Question 3

The newborn nursery staff asks you to see a 3-day-old infant born at term after a repeat cesarean section. The pregnancy was uncomplicated. He has had bilious emesis with his last feeding. His abdomen has become distended and tender to palpation. The infant passed a grossly bloody stool while being examined. An abdominal radiograph was obtained (Figure 1).

Figure 1: Midgut volvulus (from emedicine.medscape.com/article/411249-media)

After reviewing the radiograph, you discuss the pathogenesis of the neonate’s defect with the resident.

Of the following, the MOST likely cause of the infant’s lesion is:

A. abnormal midgut rotation
B. colonic hypoperistalsis
C. failure of the duodenum to recanalize
D. failure of the ventral pancreas to migrate
E. vascular ischemia to the jejunum

Incorrect:
Correct Answer: A

The neonate in the vignette has a surgical emergency complicating intestinal malrotation. Approximately two-thirds of patients with intestinal malrotation become symptomatic during the first month after birth. Acute midgut volvulus with intestinal ischemia is the most significant and potentially life-threatening complication of intestinal malrotation. Radiographic findings of a malrotation may range from a few nonspecific findings to a complete small bowel obstruction with multiple loops of dilated bowel and air fluid levels. The previously healthy infant in the vignette developed bilious emesis and had a tender and distended abdomen; radiography demonstrated a nearly gasless abdomen with a central mass effect; and displayed sufficient evidence to proceed with an emergency exploratory laparotomy in an attempt to prevent intestinal necrosis.

During the 4th to 8th week of embryonic development, the coelom cannot contain the rapidly expanding midgut (Figure 2).

Figure 2: Illustration of counterclockwise rotation of the gut during early gestation. A, the superior mesenteric artery is seen coming out into the yolk stalk with the duodenojejunal loop above and the cecocolic loop below. B, the gut has rotated 90° counterclockwise and grown considerably. C, The small intestine starts to move to the left side of the abdomen as it grows and the gut rotates another 90° counterclockwise. D, When the gut returns to the abdomen, it rotates a final 90°. With growth and the final rotation the cecum and ileum descend into the right lower quadrant anterior to the superior mesenteric artery and the third portion of duodenum is pushed up into the left upper quadrant posterior to the superior mesenteric artery (adapted from Moore and Persaud [1998]).
Because of these restrictions, the midgut loop must buckle into the yolk stalk. The axis of this loop is the future superior mesenteric artery. As the intestine buckles out of the abdomen, it begins twisting counterclockwise. The initial 180° of counterclockwise rotation occurs because: (1) the proximal duodenojejunal (duodenum, jejunum, and a majority of the ileum) loop grows faster than the distal cecocolic (terminal ileum, cecum, and right and proximal transverse colon) loop, and (2) the liver is rapidly expanding. By the 12th week of gestation, after the primary loop is considerably longer, it returns to the abdominal cavity and rotates an additional 90° counterclockwise. Most of the important rotational anomalies are thought to occur from altered or arrested development during this stage. The first segment to return is the duodenojejunal segment. The final rotation places the third portion of the duodenum posterior to the superior mesenteric artery and the ligament of Treitz in the upper left quadrant. The distal loop returns after the proximal loop and also rotates 90°. Because the cecum and terminal ileum are the final portions of the midgut to return, the transverse colon will eventually lie anterior to the superior mesenteric artery and the cecum will be positioned in the right lower quadrant. (see also http://www.indiana.edu/~anat550/gianim/gutrot/gutrot.html)

After the bowel has rotated to its final position, it becomes anchored. The process of fixation begins during the 12th week of gestation and continues after birth. Normal fixation will occur only if midgut rotation has been completed appropriately. The most proximal portion is fixed at the ligament of Treitz to the retroperitoneum early in development. Fixation of the distal portion of the midgut (colon) to the retroperitoneum is more gradual and later. The jejunum, ileum, and transverse colon remain mobile in the intraperitoneal cavity.

