NEONATAL SURGICAL EMERGENCIES

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Advances in perinatology and neonatology over the last 2 decades have improved the survival of premature, especially extremely premature (<1500 g) and critically ill, newborn babies. Most of the disorders that were considered neonatal surgical emergencies in the past no longer require immediate surgery because of new technology and new methods of treating sick neonates. This article describes the more common neonatal disorders that require semi-elective or emergency surgery, and focuses on factors that affect the anesthetic management of patients with these disorders.

PYLORIC STENOSIS

Pyloric stenosis is the most common gastrointestinal obstructive anomaly in neonates. It occurs in approximately 1 in 500 live births, and is found more frequently in males. Symptoms are usually not apparent until the second to sixth week of life, and are caused by hypertrophy of the muscular layer of the pylorus. The affected area feels like an olive-shaped mass. It is palpated most easily when the stomach is empty, and is located slightly to the right of the midline in the epigastric area. A barium swallow roentgenogram or ultrasonogram can confirm the lesion.95

When an infant is suspected of having pyloric stenosis, oral feedings
should be discontinued. Metabolic derangements associated with pyloric stenosis result from persistent vomiting, with loss of water, hydrogen, chloride, sodium, and potassium. As hydrogen ions are lost, the kidneys initially secrete potassium in exchange for hydrogen to maintain a normal serum pH. When sodium depletion becomes more severe, the kidneys secrete potassium and hydrogen in exchange for sodium, which results in a patient who is hypokalemic, hypochloremic, and alkalotic. If electrolyte loss continues, the kidneys secrete an acid urine (i.e., paradoxic acidosis), and metabolic alkalosis becomes more profound. Eventually, metabolic acidosis from dehydration and hypoperfusion occurs.

**Anesthetic Management**

Surgical correction of pyloric stenosis, pyloromyotomy, is not a procedure that should be performed emergently if the patient is dehydrated or is depleted of electrolytes. Fluid, electrolyte, and acid-base balance should be re-established before anesthetizing a neonate with pyloric stenosis, so that perianesthetic complications can be avoided. For example, if a patient is still alkalotic postoperatively, hypoxia can occur as the baby attempts to correct the alkalotic state by hypoventilating. Mild-to-moderate fluid and electrolyte problems (chloride > 90 mEq/L and urine specific gravity < 1.020) can be corrected with 5% dextrose in 0.45% sodium chloride and 20 to 40 mEq/L of potassium, infused at a rate of 10 mL/kg per hour. Severe dehydration and electrolyte imbalance (chloride < 90 mEq/L, sodium < 120 mEq/L, and no urine output) requires volume expansion with isotonic sodium chloride or colloid and blood until urinary output is re-established. Potassium chloride then can be added to the infusion. In severe cases of dehydration, restoring fluid, acid-base, and electrolyte balance may take several days.

Immediately before the induction of anesthesia, standard monitoring (e.g., blood pressure cuff, ECG leads, pulse oximeter probe, and precordial stethoscope) should be started, and a large-bore orogastric tube should be passed into the stomach several times until the amount of fluid aspirated is minimal. Endotracheal intubation after preoxygenation and a rapid-sequence induction is the method many anesthesiologists prefer for inducing anesthesia in these babies. Induction agents and doses for these patients are found in Table 1. If difficulty with intubation is anticipated, an endotracheal tube should be placed to secure the airway before anesthesia is induced. An inhalation induction of anesthesia should be avoided in these patients because they have an increased risk for vomiting and aspirating gastric contents. Even after multiple attempts to remove gastric contents by suction, a considerable amount of residual material still may be present. Anesthesia may be maintained with inhalation anesthetics or in combination with intravenous drugs. Any volatile anesthetic drug may be used for anesthesia; however, the same inspired concentration of volatile anesthetic generally causes a
higher incidence of cardiovascular instability in infants, compared with older patients. This effect can be attributed to several factors, including faster equilibration, rapid myocardial and brain uptake, increased anesthetic requirements, and sensitivity of the immature myocardium to volatile agents (especially halothane). These factors, the decrease in myocardial contractile mass, and less magnitude and velocity of fiber shortening produce the adverse cardiovascular effects. Volatile agents also depress the baroreceptor reflexes in a concentration-dependent manner in the younger compared with the older patient.\textsuperscript{76, 77, 108}

Before terminating anesthesia, measures to minimize postoperative pain should be instituted. These include rectal acetaminophen, (30–40 mg/kg),\textsuperscript{15} caudal analgesia, and infiltration of the surgical incision with a local anesthetic (e.g., 0.25% bupivacaine) before abdominal closure. After the patient is awake and ventilating adequately, the endotracheal tube can be removed.

