Intestinal Obstruction in the Newborn

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FOCUS QUESTIONS

1. What is the significance of bilious vomiting in infancy and childhood?
2. What are the distinguishing clinical signs and symptoms of congenital upper small bowel obstruction versus those of congenital large bowel obstruction?
3. What are the common and the uncommon presenting symptoms of malrotation?
4. Explain the pathophysiology of complicating volvulus, the basis for urgency in diagnosis, and the appropriate studies needed for diagnosis.
5. What is the significance of the radiographic finding of pneumatosis intestinalis?
6. What are major causes of partial or complete large bowl obstruction occurring in the neonatal period?

Obstruction of an infant's gastrointestinal (GI) tract can occur anywhere from the esophagus to the anus. For purposes of this review, the newborn infant will be defined as an infant from birth to 30 days of age. Both congenital and acquired obstructions will be addressed. In each instance, the epidemiology, pathogenesis, clinical aspects, and management of the disorder will be considered.

Esophageal Atresia

EPIDEMIOLOGY AND PATHOGENESIS

Esophageal atresia, or interruption of the esophagus, generally occurs in association with a tracheoesophageal fistula (EA – TEF). The most common anatomic arrangement is a blind proximal esophageal pouch that has a distal tracheoesophageal fistula (Figure 1). This is seen in 85% to 90% of infants who have this anomaly. Seen less commonly is pure esophageal atresia that does not have a tracheoesophageal fistula and tracheoesophageal fistula that does not have an esophageal atresia (H-type tracheoesophageal fistula). These latter two conditions occur in approximately 10% of newborns who have these types of anomalies. Other anatomic arrangements, such as an esophageal atresia that has a fistula between the upper pouch and trachea or esophageal atresia that has a fistula to both pouches, are seen in only a tiny fraction of these infants.

EA – TEF occurs in approximately 1 in 4000 live births. There have been numerous reports of siblings who have EA – TEF as well as reports of the anomaly in identical twins. Also reported, however, are many instances of identical twins in which one has the anomaly and the other has been spared. It is well recognized that esophageal atresia is a frequent component of the VATER association as well as other malformations, suggesting that the anomaly also might result from a specific teratogen in the developing fetus.

It commonly is believed that interruption of the events responsible for the elongation and separation of the esophageal and tracheal tubes during the fourth week of development leads to the development of this anomaly.

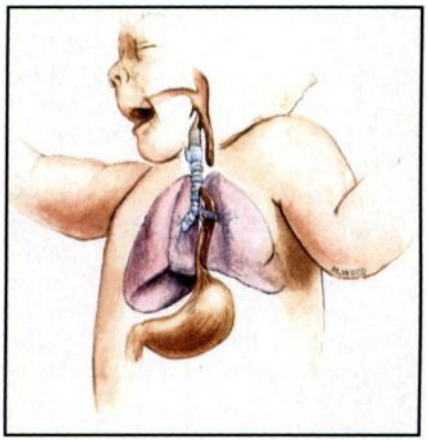

FIGURE 1. The most common type of esophageal atresia/tracheoesophageal fistula. Note the blind proximal esophageal pouch and the distal tracheoesophageal fistula.

PRESENTATION

Infants who have EA – TEF commonly will present in the nursery having an excessive amount of saliva. The saliva pools in the blind proximal esophageal pouch and is either regurgitated or continuously dribbled from the infant's mouth. The infant's first feeding will not be tolerated; the formula, which is, of course, not bile-stained, will be regurgitated immediately. Because these infants most commonly have a distal tracheoesophageal fistula, air enters their GI tracts through the fistula, and the abdomen will not be scaphoid. Indeed, the infant who is ventilated may well have air forced into his or her GI tract via the fistula, causing distension. Such an infant's respiratory difficulty, thus, may be compounded by the gastric distention leading to diaphragmatic elevation. In pure esophageal atresia that has no fistula, no air will enter the GI tract, and the infant will have a scaphoid abdomen.

DIAGNOSIS

The diagnosis of esophageal atresia can be made by attempting to pass a firm catheter through the mouth and into the esophagus. Obstruction to passage of the catheter, which should not be "forced," suggests the anomaly, and a chest radiograph usually will confirm the diagnosis via the presence of the coiled catheter sitting within the proximal esophageal pouch. Air injected into the catheter provides an excellent "contrast" agent to help confirm the diagnosis. The use of a true contrast agent generally is not recommended because the infant will be at risk of aspirating the agent and acquiring a chemical pneumonitis. In the rare instance in which contrast is required, 1 mL or less of a water-soluble agent can be injected into the pouch and then withdrawn immediately once the diagnosis has been confirmed. Air in the GI tract confirms the presence of the distal tracheoesophageal fistula.

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MANAGEMENT

Appropriate management of these babies begins at the time of diagnosis. A significant risk to the infants is the potential for gastric juice to pass upward in the distal esophagus and traverse the tracheoesophageal fistula where it may be aspirated, resulting in the development of chemical pneumonitis. These infants need to have a sump catheter placed immediately into the upper pouch and into the head up position at an angle of at least 45 degrees. This will help minimize the aspiration of saliva and the chance of gastric juice soiling the lungs.

As a general rule, all newborns whose GI tracts are obstructed should have intravenous fluids instituted and antibiotics begun. If the neonate is not at a surgical center, transport needs to be arranged as soon as possible.

In infants who have EA – TEF, immediate primary repair generally is undertaken in those weighing as little as 1200 g. An infant presenting with significant pneumonia or other major congenital anomalies will require a more individualized approach; a staged repair via an initial gastrostomy may be performed in an infant who has EA – TEF and is ill. The infant can be allowed to improve or to be evaluated for other anomalies prior to performing definitive repair.

Infants who have pure esophageal atresia generally are unable to have a primary repair performed in the newborn period because the distance between the two ends of the esophagus is too great. These infants require a gastrostomy and either exteriorization of the esophagus with a later esophageal substitution procedure (reverse gastric tube, colon interposition) or serial attempts at dilatation of the two ends of the esophagus with a later attempt at a primary anastomosis. Although conceptually more attractive, the latter option requires a prolonged initial hospitalization and has an attendant ongoing risk of aspiration.

