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

of failed extracelomic rotation. It is most commonly characterized by a duodenal-jejunal junction (DJJ) located in the right upper quadrant and a cecum located in the middle to upper abdomen, which is fixed in place by adhesive bands to the gallbladder, duodenum, and right-sided abdominal wall (Ladd bands). Most importantly, classic malrotation results in a narrowed mesenteric base, which predisposes the child to potentially fatal midgut volvulus (Fig. 1). Less commonly, the bowel makes a 90° turn clockwise, rather than counterclockwise, resulting in reverse rotation, in which the duodenum lies anterior to the SMA and the colon lies posteriorly, producing a retroarterial tunnel, which may be associated with partial mesenteric arterial, venous, and lymphatic obstruction. If the mesoderm does not fuse to the retroperitoneum during the fourth and fifth months of gestation, paraduodenal or paracolic hernias may form.

CLINICAL PRESENTATION

The reported incidence of rotation abnormalities varies widely, depending largely on how they are defined. Autopsy series suggest an incidence of 0.5%. A population-based study estimated an incidence of approximately 15 per 1 million in children less than 1 year old and 10 per 1 million in children aged 1 to 2 years,[2] with a decreasing incidence after that. Associated congenital abnormalities are found in approximately 30% to 60% of cases and may include intestinal atresia or web (the most common associated anomaly), Meckel diverticulum, intussusception, Hirschsprung disease, mesenteric cyst, and anomalies of the extrahepatic biliary system. Congenital diaphragmatic hernia and abdominal wall defects such as omphalocele and gastroschisis are typically associated with an intestinal rotation abnormality, usually nonrotation, because the intestine is in an abnormal location during the time it should be undergoing rotation. Many children with intestinal motility disorders have associated rotation abnormalities. In addition, rotation abnormalities are commonly associated with congenital heart disease, often in the context of one of the various heterotaxia syndromes (HS).

The classic presentation of malrotation is bilious vomiting in a newborn infant, and every child with bilious vomiting should be assumed to have malrotation until proved otherwise. Bilious vomiting may occur for 3 reasons (which are not necessarily mutually exclusive): obstructive compression of the duodenum by Ladd bands, an associated duodenal atresia or web, or (more ominously) midgut volvulus. Midgut volvulus occurs around the narrow-based mesenteric pedicle, causing twisting of the SMA and superior mesenteric vein (SMV). The abdomen is not distended initially because the obstruction is very proximal. As the intestine becomes ischemic, the patient may develop hematochezia, irritability, pain, abdominal distension, and peritonitis. Ultimately, abdominal wall erythema, septic shock, and death may occur as the midgut becomes necrotic. Many of these findings are similar to those found in neonates with necrotizing enterocolitis; the presence of bilious vomiting or nasogastric tube aspirates should suggest the possibility of midgut volvulus as part of the differential diagnosis. In older children, chronic, partial, or intermittent volvulus may present with crampy abdominal pain, intermittent vomiting, diarrhea, occult gastrointestinal bleeding, protein-losing enteropathy caused by lymphatic obstruction, failure to thrive, or malnutrition. Distension is frequently present, although it may be intermittent.

Although volvulus associated with malrotation represents the condition's most acute, dramatic, and urgent presentation, duodenal obstruction may also be caused by Ladd (or other congenital) bands, even without midgut volvulus. In neonates, these bands can cause incomplete obstruction with feeding intolerance with or without bilious vomiting. Older children more commonly present with intermittently colicky abdominal pain associated with bilious emesis. An internal hernia associated with

