HOSPITAL PHYSICIAN®

GENERAL SURGERY BOARD REVIEW MANUAL

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Frequently Encountered Problems in Pediatric Surgery I: Neonatal Problems

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Cover Illustration by Christy Krames

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I. INTRODUCTION

Part I of this topic addresses surgical problems in neonates. After the review of some special considerations for treating newborns, several conditions (many of them congenital) that tend to present in newborns are discussed. Part II of this topic will address commonly seen surgical problems that occur in older infants and children.

II. SPECIAL CONSIDERATIONS FOR NEONATES

A. **Transporting a neonate for surgery**
   1. **Referrals.** Outside referrals are often transported by a team from the referring hospital. The surgical service should be consulted before patients with surgical problems are transported.
   2. **Instructions.** Proper instructions are important for the safety of the patient.

a. The patient's specific problem(s) should be defined, if possible.
b. Specific instructions regarding suctioning and positioning should be given to the referring physician and the transport team.
c. Appropriate records and radiographs must accompany the patient.
d. Anticipated time of hospital arrival should be estimated.
e. The chief surgical resident should be informed by the senior pediatric resident as soon as possible.
f. The intensive care unit should be informed of the patient's expected problems and needs.
g. All other physicians who will be contributing to the patient's care (eg, neonatologist, radiologist, anesthesiologist) should be informed of the patient's problems and the anticipated arrival time.
h. If the patient is likely to require urgent operative intervention, the operating room staff should be informed.

B. **Preoperative preparation for neonates**
   1. **General considerations**
      a. Blood setup
      b. Intravenous antibiotics (ampicillin and gentamicin)
      c. Parental consent for surgery and anesthesia
      d. Patients with possible cardiac anomalies need a electrocardiogram (ECG), chest radiograph, and echocardiogram.
      e. To prevent possible subsequent disastrous bleeding, newborns should be given 1 mg of vitamin K intramuscularly if they have not already received it. Vitamin K administration sometimes is overlooked during a difficult delivery of an infant with a congenital problem.
   2. **Newborns with metabolic complications**
      a. **Hypoglycemia.** Hypoglycemia is a particular risk in infants of diabetic mothers or infants who are small for their gestational age.
         1) Symptoms can include jitteriness, seizures, apathy, hypotonia, apnea, and hypothermia, but these infants can be asymptomatic.
         2) Glucose levels should be kept above 40 mg/100 mL. Prophylactically, 4 to 8 mg glucose/kg per minute should be administered (eg, 100 mL/kg per 24 hours of 10% aqueous dextrose solution). For acute hypoglycemia, an immediate push of 25% aqueous dextrose solution, 1 to 2 mL/kg, is required.
      b. **Hypocalcemia.** Hypocalcemia is likely in low-birth-weight or stressed infants and in infants of diabetic mothers.
         1) Symptoms can include jitteriness, convulsions, and other nonspecific symptoms.
         2) The critical level is that of ionized calcium, which depends on serum total protein. Infants with acute symptomatic hypocalcemia should be started on 10% calcium chloride at 20 mg/kg per dose (0.2 mL/kg per dose) intravenously, slowly. Stop administration when clinical response is obtained. Monitor ECG continuously. Follow with calcium infusion up to 50 to 60 mg/kg per 24 hours.

III. ESOPHAGEAL ATRESIA

A. **Definition.** There are five major types of esophageal atresia and tracheoesophageal fistula (**Figure 1**). The most common type is proximal esophageal atresia with concomitant tracheoesophageal fistula (Figure 1C).

B. **Embryology.** Esophageal atresia begins prenatally at 3 to 6 weeks after conception. During this time, the trachea and lungs are developing and separating from the foregut. Thirty percent of infants with this syndrome are premature.

C. **Clinical presentation**
   1. Clinical signs include:
      a. Excessive salivation
      b. Choking during feedings
   2. Patients with esophageal atresia may also present with:
      a. Recurrent aspiration pneumonia
      b. Right upper lobe pneumonia and atelectasis