The prognosis for most babies is excellent; only a few sick infants who have serious coexisting anomalies, are of extreme low birth weight, and have persistent pulmonary disease have a diminished chance for survival. It is not at all uncommon for the infant to develop a relative narrowing or stricturing at the anastomotic site, which does not become evident until the child is advanced to solid foods. Generally, this problem is managed easily with esophageal dilatation. The infants also may have problems with esophageal motility that mimics a stricture. This is determined easily by a contrast swallow radiograph.

Pyloric Atresia/Stenosis

EPIDEMIOLOGY AND PATHOGENESIS

Whereas congenital obstructions of the gastric outlet are uncommon, infantile hypertrophic pyloric stenosis, an acquired condition, is one of the most commonly encountered surgical problems of infants. Studies have found an incidence of pyloric stenosis in about 1 of every 500 live births, and it is well recognized that male infants are affected more often than females. Pyloric stenosis is seen more often in caucasian children, and interestingly, many authors have noted that the first-born infant is affected most often. Hereditary and familial predisposition also exist in this condition, with as many as 7% of infants of affected parents also having pyloric stenosis. The chances an infant will have pyloric stenosis are greater if the mother is the parent who had the condition.

Actual atresia of the pyloric or antral areas is relatively uncommon, whereas membranes or diaphragms in this region, although also rare, are seen with more frequency. The antral/pyloric membranes and diaphragms are thought to result from some endodermal redundancy during development. Pyloric stenosis, however, results from hypertrophy of the muscularis of the pyloric channel, especially the circular muscle. Because pyloric stenosis has not been known to occur earlier than the fourth or fifth day of life and usually develops at 3 or 4 weeks of age, it is not a true congenital defect. Its pathogenesis remains unexplained.

PRESENTATION

Infants who have an antral/pyloric diaphragm generally present with complete gastric obstruction, even if the diaphragm is perforate. These infants often are thought mistakenly to have esophageal atresia because they salivate excessively and frequently have respiratory problems due to gastric distention. The abdomen, however, is scaphoid, and the emesis rarely is bilious. This problem generally is manifested within the first day of life. In contrast to the early presentation of infants having an antral/pyloric diaphragm, children who have hypertrophic pyloric stenosis have the onset of symptoms, on the average, at 3 weeks of age.

Babies who have pyloric stenosis feed eagerly but lose weight and may become dehydrated and then develop hypochloremic-hypokalemic metabolic alkalosis.

The initial presenting symptom of pyloric stenosis is vomiting; the vomitus is not bile-stained. Although occasionally mistaken for gastroesophageal reflux, usually one is able to elicit a history of absolutely no reflux-type symptoms until the first episode of emesis, which then becomes more frequent. As the obstruction increases, the vomiting becomes more constant and "projectile." It occasionally can have a coffee ground appearance or blood streaking within it. The babies continue to feed eagerly but lose weight and may become dehydrated. Should the problem continue, they develop a hypochloremic-hypokalemic metabolic alkalosis.

The abdominal examination of those who have pyloric stenosis almost always is remarkable for the presence of a firm, small, movable mass that sits within the right upper

quadrant. Gastric decompression with a nasogastric tube (NG) tube may make this physical finding more easily demonstrable. Often times, visible waves of gastric peristalsis can be seen, and for reasons that are not clearly understood, jaundice is associated in a small percentage of the infants. This jaundice resolves following an operation.

DIAGNOSIS

The pyloric mass or "olive" virtually is pathognomonic, and no further studies need be performed if it is a definite, reproducible finding made by an experienced examiner. Should there be any doubt as to the diagnosis, abdominal ultrasonography and/or an upper GI series can confirm the diagnosis.

MANAGEMENT

Once diagnosed, these infants are admitted to the hospital and given intravenous fluid rehydration. The metabolic alkalosis, due to chloride loss in the vomitus and renal "compensation," must be corrected. The babies do not require emergency surgery and can be returned to normal metabolic balance before pyloromyotomy. These infants generally recover rapidly and then live a complete and normal life. They occasionally will have some "reflex" vomiting for several days following the operation, but this will subside rapidly. Similarly, infants who have pyloric/antral webs are expected to recover rapidly and do well following surgery.

Duodenal Obstruction

Obstructions of the duodenum are caused by congenital anomalies, including atresia/stenosis, annular pancreas, and a preduodenal portal vein.

EPIDEMIOLOGY AND PATHOGENESIS

Most obstructions of the duodenum are congenital anomalies, including atresia/stenosis, annular pancreas, and a preduodenal portal vein.

Malrotation also can cause obstruction of the duodenum, which is addressed later in this review.

Congenital duodenal obstruction is seen in approximately 1 of every 10 000 births. There frequently is a history of polyhydramnios, and these infants often are either preterm or small for gestational age. There is a significant coincidence of other anomalies, including a 30% incidence of Down syndrome. Children whose duodenal obstruction is a result of an annular pancreas have as much as a 70% incidence of other anomalies.

The most common causes of duodenal obstruction in the newborn are duodenal atresia and stenosis. Whereas atresia produces symptoms shortly following birth, duodenal stenosis may take several weeks before its presence becomes obvious. Duodenal mucosal proliferation begins about the fourth week of gestation, and the cellular proliferation is so abundant that by 5 to 6 weeks of gestation, the duodenum occludes completely. If the lumen does not reform, an atresia of the duodenum will result. These atresias commonly occur in the second or third portion of the duodenum, with the site of the atresia located just distal to the opening of the bile duct. If the recanalization is incomplete, a duodenal stenosis may result. Obstruction in this area may present as a partial or complete web.

