Malrotation and Volvulus

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Incidence
From autopsy studies it appears that the incidence of malrotation in the general population is between 0.2-0.5%. However, the generally accepted incidence based on clinical presentation at the major children's hospitals is one per every 6000 live births. It is clear that this condition can be present throughout life without causing any clinical problems. However, those afflicted generally present within the first six months of life with smaller numbers occurring each year throughout childhood. Presentation after teenage years or in adult life is quite rare.

Etiology
The growth rate of the gastrointestinal tract in early gestation is faster than that of the body. Therefore, intestinal development occurs through the umbilical ring within the physiological umbilical hernia (fourth week of gestation). During the tenth week of gestation the intestine begins an orderly migration back into the abdominal cavity. This includes a counterclockwise rotation of two intestinal segments: the duodenojejunal segment and the cecolic segment.

The duodenojejunal limb returns first and rotates 270° to the right of the superior mesenteric artery (SMA), passing beneath the artery into the left upper quadrant. At the completion of this process, the duodenojejunal junction is fixed to the retroperitoneum at the ligament of Treitz.

The rotation of the cecolic limb is also counterclockwise. It rotates from a position left of the SMA, around SMA axis to attain its final position in the right lower quadrant. By the twelfth week of gestation this process is complete, and the colon becomes fixed to the retroperitoneum.

A broad based mesentery provides a stable position for the small intestine within the peritoneal cavity. It begins in the right iliac fossa at the ileocecal junction and runs obliquely upward across the abdomen to the ligament of Treitz. The duodenum is fixed securely to the retroperitoneum as the C-loop. The ascending and descending colon are fixed to the right and left retroperitoneum, and the transverse colon is fixed at either end as it drapes across the superior mesentery vessels. If fixation does not take place, the intestine is suspended by a thin stalk containing the superior mesenteric vessels and is susceptible to midgut volvulus.

Children with congenital diaphragmatic hernia, omphalocele or gastroschisis will have malrotation as a matter of course since the bowel cannot undergo normal
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rotation and fixation. However, other gastrointestinal problems, including intestinal atresias and Hirschsprung's disease, have occurred in association with malrotation.

**Classification**

During the course of rotation and fixation, three conditions develop that make the gut susceptible to volvulus:

**Nonrotation**

This is the most common anomaly. It occurs when neither duodenojejunal or cecocolic limb have undergone correct rotation. The duodenojejunal and ileocecal junctions lie close together while the midgut hangs on a thin stalk containing the superior mesenteric vessels. The entire gut is poorly stabilized due to the narrow base of the mesentery.

**Abnormal Rotation of the Duodenojejunal Limb**

Nonrotation of the duodenojejunal limb followed by normal rotation and fixation of the cecocolic limb results in duodenal obstruction caused by abnormal mesenteric (Ladd's) bands extending from the colon across the anterior duodenum. The risk of midgut volvulus in this condition is lower because there is a relatively broad mesenteric base between the duodenojejunal junction and the cecum.

**Abnormal Rotation of the Cecocolic Limb**

Normal rotation of the duodenojejunal limb with nonrotation of the cecocolic limb results in the same potential for midgut volvulus as complete nonrotation. In this condition, the cecum and the first part of the colon lie close to the midline against the third portion of the duodenum over the superior mesenteric vessels. The mesenteric base is narrow and stability is poor. Volvulus easily develops.

Reversal rotation of either limb create rarer forms of malrotation presenting as colonic obstruction, entrapment in a paraduodenal hernia, or midgut volvulus.

**Clinical Presentation**

Presentation varies depending on the specific mechanism of obstruction and the extent of vascular compromise. Symptoms may be either mild and intermittent or severe and catastrophic with complete obstruction and vascular occlusion.

The most common symptom of malrotation with volvulus is vomiting (95%). Initially, the vomitus is gastric or bilious in nature but may become grossly bloody if bowel compromise is present. Abdominal distention follows with bloody diarrhea (28%) indicating bowel ischemia or necrosis. Children with volvulus appear severely ill and complain of generalized abdominal pain if they can speak. Lethargy, grunting respirations, dehydration, peritonitis and shock follow as the bowel ischemia persists or worsens.