D. **Associated anomalies.** Significant associated anomalies occur in 30% of patients with esophageal atresia.
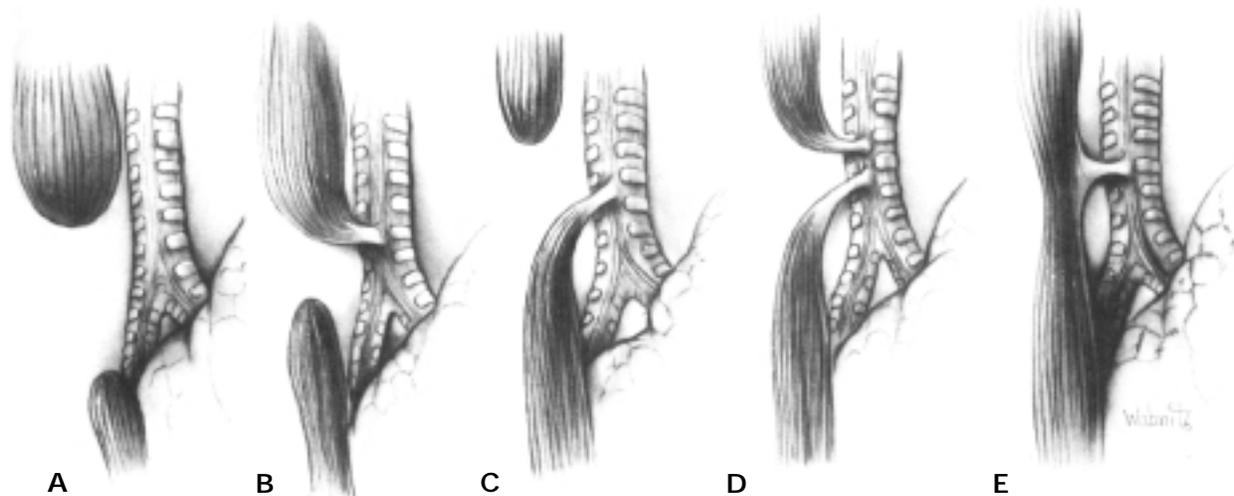   1. Esophageal atresia is frequently associated with

Figure 1. Five major varieties of esophageal atresia and tracheoesophageal fistula. (A) Esophageal atresia without associated fistula. (B) Esophageal atresia with tracheoesophageal fistula between proximal segment 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). Adapted with permission from Guzzetta PC, Anderson KD, Altman RP, et al: Pediatric Surgery. In *Principles of Surgery*, 6th ed. Schwartz SI, Shires GT, Spencer FC, eds. New York: McGraw-Hill, 1994:1691.

the VACTERRL (Vertebral, Anal, Cardiac, TracheoEsophageal, Renal, Radial, and Limb) pattern of anomalies.
2. Cardiac anomalies are the most serious and contribute significantly to the mortality rate.
E. **Diagnosis**
　1. Diagnosis is made by observing a coiling nasogastric tube in a proximal pouch on radiograph.
　2. Barium (0.5 mL) may be carefully instilled in the proximal pouch.
　3. Radiographs should include the abdomen.
　　a. If air is seen in the intestine, the diagnosis is esophageal atresia with distal tracheoesophageal fistula (Figure 1C).
F. **Treatment**
　1. **Preoperative management**
　　a. If present, right upper lobe pneumonia and atelectasis should be corrected with antibiotics before surgery.
　　b. The baby should be kept in a reverse Trendelenburg's position.
　2. **Operative management.** A gastrostomy should be performed as soon as possible.
　　a. Vigorous chest physical therapy and suctioning should be performed.
　　　1) The gastrostomy tube should be placed to gravity.

　　　2) Saliva should be suctioned from the blind proximal pouch either by continuous sump tube (Replogle) or by oral suctioning every 15 minutes.
　　b. An extrapleural division and closure of the tracheoesophageal fistula with end-to-end anastomosis without undue tension should be performed. In certain types of tracheoesophageal fistula with esophageal atresia or in premature infants, a delayed anastomosis may be considered.
　3. **Outcome.** Mortality should be close to zero in a full-term infant without associated major anomalies.

IV. HYPERTROPHIC PYLORIC STENOSIS

A. **Definition.** Pyloric stenosis is obstruction of the pyloric orifice of the stomach.
B. **Epidemiology.** Pyloric stenosis usually occurs in the first 3 to 6 weeks of life. It is extremely rare during the first week of life.
C. **Clinical presentation.** Nonbilious vomiting (becoming projectile), cannot hold down water, leading to severe dehydration (metabolic alkalosis, decreased potassium and chloride ions). Serum pH is increased.

