# Intestinal Rotation Abnormalities and Midgut Volvulus

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**KEYWORDS**

• Malrotation • Nonrotation • Heterotaxia • Intestinal obstruction • Bilious vomiting

**KEY POINTS**

- Rotation abnormalities represent a spectrum from non-rotation to normal rotation.
- Malrotation may result in lethal midgut volvulus. Any child with bilious vomiting must be assumed to have midgut volvulus until proven otherwise.
- The gold standard for the diagnosis of a rotation abnormality is an upper gastrointestinal contrast study looking for the location of the duodenojejunal junction.
- A laparoscopic approach is useful for children without midgut volvulus. Infants, and older children with suspected midgut volvulus should undergo laparotomy.

## INTRODUCTION: NATURE OF THE PROBLEM

Intestinal rotation abnormalities constitute a spectrum of conditions that occur during the normal embryologic process of intestinal rotation. In some patients the rotation abnormality is asymptomatic, but others experience a variety of symptoms, including obstruction, lymphatic and venous congestion, and misdiagnosis of appendicitis in an abnormally positioned appendix **Box 1**. The most important form of obstruction is midgut volvulus, which may be fatal. For this reason, it is important for all surgeons to have an understanding of rotation abnormalities, and to have a high index of suspicion in any patient with signs and symptoms of intestinal obstruction.

## RELEVANT EMBRYOLOGY AND ANATOMY

Intestinal rotation occurs during the fourth through twelfth weeks of gestation.[1] During the fourth to fifth week postconception, the straight tube of the primitive embryonic intestinal tract begins to elongate more rapidly than the embryo, causing it to buckle ventrally and force the duodenum, jejunum, ileum, and the ascending and transverse colon to extend into the umbilical cord. The duodenum curves downward and to the right of the axis of the artery, initially completing a 90° counterclockwise turn. Over the next 3 weeks, the duodenum continues to rotate so that, by the end of 8 weeks, it has

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**Box 1**
**Signs and symptoms of intestinal rotation abnormalities**

*Nonrotation*
- Asymptomatic
- Associated motility disorder
- Associated condition (eg, abdominal wall defect, diaphragmatic hernia, heterotaxia)
- Appendicitis in abnormal location

*Malrotation without volvulus*
- Asymptomatic
- Bilious vomiting caused by Ladd bands or associated duodenal web
- Associated medical condition (eg, heterotaxia syndrome)
- Appendicitis in abnormal location

*Malrotation with volvulus*
- Bilious vomiting
- Abdominal pain
- Hematochezia
- Peritonitis
- Death

*Malrotation with partial or intermittent volvulus*
- Protein-losing enteropathy
- Abdominal pain
- Failure to thrive
- Malnutrition
- Occult gastrointestinal bleeding

undergone 180° rotation. During the tenth gestational week the intestines return to the abdomen. The cecum is the final portion of the intestine to return and does so by rotating superiorly and anteriorly around the superior mesenteric artery (SMA). This sequence of return causes the duodenum and proximal jejunum to be pushed superiorly and to the left posterior to the SMA so that they become fixed in a 270° rotation from their initial position. Fixation of the intestines in this position takes place over the fourth and fifth months of gestation.

## DEFINITIONS OF INTESTINAL ROTATION ABNORMALITIES

The spectrum of intestinal rotation abnormalities arises from perturbations in the sequence of herniation, rotation, and fixation of the midgut. If the cecocolic loop returns to the abdomen before the return of the proximal foregut, the duodenum and jejunum are not pushed superior-laterally and undergo only 180° of rotation. In this scenario, the cecum does not undergo proper fixation and the colon remains on the left side of the abdomen, whereas the midgut fills in the space on the right and the duodenum descends directly along the course of the SMA. This condition is termed nonrotation and, because it is associated with a wide-based mesentery, does not put the child at risk for midgut volvulus. Classic malrotation occurs as a result