Postoperatively, the child may exhibit signs of drowsiness or lethargy. Respiratory depression (e.g., apnea) also may be present.\textsuperscript{5} If total body potassium is decreased (which may occur despite a normal serum potassium), there may be altered electrical forces across cell membranes. This causes an unpredictable response (i.e., potentiation) to nondepolarizing neuromuscular blocking agents and weakening of the muscles, including the muscles of respiration, creating a need for postoperative mechanical ventilation. If hypokalemia is severe, rhabdomyolysis and impairment of smooth muscle function can occur. Bladder dysfunction, ileus formation from lack of peristalsis, poor peripheral vascular tone with orthostatic hypotension, and a poor pressor response to catecholamines are possible. Cardiac dysrhythmias also may be present because

Table 1. DRUGS AND INTRAVENOUS DOSE

<table>
<thead>
<tr>
<th>Drugs</th>
<th>Dose</th>
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<tbody>
<tr>
<td>Anticholinergic drugs</td>
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<tr>
<td>Atropine</td>
<td>0.01–0.02 mg/kg</td>
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<tr>
<td>Glycopyrrolate</td>
<td>0.005–0.01 mg/kg</td>
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<td>Anesthetic induction drugs</td>
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<tr>
<td>Thiopental</td>
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<tr>
<td>Propofol</td>
<td>2.5–3 mg/kg</td>
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<tr>
<td>Etomidate</td>
<td>0.3 mg/kg</td>
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<tr>
<td>Muscle relaxants</td>
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<tr>
<td>Depolarizing</td>
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<tr>
<td>Succinylcholine</td>
<td>1–2 mg/kg</td>
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<tr>
<td>Nondepolarizing</td>
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<tr>
<td>Cisatracurium</td>
<td>0.2 mg/kg</td>
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<tr>
<td>Rocuronium</td>
<td>0.6–1 mg/kg</td>
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<tr>
<td>Vecuronium</td>
<td>0.1 mg/kg</td>
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<tr>
<td>Pressors</td>
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<tr>
<td>Catecholamines</td>
<td>3–5 μg/kg/min</td>
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<td>Dopamine</td>
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hypokalemia enhances the intrinsic automaticity, sustains dysrhythmias, and alters conductivity.\textsuperscript{14}

\textbf{CONGENITAL DIAPHRAGMATIC HERNIA}

Approximately 1 in 2500 babies is born with a congenital diaphragmatic hernia.\textsuperscript{85} The disorder is characterized by failure of the pleural and peritoneal canal to close at approximately the eighth week of gestation, resulting in herniation of the abdominal viscera into the thoracic cavity and pulmonary hypoplasia from compression by the viscera on the developing lungs. Hernias through the posterolateral portions of the diaphragm (Foramen of Bochdalek) account for approximately 80\% of diaphragmatic hernias; herniation on the left side is five times more common than on the right side. These Bochdalek hernias are the largest diaphragmatic hernias, and pulmonary hypoplasia is more extensive in these patients. Hernias through the anterior part of the diaphragm (Foramen of Morgandi) account for only 2\% of all diaphragmatic hernias; esophageal hiatus hernias account for 18\%.

The prevalence of posterolateral hernias may be related to the development of the diaphragm. During the second month of gestation, a pleuroperitoneal membrane is formed. This membrane eventually divides the common pleuroperitoneal cavity into two compartments, the pleural cavity and the peritoneal cavity. The last areas of the membrane to develop are the posterolateral portions, with the right side closing before the left. At approximately the ninth gestational week, the gut, which was in the yoke sac, migrates to the peritoneal cavity. If this migration occurs before the pleuroperitoneal membrane closes off the pleural cavity, the stomach, spleen, liver, and bowel can migrate into the pleural cavity and compress the developing lungs. Compression of the lungs affects their growth. Although the lung on the side of the hernia is more severely affected, the lung on the contralateral side is also hypoplastic. Severe pulmonary hypoplasia with arrest of bronchial and bronchiolar development occurs if a large volume of viscera is present in the pleural cavity before the 12th week of gestation.\textsuperscript{86} Compression of the lungs, especially when airways are dividing and pulmonary arteries are forming, causes a decrease in lung volume, number of bronchi and alveoli, and cross-sectional area of pulmonary arterial branches.\textsuperscript{86} The pulmonary artery is smaller, arterial branching is diminished, and the arteries are more muscular. Although significant lung growth at the alveolar level and remodeling of the vasculature (i.e., in larger, less muscular arteries) occurs after birth, the time this takes exceeds the current limitation of supportive measures, such as extracorporeal membrane oxygenation.\textsuperscript{10} Severe impairment of normal pulmonary development often results in a critically ill neonate. Persistent pulmonary hypertension with increased vascular resistance frequently occurs; in many cases, these conditions lead to hypoxemia, acidosis, and death.\textsuperscript{39, 45, 66}