D. **Diagnosis**
  1. **Palpation of the pyloric olive.** Contrary to the textbook description of its location in the right upper quadrant, the pyloric olive is more commonly found in the midline. If the pyloric olive can be felt, no further diagnostic tests are necessary.
      a. In an infant with a history that strongly suggests pyloric stenosis, emptying the stomach with a nasogastric tube to make the olive easier to feel is recommended.
      b. Palpating the olive is impossible if the infant is crying.
          1) Crying can be suppressed by giving the infant a pacifier or a small amount of an oral electrolyte maintenance solution.
          2) Patience on the part of the physician is important in this circumstance.
      c. When the infant is not crying, the physician should stand at the infant's left side and hold up the baby's feet with his or her left hand to relax the infant's belly. The physician should then gently palpate the epigastrium with the extended middle finger of the right hand, being careful not to dig into the baby's abdomen.
  2. **Barium study.** Typical findings on barium study indicating pyloric stenosis are the "string" sign and the "double tract" sign.
  3. **Ultrasonography** is the gold standard for diagnosis of pyloric stenosis.
      a. If the history is strongly suggestive of pyloric stenosis but a mass is not palpable, an ultrasound is a good diagnostic test in experienced hands.
      b. If pyloric stenosis is not the cause of vomiting, gastroesophageal reflux may also be diagnosed by ultrasound.
E. **Treatment** is surgical.
  1. **Preoperative management.** A clinical assessment of the patient's hydration should be made, and serum electrolyte levels should be checked immediately on admission to rule out a serious hypokalemic hypochloremic metabolic alkalosis. This should be corrected with appropriate potassium- and chloride-containing intravenous fluids before elective pyloromyotomy.
  2. **Pyloromyotomy** (**Figure 2**)
  3. **Postoperative management**
      a. **Feeding regimen**
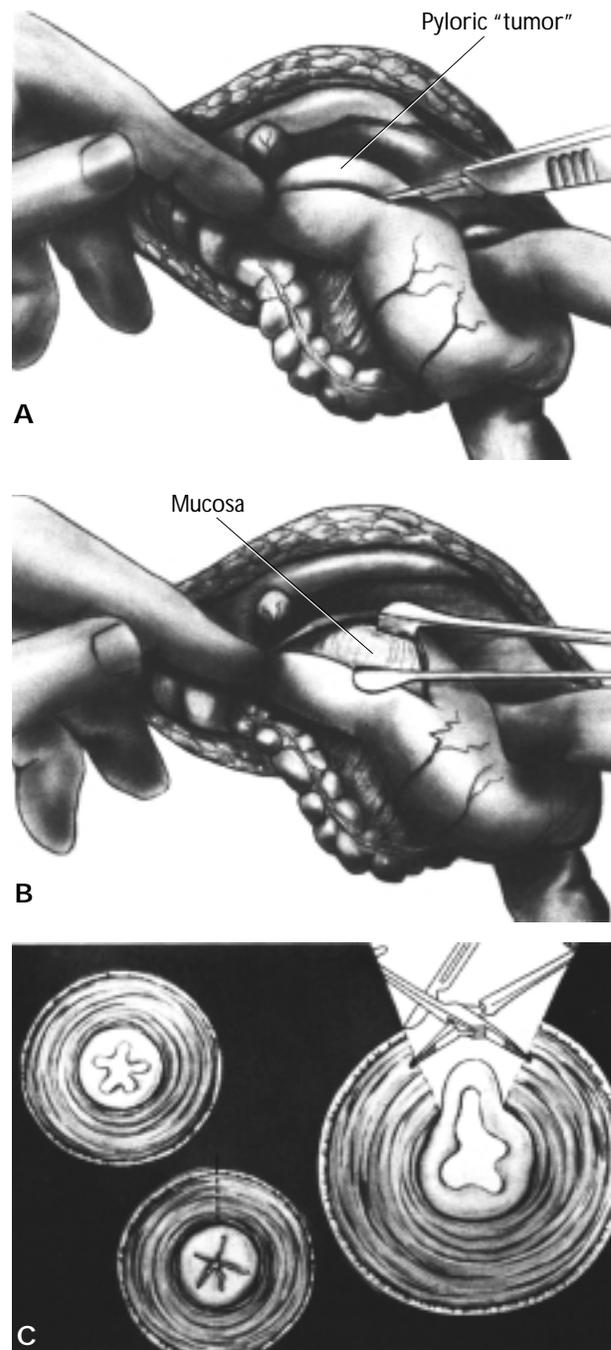
Figure 2. Fredet-Ramstedt pyloromyotomy. (A) Pylorus delivered into wound and seromuscular layer incised. (B) Seromuscular layer separated down to submucosal base to permit herniation of mucosa through pyloric incision. (C) Cross section demonstrating hypertrophied pylorus, depth of incision, and spreading of muscle to permit mucosa to herniate through incision. Adapted with permission from Guzzetta PC, Anderson KD, Altman RP, et al: Pediatric Surgery. In *Principles of Surgery*, 6th ed. Schwartz SI, Shires GT, Spencer FC, eds. New York: McGraw-Hill, 1994:1695.

