



# Malrotation

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Malrotation is one of the most challenging disease processes encountered in the field of pediatric radiology. Proper diagnosis is critical as malrotation may progress to volvulus. Volvulus is life-threatening. Unfortunately, distinction of the normal from abnormal may be difficult. A good understanding of the radiologic findings of malrotation and volvulus is thus of paramount importance.

## Embryology

To distinguish normal from abnormal and properly diagnose malrotation, a review of the intestinal development is helpful. Development of the gut is a complex process.<sup>1-4</sup> Frazer and Robbins first described three stages of development.<sup>1</sup> In Stage 1, from week 5 to week 10, midgut herniates into the umbilical cord. In Stage 2, at weeks 10 to 11, midgut returns to the abdomen. In Stage 3, from the conclusion of stage 2 until birth, the gut undergoes fixation. While visualization of the three stages of midgut development aid in understanding, it is important to recognize that the process is a continuous one.

In the early embryo, the midgut starts as a straight tube. Blood supply is derived from the superior mesenteric artery (SMA), which, with the vitelline duct, divides the midgut into cephalad, prearterial, and caudal, postarterial portions. The prearterial segment rotates 180° counterclockwise around the axis of the SMA as it herniates into the umbilical cord, whereas the postarterial segment rotates 90° counterclockwise. As the midgut reenters the abdomen, the prearterial segment enters first, undergoing an additional 90° counterclockwise rotation. The postarterial segment follows, undergoing an additional 180° rotation. The result of these rotations is the normal C-loop configuration of the duodenum with the distal duodenum passing posterior to the SMA, a peripheral "picture frame" location of the colon with the transverse colon passing anterior to SMA, and the cecum located in the right lower quadrant. Small bowel thus courses from the duodenal–jejunal junction in upper left abdomen to

ileocecocolic valve in lower right abdomen. Fixation tethers the distal duodenum, the ascending colon, and the descending colon within the retroperitoneum and produces a long attachment of small bowel mesentery extending from upper left to lower right.<sup>5</sup> This broad root of the mesentery tethers the small bowel in place. The ligament of Treitz, fixating the duodenal–jejunal junction and marking the upper limit of the root of the mesentery, is a poorly defined extension of the right diaphragmatic crus and fibrous tissue from around the celiac artery.<sup>6</sup>

## Maldevelopment

Errors in development can occur at any point in the complex process of midgut development.<sup>3-5,7</sup> Timing of the error will affect the resultant defect and its potential morbidity. The resultant errors are termed "malrotation." Malrotation thus is not a single entity, but rather a spectrum of abnormalities. Malrotation, in itself, simply implies that the bowel is abnormal in location. Abnormal location may not be problematic; however, malrotated bowel is also malfixated. It is lack of proper fixation which predisposes the midgut to volvulus.

Anatomically, the arrangement of bowel in malrotation reflects the stage of development at which the embryological error occurred.<sup>3,5</sup> Some authors have attempted to categorize malrotation based on the anatomic arrangement of bowel in the abdomen.<sup>2,3,8,9</sup> The risk for volvulus varies dependent on the anatomical location of the bowel and the length of the midgut mesenteric root attachment. In general, shorter distance between the duodenal–jejunal junction and the cecum creates greater the risk of volvulus.

Arrest early in development yields "nonrotation" with small bowel located at left and colon at right. In this entity, both the prearterial and the postarterial segments have undergone a 90° rotation but fail to undergo the final 180° rotation. The term "nonrotation" is a misnomer because the bowel has undergone some rotation, much short of complete. "Incomplete rotation" represents a failure occurring in the final 180° of small bowel and/or colonic rotation. The resultant defect ranges from near normal to frank "nonrotation." In "reversed rotation," the postarterial segment is thought to have returned to the abdomen first, resulting in the duodenum anterior to SMA and colon posterior to SMA. Unde-

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scended cecum and congenital internal hernias also represent anomalies of midgut rotation and fixation.<sup>4</sup>

Malrotated gut is malfixated. Greater lack of fixation creates greater risk for volvulus developing. Malfixated gut with short mesenteric root may easily twist on itself, producing volvulus. With volvulus, the midgut twists around the axis of the SMA. Increasing degrees and duration of volvulus produce obstruction of lymphatics, bowel lumen, venous drainage and, finally, arterial supply. Volvulus thus threatens viability of the midgut and therefore viability of the patient. Volvulus is a true surgical emergency.

Malfixated gut attempts to fixate itself by forming peritoneal bands, often running from cecum or ascending colon to the right upper quadrant.<sup>10</sup> Peritoneal bands, also known as Ladd bands, often cross the descending duodenum, causing variable degrees of obstruction. With complete obstruction, clinical and imaging findings mimic duodenal atresia. Malrotation with an obstructing Ladd band may also be found concomitantly with duodenal atresia.