With normal rotation of the intestinal tract, a wide-based mesentery develops, extending from the ligament of Treitz in the upper left quadrant of the abdomen to the ileocecal valve in the lower right quadrant. The normal broad-based mesentery prevents significant rotation of the mobile intestine. Incomplete rotation of intestine (approximately 180°), the most common variant of malrotation or nonrotation (<90°), and subsequent fixation will produce narrow mesenteric variations that are prone to cause intestinal obstruction. The narrow mobile mesenteric pedicle is prone to clockwise rotation around the superior mesenteric artery axis which can result in obstruction of the intestinal tract and mesenteric vessels. The shortened fused mesentery created by nonrotation or incomplete intestinal rotation can also cause extrinsic compression and obstruction of the duodenum. These shortened fused bands, known as Ladd’s bands, extend from the retroperitoneum in the right upper quadrant and attaches to the right colon compressing the duodenum (Figure 3).

Figure 3: Illustration of Ladd’s band producing duodenal obstruction (adapted from Oldham [2005])
Congenital duodenal atresia can be complete or partial and intrinsic or extrinsic. Intrinsic atresia likely stems from a failure of the duodenum to recanalize during the 8th to 10th week of gestation following obliteration of the lumen by epithelial proliferation during the 6th and 7th weeks. Neonates with complete duodenal atresia present with gastric distention and bilious emesis. An abdominal radiograph will show a double-bubble sign because of dilation of the stomach and proximal duodenum. Distal gas is absent. Abdominal tenderness and hematochezia are uncommon in neonates with duodenal atresia.

The pancreas becomes annular when the ventral bud of the developing pancreas fails to rotate behind the duodenum and fuse with the dorsal portion of the pancreas before the 7th week of gestation. Pancreatic tissue is thus able to fully surround and compress the second aspect of the duodenum. More than two-thirds of children with annular pancreas present during the neonatal period with features of gastric outlet obstruction. Symptoms may include bilious vomiting and gastric distention. In symptomatic neonates with an annular pancreas, abdominal radiographs will show the classic “double-bubble” sign that is suggestive of duodenal obstruction. Abdominal tenderness and hematochezia are uncommon in neonates with an annular pancreas.

Jejunal or ileal atresia is an acquired lesion thought to be the result of vascular disruption to the intestine that causes ischemic necrosis. The necrotic tissue is reabsorbed, leaving blind
ends, often with a gap in the mesentery. Causes of vascular disruption may include volvulus, intussusception, internal hernia, and interruption of the segmental mesenteric blood supply. Affected neonates with jejunal or ileal atresia develop abdominal distention, often with palpable loops of bowel. Vomiting occurs within the first 2 days of birth. Hematochezia is rare. Abdominal radiographs of neonates with jejunal atresia usually have multiple loops of dilated bowel with air fluid levels. The radiograph of the infant in the vignette is not compatible with jejunal atresia.

Meconium plug syndrome refers to a spectrum of clinical presentations that are characterized by obstruction of the neonatal colon. Various terms have been used to describe meconium plug syndrome including small left colon syndrome, left-sided microcolon, and functional colonic inertia of prematurity. Infants with meconium plug syndrome are generally healthy. Infants born to diabetic mothers can present with meconium plug syndrome that is believed to be associated with hypoperistalsis caused by increased endogenous glucagon production. Meconium plug syndrome can be associated with impressive abdominal distention, bilious emesis, and failure to pass meconium during the first 24 to 48 hours after birth. An abdominal radiograph may show intestinal distention with multiple dilated loops of bowel and air fluid levels. The radiograph of the neonate in the vignette is not compatible with meconium plug syndrome. Hematochezia is rare with meconium plug syndrome.

References:


American Board of Pediatrics Content Specification(s):

11_Gastroenterology: Know the pathogenesis of atresias, stenosis, diverticulae, and duplications of the small intestine including those associated with annular pancreas

11_Gastroenterology: Know the clinical manifestations of atresias, stenosis, diverticulae, and duplications of the small intestine including those associated with annular pancreas

11_Gastroenterology: Know the pathogenesis and clinical manifestations of infants with malrotation and/or volvulus of the small intestines