The results of recent animal models have challenged current think-
ing regarding the cause of congenital diaphragmatic hernia. When the drug nitrofen (2,2 dichloro-phenyl-p-nitrophenyl ether) was used, the left lung failed to develop, leading to a left diaphragmatic hernia. Congenital diaphragmatic hernia may be secondary to the failure of normal lung development rather than being the cause of it.55

With the increased use of prenatal ultrasonography, prenatal diagnosis of congenital diaphragmatic hernia has become more common.1, 47, 56 Neonates with congenital diaphragmatic hernias usually have scaphoid abdomens and barrel chests. Children who are in respiratory failure within the first 6 hours of life have the highest mortality, whereas those diagnosed later generally have a good prognosis.56, 111 Approximately one third of the babies who are symptomatic in the first 6 hours of life have an associated anomaly, with cardiac lesions predominating.39, 40 The babies with associated anomalies, especially those with cardiac anomalies, have significantly lower APGAR scores. Their best postductal Po2 before extracorporeal membrane oxygenation or surgery is lower than that in those children with hernia with no other anomalies.40 The literature indicates that a sustained alveolar-to-arterial oxygen tension gradient of greater than 600 mm Hg and an oxygenation index over 40 for 8 to 12 hours still are associated with mortality.99 Some clinicians suggest that the persistence of a postductal Pao2 below 100 mm Hg and of a Paco2 above 40 mm Hg despite optimal conventional therapy indicates poor prognosis.

Data extrapolated from Boix-Ochoa16 and Bohn16, 17 indicate that some babies who have a persistently low postductal Pao2 should have their preductal Pao2 measured because it is a directly measured physiologic value. An infant with congenital diaphragmatic hernia with a persistent Pao2 below 8046 or who cannot generate a preductal Pao2 of at least 100 mm Hg at some point has pulmonary hypoplasia to a degree incompatible with life despite extracorporeal life support (ECMO).99

Before Ladd and Gross’s series in 1940 of long-term survivors, operative repair of neonates with congenital diaphragmatic hernia was considered impossible.68 For years later, the survival rate of these patients remained at approximately 50%.50, 75, 91 Despite improvements in the clinical management of neonates, diagnostic tools, and surgical techniques, the multicenter Congenital Diaphragmatic Hernia Study Group reported, in 1998, a survival rate of only 63%.27

In the past, babies with congenital diaphragmatic hernia were rushed to the operating room immediately after birth, without taking time for stabilization. The change in practice to delay surgery until the respiratory problems are stabilized reflects the present belief that surgery cannot correct respiratory insufficiency. The primary problem in these babies is not respiratory failure from lung compression by the herniated viscera but respiratory failure from severe pulmonary hypoplasia and pulmonary hypertension.9 Currently, neonates with congenital diaphragmatic hernia are managed medically until their respiratory and cardio-

*References 6, 13, 17, 19, 28, 54, 59, 72, 93, and 107.
vascular statuses are stabilized. Only after they are optimized are they scheduled for semi-elective surgery.

Because pulmonary hypoplasia and associated anomalies are the leading causes of death in neonates with congenital diaphragmatic hernia, treatments now are directed at correcting problems caused by these conditions, with concurrent improvement in outcome. Lung rupture from barotrauma easily can occur at any time during the perioperative period. Ventilatory strategies that minimize airway pressure and reduce barotrauma to the severely hypoplastic lungs improve survival. Gentle ventilation that allows arterial CO₂ to be slightly higher than normal (i.e., permissive hypercapnia) and marginal postductal oxygen saturation are associated with a higher survival rate than conventional ventilation with or without extracorporeal membrane oxygenation.

High-frequency ventilation has been advocated to reduce barotrauma and to produce alkalosis to improve pulmonary blood flow. This artificial ventilation method can be used before, during, and after surgical repair. The major disadvantage of using high-frequency ventilation during anesthesia is that end-tidal CO₂ is difficult to monitor. Frequent monitoring of end-tidal CO₂ and O₂ saturation and arterial blood gas determinations are essential to ensure effective alveolar ventilation and oxygenation. If CO₂ tension is allowed to rise too high, pulmonary vascular resistance, which is generally high, increases further.