## Clinical Presentation

The classic presentation of malrotation is a newborn infant with bilious vomiting. The vomiting is bilious as the point of obstruction is distal to the ampulla of Vater. Approximately 80% of patients who present with malrotation do so in the first month of age.<sup>10-12</sup> Most do so in the first week of life. While bilious vomiting appropriately raises concern for malrotation, any congenital or acquired obstruction distal to the ampulla of Vater may cause bilious vomiting. Moreover, most infants with “bilious vomiting” prove not to have an anatomic obstruction.<sup>13</sup> Given its potential deleterious consequences, however, exclusion of malrotation is usually pursued.

Malrotation may present at any age; however, presentations beyond the neonatal period are usually less specific and are often cryptic.<sup>14</sup> Patients may present with nonbilious vomiting, intermittent or acute abdominal pain, diarrhea, constipation, or hematochezia. Presentation with an “acute abdomen” or shock is rare and portends a poor prognosis as gut ischemia is usually present.<sup>12</sup> Symptoms may occasionally be insidious, with chronic pain, intermittent vomiting, or failure to thrive. Not infrequently, the diagnosis of malrotation is made incidentally in an older child or adult who is seemingly asymptomatic relative to the defect.<sup>15,16</sup>

Patients with congenital defects of abdominal compartmentalization—congenital diaphragmatic hernia, gastroschisis, omphalocele—have malrotation.<sup>17</sup> The risk for volvulus is low in these patients, in part due to fixation of the gut by adhesions when the abdominal defect is repaired. An overwhelming majority of infants with heterotaxy—asplenia, polysplenia—have malrotation.<sup>18-21</sup> Upper gastrointestinal (GI) studies are performed for confirmation. Some controversy exists as to the risk for volvulus in these patients. At the author’s institution, a patient with asplenia developed a fatal volvulus while recovering from heart surgery. As many patients with heterotaxy have coexistent heart disease, often severe, particularly with asplenia, timing of workup for malrotation and treatment (ie, Ladd procedure) must be tem-

pered by the need for cardiac surgery and subsequent recovery. Evaluation of the child with heterotaxy by upper GI may be challenging when the stomach is right-sided. In such cases, the normal (not malrotated) duodenum should be the mirror image of normal, with the second portion descending on the left and crossing to the right to ascend to the duodenal–jejunal junction.

## Radiography

Imaging evaluation of the infant or child with suspected malrotation begins with radiographs, particularly in the infant. Radiographs aid in excluding other diagnoses, namely more distal obstructions in the neonate.

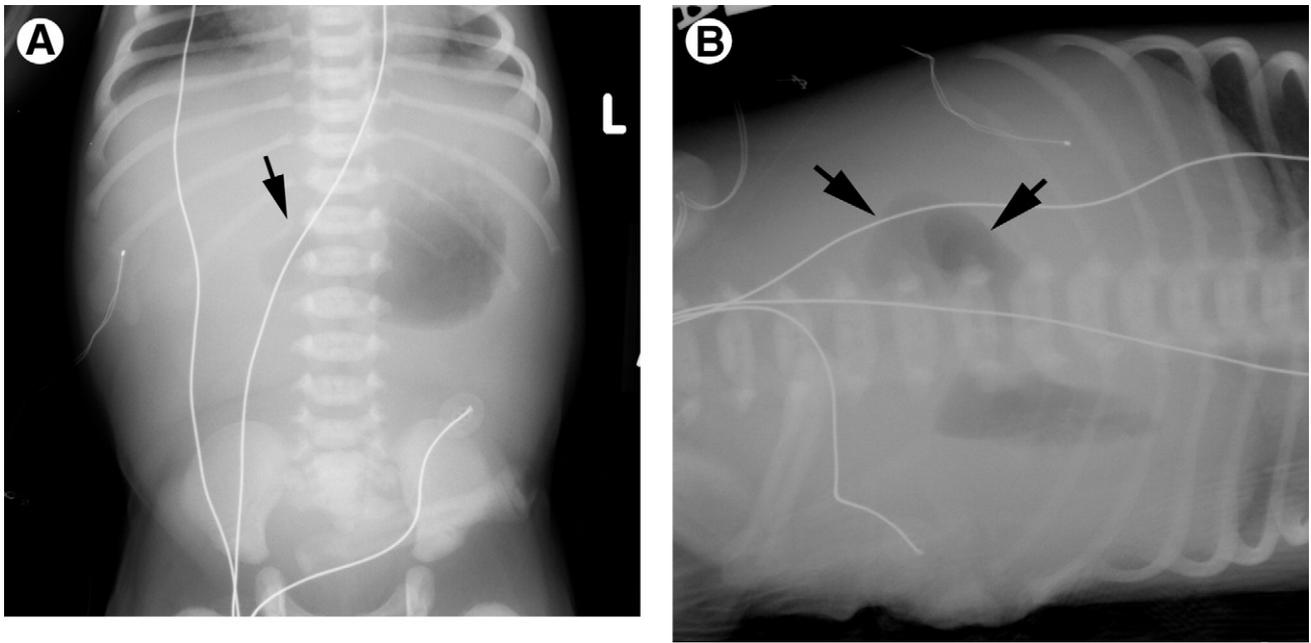
A variety of radiographic appearances may be seen with malrotation, but, unfortunately, the most common radiographic appearance of malrotation and malrotation with volvulus is that of a “normal bowel gas pattern.” A normal radiograph therefore should not preclude additional workup as it does not exclude malrotation.<sup>10</sup> Radiographic findings which may be seen with malrotation include malposition of the bowel (small bowel all to the right, colon all to the left), lack of bowel gas distal to the duodenum (Fig. 1), disproportionate dilation of the duodenal bulb (“double bubble”), a gasless abdomen (nonspecific), mass effect (nonspecific), a “whirled” appearance of bowel in the mid-abdomen (rare), or thick-walled, “tubular”-appearing loops with fold thickening or thumbprinting. The latter findings are grave prognostic signs indicative of gut ischemia. Fortunately, such cases are rare. Intramural gas, free intraperitoneal gas, and diffuse gaseous distension of bowel are also poor prognostic signs indicative of ischemic gut.<sup>22,23</sup> Such signs are also nonspecific and may not readily suggest the proper diagnosis.

