

Imaging of Malrotation in the Neonate



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Intestinal malrotation is a congenital abnormal fixation of the bowel that predisposes to life threatening midgut volvulus. Most events of midgut volvulus occur in the first month of life with decreased risk with age. The most typical presentation is bilious vomiting. Upper gastrointestinal series is the study of choice for diagnosis of malrotation. For optimal results, the study should be well planned with meticulous attention to technique. In equivocal cases, small bowel follow-through study and ultrasound can be performed.

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Intestinal malrotation is a congenital abnormal position of the bowel with a short mesenteric small bowel root that may be accompanied by obstructing peritoneal bands. It may lead to a life-threatening midgut volvulus, for which newborns have the highest risk. The most typical presentation of midgut volvulus is new onset of bilious emesis. As delay in the diagnosis of midgut volvulus can have catastrophic consequences, any infant with a sudden onset of bilious emesis must be urgently evaluated for malrotation with midgut volvulus. The upper gastrointestinal series (UGIS) is the examination of choice in the diagnosis of malrotation. The accuracy of the study depends on meticulous technique and positioning of the child. In challenging cases, a small bowel follow-through (SBFT) study, ultrasound (US), and repeated examination should be considered.¹

Background

Malrotation is estimated to be present in 1 in 500 live births.² The failure of normal embryologic rotation results in a short mesenteric attachment from the duodenojejunal junction (DJJ) to the cecum, making the gut prone to twist counterclockwise around the superior mesenteric artery (SMA) and vein (SMV), resulting in midgut volvulus. Malrotation may also be accompanied by obstructive Ladd (mesenteric) bands.¹

The classic clinical presentation of malrotation in newborns is new-onset bilious emesis. However, the clinical picture is not always clear and the child may present with abdominal pain, nonbilious emesis, shock, or gastrointestinal bleeding. Intermittent midgut volvulus or partial upper intestinal tract obstruction by Ladd bands is more typical in older children and produces a broad spectrum of symptoms that are less dramatic. These children may present with nonspecific symptoms of postprandial bloating, intermittent cramping, and occasional emesis.³

Approximately 90% of cases of midgut volvulus occur within the first year of life, including up to 75% in the newborn period.^{2,4} The risk of midgut volvulus decreases with age.^{5,6} Treatment is the Ladd surgical procedure in which the mesenteric bands are lysed, the small bowel is positioned in the right hemiabdomen, the colon in the left, and an appendectomy is performed.⁷

Embryology and Anatomy

Beginning at gestational week 5 and continuing into the postnatal period, the intestines go through a series of complex development, including elongation, herniation into the umbilicus, and rotation and fixation within the peritoneal cavity (Fig. 1).

The embryonic gut begins as a straight tube. The midgut extends from the middle third of the duodenum to the distal transverse colon and is supplied by the SMA.⁸ As the midgut lengthens, it herniates through the umbilicus at 6 weeks of gestation before returning to the abdominal cavity at 9-10 weeks of gestation and becoming fixed to the retroperitoneum. During that process, the bowel continues to grow and undergoes 270° of counterclockwise rotation around the SMA and

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the SMV (Fig. 1). In normal fixation of the bowel, the mesenteric root is broad and extends from the DJJ in the left upper abdomen to the cecum in the right lower quadrant.⁸

The fixation of the cecum to the right lower quadrant continues after birth and, therefore, a mobile cecum positioned in the right upper quadrant is a common normal variation in newborns.⁹ In addition, abnormal

fixation of the small bowel may occur without affecting the normal fixation of the colon. Up to 32% of infants with malrotation have a normal position of the cecum.¹⁰ This had a major influence on imaging, as the enema study was the preferred imaging for evaluation of malrotation before 1960.^{11,12} UGIS is now the examination of choice given its increased sensitivity and ability to directly visualize upper intestinal tract obstruction.¹³⁻¹⁵