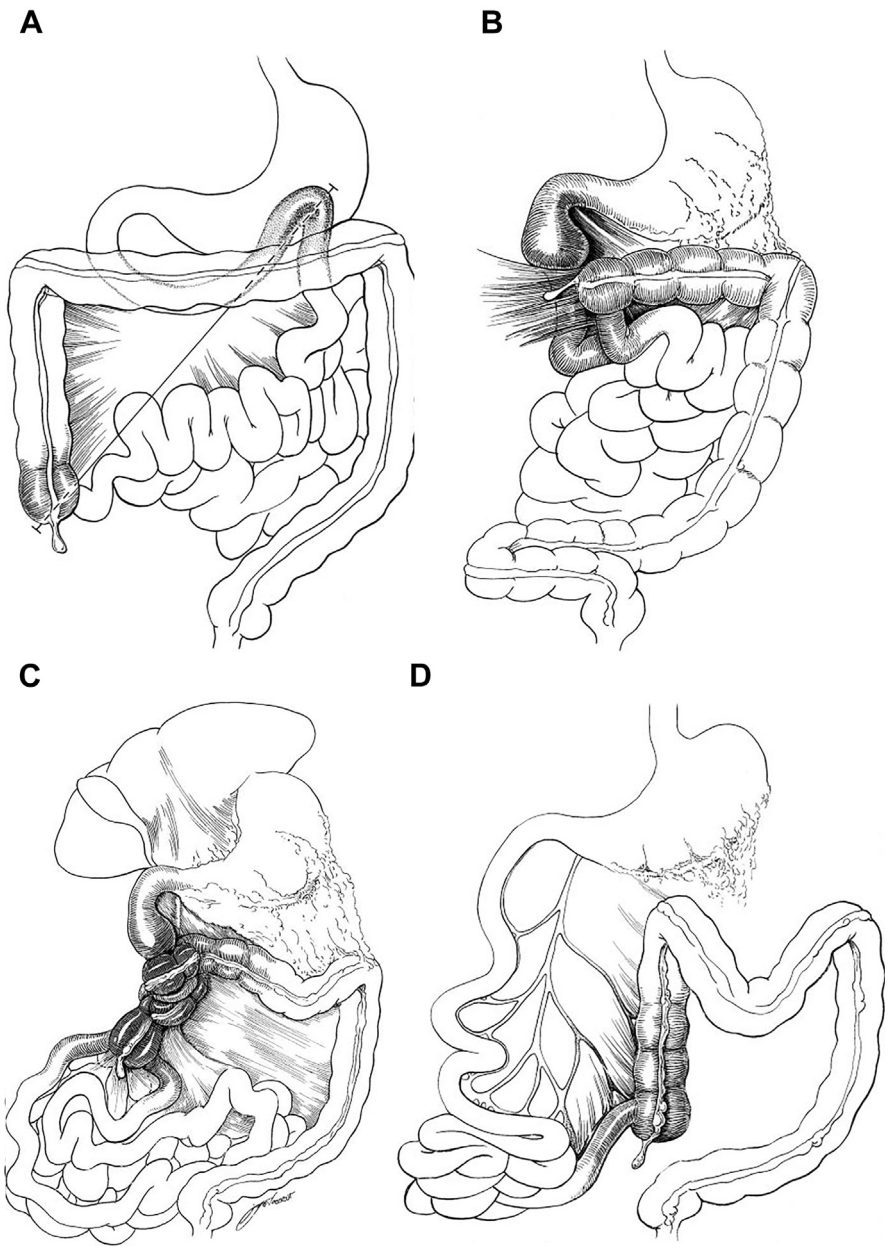

Fig. 1. (*A*) Normal intestinal rotation, (*B*) malrotation without volvulus, (*C*) malrotation with volvulus, (*D*) nonrotation.

inappropriate intestinal fixation may present in a similar fashion. Chronically symptomatic patients commonly present with recurrent respiratory symptoms, including asthma and aspiration. In addition, it is important to remember that children with a rotation abnormality may also have duodenal or jejunal atresia or web, which may be missed if the surgeon is not specifically looking for it.

RADIOLOGIC DIAGNOSIS

Prenatal diagnosis of isolated rotational abnormality is very uncommon, but fetal ultrasonography may show the sequelae of prenatal midgut volvulus, such as bowel dilatation, meconium peritonitis, and/or fetal ascites.

Postnatally, most children with vomiting have plain abdominal radiography, which is nonspecific for the diagnosis of rotation abnormalities. Proximal obstruction caused by Ladd bands, incomplete volvulus, or associated duodenal atresia or web may present with a double bubble and a paucity of distal air. Infants with established intestinal ischemia may have pneumatosis intestinalis, which may lead to confusion with a diagnosis of necrotizing enterocolitis. Although unusual, a pattern of distal bowel obstruction consisting of multiple dilated bowel loops with air-fluid levels may be seen. Most importantly, children with rotation abnormalities, including malrotation with volvulus, may initially present with a normal bowel gas pattern (**Fig. 2**).

The current gold standard for the diagnosis of a rotation abnormality is upper gastrointestinal contrast radiography (UGI) to evaluate the position of the DJJ, which should be located to the left of the vertebral body at the level of the inferior margin of the duodenal bulb on an anteroposterior projection and must travel posteriorly on the lateral projection.[3] If the DJJ does not show these radiographic characteristics, a diagnosis of rotation abnormality should be entertained. However, conditions such as splenomegaly, renal or retroperitoneal tumors, gastric overdistension, liver transplant, small bowel obstruction, and scoliosis may cause the DJJ to be medially or inferiorly displaced. The group at the University of Arkansas attempted to define risk of malrotation, ischemic volvulus, and internal hernia in a group of consecutive patients undergoing operation for rotation abnormalities based on the positioning of the DJJ on initial UGI series.[4] The rotation abnormality was described as typical if the DJJ was positioned to the right of the midline or if it was absent, and atypical if the DJJ was at or to the left of the midline and the DJJ was low-lying. At the time of operation, volvulus had occurred in 12 of 75 typical patients versus 2 of 101 atypical patients. Internal hernias were also more common in typical than in atypical patients. Moreover, this group found that 11% to 13% of atypical patients had persistent postoperative symptoms compared with 0% of typical patients. Given the cited postoperative bowel obstruction rate following Ladd procedure (8%–12%) and the fairly high incidence of continued symptoms, the investigators advocated careful discussion in patients with atypical radiological findings. In this group a laparoscopic approach might be particularly useful (discussed later).