A pneumothorax should be considered if sudden unexplained hypotension or desaturation occurs. A pneumothorax that is not treated promptly can lead to significant hypotension and hypoxia. A pneumothorax on the contralateral side can affect ventilation more significantly than one on the same side as the hernia, because the lung on the contralateral side is generally the better developed. In patients with chest tubes, an increase in lung compliance and abdominal distension may be caused by a pneumoperitoneum. If the air leak into the chest is large and the chest tube cannot evacuate the gas from the thorax adequately, gas could flow through the diaphragmatic defect into the peritoneal cavity. Significant abdominal distension can result, causing diminished lung expansion and hypoxia. In this case, decompressing the abdomen with a large-bore needle allows the gas to escape so the lungs can expand.

New modes of therapy have led to some improvement in the survival rate of neonates with congenital diaphragmatic hernia. Extracorporeal membrane oxygenation is one treatment that has improved the survival rate of infants who originally had a predicted mortality of 80%. Inhaled nitric oxide, a selective pulmonary vasodilator, does not improve survival of neonates with congenital diaphragmatic hernia. The efficacy of surfactant replacement therapy in these patients is currently unclear. Further discussion of these treatment modalities is beyond the scope of this article, but can be found in the literature.

Surgical repair of congenital diaphragmatic hernia in neonates can be performed through an abdominal incision, but some surgeons prefer
a thoracoabdominal approach. Small defects are closed by suturing existing diaphragmatic tissue together. With large defects, an artificial diaphragm is constructed. If the artificial diaphragm material is not permeable to gases, abdominal distension from a pneumothorax is minimized and the need for a peritoneal drain is reduced. Occasionally, abdominal closure may not be possible without compromising ventilation. In these cases, an abdominal silo is created to house the viscera until the abdominal wall stretches enough to allow abdominal closure at a later stage.

**Anesthetic Management**

The anesthetic management of neonates with congenital diaphragmatic hernia involves a thorough assessment of the child. Management of these patients depends on their physiologic condition and anatomic pathology. A review of laboratory, radiographic, and physical findings and optimization of any anomalies should be completed before anesthesia is administered. Listening to breath sounds beforehand and knowing whether the hernia is on the left or right side are important. For example, a decrease in breath sounds over the right chest in a child with a right-sided hernia may not be caused by the intubation of the left main stem bronchus.

In the past, babies with congenital diaphragmatic hernia were operated on immediately after birth. Surgery now is delayed until the patients are medically stabilized. Consequently, many babies already are intubated, and have indwelling intravenous and arterial lines in place by the time they arrive at the operating room. Children who are not intubated and mechanically ventilated before arriving at the operating room generally fare best if they are nursed in a semirecumbent position with their affected side down. An orogastric or nasogastric tube often is inserted to decrease and prevent distension of the stomach and intestines. Neonates who are not intubated before arrival at the operating room generally are intubated awake or after a rapid-sequence induction (see Table 1). An anticholinergic drug, such as atropine, 0.02 mg/kg administered intravenously, is often the only drug used for premedication. This drug is administered immediately before the induction of anesthesia to prevent bradycardia and a drop in cardiac output during induction. If the baby is stable and an awake intubation is planned, analgesics may be administered to decrease the stress of airway instrumentation. Although an inhalation induction of anesthesia in a spontaneously breathing child is possible, assisted or controlled ventilation generally is required to ventilate the child adequately before a satisfactory level of anesthesia is achieved for laryngoscopy and intubation. If the stomach is in the chest, mask ventilation with high inflation pressures should be avoided to prevent gastric distension. High inflation pressures also should be avoided because many of these children develop lower
esophageal sphincter incompetence and are at risk for reflux and aspiration of gastric content.\textsuperscript{80}

The choice of drugs for anesthesia should be determined by the patient's general condition. In infants and children with small veins, pentothal is preferred to propofol because of the high incidence of pain when propofol is administered into small veins. High doses of narcotics (e.g., fentanyl, 15–25 \(\mu\)g/kg) can be used if hemodynamically tolerated. Nitrous oxide should be avoided because it defuses into the bowel lumen, causing further bowel expansion, and may compromise lung expansion, especially if the intestines are in the chest. The risk and benefits of the various inhalation anesthetics should be considered before deciding on their use. Compared with other volatile anesthetics, halothane produces a greater decrease in inotropy and chronotropy of the heart at equipotent doses.

Vigilant monitoring is mandatory in neonates with congenital diaphragmatic hernias. Monitoring blood pressure, ECG, pulse oximetry, capnography, temperature, and heart rate is essential during anesthesia for all babies with congenital diaphragmatic hernias. Following preductal and postductal oxygen saturation with pulse oximeter and probes on the right upper extremity and on a lower extremity allows early detection of right-to-left shunting. Although invasive monitoring is not necessary in all patients, it may be indicated in those who are in respiratory distress. Cannulation of the right radial artery will allow measurement of preductal blood gases. In addition to providing continuous blood pressure readings, preductal blood gases can be measured. Conditions that can cause pulmonary vasoconstriction, such as hypoxia, acidosis, and hypothermia, should be avoided. If possible, oxygen saturation should be kept above 80 mm Hg, and CO\(_2\) should be kept within normal or only slightly above the normal range (i.e., permissive hypercapnia). Metabolic acidosis should be treated immediately, and any significant blood loss should be replaced.