Table 1. Causes of Intestinal Obstruction

Age	Causes
Newborn	Intestinal atresias and stenoses (including imperforate anus and pyloric atresia), anomalies of rotation and fixation (including midgut volvulus), Hirschsprung's disease, meconium ileus, pseudocyst, meconium plug, abscess or adhesions from peritonitis, peritoneal bands, segmental volvulus, incarcerated hernia (inguinal, internal, or diaphragmatic)
Infants to 24 months	Pyloric stenosis, incarcerated inguinal hernia, intussusception, Hirschsprung's disease, intestinal stenosis, congenital bands, duplications, cyst, omphalomesenteric duct remnant, internal hernia, midgut volvulus, trauma
24 months and older	Incarcerated inguinal hernia, appendicitis, adhesions from prior surgery, duplications, cysts, anomalies of rotation and fixation, trauma, granulomatous disease, tumors

1) The patient should be given nothing orally for 6 hours after surgery.
2) Feeding can usually be initiated 6 to 8 hours postoperatively. Sugar water is generally given first, followed by formula or breast milk, using the following guidelines. This regimen can be advanced more rapidly or slowly depending on how the baby does.
   a) **Sugar water,** 30 mL every 2 hours, two times
   b) **Formula or breast milk**
      i) If sugar water is tolerated, the baby may be given half-strength formula, 30 mL every 2 hours, two times. This is followed by full-strength formula every 4 hours at liberty.
      ii) Breast milk may be substituted for formula but must be measured and fed by bottle.
3) If the infant vomits, feedings should cease for 2 hours.
4) All routine procedures (eg, taking vital signs, diaper changing, sponge bathing) should be completed before each feeding begins.
   b. **Hospital discharge.** Most infants may be discharged 24 hours after surgery.

c. **Surgical complications.** If the duodenum is inadvertently entered during the pyloromyotomy, the infant should remain on both nasogastric suction and intravenous antibiotics postoperatively for a minimum of 2 days.

V. CONGENITAL INTESTINAL OBSTRUCTION

A. **Causes. Table 1** lists the causes of intestinal obstruction in various age groups.
B. **Intestinal atresias**
   1. **Duodenal atresia**
      a. **Etiology.** The condition is probably the result of canalization failure.
      b. **Incidence.** Duodenal atresia is the most frequent type of intestinal obstruction, followed by jejunal atresia and then ileal atresia.
      c. **Associated conditions.** High incidences of low birth weight (50%), Down syndrome (30%), and other major anomalies (30% to 50%) are associated with duodenal atresia. Most duodenal atresias are distal to the ampulla of Vater.
      d. **Clinical presentation.** The patient usually presents with bilious vomiting shortly after birth.
      e. **Diagnosis.** Radiographs show a stomach and duodenum with a gasless abdomen ("double bubble"). If a delay in surgery is anticipated it is imperative to differentiate atresia from midgut volvulus, which requires immediate intervention.
      f. **Management.** Management is by duodenojejunostomy, duodenoduodenostomy, and, occasionally, gastrostomy.
   2. **Small bowel atresia**
      a. **Etiology.** Small bowel atresia is almost certainly the result of vascular occlusion (ie, intrauterine volvulus, intussusception) with aseptic necrosis and resorption of the gangrenous segment.
      b. **Associated conditions.** A high incidence of low birth weight (40%) and low incidence of other anomalies are noted. Small bowel atresia is associated with meconium ileus.
      c. **Clinical presentation.** Patients present with bilious vomiting and abdominal distention.

d. **Diagnosis.** Radiographs show many dilated loops with air-fluid levels.

e. **Differential diagnosis** (*see* Table 1)

f. **Management**
   1) Dehydration, along with acid-base and electrolyte imbalances, should be corrected.
   2) Laparotomy may be performed with resection of the proximal dilated end. End-to-end anastomosis usually is possible.