Annular pancreas, another common obstruction, results from an aberration in the rotation of the ventral anlage of the pancreas, which normally fuses with the dorsal anlage. The result is a 270-degree (or greater) "ring" of pancreas around the duodenum. The duodenum also may be obstructed by a preduodenal portal vein that is a result of an abnormal development of vitelline vein anastomoses. Usually this phenomenon is found in association with other serious malformations of a rotational variety. A preduodenal portal vein often is encountered in situs anomalies, where it may be associated with duodenal obstruction.

DIAGNOSIS AND PRESENTATION

A duodenal obstruction may be diagnosed by prenatal ultrasonography, which will demonstrate a dilated stomach and proximal duodenum as a large, fluid-filled, cystic mass.

Because absorption of the amniotic fluid by the fetus is impeded if the duodenum is obstructed, many of these fetuses will have polyhydramnios. The typical presentation of duodenal obstruction is an infant who has bilious vomiting shortly after birth. Nonbilious emesis, however, does not rule out the diagnosis of duodenal obstruction. A flat and erect (or lateral decubitus) abdominal radiograph is sufficient to diagnose the duodenal obstruction because the typical "double-bubble sign" of duodenal atresia is diagnostic. Infants who have complete duodenal obstruction generally have a scaphoid abdomen due to the absence of gas in the intestine. It is possible, however, that the abdomen will appear distended initially due to the greatly dilated stomach. Passage of an NG tube should decompress the dilated stomach and leave the infant with a scaphoid abdomen.

Should the child have a duodenal stenosis that is not high grade, the diagnosis may not be made during infancy. These children occasionally can present with long-term feeding problems and not have significant symptoms until later in childhood. Although the double-bubble usually is pathognomonic of duodenal obstruction, occasionally one is not seen on a preliminary radiograph. We find that instilling 50 mL of air often will provide a sufficient amount of "contrast" to establish the diagnosis. Should there be any question, a radiopaque agent contrast study can be performed. We believe that there is little role for a contrast study, though, unless one sees the suggestion of distal air within the GI tract. Although this finding could be due to a duodenal stenosis, the possibility exists that malrotation is present, and this diagnosis must be established (or

ruled out) immediately because the potential problem of midgut volvulus is life-threatening.

MANAGEMENT

If the infant whose duodenum is obstructed has no significant respiratory or metabolic problems or other life-threatening anomalies, he or she is a candidate for immediate operative correction. On the other hand, in the presence of any of these complicating factors, the infant should undergo appropriate evaluation, stabilization, or both before transport to the operating room. The major exception to this rule is the infant in whom the evaluation determines that the duodenal obstruction is due to malrotation; this represents an emergency because of the potential for volvulus, and no undue delays in transport to the operating room are acceptable.

Generally, either duodenoduodenostomy or duodenojejunostomy is the procedure of choice for duodenal atresia/stenosis and annular pancreas. Children whose duodenums are obstructed often are "slow" to open up and tolerate full enteral feeds. This is why some place gastrostomy tubes at the time of operation, especially in small, sick infants who tend to take longer to progress to full "feeds." Otherwise, their prognosis generally is excellent unless they have associated congenital anomalies of a limiting nature.

Malrotation

EPIDEMIOLOGY AND PATHOGENESIS

Anomalies of intestinal rotation may be life-threatening or discovered incidental to other conditions. These anomalies result from disruption of the normal rotational process of the midgut during its return from its extracoelomic phase of development to the abdominal cavity during the fourth to tenth week of embryogenesis (Figure 2). Infants who have malrotation often will have incomplete duodenal obstruction created by peritoneal (Ladd) bands compressing the duodenum. Because the mesentery is not broadly fixed and attached, these infants are at risk for developing a midgut volvulus.

Because there are so many types of rotational anomalies, it is difficult to know their exact frequency in live births. However, the majority of patients who develop midgut volvulus as a result of malrotation do so within the first year of life; indeed, 50% of such patients present within the first month of life. Incomplete rotation is seen in association with congenital diaphragmatic hernia as well as abdominal wall anomalies such as omphalocele and gastroschisis. Malrotation also has been noted "incidentally" in many children being treated for Hirschsprung disease and intussusception. There does not seem to be a hereditary/familial form of malrotation.

PRESENTATION

Because the majority of symptomatic children present before 1 month of age and manifest bilious vomiting,

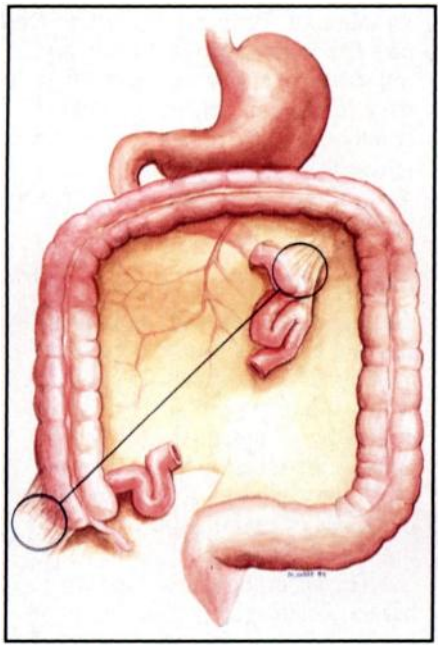

FIGURE 2A. Normal complete rotation of the bowel with fixation of the duodenojejunal junction (ligament of Treitz) in the left upper quadrant and fixation of the cecum in the right lower quadrant. The fixation results in a broad-based mesentery. The potential of a midgut volvulus developing is very unlikely.

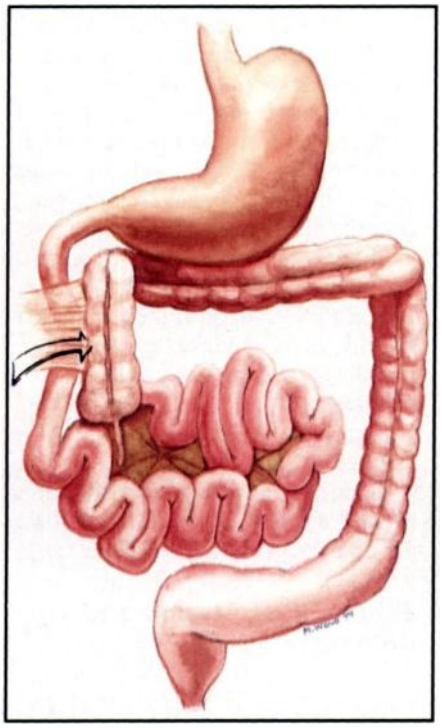

FIGURE 2B. Malrotation. The lack of fixation results in the potential for midgut volvulus. Note Ladd bands covering partial duodenal obstruction.