Familiarity with the different therapeutic modalities that may be needed during anesthesia is essential to provide optimal anesthetic care. Understanding the principles of high-frequency ventilation and ECMO is essential. Occasionally, the hernia is repaired while a child is still on ECMO, despite hemodynamic and pulmonary instability.\textsuperscript{29} For these patients, an opioid and a nondepolarizing muscle relaxant may cause less hemodynamic instability than an inhalation anesthetic. A volatile anesthetic agent, such as isoflurane, can be used for these patients by administering it through the ECMO circuit (Fig. 1).\textsuperscript{7} For children on ECMO, drugs may be administered directly to them, or into the ECMO circuit. Because patients on ECMO are heparinized, intraoperative bleeding can be problematic because hemostasis is abnormal.\textsuperscript{106}

**ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA**

Approximately 1 in 4000 babies are born with esophageal atresia, and 90% of them have an associated tracheoesophageal fistula (i.e.,
Figure 1. Isoflurane during extracorporeal membrane oxygenation.
cardiac, gastrointestinal, genitourinary, musculoskeletal, or craniofacial anomalies) are present in 30% to 50% of newborns with esophageal atresia and tracheoesophageal fistulas. One form of the associated anomalies is the VATER (i.e., vertebral and vascular anomalies, imperforate anus, tracheoesophageal fistula, radial aplasia, and renal abnormalities) or VACTERL (i.e., vertebral anomalies, imperforate anus, cardiac anomalies, tracheoesophageal fistula, renal abnormalities, radial limb aplasia) association.

The diagnosis of esophageal atresia is usually made when a tube cannot be passed into the newborn's stomach after delivery, after the first feeding when coughing and choking occurs, or after recurrent pneumonia associated with feedings. Prenatally, the diagnosis should be considered if polyhydramnios is present. After birth, the diagnosis is confirmed by the inability to pass an orogastric tube into the stomach. Location of the atretic segment is confirmed by a radiograph showing the tip of a radiopaque catheter that is passed into the esophagus until it stops. Air in the stomach and intestines usually is seen in abdominal radiographs of neonates with tracheoesophageal fistula. The absence of air usually indicates esophageal atresia without a fistula. Occasionally, the diagnosis of a tracheoesophageal fistula is not made until later in the child's life.

Although Thomas Gibson first described esophageal atresia in 1697, no baby survived until Dr. Logan Levin performed the first successful staged repair in 1939. Four years later, the first primary repair was performed. Because of subsequent advances in neonatal and perioperative care, nearly all babies survive. The exceptions are those who are very premature (< 1500 g) and those who have severe cardiac or chromosomal anomalies.

The most common form of esophageal atresia and tracheoesophageal fistula (80%-90% of patients) is an upper esophagus ending in a blind pouch and the distal esophagus forming a tracheoesophageal fistula. In most cases, anastomosis of the two ends of the esophagus is possible. The challenge occurs when a long segment of atresia is present. A staged repair with traction, myotomies, or multiple esophagotomies on the segments is possible in some cases. In others, a portion of the colon or jejunum or a tube made from the stomach was interposed. Gastric interposition is another procedure. Although survival is improved with these interposition procedures, the mortality rate in these patients is high.

Morbidity and mortality in neonates with esophageal atresia and tracheoesophageal fistula generally are related to pulmonary complications. The goal of preoperative management should be to minimize these complications. Once the disorder is diagnosed, attempts should be made to prevent aspiration pneumonia. Oral feedings should not be given, and the baby should be nursed in the semirecumbent position to minimize the possibility of aspiration of gastric and nasopharyngeal contents. A tube should be passed into the esophagus and suctioned frequently to minimize the accumulation of nasopharyngeal
secretions in the blind pouch. For an extremely premature or critically ill infant, a staged approach should be considered. After the diagnosis is made, a gastrostomy is placed to prevent excess gastric distension from impairing diaphragmatic excursion, and definitive repair is postponed until the baby is more stable.4,52

Anesthetic Management

Atropine generally is the only drug used for premedication of infants with esophageal atresia and tracheoesophageal fistula. It is given to decrease the incidence of bradycardia from vagal stimulation during intubation and to maintain cardiac output during the induction of anesthesia. Monitors should include devices to measure blood pressure, ECG, heart rate, temperature, pulse oximetry, and capnography. A precordial stethoscope placed on the left chest under the axilla allows monitoring of heart and breath sounds.