3. **Colon atresia**
   a. **Incidence.** Rare
   b. **Management.** Colostomy at the point of atresia

C. **Hirschsprung's disease** (congenital aganglionic megacolon)
1. **Incidence.** Hirschsprung's disease is a frequent cause of neonatal intestinal obstruction. It may also present during the first few years of life.
2. **Etiology.** The most common form is the absence of ganglion cells in the lower rectum.
   a. This leads to ineffective conduction of peristalsis, resulting in a functional obstruction.
   b. The aganglionic segment may extend more proximally and can involve the entire colon.
3. **Clinical presentation.** Symptoms are nonspecific and may include episodic abdominal distention, diarrhea, and obstipation (which is not ordinarily seen in neonates) or constipation.
4. **Diagnosis**
   a. **Radiologic examination**
      1) A barium enema shows a narrow rectum with a dilated colon proximally. However, this finding is often absent in infants.
      2) If the barium enema is normal and there is a high suspicion for Hirschsprung's disease, a plain radiograph of the abdomen should be obtained on the following day. Retained barium in the colon on this follow-up film is highly suggestive of Hirschsprung's disease.
   b. **Biopsy.** The diagnosis is confirmed by rectal biopsy (suction mucosal or full-thickness) showing an absence of ganglion cells in the submucosal plexus and hypertrophied nerve endings.

5. **Management.** Hirschsprung's disease may be initially managed with a temporary colostomy above the aganglionic segment. Recently, pediatric surgeons have performed primary pull-through procedures in the neonate.

D. **Meconium ileus**
1. **Incidence.** Meconium ileus accounts for almost one third of obstructions of the small intestine in neonates.
2. **Epidemiology.** The disorder occurs in approximately 15% of infants with cystic fibrosis.
3. **Clinical presentation**
   a. The diagnosis should be suspected in an infant who develops generalized abdominal distention, bilious vomiting, and failure to pass meconium in the first 24 to 48 hours after birth.
   b. A family history of cystic fibrosis is not uncommon; a maternal history of polyhydramnios is present in 20% of patients.
   c. The meconium may be palpable as a doughy substance in the dilated loops of distended bowel. The anus and rectum are typically narrow.
4. **Imaging studies**
   a. Plain abdominal radiograph demonstrates bowel loops of variable size.
      1) Bowel contents have a soap-bubble appearance.
      2) Calcifications usually indicate meconium peritonitis resulting from an intrauterine intestinal perforation.
   b. A barium enema demonstrates a microcolon with inspissated meconium proximally.
5. **Management**
   a. **Initial treatment.** Initial treatment is meglumine diatrizoate enemas and no surgery.
      1) The patient should be intravenously hydrated.
      2) Under fluoroscopic control, a 50% solution of meglumine diatrizoate and water should be infused into the rectum and colon through a catheter.
      3) This procedure usually results in a rapid passage of semiliquid meconium that continues during the next 24 to 48 hours.
      4) Multiple enemas may be required.

    b. **Surgical treatment.** Surgery is indicated if:
      1) The meglumine diatrizoate enemas do not relieve the obstruction.
      2) The infant appears too ill to delay operation.
      3) The diagnosis of meconium ileus is uncertain.
    c. **Postsurgical management**
      1) All infants diagnosed with meconium ileus require an iontophoresis test to confirm a diagnosis of cystic fibrosis. This test is usually not practical before operation.
      2) All infants require vigorous postoperative pulmonary therapy.
      3) When oral feedings are begun, a pancreatic enzyme preparation is given with each feeding.

E. **Malrotation**
  1. **Etiology.** The infant has compression of the second portion of the duodenum by Ladd's bands, which can potentially cause obstruction.
  2. **Incidence.** Malrotation is a very common cause of intestinal obstruction in infants.
  3. **Clinical presentation**
    a. Sudden onset of bilious emesis is the primary presenting sign; malrotation must be considered in every infant with bilious emesis.
    b. Abdominal distention is common but may be absent.
    c. Abdominal tenderness varies.
    d. On rectal examination, stool, if present, is guaiac positive.
  4. **Diagnosis**
    a. Midgut volvulus is one of the most serious emergencies seen in these neonates or infants, and delay in diagnosis can result in loss of the entire midgut.
    b. Plain films of the abdomen are variable; a definitive diagnosis requires a contrast study.
      1) An upper gastrointestinal test is the preferred study and should be done in most cases.
      2) Occasionally, a barium enema is also helpful.
      3) These studies should be performed expeditiously because a few hours may be the difference between a totally reversible condition and loss of the entire midgut.

      4) Contrast studies may be dispensed with in cases of shock or clear indication for exploration.