## Upper GI

A fluoroscopic upper GI study is the preferred method of diagnosing malrotation or excluding the diagnosis. For greatest accuracy, the upper GI examination must be performed with careful attention to the anatomy of the duodenum. As a rule, unless previously documented, every upper GI examination performed in a child should include documentation of duodenal anatomy. Barium is utilized, unless the infant is critically ill or perforation is suspected. In such cases, nonionic water-soluble contrast is utilized.

The distal duodenum should extend upward and to the left. The duodenal–jejunal junction (“ligament of Treitz”) is normally located to the left of the left spinal pedicle and at approximately the same level as the pylorus. As the second, third, and fourth portions of the duodenum are fixated in the retroperitoneum, they are located posteriorly with the abdomen. In a true lateral projection, the second and fourth portions overlap posteriorly, near the spine (Fig. 2).<sup>4,24,25</sup> The duodenal–jejunal junction is thus also located posteriorly.

Care must be taken to fluoroscopically observe the first passage of barium through the duodenum. Barium reaching the proximal jejunum may obscure the duodenum, preventing documentation of the duodenal anatomy. Once barium



**Figure 1** Newborn boy with malrotation and volvulus. The infant developed bilious vomiting shortly after birth. (A) Anteroposterior radiograph shows air only in the stomach and duodenum (arrow). (B) On a left-side down decubitus view, the duodenum (arrows) is better seen. The duodenum is mildly prominent but not dilated to the extent usually seen with duodenal atresia. An air–fluid level is seen dependently in the stomach. Ten centimeters of infarcted proximal small bowel were resected at surgery.

reaches the duodenal–jejunal junction, turning the patient into a lateral position allows for confirmation of the posterior position of the duodenal sweep. Turning the patient from anteroposterior to lateral under direct fluoroscopic observation aids in identifying the distal duodenum in the lateral projection. As the normally located fourth portion of the duodenum is partially obscured by the second portion, slight obliquity may help delineate the anatomy.

Sometimes barium is slow to traverse the duodenum, hanging up in the second portion. In this case, turning the patient briefly to the left elicits passage of barium into the distal duodenum. Once barium is seen to be passing distally, the patient is turned back supine and the duodenal course will be nicely visualized. An added benefit of this maneuver is dumping of barium from the gastric antrum back into the gastric fundus. The distal duodenum thus overlies a gas-filled, barium-coated, “double-contrast” antrum, allowing for better visualization. Once the duodenum is documented in the anteroposterior projection, the patient is turned to either lateral projection to document the posterior location of the distal duodenum.