If a difficult intubation is anticipated, if gastric distension is present, or if the baby is vomiting, endotracheal intubation should be performed while the child is still awake. Otherwise, intubation usually is performed after a rapid-sequence induction of anesthesia (see Table 1). If an inhalation induction of anesthesia is chosen before the endotracheal tube is placed distension of the stomach will be minimized if the child is allowed to breathe spontaneously. To prevent gas from transiting through the fistula and distending the stomach when ventilation is assisted or controlled before the endotracheal tube is inserted airway pressure should be minimized. Because a tracheoesophageal fistula generally is located on the posterior aspect of the trachea and just proximal to the carina, placing the endotracheal tube into a bronchus and withdrawing it until breath sounds are equal may prevent ventilation of the fistula.94 Rotating the endotracheal tube during intubation so the bevel faces posteriorly may prevent intubation of the fistula. After the endotracheal tube is placed, positive-pressure ventilation should be instituted, and the size of the stomach should be assessed before administering a muscle relaxant. If gastric distension becomes a problem, it may be necessary to allow the child to breathe spontaneously until a gastrostomy tube is placed or until the surgeon can obstruct the fistula manually. After a gastrostomy, ventilating the patient adequately may be difficult if the volume of gas flowing through the fistula is high. In these cases, a balloon Fogarty catheter placed in the fistula through a bronchoscope or insertion of this catheter under fluoroscopic guidance through the gastrostomy into the fistula will occlude the fistula and allow for easier ventilation of the child.41,58

In most cases, the esophagus is anastomosed primarily, and the fistula is ligated through a right thoracotomy. During surgery, airway obstruction can occur if the trachea is compressed, secretions become inspissated or a blood clot forms in the airway. The surgeon must be
informed of the problem so that he or she can correct the situation or help change the endotracheal tube.

At the end of the surgical procedure, most infants are extubated if they are awake and are not at risk for postoperative respiratory problems. Those who are at risk for respiratory problems (e.g., postanesthetic apnea\textsuperscript{31, 73, 110}) should remain intubated until they demonstrate that they can sustain adequate ventilation postoperatively. Premature extubation also may be dangerous in these babies because the recently closed fistula may be ruptured by the endotracheal tube if it needs to be reinserted.

Neonates with a tracheoesophageal fistula frequently have a reduction in tracheal cartilage (i.e., tracheomalacia).\textsuperscript{33, 48, 105} In babies with tracheomalacia, ventilation may be adequate immediately after extubation. In some patients, ventilatory problems may not be apparent until later in the postoperative period. Dynamic collapse of the trachea during inspiration, may be magnified as the child’s respiratory efforts increase. Tracheopexy may be necessary, or a tracheotomy may be required.\textsuperscript{22, 30, 65, 100}

**OMPHALOCELE AND GASTROSCHISIS**

Omphalocele and gastroschisis are abdominal wall defects that can be diagnosed by fetal ultrasound in the first trimester.\textsuperscript{9, 42} Salient features of these conditions are shown in Table 2. Omphalocele occurs in 1 in 6000 births, and is caused by a failure of the gut to migrate from the yolk sac to the abdomen during gestation.\textsuperscript{36, 62} Neonates with an omphalocele have a high incidence of other anomalies, and frequently are premature at birth. Associated anomalies include cardiac, gastrointestinal (e.g., malrotation, Meckel’s diverticulum, intestinal atresia), genitourinary (e.g., extrophy of the bladder), metabolic (e.g., Beckwith-Wiedemann syndrome: macroglossia, hypoglycemia, organomegaly, gigantism), and chromosomal abnormalities. Cardiac and thoracic defects are more frequent in neonates with epigastric omphaloceles, whereas cloacal anomalies and extrophy of the bladder more frequently are associated with hypogastric omphaloceles.

Gastroschisis results from occlusion of the omphalomesenteric artery during gestation, and involves herniation of the viscera through the lateral abdominal wall defect.\textsuperscript{53} Gastroschisis occurs in 1 in 15,000 births, and is associated with a low incidence of other anomalies. Intestinal

<table>
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<th>Table 2. COMPARISON OF OMPHALOCELE WITH GASTROSCHISIS</th>
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<tr>
<td><strong>Omphalocele</strong></td>
</tr>
<tr>
<td>Incidence</td>
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<tr>
<td>Gestation</td>
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<tr>
<td>Peritoneal covering</td>
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<td>Incidence of other anomalies</td>
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atresia, however, is higher in neonates with gastroschisis, compared with omphaloceles.