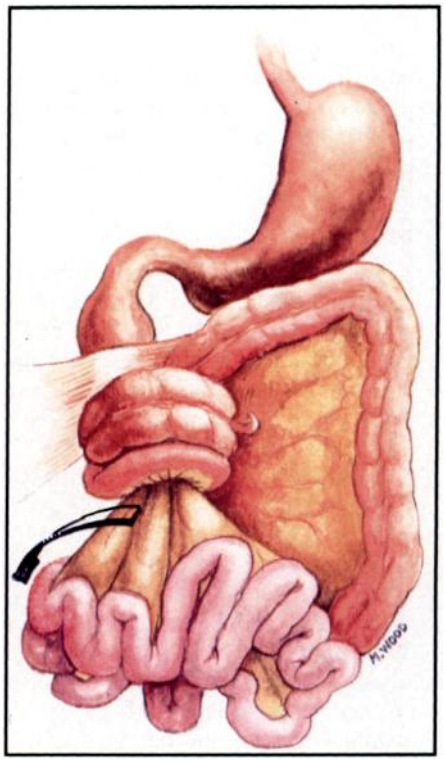

FIGURE 2C. Midgut volvulus. The entire distribution of the superior mesenteric artery (duodenal-jejunal junction to mid-transverse colon) is at risk. Operative reduction is in a counterclockwise direction.

we maintain a rule that "bilious vomiting in the neonate is malrotation until proven otherwise." The symptoms of malrotation may appear in several different ways, and the patient may present having either intermittent abdominal pain, vomiting, or an extreme abdominal emergency related to the development of a midgut volvulus. The most common presentation is that of intermittent duodenal obstruction by Ladd bands occurring as bilious vomiting in the otherwise normal infant. A flat and upright abdominal film may demonstrate evidence of an incomplete duodenal obstruction with an enlarged duodenum. Often the distribution of intestinal gas is seen to be mostly in the right half of the abdomen; gas is seen in the distal intestine. This finding represents a very important distinction between the radiographic appearance of a child who has malrotation and the child who has duodenal obstruction due to another entity.

Bilious vomiting in the neonate is an indication of malrotation until proven otherwise.

DIAGNOSIS

A fair caveat is that the baby who has green vomitus and duodenal obstruction will need an operation and should be referred immediately. The child should be made nothing per os (NPO), with the placement of an NG tube and administration of intravenous fluids and antibiotics. Contrast studies often are employed to help make the diagnosis of malrotation; some pediatric radiologists prefer an upper GI series, others a barium enema. We, and most others, prefer the upper GI series to document whether the ligament of Treitz is positioned properly in the left upper quadrant and whether the duodenum is obstructed. In any child who has bilious emesis and evidence of a rotational anomaly, the possibility of volvulus accentuates the urgency of the diagnosis; expeditious management on the part of the parents, referring physician, and surgeon may make the difference between a condition that is completely reversible and one that, in a few hours, results in loss of a large segment of intestine. Precious time should not be lost in trying to correct a metabolic imbalance that is not correctable until the volvulus has been addressed.

MANAGEMENT

The procedure of choice in infants who have malrotation is the Ladd procedure. This operation consists of dividing the peritoneal (Ladd) bands, which partially obstruct the duodenum, and then placing the colon on the infant's left and the duodenum and jejunum on the right so as to "broaden" the mesentery. An appendectomy always is performed to avoid future confusion when the child has abdominal pain.

The return of intestinal function can be delayed in those who have had a severe ischemic insult from a midgut volvulus. On the other hand, those who have few presenting symptoms and undergo management before the appearance of a volvulus do well and generally return to normal function within a few days. From a prognostic standpoint, these children should do well unless they have had a volvulus that has required a massive resection of small bowel. Such children require long-term intravenous nutrition. Occasionally, the volvulus results in the irreversible loss of such a large segment of bowel that the child's prognosis is hopeless.

Jejunal/Ileal Atresia and Stenosis

EPIDEMIOLOGY AND PATHOGENESIS

Atresia or stenosis of the jejunoileal area is the diagnosis in a significant percentage of neonates who have intestinal obstruction. Atresia, or a complete congenital obstruction, is far more common than is stenosis or a partial occlusion. Reports of its incidence would seem to be on the average of 1 in 750 live births. Coexisting anomalies and familial tendencies are uncommon; the incidence of associated extraintestinal anomalies is thought to be only about 5%. Whereas atresia and stenosis in the more proximal duodenum has been attributed to the failure of recanalization of the proximal small bowel, atresias and stenoses of the jejunoileal region likely are due to late intrauterine mesenteric vascular accidents. Jejunoileal atresia is seen in association with malrotation, meconium ileus, and gastroschisis; each of these could have led to a late intrauterine mesenteric vascular accident followed by atresia.

PRESENTATION

A cardinal sign of jejunoileal atresia is abdominal distention, but it is rarely, if ever, present immediately at birth. Distention in the infant who has jejunoileal atresia will develop 12 to 24 hours after birth, following the initiation of feedings. The infant who presents with abdominal distention immediately at birth is more likely to have meconium peritonitis. Other common findings in those who have jejunoileal atresia are polyhydramnios, bilious vomiting, and failure to pass meconium.