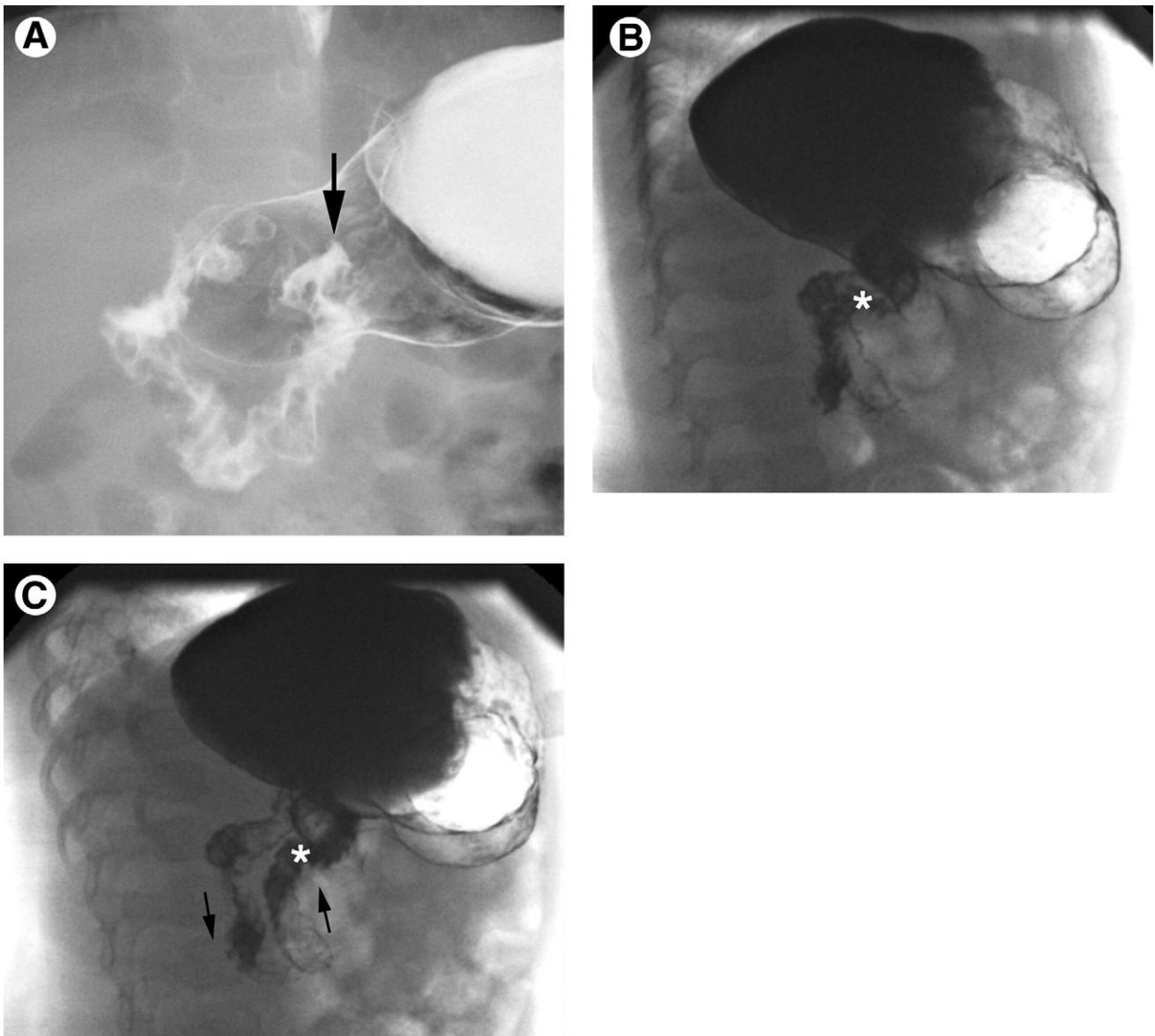
In cases of malrotation, the distal duodenum fails to extend to the left and fails to ascend. In the lateral projection, the malrotated distal duodenum often projects anteriorly, rather than maintaining the normal posterior course (Fig. 3). If the distal duodenum stays to the right of midline and is properly documented on the first pass of barium, the diagnosis of malrotation is easily made.

Although most cases will be unequivocally normal or abnormal, there is considerable variation in the course of a normal duodenum and subtle cases of malrotation may not

differ considerably from variants of normal.<sup>4,7,26,27</sup> Some authors state that up to 15% of upper GI examinations will potentially demonstrate equivocal positioning of the duodenum.<sup>7</sup> The duodenum may demonstrate moderate redundancy in its course. Slight variation in height of the duodenal–jejunal junction may also be seen. In these cases, demonstration of the normal posterior location of the distal duodenum offers confirmatory evidence of normalcy. If the duodenum is excessively redundant, fails to extend to at least the left pedicle of the spine on a well-positioned anteroposterior view, or fails to ascend to the duodenal–jejunal junction, then malrotation is likely present.<sup>4,7,26,27</sup> In such cases, the lateral view is usually confirmatory as the distal duodenum fails to maintain a normal posterior course.<sup>4,7,24</sup>

The classic upper GI finding in volvulus is a “corkscrew” downward path of the distal duodenum and proximal jejunum in the mid-abdomen. Typically, the distal duodenum is beaked entering the corkscrew with a partial obstruction (Fig. 4). Although the duodenum may extend to the left of midline in some of these cases, it takes a very characteristic downward turn into the volvulus. In some patients, the obstruction is complete with the distal duodenum having a beaked, downward point. In a newborn, care must be taken not to mistake the findings for duodenal atresia. In duodenal atresia, the duodenum is dilated and round in contour. In acute malrotation with volvulus, the duodenum is relatively normal in caliber and not round in contour but pointed or beaked at the point of obstruction.