VI. OMPHALOCELE AND GASTROSCHISIS

A. **Definition.** Omphalocele and gastroschisis are abdominal wall defects.
B. **Associated anomalies.** Associated anomalies should be ruled out, particularly in neonates with an omphalocele. The VACTERRL constellation is often found in patients with omphalocele.
C. **Management.** Treatment begins immediately following delivery.
  1. **Medical treatment**
    a. Hypothermia is usually the immediate life-threatening problem.
    b. Systemic intravenous antibiotics (ampicillin/gentamicin) are given to protect contaminated amnion and viscera. Infection can be devastating if a mesh closure is necessary.
    c. Intravenous hydration with balanced salt solution and colloid is essential.
  2. **Surgical treatment**
    a. The sac or exposed intestines should be covered by a barrier-type dressing. A large circumferential dressing is applied last.
    b. With gastroschisis in particular, it is essential that the bowel be supported, usually with the patient on his or her side and the bowel supported by towels, to prevent strangulation of the bowel and consequent bowel ischemia.
    c. Gastrointestinal decompression by nasogastric tube is imperative to minimize further gastrointestinal distention and prevent aspiration of gastric contents.

VII. HERNIAS

A. **Incarcerated inguinal hernia**
  1. **Definition.** Incarcerated inguinal hernia in a child is a patent processus vaginalis with an intra-abdominal organ within it. This condition is age related, occurring most often in infants during the first year of life.
    a. Incarcerated inguinal hernia in boys often contains bowel.

b. Incarcerated inguinal hernia in girls often contains ovary and tube.
2. **Etiology** is not known.
3. **Clinical presentation.** Both boys and girls invariably are noted to have a lump in the groin.
4. **Differential diagnosis**
   a. **In boys,** differentiating an incarcerated hernia from a hydrocele of the cord is imperative.
      1) **External examination**
         a) A hydrocele of the cord is often tense.
         b) The end of the hydrocele can be distinguished from the testis itself.
         c) The proximal end of the cord can be detected.
      2) **Rectal examination** can also distinguish the two conditions.
         a) The inside of the abdominal wall at the level of the internal ring should be palpated.
         b) If the vas and the ring are easily palpable, an incarceration cannot be present.
         c) If the physician is still unsure of the diagnosis, palpation on the other side can be compared.
   b. **In girls,** differential diagnosis is hydrocele of the canal of Nuck.
5. **Treatment**
   a. **Treatment of boys**
      1) **Manual reduction.** Most incarcerated inguinal hernias can be reduced manually, obviating emergency surgery. Some hernias reduce easily; others require several attempts.
         a) If necessary, the patient should be sedated with an appropriate barbiturate (eg, pentobarbital sodium 2 to 3 mg/kg body weight, morphine 0.1 mg/kg).
         b) Occasionally, simply holding the patient in a very steep Trendelenburg's position reduces the hernia because of the pull of the mesentery.
         c) If the hernia does not spontaneously reduce with the patient in Trendelenburg's position, an assistant should hold the infant

above the knees in a frog-leg position to relax the abdominal wall.
         d) The physician should use the fingers of one hand to attempt to fix the hernia while gradually pressing the incarcerated mass with the other hand.
            i) The physician should try to milk the bowel contents out of the incarcerated bowel until it "pops" back within the abdomen.
            ii) A considerable length (ie, 5 minutes) of steady pressure may be required to produce the desired reduction, so the physician should be in a comfortable position.
      2) **Postreduction management**
         a) If manual reduction of the hernia is successful, the patient is always admitted to the hospital, and the repair is performed electively within 24 to 48 hours after the inguinal edema has resolved.
         b) High ligation of the sac is the operation performed in both boys and girls.
      3) **Surgical management.** Emergency surgical intervention is required if the hernia cannot be manually reduced or if there is postreduction evidence of persistent intestinal obstruction or nonviable bowel.
   b. **Treatment of girls.** Because the blood supply to the ovary is usually not impaired, these hernias can generally be repaired on a semielective basis. As with boys, treatment consists of manual reduction followed by high ligation of the sac.
6. **Complications.** Hemorrhagic infarction of the testicle is an unfortunate complication of an incarcerated hernia. Reduction will usually reinstitute blood flow to the testis. Incarceration of an ovary in a girl generally has no sequelae.
7. **Incarcerated inguinal hernia in a premature infant**
   a. An inpatient premature infant with multiple problems can have a hernia repaired just before discharge home.

b. An infant with a history of prematurity (gestational age less than 36 weeks at birth) should be admitted overnight after hernia repair for apnea monitoring. This pertains until the child reaches a post-conceptual age of at least 50 weeks.

