newborn with suspected EA, a 10-gauge French catheter introduced into the mouth will not pass into the stomach as seen on a radiograph (Fig 8). The tube will be in a blind pouch in the upper mediastinum. In neonates with EA without a fistula or atresia with a proximal fistula, radiographs will
show absence of gas in the abdomen. A contrast study or bronchoscopy will show the proximal fistula.

Emergency treatment includes support of the infant's airway, breathing, and circulation (ABCs), while minimizing crying (which fills the stomach with excessive air). Allow the infant to sit upright, suction the upper esophageal pouch frequently, give intravenous fluids but no oral fluids, and avoid bag-valve-mask ventilation. Occasionally, assisted ventilation by endotracheal intubation is necessary. Definitive surgical treatment depends on the type of EA. In general, the connection of the distal esophagus and trachea is identified and divided. The upper esophagus is mobilized to allow an end-to-end anastomosis between the upper pouch and lower esophageal segment.

Several complications can occur after repair of EA, including an anastomotic leak, recurrent TEF, anastomotic stricture resulting in feeding difficulties, and gastroesophageal reflux. Most infants will also have significant respiratory symptoms during the first year after repair. Symptoms include wheezing, persistent coughing, stridor, choking, and even life-threatening episodes of apnea and cyanosis. Neonates over 1,500 grams without major congenital cardiac malformations do very well. Excluding those with major life-threatening congenital anomalies, survival exceeds 90%.

References

19. Meurs KV, Short BL: Congenital diaphragmatic