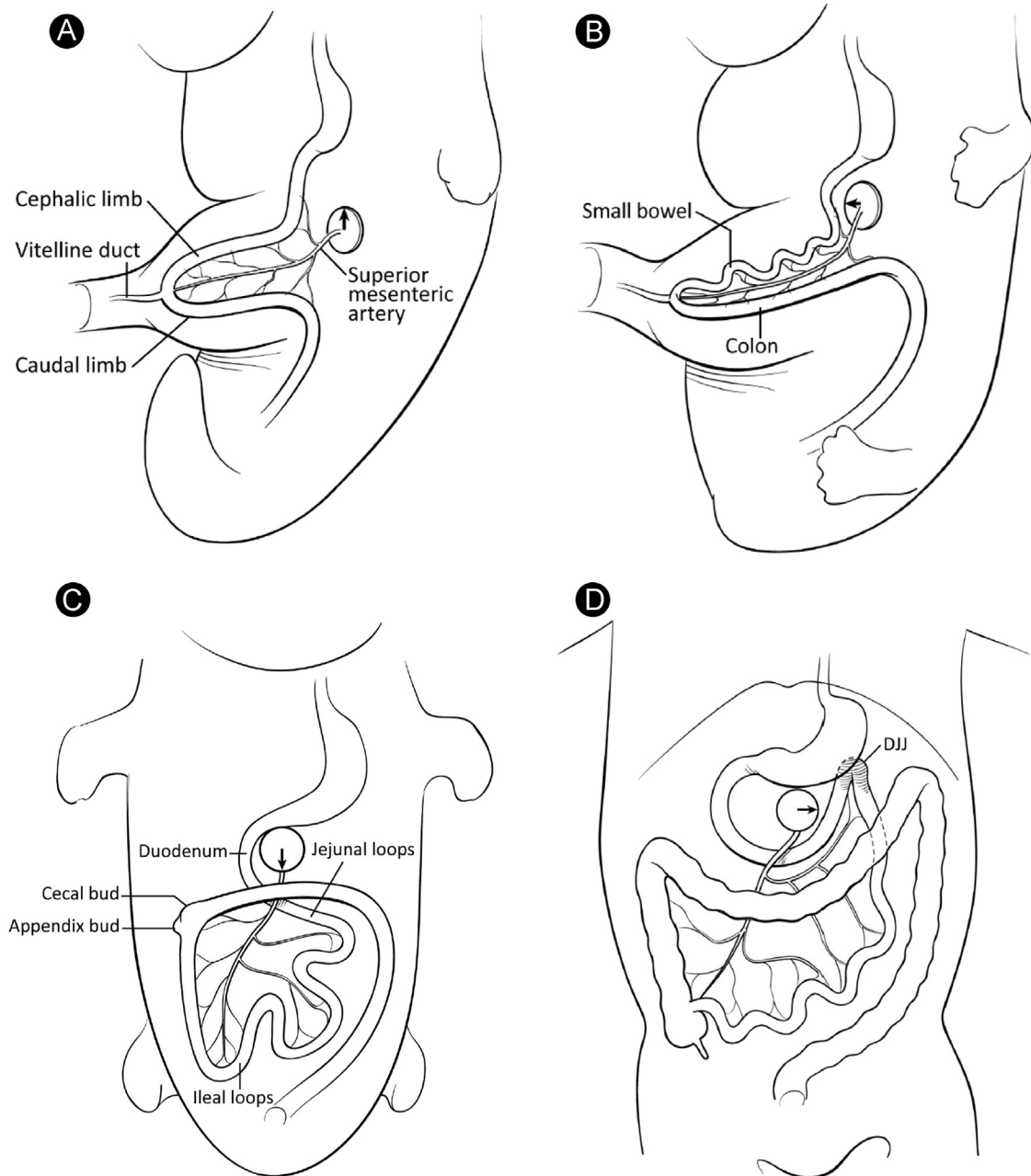


Figure 1 Normal intestinal rotation and fixation. (A) At week 6 of gestation, the bowel loops elongate and herniate in a U-shaped loop through the umbilicus (U) to the primitive extraembryonic celom. (B) During weeks 6-10 of gestation, the bowel loops (mainly the cephalic limb of the primitive gut) rotate counterclockwise 90°. (C) In week 10 of gestation, the cecum can be appreciated and the intestine reenters the abdominal cavity completing 180° of counterclockwise rotation. (D) At 12 weeks of gestation, the counterclockwise rotation (270°) of the intestine is complete. The duodenum extends retroperitoneally behind the SMA, and the duodenojejunal junction (DJJ) is fixated by the ligament of Treitz to the left of the spine at the level of the pylorus. Throughout the remainder of gestation and first few months of life, the colon continues to elongate and the cecum descends into the right lower quadrant.

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