**Diagnosis**

The diagnosis of malrotation with or without volvulus usually rests upon radiographic confirmation. Plain abdominal radiographs may demonstrate obstruction and abdominal distention. The radiographic appearance of a "gasless" abdomen occurs when the volvulus has created a closed-loop obstruction from which the intraluminal air has been absorbed and replaced with fluid.
Contrast radiographic studies can easily demonstrate the site of obstruction and the presence of the malrotation. In simple malrotation, the upper gastrointestinal series shows the incomplete rotation of the duodenojejunal loop (Fig. 58.1). If volvulus has occurred, there is frequently an abrupt cut-off to passage of barium described as a “bird’s beak” in the third portion of the duodenum. Alternatively, duodenal obstruction may be only partial and have a spiral or corkscrew appearance.

Barium enema can identify the position of an abnormally placed cecum suggesting malrotation. However, the position of the cecum is highly variable in small children, and about 15% of children with malrotation will have a normally placed colon.

Ultrasonography has proven to be very reliable in making the diagnosis of malrotation at some centers. The relative position of the superior mesenteric artery and vein is normally quite constant and characteristic with the vein to the right of the artery. Absence of this normal relationship strongly suggests malrotation.

Treatment
Midgut volvulus is a surgical emergency. A child with this condition requires intravenous access, a nasogastric tube, expeditious fluid resuscitation followed by an emergent laparotomy. Malrotation without volvulus is a relatively nonemergent condition that allows more time for preoperative decision making.

The operative management (Ladd procedure) of malrotation involves six principles. These can be applied to those with uncomplicated malrotation as well as to those with midgut volvulus.

Evisceration
Generally a supra-umbilical transverse incision is made. The bowel is eviscerated and the malrotation is confirmed (Fig. 58.2). Ascites (chyrous from obstruction or bloody from necrosis) is frequently encountered and drained.

Untwisting of the Volvulus
The volvulus is untwisted by rotating it counterclockwise. If there is no bowel compromise, the operation is continued. If the bowel is edematous and hemorrhagic, improvement may occur with untwisting. If the bowel appears necrotic or nonviable, a second look operation 24-36 hours later may help to determine viability and preserve bowel.

Division of Ladd’s Bands
Ladd’s bands create a duodenal obstruction as they pass over the malpositioned second and third portion of the duodenum. These peritoneal folds are divided freely mobilizing the cecum.

Widening the Mesenteric Base
After untwisting the volvulus and lysis of Ladd’s bands, it is generally possible to widen the base of the mesentery. This is achieved by placing the small bowel along the right gutter and positioning the colon to the left with the ileocecal valve facing in the opposite direction.
Fig. 58.1. Upper gastrointestinal series x-ray demonstrating failure of the duodenum to complete rotation and reach a proper position in the left upper quadrant at the ligament of Trietz.

Fig. 58.2. Intraoperative photograph of malrotation. Large arrow indicates the cephalad direction. Smaller arrows demonstrate the backward orientation of the ileocecal valve with the cecum and appendix clearly unfixed and in the wrong position.
Relief of Duodenal Obstruction
Lysis of Ladd’s bands will relieve most obstructions but occasionally further dissection is needed to fully mobilize and straighten the duodenum. In addition, intraluminal duodenal obstruction (i.e., atresia, stenosis) may coexist and must be identified and corrected. A soft tube can be passed through the bowel along its entire length to verify its patency.

Incidental Appendectomy
Since the colon and the attached appendix generally lie in an abnormal position both before and after an operation for malrotation, it is advisable to remove the appendix to prevent confusion if the child were to develop appendicitis in the future.

Outcomes
Recurrence volvulus has been reported in up to 10% of children having the Ladd procedure. In addition, these children have the 5-6% chance of postoperative bowel obstruction secondary to adhesions. Complications of atelectasis, wound infection, postoperative bleeding, and prolonged ileus are all common. Many children have protracted hospital stays, particularly if a volvulus progressed to long segments of intestinal necrosis requiring resection. In fact, 18% of children with short bowel syndrome have this condition secondary to malrotation with midgut volvulus. In children who present with peritonitis, shock and sepsis, multiple organ failure with death may occur despite aggressive resuscitation and surgical intervention.

Selected Readings