The most common pitfall of an upper GI examination is missing the first pass of barium through the duodenum. Rotation on the anteroposterior view may distort the position of

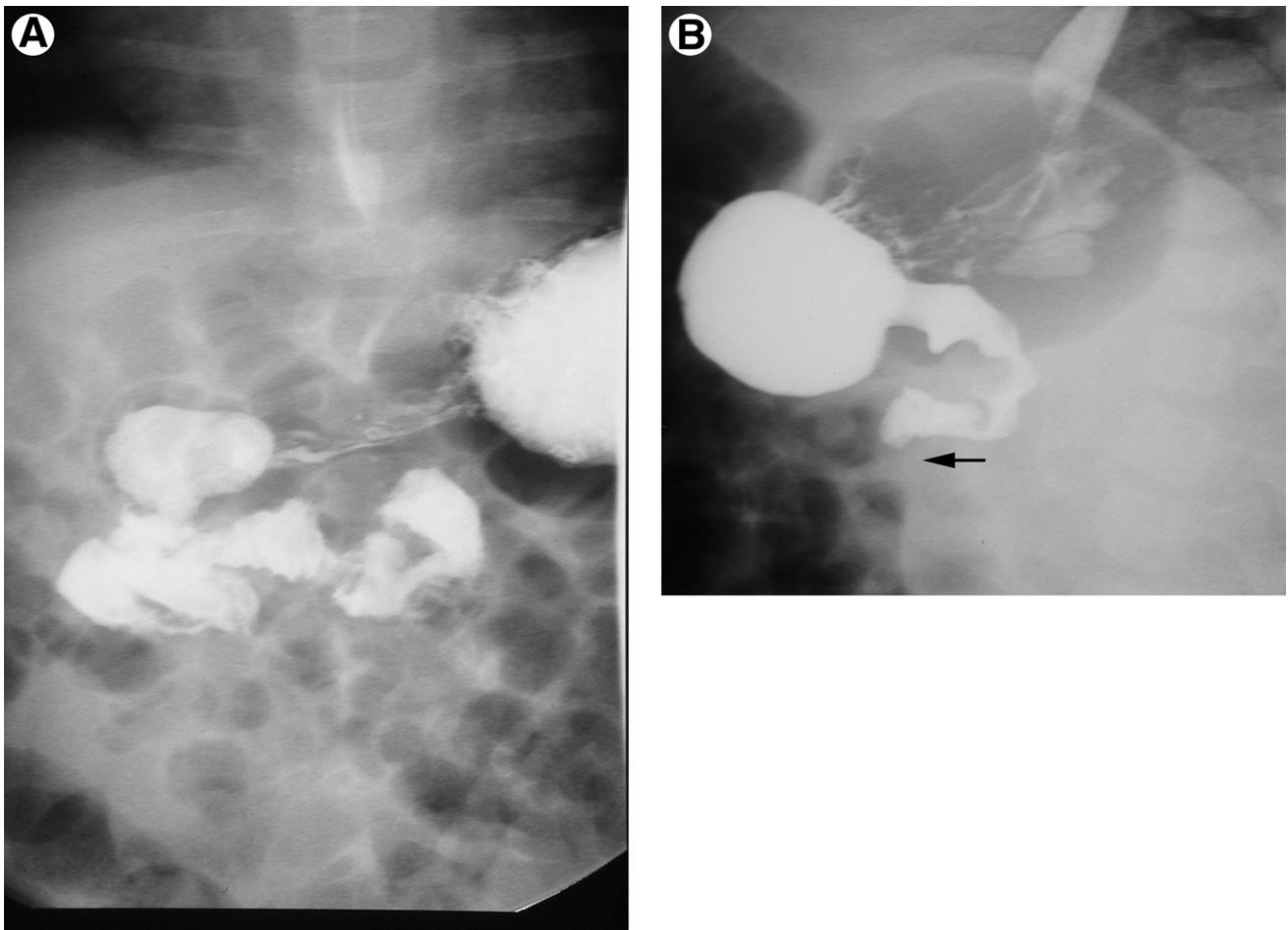


**Figure 2** A 10-month-old girl with a normal upper GI study. (A) Normal appearance of the duodenum in the anteroposterior projection. The distal duodenum ascends to the level of the pylorus and the duodenal-jejunal junction (arrow) is to the left of the left spinal pedicle. (B) In a true lateral projection, the second and fourth portions of the duodenum overlie each other at posterior. \*, duodenal-jejunal junction. (C) With slight obliquity the duodenal course is better seen. ↓, second portion of duodenum; ↑, = fourth portion of duodenum; \*, duodenal-jejunal junction.