DIAGNOSIS

The diagnosis generally is implied by flat and erect (or lateral decubitus) abdominal radiographs. Large dilated loops of bowel with air fluid levels generally are noted; the lower the atresia, the greater the number of such distended loops. A differential diagnosis of malrotation with or without volvulus, meconium ileus, and Hirschsprung disease exists. A barium enema can by helpful by allowing one to determine whether the colon is used or unused (ie, microcolon) as well as by locating the position of the cecum. Additionally, the barium enema will be able to differentiate between small bowel and colon distention. This will help differentiate conditions in need of sur-

gical intervention (malrotation, Hirschsprung disease, atresia/stenosis) from meconium ileus, which often can be managed nonoperatively (see section on meconium ileus).

MANAGEMENT

Babies who have atresia/stenosis are managed best with placement of an NG tube and initiation of intravenous fluid therapy and antibiotics. The atresia may be corrected immediately, provided that no other life-threatening anomalies exist. Most children have not had severe fluid and electrolyte losses and can be prepared rapidly for the operating room. Occasionally, however, children present with prolonged vomiting and marked electrolyte losses. They should have adequate fluid replacement provided and normal urine output restored before surgery.

Postoperative management includes continued intestinal decompression and administration of appropriate intravenous nutrition until the infant has recovered from his or her period of postoperative ileus and anastomotic dysfunction. This often will last as long as 2 to 3 weeks and, occasionally, longer. These children otherwise tend to do well. Occasionally, those who have small bowel atresias have markedly foreshortened lengths of small bowel due to the in utero vascular accident; they will be limited by their "short gut" syndrome.

Necrotizing Enterocolitis

EPIDEMIOLOGY AND PATHOGENESIS

Neonatal necrotizing enterocolitis (NEC) is a problem that affects preterm infants predominantly. It is unclear why some children are affected and others are spared, but it may be related to compromise of the sick, stressed preterm infant's mesenteric blood flow with an ischemia-reperfusion type of injury sustained by the gut. Epidemiologic data suggest that the presence of enteric feeds has a role in the pathogenesis of necrotizing enterocolitis; in most studies, the affected patients were fed by mouth prior to the onset of the disease. Additional workers have shown that necrotizing enterocolitis has occurred in clustered epidemics, implicating an infectious agent. The most significant risk factor for children to develop necrotizing enterocolitis is prematurity. It is believed by most that in such infants the coexistence of circulatory instability, enteral feeding, and infection in some combination plus the presence of inflammatory mediators function as a sufficient inciting event to create mucosal injury, with bacterial overgrowth and translocation allowing the presence of endotoxin and gas production by bacteria within the gut wall.

The most common site of involvement of necrotizing enterocolitis is the terminal ileum; the colon is the second most common location. The disease can involve single or multiple segments of intestine, although occasionally a fulminating form of necrotizing enterocolitis is characterized by necrosis of the entire gut.

Pneumatosis intestinalis is the hallmark of necrotizing enterocolitis.

PRESENTATION

Children who have necrotizing enterocolitis generally will present having abdominal distention and either a marked gastric residual or vomiting. Occult positive or even grossly bloody stools may become apparent as the abdominal distention increases. Early on, the abdomen is soft, but as the necrotizing enterocolitis progresses, the abdomen tends to become more firm; occasionally, visible loops of bowel are present. Erythema of the abdominal wall occasionally may be noted, suggesting underlying peritonitis. Often, these babies have nonspecific signs of sepsis, such as lethargy and temperature instability, along with some abdominal distention, which suggests NEC.

DIAGNOSIS

The diagnosis of necrotizing enterocolitis often can be made via abdominal radiographs. The gas-forming organisms within the bowel wall produce pneumotosis intestinalis or intramural gas; gas within the portal venous system also may be seen. The presence of intramural gas may be the most important radiologic feature of necrotizing enterocolitis, and it is seen commonly, but not always. Some believe that the presence of portal venous gas portends a more virulent form of the disease, but we have not found this always to be true. Lateral decubitus radiographs may reveal the presence of a pneumoperitoneum, and serial radiographs may demonstrate the presence of a fixed, persistently dilated intestinal loop.

MANAGEMENT

The initial management of necrotizing enterocolitis is nonoperative unless there is good evidence of intestinal necrosis or perforation. All children need to have NG decompression and broad-spectrum antibiotics instituted immediately. The children are monitored by following their white blood cell and platelet counts carefully. Persistent and/or progressive thrombocytopenia commonly is associated with severe cases of necrotizing enterocolitis. Similarly, progressive acidosis suggests significant disease.

Aside from pneumoperitoneum, there is no single criterion that can predict the presence of bowel necrosis invariably. Thus, children suspected of having NEC who develop pneumoperitoneum are operated on without hesitation, but it is difficult to know which children have developed full-thickness necrosis of a segment of bowel but no perforation. Children who have necrotic intestine, which serves as their ongoing source of sepsis, require operation. Many studies have been undertaken to determine how to diagnose intestinal necrosis prior to perforation; the positive findings on abdominal paracentesis have been used by many. We

have felt that infants suspected of having NEC who continue to deteriorate clinically despite adequate supportive therapy are managed best with an early operation. We also have been reasonably aggressive about operating on children who have significant abdominal wall erythema and a progressive thrombocytopenia. More than many other problems of the newborn, NEC requires early surgical consultation and a tremendous amount of judgment as to the appropriate time of operative intervention.

The ultimate outcome of children who have had NEC seems to depend on the severity of disease. The late outcome parallels the late outcome of otherwise sick, stressed, preterm newborns in terms of growth and developmental parameters, but these children will have GI sequelae that depend greatly on the extent of disease and especially on the amount of bowel resected. A child who has severe NEC and resection may have great difficulty with the short gut syndrome and its nutritional sequelae. Some children who did not require operative intervention will develop strictures at their site of disease and subsequently may require an intestinal resection.