B. **Diaphragmatic hernia**
  1. **Definition.** Diaphragmatic hernia is a failure of diaphragmatic development.
  2. **Embryology.** Formation of the diaphragm occurs at 8 to 10 weeks in the fetus. During this time, the intestines returning into the abdomen will enter the chest if the diaphragm is not formed (ie, persistent pleuroperitoneal canal) and will prevent normal lung development. Intestines are also malrotated and subject to possible midgut volvulus postoperatively.
  3. **Nature of defects**
      a. There is a left-side to right-side predominance of 4:1.
      b. Most defects are posterolateral (Bochdalek's hernia).
      c. Anteromedial defects (Morgagni's hernia) are rare.
      d. Pulmonary hypoplasia occurs on the ipsilateral side and also commonly occurs on the contralateral side.
  4. **Clinical presentation**
      a. The patient presents with respiratory distress. If extreme hypoplasia is present, respiratory distress is present early after delivery.
      b. Most patients present with respiratory acidosis and metabolic acidosis secondary to hypoxia and hypothermia.
  5. **Diagnosis**
      a. **Physical examination.** On examination, the patient's abdomen is scaphoid.
      b. **Radiography.** Radiographs reveal a bubbly bowel pattern in the chest and a lack of normal intestinal gas.
  6. **Treatment**
      a. **Preoperative management**
          1) Planned resuscitation
              a) The patient should not be ventilated by mask.
              b) The patient should be kept on oxygen and prepared for immediate orotracheal intubation.
          2) The patient should be kept warm to prevent hypothermia. Warming lamps should be used.

3) An orogastric tube should be placed.
4) Determine pH and blood gas, type, and crossmatch. Cutdown should be performed as needed.
5) Intravenous bicarbonate (1 to 2 mEq/kg) is almost always needed.
6) Extracorporeal membrane oxygenation may be necessary if blood gas values cannot be restored to near normal.

  b. **Operative management**
      1) The bowel is reduced, a chest tube is inserted, the defect is repaired, and a gastrostomy is performed.
      2) The small hypoplastic lung is not expanded.
  c. **Postoperative management**
      1) Ventilatory support at the lowest possible pressure is usually needed but should be discontinued as soon as possible.
      2) The chest tube should be kept at underwater seal and at 2 cm H_2O suction.
      3) The chest tube can usually be removed by the fifth postsurgical day if the patient is off ventilatory support.
      4) pH and blood gas levels should be determined frequently.
  7. **Mortality**
      a. When symptoms of diaphragmatic hernia present early (ie, less than 12 hours after birth), the mortality rate is 50%.
      b. When symptom onset is within 1 to 2 hours after birth, the mortality rate is 90%.

VIII. NECROTIZING ENTEROCOLITIS

A. **Definition.** Necrotizing enterocolitis is a highly lethal disease in newborns characterized by ischemic necrosis of the gastrointestinal tract that frequently leads to perforation. The most commonly involved sites are the distal ileum and colon.
B. **Epidemiology**
  1. Usual onset is within the first 5 days of life but may occur in babies up to 3 weeks of age.
  2. Primarily occurs in low-birth-weight infants, especially those with perinatal complications (**Table 2**).

C. **Etiology**
1. The disease is thought to evoke a primitive reflex whereby blood is shunted away from gastrointestinal tract to the heart and brain.
2. This shunting leads to mucosal ischemia, decreased mucous production, ulceration, and bacterial invasion.

D. **Clinical presentation.** Almost all patients have been fed. There may be very few signs. Usually, mild to increasing ileus is present, manifesting as distention.
1. Possible presenting signs and symptoms include:
   a. Bile-aspirates
   b. Blood in stool
   c. Abdominal distention
   d. Poor feeding
   e. Apneic episodes
   f. Jaundice
2. Later in the course of the disease, signs of peritonitis occur.

E. **Diagnosis**
1. **Radiographs** show:
   a. Dilated loops of bowel
   b. Intramural streaks or bubbles of gas (pneumatosis)
   c. Portal vein gas
2. In the absence of signs of perforation requiring surgery (or autopsy), the diagnosis must be made by the presence of pneumatosis.

F. **Complications**
1. Perforation may occur with sudden clinical deterioration. A radiograph shows signs of pneumoperitoneum, extraluminal bubbles of gas, or ascites.
2. Gangrenous bowel is often heralded by a sudden drop in serum pH and sodium and platelets to fewer than 100,000/mm³.
3. Obstruction is secondary to perforation and abscess formation, leading to cicatricial stenosis 1 to 3 weeks following onset.