the duodenal-jejunal junction. Midline positioning of the spinous processes and symmetry of the ribs are helpful in confirming proper positioning. An inadequate quantity of barium may make it difficult to appreciate duodenal anatomy. Conversely, too much barium may lead to uncontrolled rapid passage of barium through the duodenum or simply obscure visualization of the duodenum. The duodenal-jejunal junction is normally somewhat mobile in infants. Factors such as prior surgery (ie, liver transplantation),<sup>28</sup> intestinal or gastric dilation,<sup>29</sup> and enteric tubes<sup>30</sup> may cause displacement of the duodenal-jejunal junction, mimicking malrotation (Fig. 5). In young infants, the normal distal duodenum tends to be slightly low in position,<sup>7</sup> often accentuated by a transverse orientation of the stomach. In such patients, documen-

tation of the posterior location of the distal duodenum is reassuring. Similarly, in “duodenum inversum” a mildly redundant duodenum ascends to the right of midline before crossing midline.<sup>26</sup> Again, documentation of posterior location of the distal duodenum is reassuring.

If an initial upper GI examination is equivocal, further investigation is warranted.<sup>4,7,26</sup> Documentation of cecal position is valuable. This is discussed below. Unfortunately, documentation of normal cecal position does not exclude malrotation. A repeat upper GI with greater attention to documentation of the duodenal position may prove successful. Placement of an enteric tube aids in control of barium. Advancement of the tube past the pylorus is of particular aid in achieving good duodenal opacification. Obtaining the as-



**Figure 3** A 4-month-old girl with malrotation, without volvulus. The child presented with wheezing and an upper GI study was performed to assess for reflux. (A) In the anteroposterior projection, overly redundant duodenum is seen to the right of midline. Bowel extends to the left; however, this is jejunum, not the distal duodenum. (B) An earlier image in the lateral projection shows an anomalous anterior course of distal duodenum (arrow).

sistance of an experienced pediatric radiologist may be invaluable.

## Cecal Position

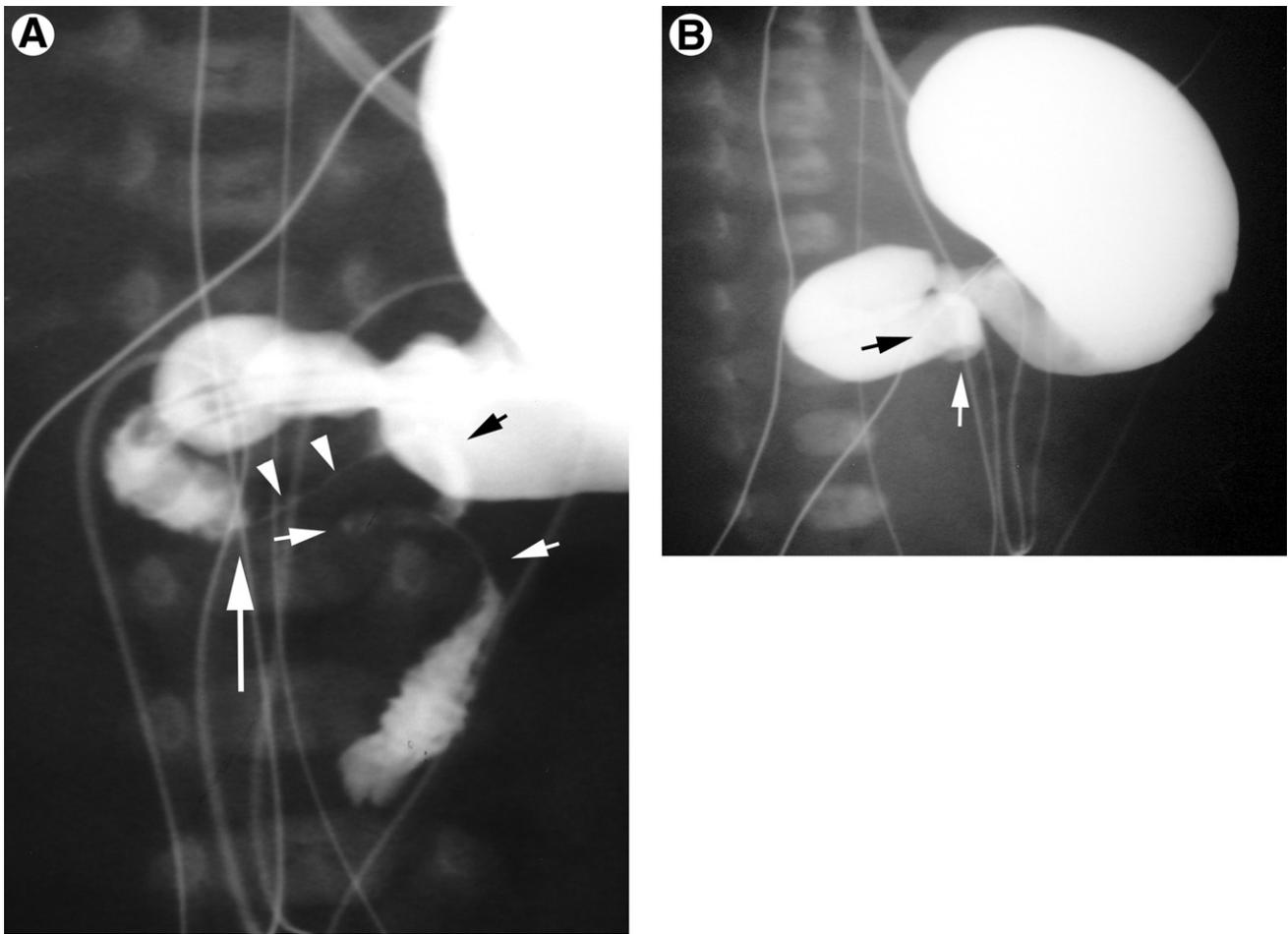
The normally rotated cecum is in the right lower quadrant of the abdomen, often descending into the right hemipelvis. The malrotated cecum may be found at varied locations throughout the abdomen.