Meconium Ileus

EPIDEMIOLOGY AND PATHOGENESIS

Meconium ileus is an intestinal obstruction in the newborn due to the presence of mucoviscidosis (cystic fibrosis), which is inherited as an autosomal recessive trait. The genetic defect responsible for cystic fibrosis (CF) has been identified recently with the gene located on the long arm of chromosome 7. Whereas the diagnosis of CF previously was established via sweat tests and stool trypsin analysis, its diagnosis now is possible prenatally via the use of restriction fragment length polymorphism analysis of amniotic fluid. The body's exocrine gland secretions are abnormal in CF. Approximately 10% to 15% of such infants will develop meconium ileus, with obstruction of varying lengths of their distal ileum and colon by inspissated meconium. CF almost exclusively is a caucasian disease that presents in approximately 1 in every 2000 live caucasian births.

PRESENTATION

Meconium ileus is classified as either simple or complicated. In simple meconium ileus one finds that the distal ileum and proximal colon are impacted with inspissated meconium and that a resultant obstruction is in the mid-ileum, which is markedly dilated and filled with a thick, tenacious, dark green, "tarry-like" meconium. The wall of the small bowel can be congested and hypertrophied; the amount of distention as one proceeds proximally becomes progressively less. Distal to the obstruction the ileum is narrow, with the inspissated meconium appearing as pellets of gray, beaded putty. Most of the colon is narrow and empty, presenting as a microcolon. The meconium ileus may become complicated by having the dilated proximal intestinal segment undergo a volvulus in utero such that the ischemic segment resorbs, resulting in the formation of an intestinal atresia. If volvulus and intestinal necrosis occur late enough in gestation, meconium perforation occurs with the presence of meconium peritonitis; occasionally, the abdomen is filled with a large pseudocyst. All of these "complications" are mechanical in nature and occur before birth, so evaluation and management of the newborn who has meconium ileus requires a distinction between the simple and complicated varieties.

DIAGNOSIS

Meconium ileus is the most common cause of neonatal intraluminal intestinal obstruction. An important differential diagnostic feature of meconium ileus is that affected infants often are born having marked abdominal distention (as compared with children who have jejunoileal atresia who develop distention) and may vomit biliously, fail to pass meconium, or both. The impacted intraluminal meconium usually is palpable as a doughy, rubbery substance, and the distended abdomen will have a characteristic feel to it.

If abdominal radiographs are performed, the children who have simple meconium ileus demonstrate distended intestinal loops without air fluid levels and with a markedly disparate degree of distention. Also, the presence of the meconium gives the radiograph a coarse, granular, "soap bubble" appearance, which may aid in the diagnosis. Patients who have complicated meconium ileus often will have scattered calcifications evident on abdominal radiographs, which reflect the presence of meconium peritonitis from in utero intestinal perforation.

Children who have simple meconium ileus demonstrate distended intestinal loops without air fluid levels.

MANAGEMENT

Management of these infants depends initially on whether the meconium ileus is simple or complicated. Infants who have a simple obstruction may be both diagnosed and treated by performing a contrast enema under fluoroscopic control, using gastrografin. This hyperosmolar enema often can relieve the obstruction of the meconium ileus by drawing fluid into the intestinal lumen and allowing the mass of meconium to become softened and disimpacted. These children will require NG decompression and intravenous fluids because the hyperosmolar contrast agent will deplete their intravascular space as the fluid shifts into the intestine. Success of the gastrografin enema is gauged by free reflux of the contrast agent into the proximal dilated small bowel. Occasionally, two to three such enemas are needed before success is achieved. In infants in whom the contrast enema fails to relieve the obstruction or in those whose meconium ileus is complicated, an operation should be undertaken.

Once recovered from meconium ileus, the infant's long-term outlook

will depend on the severity and rate of progression of the CF. The degree of pulmonary disease is likely to be the most limiting factor in his or her prospect for long-term survival.

Colonic Atresia/Stenosis

EPIDEMIOLOGY AND PATHOGENESIS

Atresia or stenosis of the colon is a rare anomaly occurring in approximately 1 of every 20 000 live births; only gastric atresia is rarer than colonic atresia. When encountered, colonic atresia frequently is associated with skeletal anomalies such as syndactly, polydactly, absent radius, and club foot. There also is an association of ocular and cardiac anomalies. As with the jejunoileal forms, colonic atresia and stenosis are widely considered to be due to in utero vascular compromise.

PRESENTATION

In colonic atresia/stenosis, the children present in the early newborn period having findings of a very distal intestinal obstruction. The infants will develop marked abdominal distention with bilious vomiting. Stools usually are absent, making much of their initial presentation not greatly dissimilar from those children who have jejunoileal atresia. However, in children who have colonic atresia, the abdominal radiograph may demonstrate more and larger dilated loops than are seen in a small bowel obstruction. It is not uncommon for children who have colonic atresia to present having colonic perforation and pneumoperitoneum. A definitive diagnosis of colonic atresia can be established via a barium enema, which reveals a microcolon with incomplete colonic filling.

MANAGEMENT

These infants are managed via NG decompression, intravenous fluids, and antibiotics prior to surgery. Once the diagnosis has been established and the intestines are decompressed, the operation can be timed according to the infant's well being and need for other evaluations.

Because colonic atresia/stenosis is so rare, there are no large series to provide a full prognosis of this anomaly, but it seems that reported deaths in modern times have occurred primarily from critical associated anomalies or prolonged delays in recognizing the problem. These infants should fare well once surgically corrected.

Meconium Plug Syndrome/ Small Left Colon Syndrome

From a clinical standpoint, these two entities are distinct, but they seem to represent a continuum of transient neonatal colonic dysfunction. Children who have meconium plug syndrome will present with evidence of a low intestinal obstruction. Following administration of a contrast enema and passage of a large meconium stool, they seem to be relieved of difficulties. This problem commonly is seen in preterm infants. Regardless of the age of presentation, should the children have further stooling abnormalities after the enema, they need to be evaluated for Hirschsprung disease.

Infants of diabetic mothers also may present having the appearance of a distal GI obstruction, and they may have abdominal distention, bilious emesis, and no stool in the immediate newborn period. Such infants are managed with NG decompression and control of hypoglycemia. The use of a contrast enema will document the presence of a small left colon and a dilated proximal colon filled with meconium. Generally, these infants are "relieved" with administration of the contrast enema, but it may need to be repeated. Many clinicians, even in the presence of a "classic" history, will obtain a suction rectal biopsy to be sure that Hirschsprung disease is not present.