G. **Management**
1. **Indications for surgery** include the following:
   a. Pneumoperitoneum
   b. Failure of medical management
2. Surgery for necrotizing enterocolitis must be tailored to findings in the operating room.
3. For stenosis, an end-to-end anastomosis or gastrostomy can usually be performed.

H. **Mortality.** In the past 5 years, the mortality rate has dramatically decreased (from 80% to 20%),

Table 2. Perinatal Complications Predisposing to Necrotizing Enterocolitis

Complications of respiratory distress syndrome

Apneic spells

Cyanosis

Hypothermia

Low Apgar score

Resuscitation in the delivery room

Umbilical vessel catheterization

Exchange transfusions

Premature rupture of membranes

Breech delivery

Delivery by cesarean section

Amnionitis during pregnancy

owing largely to early recognition and prompt treatment.

BOARD REVIEW QUESTIONS

Choose the single best answer for each question.

1. **In Hirschsprung's disease, rectal biopsy typically shows which of the following patterns?**
   A) Absence of ganglion cells and nerve fibers
   B) Absence of ganglion cells and presence of normal nerve fibers
   C) Absence of ganglion cells and presence of hypertrophied nerve fibers
   D) Presence of ganglion cells and absence of normal nerve fibers
   E) Presence of hypertrophied ganglion cells and normal nerve fibers

2. **Which of the following statements concerning congenital esophageal atresia is FALSE?**
   A) The lesion should be immediately suspected when a newborn infant aspirates the first feeding.
   B) In the most common form of the anomaly, air is absent from the gastrointestinal tract.
   C) More than 10% of affected infants have an imperforate anus.
   D) In affected infants who have pneumonia or cardiac difficulties, gastrostomy under local anesthesia is the initial surgery of choice.
   E) Stenosis at the site of anastomosis is the most common late complication.

3. **A 19-day-old, full-term, previously healthy infant develops sudden onset of bilious vomiting at home. On examination in the emergency department, the infant appears ill. His abdomen is mildly tender but not distended, and he passes blood in his stool. The most likely diagnosis is:**
   A) Pyloric stenosis
   B) Gastroenteritis
   C) Malrotation with midgut volvulus
   D) Necrotizing enterocolitis
   E) Jejunal atresia

4. **A 26-day-old boy presents with a history of nonbilious vomiting. The child has lost 500 grams over the past week and appears clinically dehydrated. A palpable olive-sized mass is present in the midepigastrium. Laboratory data reveal serum sodium, 131 mEq/L; serum potassium, 2.8 mEq/L; serum chloride, 82 mEq/L; serum bicarbonate, 42 mEq/L; and a pH of 7.5. The most likely diagnosis is:**
   A) Malrotation with a midgut volvulus
   B) Pyloric atresia
   C) Esophageal duplication
   D) Hypertrophic pyloric stenosis
   E) Appendicitis

5. **The most appropriate maintenance fluid for a 9-kg infant is:**
   A) 5% dextrose in 0.2 normal saline + 30 mEq/L of KCl at 36 mL per hour
   B) 5% dextrose in 0.5 normal saline + 30 mEq/L of KCl at 36 mL per hour
   C) Isolyte at 20 mL per hour
   D) 5% dextrose in 0.2 normal saline + 20 mEq/L of KCl at 45 mL per hour
   E) 5% dextrose in 0.5 normal saline + 20 mEq/L of KCl at 45 mL per hour

ANSWERS

5. A
4. D
3. C
2. B
1. C

SUGGESTED READINGS

Benson CO: Infantile Hypertrophic Pyloric Stenosis. In *Pediatric Surgery,* 4th ed. Welch KJ, et al, eds. Chicago: Year Book, 1986.

Ein SH, Stephens CA: Intussusception: 354 cases in 10 years. *J Pediatr Surg* 1971;6:16–27.

Grosfeld JK, Ballantine TV, Shoemaker R: Operative management of intestinal atresia and stenosis based on pathologic findings. *J Pediatr Surg* 1979;14:368–375.

Leape LL: *Patient Care in Pediatric Surgery.* Boston: Little, Brown, 1987.

Ravitch MM: The story of pyloric stenosis. *Surgery* 1960;48:1117.

Williamson RC: Death in the scrotum: testicular torsion [Editorial]. *N Engl J Med* 1977;296:338.

Wilmore DW: Factors correlating with a successful outcome following extensive intestinal resection in newborn infants. *J Pediatr* 1972;80:88–95.