If the UGI is confusing or equivocal, a small bowel follow-through or a contrast enema to visualize the cecum should also be done. A short distance between the DJJ and the cecum strongly suggests the presence of malrotation with a narrow-based mesentery, and therefore a risk of midgut volvulus. However, there is a wide range of variability in normal cecal positioning and fixation, especially in neonates, and therefore a cecum located in the right lower quadrant cannot definitively rule out malrotation, and a cecum located in the right upper quadrant or epigastrium is not diagnostic of malrotation. Examples of contrast studies in the diagnosis of intestinal rotation abnormalities are shown in **Fig. 3**.

More recently, identification of an abnormal orientation of the SMA and SMV on ultrasonography has been advocated as a noninvasive way to screen for malrotation.[5] In addition, the presence of a whirlpool sign on Doppler ultrasonography has been correlated with the presence of midgut volvulus. However, the sensitivity and specificity of ultrasonography are not sufficient for it to replace UGI for a definitive diagnosis. In children with peritonitis who are too unstable to have UGI, in which midgut volvulus is part

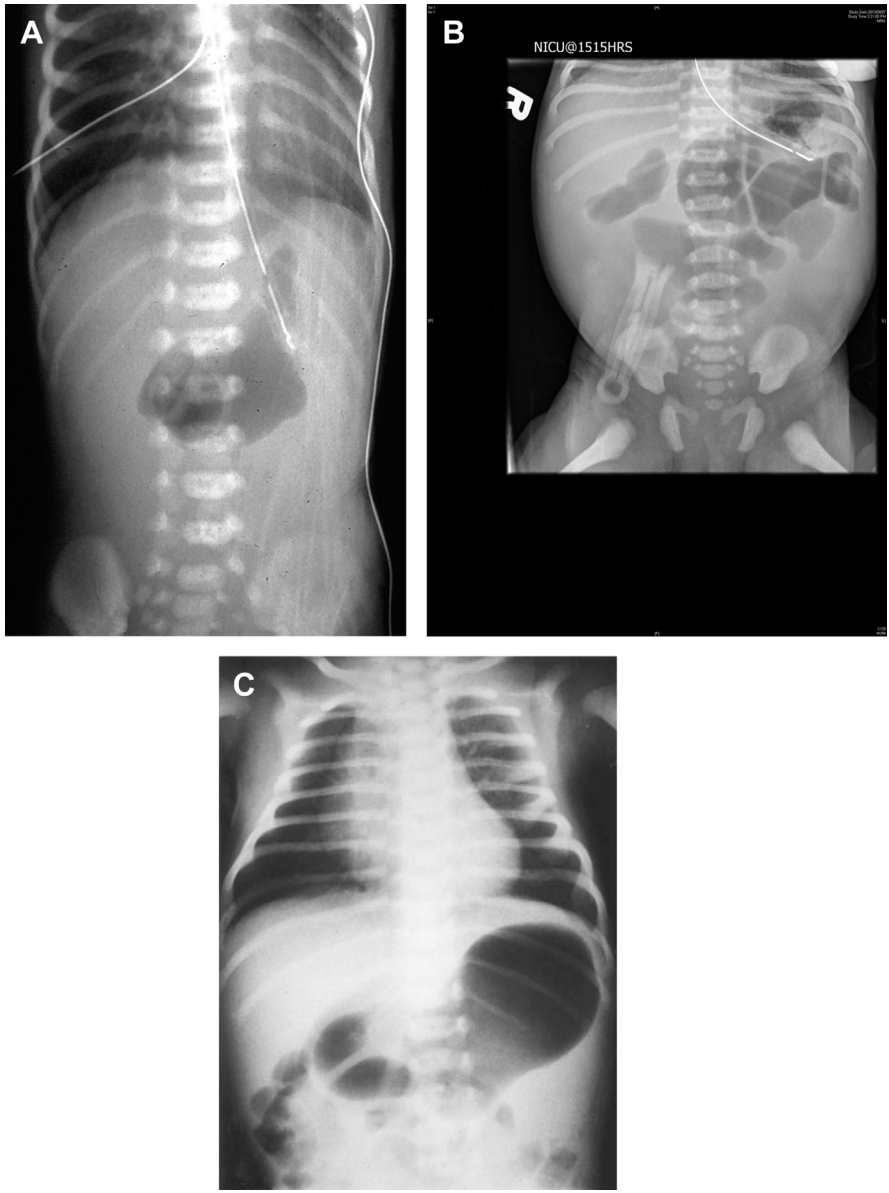

Fig. 2. The varying appearances of malrotation on plain film. (*A*) Gasless abdomen with dilated gastric bubble; (*B*) dilated small bowel suggestive of a distal obstruction; (*C*) normal film with slightly dilated duodenum.

of the differential diagnosis, a normal SMA/SMV orientation and absence of a whirlpool sign may provide reassurance that the diagnosis is not midgut volvulus.