Hirschsprung Disease

EPIDEMIOLOGY AND PATHOGENESIS

Hirschsprung disease is a congenital anomaly that can present as complete intestinal obstruction in the newborn. Interestingly, though, Hirschsprung disease seems to have varying degrees of severity, and it is not uncommon for the diagnosis not to be made until the child is older and experiencing severe problems of constipation.

Hirschsprung disease is characterized by a congenital absence of ganglion cells in the myenteric and submucosal plexuses of the bowel. Its exact etiology is unknown, but it is suggested that the migration of neuroblasts during their cranial caudad descent in the intestine has ceased. The length of aganglionic bowel varies according to the time of the arrest of the migration. Approximately 80% of Hirschsprung disease cases involve only the rectosigmoid region; 3% of cases involve the entire colon, and even fewer have been reported to have total intestinal aganglionosis.

Hirschsprung disease is three times more common in males than in females.

The incidence of Hirschsprung disease seems to be approximately 1 in 5000 live births and is three times more common in males than in females; a pronounced difference in sex incidence occurs in the long segment disorder. Some familial occurrences of Hirschsprung disease have been reported, especially where long segment involvement is present.

Peristalsis is abnormal or absent in the aganglionic segment of the child's colon. Failure of the involved bowel and the internal anal sphincter to relax produce the intestinal obstruction and/or constipation found in Hirschsprung disease. The intestine proximal to the aganglionic segment becomes dilated and hypertrophied. When Hirschsprung disease presents in the newborn, there often is evidence of complete intestinal obstruction, with abdominal distention and absence of passage of meconium.

PRESENTATION

The presentation may vary from being mildly constipated to having an initial, fulminant toxic enterocolitis,

which occasionally can be fatal. Such severely affected children develop tense abdominal distention over a few hours and vomit profusely while passing large amounts of foul-smelling gas and putrid loose stools. The spectrum of presentation ranges from complete obstruction at birth, with vomiting, abdominal distention, and failure to pass meconium, to delayed passage of meconium and repeated episodes of constipation. One must always be suspicious of the diagnosis of Hirschsprung disease in the baby who has evidence of a partial or complete distal bowel obstruction.

DIAGNOSIS

The differential diagnosis includes meconium ileus, jejunoileal atresia, meconium plug syndrome, and small left colon syndrome as well as nonsurgical sources such as neonatal sepsis, adrenal insufficiency, and hypothyroidism. The obstruction can be ascertained as mechanical and not functional by the performance of a barium enema, which should demonstrate a transition zone at the junction of aganglionic and ganglionated bowel. Any child who presents having colonic distention and the absence of rectal air on a plain abdominal radiograph, therefore, should be examined.

The diagnosis may be difficult to make in the newborn because the transition zone between the aganglionic segment and the proximal dilated colon may be subtle or absent. In this instance, obtaining follow-up radiographs 24 and 48 hours after the study can be helpful because if barium is being retained, this can be interpreted as a relatively "positive" diagnostic sign. Although some have used manometric studies of the colon to diagnose Hirschsprung disease, these rarely are useful in the neonate. The best and most definitive means of diagnosis is a rectal biopsy, which will confirm aganglionosis histologically. Although a preoperative biopsy may not be necessary if the child's history and findings from the barium enema are convincing, even in the smallest of babies, suction rectal biopsy can be performed at the bedside.

MANAGEMENT

Management of these children, as any child who evidences distal bowel obstruction, should include the placement of an intravenous catheter as well as NG decompression and the use of intravenous antibiotics. It is important that cleansing enemas and rectal examinations not be done prior to the performance of the contrast study because the characteristic configuration of the bowel could be lost if the colon is evacuated. Once the diagnosis of Hirschsprung disease has been established, prompt colostomy should be undertaken and the level of ganglionosis is established. The colostomy then is placed in a segment of bowel that is ganglionated, and the infant is allowed to grow and thrive. The child may be managed further via a definitive pull-through procedure at approximately 8 to 10 months of age.

Imperforate Anus

EPIDEMIOLOGY AND PATHOGENESIS

The term imperforate anus encompasses a broad spectrum of anorectal malformations that occur in 1 in 5000 live births, with a slight preponderance in males. This is a disorder of embryogenesis involving the hind gut and results from the abnormal development of the urorectal septum, with incomplete separation of the cloaca into its urogenital and anorectal components (Figure 3). As a result of this abnormal development, the anal canal may end blindly or there may be an ectopic opening on the perineum that virtually always is directed anteriorly. This canal may terminate in the vulva/vagina or in the male urethra.

Imperforate anus generally is categorized as either low or high; this classification is determined by whether the blind end of the rectum ends above or below the level of the levator musculature. A high type of lesion is more common than the low type and, in general, is more complex; rectourinary and rectovaginal fistulas commonly are associated. There is a greater male-to-female predominance in patients who have the high type of lesions, and more than 80% of such patients will demonstrate such fistulas. The male-to-female ratio in low imperforate anus is closer to 1:1, and in most of these children, a discernible external perineal fistula is noted at or shortly following birth. There does not seem to be a strong genetic predisposition, but reports of familial cases do exist.