Up to 20% of patients with malrotation have a normally located cecum.<sup>16,27,31</sup> A contrast enema is therefore not the preferred method of diagnosing malrotation as it will fail to make the diagnosis in too many patients. In young infants, the cecum is often slightly high in position; thus, the contrast enema may lead to confusion or over diagnosis of malrotation.<sup>26</sup> Up to 15% of patients of all ages will have a mobile cecum as a normal variant, potentially mimicking malrotation.<sup>3,5,25</sup> The other disadvantage of a contrast enema in the setting of suspected malrotation is that it is much less effective at demonstrating volvulus than an upper GI examination. The diagnosis of volvulus indicates a need for urgent surgical management, as opposed to malrotation, in itself,

which approached with semielective surgery in the absence of volvulus. Upper GI findings of volvulus may thus lead to greater expediency of treatment than the enema findings in the same patient. In a patient presenting acutely, enema findings of malrotation should prompt upper GI evaluation for volvulus, unless volvulus is shown by enema and/or the expedient surgery is already planned.

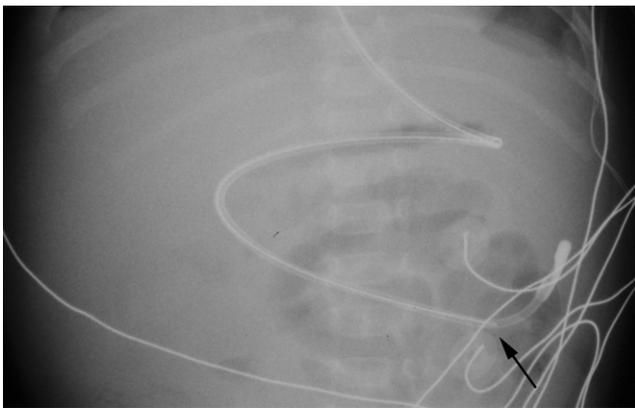
The diagnosis of malrotation may also be made on an enema study performed for another reason. More importantly, documentation of cecal position can be performed as an adjunct to upper GI evaluation of the duodenum.<sup>7,8</sup> In patients with an equivocal duodenal course, or in whom the duodenal course is suspected to be a variant of normal, documentation of cecal position can be performed. An abnormal cecal position supports the diagnosis of malrotation. A normal cecal position supports the diagnosis of normalcy. In general, if there is any deviation from the norm of the course of the duodenum, documentation of cecal position should be performed.

After an upper GI, the barium can be followed through the small bowel with serial images until it reaches the cecum. Additional fluoroscopy may be required to document the



**Figure 4** One-day-old boy with malrotation and volvulus. (A) In the anteroposterior projection there is a partial obstruction of the distal duodenum (large arrow). The duodenum distal to the point of obstruction is markedly narrow (arrowheads) and then takes a “corkscrew” downward course (small arrows). (B) An earlier image in the lateral projection shows an anomalous anterior course of the distal duodenum (black arrow). The duodenum appears beaked at the point of obstruction (white arrow). At surgery, the midgut was ischemic, but no resection was required.

terminal ileum and cecum. While this approach works in most patients, it requires time for the barium to progress to



**Figure 5** Three-month-old boy with normal anatomy. This abdominal radiograph was obtained to assess feeding tube placement. A wire is present in the feeding tube, stiffening it. As a result, the distal duodenum appears low in position. Arrow, tip of wire in feeding tube.

the cecum. An inadequate quantity of barium or poor timing of images may lead to a failure to visualize cecal position. Alternatively, an enema may be performed. The advantage of performing an enema is that cecal position is determined quickly. In infants and young children, water-soluble contrast media can be used for the enema, allowing distinction from barium administered orally for the upper GI. The decision of whether to perform a small bowel follow-through or enema depends on several factors, including the upper GI findings, the practicality of performing the small bowel follow through based on the time of day and available personnel, and personal/institutional preference. Most cases of volvulus will not be equivocal on upper GI; however, if volvulus is a consideration, waiting for barium to traverse the small bowel is not an acceptable option.