**Anesthetic Management**

Preoperative management of neonates with gastroschisis and omphalocele is similar, and is directed at preventing infection and minimizing fluid and heat loss. Covering the exposed viscera or membranous sac with sterile saline-soaked dressings and wrapping these dressings with plastic wrap decreases evaporative fluid and heat loss.

Although surgical correction of an omphalocele or gastroschisis is urgent, surgery usually is delayed until the patient is properly prepared for anesthesia. Preoperative preparation involves a thorough evaluation because many of these babies have associated anomalies. Preoperative management should be individualized. Fluid and electrolyte abnormalities should be corrected preoperatively.

Anesthetic management involves volume resuscitation and the prevention of hypothermia. Even when the abdomen is relaxed by muscle relaxants and the viscera are not distended, primary closure may not always be possible without markedly increasing intra-abdominal pressure. Stretching the abdominal wall and pushing bowel contents toward the stomach and rectum may help decrease intra-abdominal pressure. Ventilatory compromise and decreased organ perfusion are the major problems that result when intra-abdominal pressure is increased markedly. Other problems include bowel edema, anuria, and hypotension. A pulse oximeter probe on the lower extremity will detect a decrease in oxygen saturation that could be caused by congestion of the lower extremities due to obstruction of venous return. Measurement of intra-gastric pressure, central venous pressure, or cardiac index can aid in determining whether primary closure is appropriate. If primary closure is not reasonable, a staged repair can be done. This involves leaving the bowel extraperitoneally encased in a synthetic mesh (silon chimney) or plastic pouch, that is reduced in stages over several days until the abdominal cavity can accommodate the viscera without compromising organ perfusion or ventilation. Most neonates do not require anesthesia for the daily reduction in size of the chimney, but they do need anesthesia for the final stage of repair, which involves removing the synthetic material and closing the abdomen. Postoperative management depends on the type of repair and whether or not the child has associated anomalies. Fluid resuscitation should continue postoperatively because fluid loss through the viscera continues, especially in a staged repair, where the viscera are left extraperitoneally.

**NECROTIZING ENTEROCOLITIS**

Necrotizing enterocolitis is a condition that results from hypoperfusion of the gut. It occurs predominantly in premature infants with a
gestational age of less than 32 weeks and with a weight of less than 1500 g. The pathogenesis of this condition is elusive. Immaturity of the gastrointestinal mucosal barrier and the immune system and premature oral feeding are conditions frequently implicated in causing this condition. Early signs of necrotizing enterocolitis include feeding problems, lethargy, hyperglycemia, bloody stools, and fever. The abdomen may be distended and tender. Radiographic studies that may initially suggest an ileus with edematous bowel show free air in the abdomen if intestinal perforation occurs.

Necrotizing enterocolitis frequently is treated medically if the diagnosis is made early. Enteral feedings are discontinued, the abdomen is decompressed with a nasogastric or orogastric tube that is attached to intermittent low-pressure suction, and blood volume is replenished with intravenous fluids. Broad-spectrum antibiotics usually are administered, although no specific organism has been implicated in causing necrotizing enterocolitis. Dopamine is given to improve renal and intestinal perfusion and cardiac output.

Indications for surgery are usually refractory sepsis or intestinal perforation. As a result, babies with necrotizing enterocolitis are usually severely ill when they are scheduled for surgery. They may be hypotensive, anemic, thrombocytopenic, and coagulopathic, and have prerenal azotemia and a metabolic acidosis.

**Anesthetic Management**

Patients with necrotizing enterocolitis frequently arrive in the operating room with an endotracheal tube in place. If they are not intubated, a rapid-sequence or awake intubation is performed after the standard monitors are placed. Drugs used for anesthesia depend on the patient’s condition. A narcotic (e.g., fentanyl, 1-5 µg/kg) and a relaxant (e.g., cisatracurium, 0.1-0.2 mg/kg) frequently are the only drugs used for anesthesia, because potent inhalation anesthetics may decrease blood pressure below acceptable levels. Nitrous oxide generally is avoided to prevent further intestinal distension. Other drugs may be required to treat cardiovascular instability or electrolyte abnormalities. Dopamine may be indicated if renal perfusion and cardiac output are low; bicarbonate may be indicated if the base deficit is significant.

Volume resuscitation is crucial when managing neonates with necrotizing enterocolitis. Third-space fluid loss is high, and multiple blood volumes of lactated Ringer’s solution or albumin may be needed to replenish intravascular volume. Blood, fresh frozen plasma, and platelets also may be needed to improve oxygen-carrying capacity or to treat factor deficiencies.