DIAGNOSIS

The distinction between low and high imperforate anus occasionally can be hard even for experts to make. In general, though, the distinction is best made by using the combination of physical and radiographic examinations. Whether the child is male or female, a visible fistula through which meconium is passed very likely has a low lying rectal pouch. If there is any doubt, however, a contrast injection through the fistula can determine the diagnosis with certainty. A helpful means of diagnosing the level of the rectal pouch is the so-called invertogram, first described by Wangenstein and Rice, which when properly performed, often can show the level of the pouch relative to the levator musculature. Some currently are employing ultrasonography to help determine the level of the blind rectal pouch. Most believe that it can take as long as 24 hours for air to descend far enough

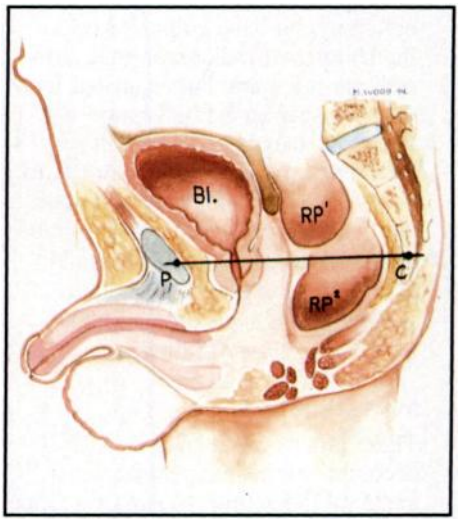

FIGURE 3. Imperforate anus. Rectal pouch 1 (RP^1) sits above the pubococcygeal line (PC) and would be classified as a "high type" anomaly. Rectal pouch 2 (RP^2) sits below the PC line and represents a "low type" anomaly. The level of the rectal pouch is crucial in decisions of management. Bl indicates the bladder.

down in the pelvic colon to determine the depth of descent of the rectal pouch accurately. Children generally are studied at this point unless they become distended or demonstrate the presence of a perineal fistula. Such patients should be made NPO and begin intravenous antibiotics. Although NG tubes are not placed routinely, if the child needs to be transferred to a referral center for appropriate diagnosis and management, we believe that the NG tube should be used during transport.

Children who have imperforate anus often will manifest the VATER associations and need to be investigated carefully for other anomalies. A significant feature of this anomaly is the absence of sacral vertebral segments in some children who have high type defects; these children can be predicted to have some deficiencies in their neurologic control of defecation. Almost 50% of children who have high imperforate anus also will have genitourinary malformations; an ultrasonographic study of the kidney and early assessment of the renal excretory status is important in the preoperative evaluation.

MANAGEMENT

In general, most low type imperforate anus problems can be managed via a perineal anoplasty performed during infancy. High imperforate anus, however, requires a more extensive operative repair as well as division of the likely rectourinary fistula. Therefore, these infants generally are managed via a colostomy in the newborn period. In addition to stool diversion, the properly constructed colostomy will avoid urinary tract soiling via contamination of the rectourinary fistula. Definitive reconstructive pull-through surgery then is undertaken at about 8 months of age.

Children who have low type imperforate anus do very well; those who have high type imperforate anus tend to have difficulty with fetal continence, which can vary in severity. This especially is true of infants who have absent sacral segments, so this problem should be identified early for the family. Other limitations of children who have imperforate anus depend greatly on the coassociation with other VATER anomalies.

SUGGESTED READING

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PIR QUIZ

1. Among the following, the *most important* symptom or finding in an infant that is suggestive of intestinal obstruction is:
   A. Distention of intestinal loops on radiograph.
   B. History of oligohydramnios.
   C. Hyperactivity of bowel sounds.
   D. Passage of bloody stools.
   E. Vomiting of bile.

2. A major difference in the clinical picture of congenital upper small bowel obstruction versus congenital large bowel obstruction is the finding of:
   A. Absence of bowel sounds.
   B. Bilious vomiting.
   C. Failure to pass meconium.
   D. Generalized abdominal distention.
   E. Metabolic acidosis.

3. A 2-day-old male infant has been transferred from a community hospital with the complaint of repeated emesis since first feeding. His mother is primiparous with a history of crack cocaine use in the third trimester. Birth weight was 7 lb. At birth the abdomen was noted to be "unusually full." Findings on the admission physical examination included a distended abdomen with hyperactive bowel sounds. On rectal examination, the ampulla is empty. On radiographic examination, air is present in multiple loops of bowel, no air fluid levels are seen, and flecks of calcium are scattered throughout the abdomen. The *most likely* diagnosis is:
   A. Annular pancreas.
   B. Bacterial peritonitis.
   C. Drug withdrawal.
   D. Meconium ileus.
   E. Neonatal toxoplasmosis.

4. A 6-day-old full-term female infant develops repeated vomiting, abdominal distention, and cessation of passage of stool. According to her history, meconium passage was delayed. Temperature is normal. Rectal examination is followed by a large quantity of liquid feces with much gas. Of the following, the *most appropriate* next step in management is to:
   A. Order a gastrografin (oil-based contrast) enema.
   B. Order a barium enema after saline cleansing enema.
   C. Refer for anorectal manometry.
   D. Refer for suction rectal biopsy.

5. A 10-day-old preterm female infant develops repeated episodes of vomiting with abdominal distention. Birthweight was 1400 g, and her course was uneventful until the present symptoms. Nasojejunal feedings were well tolerated initially. Platelet count and coagulation studies are normal. Stools now are liquid and guaiac positive. The *most correct* statement regarding this clinical picture is:
   A. A previously unsuspected congenital anomaly of the gastrointestinal tract is the basis for the problem.
   B. Continuation of normal platelet counts predicts a benign course.
   C. Prompt antibiotic therapy is required to avoid severe dehydration from enterotoxigenic *Escherichia coli* infection.
   D. The possibility of intussusception necessitates performing a barium enema.
   E. The presence of pneumatosis intestinalis on radiograph will indicate the underlying pathology.

6. A 3-week-old male infant presents with a 1-day history of recurrent emesis. The vomitus is bile-stained. A similar episode occurred at 5 days of age and spontaneously resolved in 1 day. Findings on physical examination include a normal temperature, flat abdomen, and occasional bowel sounds. On rectal examination, normal stool is present. The *most appropriate* next step in management is:
   A. Intravenous fluid replacement until alkalosis is corrected.
   B. Observation until preliminary results of septic evaluation are available.
   C. Prompt upper GI radiographic study.
   D. Trial of thickened feedings with hypoallergenic formula.

Intestinal Obstruction in the Newborn

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