### Cross-Sectional Imaging

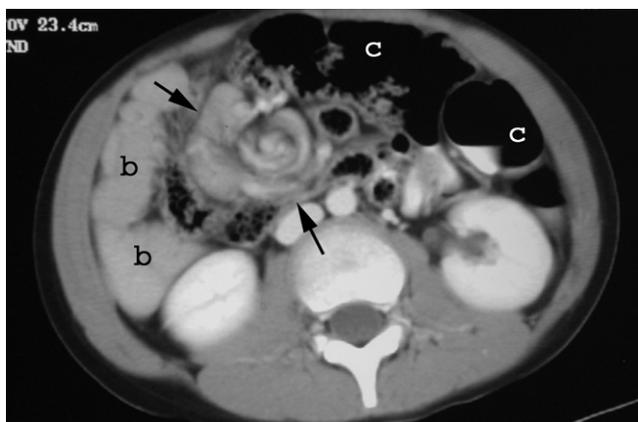
Increasing utilization of cross-sectional imaging modalities has led to the increased incidence of the diagnosis of malrotation by modalities other than fluoroscopic study. Cross-

sectional imaging findings of malrotation include dilation of the proximal duodenum, absence of the crossing duodenum posterior to the SMA, malposition of small and/or large bowel, and inversion of the normal SMA/superior mesenteric vein (SMV) relationship.<sup>4,32-34</sup>

Normally, the SMA is to the left of the SMV. If this relationship is reversed, then malrotation is suspected.<sup>32-34</sup> Unfortunately, this finding is neither fully sensitive nor specific; however, unless another reason for inversion is present (ie, a mass, prior surgery), then an upper GI should be performed to assess for malrotation. Conversely, demonstration of a normal oriented SMA and SMV does not exclude malrotation and does not preclude performing the upper GI if malrotation is a consideration.

When malrotation is suggested by a cross-sectional imaging study, an upper GI examination should be performed for confirmation. An exception to this rule occurs when there is evidence of volvulus or gut ischemia, in which case expedient surgery is preferred to further imaging. With volvulus, a whorled mass is seen in the mid-abdomen with vasculature and bowel wound around the axis of the SMA (Fig. 6).<sup>35,36</sup> When the volvulus has advanced to the point of vascular occlusion, computed tomography may demonstrate a striking lack of midgut enhancement (Fig. 7).<sup>37,38</sup> The volvulus, itself, may be obscured due to the lack of enhancement and abundant fluid. Enhancement of foregut and hindgut, with different vascular supply, is preserved. The findings of midgut ischemia portend a poor outcome due to gut ischemia; however, survivors are reported.

Cross-sectional imaging is not the method of choice for diagnosing malrotation. Nevertheless, increasing reliance of clinical physicians on imaging and the oftentimes confusing presentation of children with malrotation means that many cases will be initially detected by ultrasound, computed tomography, or even magnetic resonance imaging. The radiologist must be prepared and know the findings of malrotation and malrotation with volvulus on these modalities.



**Figure 6** Six-year-old girl with malrotation and volvulus. Axial computed tomography image shows small bowel and vasculature has a whorled appearance in the right mid-abdomen (arrows), consistent with volvulus. Small bowel (b) is at right and colon (c) is at left. At surgery, no ischemia was evident.



**Figure 7** Five-year-old boy with malrotation and volvulus. The superior mesenteric artery (arrow) is deviated to the right and appeared truncated beyond this point. There is a striking lack of enhancement of bowel and mesenteric vasculature. Bowel is distended with fluid and abundant free intraperitoneal fluid is present. Enhancement of the duodenum (d) is preserved. At surgery, most of the ileum and proximal half of the colon were infarcted requiring resection. The proximal midgut was ischemic but was preserved. The child has manifestations of short-gut syndrome.

## Conclusion

If not promptly diagnosed and treated, malrotation with volvulus may be fatal to the affected child. Unfortunately, and despite the best efforts of frontline physicians (pediatricians and emergency room doctors), radiologists, and pediatric surgeons, fatal cases of malrotation with volvulus are occasionally seen. Fortunately, an overwhelming majority of affected children are promptly diagnosed and treated and do survive without significant sequelae. The role of the radiologist is paramount to this good outcome. There are few other diagnoses in medicine in which the prepared and skilled radiologist can have such a profound bearing on patient outcome.

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