After surgery, babies are transported with their endotracheal tube in place to the ICU, and are placed on a ventilator until extubation criteria are met. Fluid and drug resuscitation may need to be continued until vital signs are stable.
INTESTINAL OBSTRUCTION

Intestinal obstruction is a surgical emergency in the newborn that requires swift intervention. Delays in diagnosis and treatment may lead to intestinal perforation, bowel necrosis, and septicemia.

DUODENAL OBSTRUCTION

The incidence of duodenal obstruction in the neonate is 1 in 10,000 to 40,000 births. It frequently is associated with other congenital anomalies, such as Down syndrome, cystic fibrosis, renal anomalies, intestinal malrotation, and midline defects, such as esophageal atresia and imperforate anus. Obstruction of the duodenum can be due to an intraluminal diaphragm or membranous web. Infants with obstructions may or may not pass meconium in the first day of life, depending on whether the obstruction is partial or complete. The infant presents with vomiting of bile or bile-stained gastric contents and minimal abdominal distension.

JEJUNOILEAL ATRESIA

Jejunoileal atresia causes complete intestinal obstruction. The incidence is 1 in 5000 live births. This lesion, in contrast to duodenal obstruction, rarely is associated with other congenital anomalies. Fifty percent of infants with this lesion are premature. Jejunoileal atresia is believed to be secondary to an intrauterine vascular accident.

MECONIUM ILEUS

Meconium ileus is an intraluminal obstruction of the distal small bowel caused by abnormal meconium. This entity is found exclusively in patients with cystic fibrosis. Approximately 10% to 20% of the patients with cystic fibrosis are born with meconium ileus. In some patients, the obstruction is managed medically; in others, surgical intervention is necessary. During surgery the obstruction is located, and the meconium is massaged into the colon. Occasionally, the meconium may need to be softened by agents such as acetylcysteine before it can be massaged into the colon. If the meconium cannot be pushed into the colon, an enterostomy is performed, and the meconium obstruction is evacuated from the bowel.

MALROTATION AND VOLVULUS

The true incidence of malrotation and volvulus of the intestines is not known because many patients are undiagnosed, and is believed to
occur because of abnormalities or arrests in the rotation of the bowel. Areas of ischemia and atresia develop, resulting in bowel strangulation, bloody stools, peritonitis, and hypovolemic shock. Malrotation frequently is associated with major cardiac, esophageal, urinary, and anal anomalies, congenital diaphragmatic hernia, and abdominal wall defects. Malrotation is two times more common in male infants, and frequently presents in the neonatal period. This problem also can present in adulthood.

**IMPERFORATE ANUS**

The incidence of anal atresia is 1 in 5000 live births. The presence of this entity is usually obvious at birth, and can range from a mild stenosis to a complex syndrome with other associated congenital anomalies. In general, the higher the location of the anomaly, the greater the incidence of associated anomalies. In male infants, an operative procedure to relieve the obstruction may be required soon after birth. In female infants, the usual presence of a rectovaginal fistula in association with the imperforate anus prevents the development of distension; operative intervention may be postponed for several weeks.

**Anesthetic Management**

Anesthetic considerations for patients with intestinal obstruction include airway management, fluid and electrolyte replacement, treatment of sepsis, and postoperative pain management.

Complete intestinal obstruction increases the risk for aspiration of gastric or intestinal contents. Diaphragmatic excursion is impeded by abdominal distension and may cause respiratory distress. To prevent further abdominal distension in a patient with a suspected difficult airway, the patient should be intubated while awake; a rapid-sequence induction with cricoid pressure might be considered in a patient with no apparent airway problems.

Patients with intestinal obstruction may be volume depleted secondary to dyes used in diagnostic tests and to peritonitis, ileus, bowel manipulation, and sepsis. Volume resuscitation with at least 10 mL/kg per hour of normal saline or colloid may need to be given before anesthesia can be induced. The quality of heart tones, urine output, blood pressure, skin color, and turgor will aid in fluid management. In some cases invasive monitoring with arterial and central venous pressures are necessary.

The choice of anesthetic agents depends on the patient's condition. Nitrous oxide should be avoided to prevent further distension of the bowel. Regardless of the anesthetic drugs used, an adequate circulating blood volume must be maintained to ensure perfusion of vital organs.
SUMMARY

Improvements in the diagnosis and treatment of congenital disorders have resulted in a change in surgical practice. Many conditions that formerly required corrective surgery immediately after birth are no longer surgical emergencies. Most babies with congenital anomalies that can be corrected by surgery are now stabilized and optimized before the procedure. This article focused on the more common conditions that require semi-elective or urgent surgery in the neonatal period. Salient features of each of these disorders were described. Factors unique to each of these conditions that can affect the anesthetic course of these children were discussed. Methods and techniques that may aid in the anesthetic management of these children were delineated.

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