Although radiographic techniques are important diagnostic aids in the nonacute setting, infants who present in extremis with a history of bilious emesis and findings of peritonitis should be aggressively resuscitated, decompressed via nasogastric

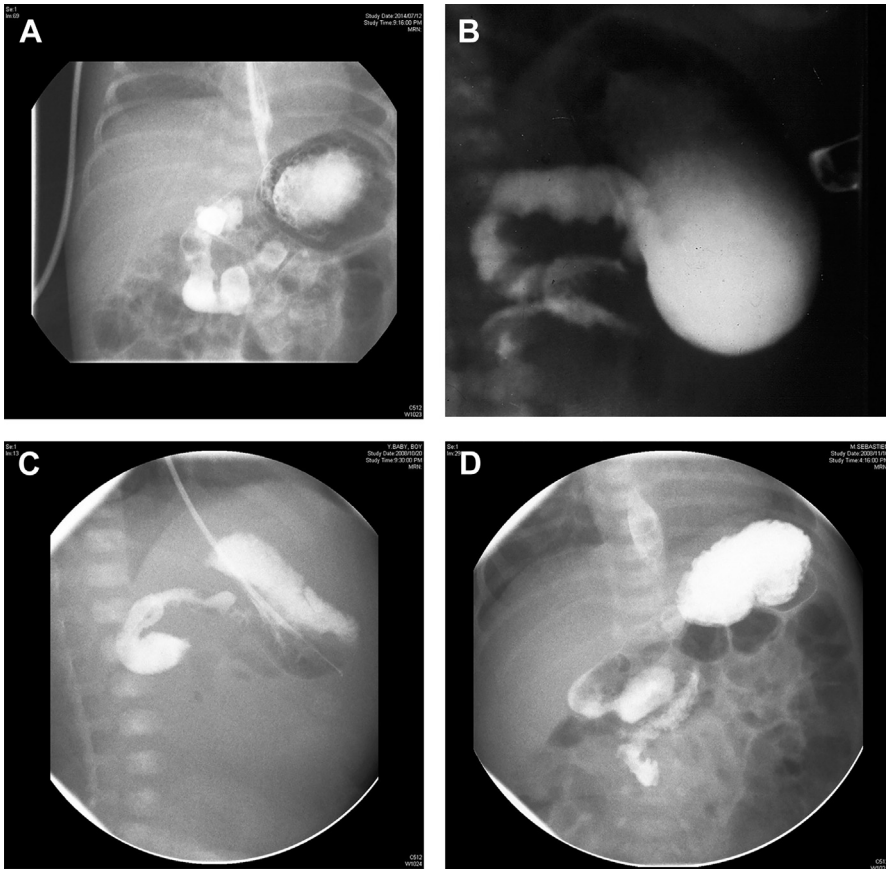

Fig. 3. UGI series with small bowel follow-through. (*A*) Normal contrast study showing the duodenal C loop crossing the midline. (*B*) Lateral view of malrotation, showing corkscrew appearance of jejunum. (*C*) Lateral view suggestive of duodenal obstruction secondary to Ladd bands or volvulus. (*D*) False-positive study; duodenal-jejunal junction is pushed rightward by large multicystic kidney.

tube, and taken for an emergent exploratory laparotomy even without a radiological diagnosis.

SURGICAL TECHNIQUE

The operative correction of malrotation was initially described by William Ladd, and has changed little since then. Most surgeons begin an open approach to a Ladd procedure via a transverse supraumbilical incision with the patient placed in the supine position. In neonates a circumumbilical omega incision affords the same access to the midgut and mesentery with a considerable cosmetic benefit.[6] On entering the abdomen, rotation and fixation of the bowel are assessed by delivering the entire midgut into the operative field. The presence of chylous ascites may indicate chronic lymphatic obstruction caused by partial midgut volvulus. If volvulus is encountered, the involved loops of bowel are gently detorsed by counterclockwise rotation until the mesentery is unfurled. At this time, the bowel is assessed for viability. Reperfusion

and delineating of viable from nonviable bowel may take several minutes. During this period, the bowel should be covered with a warm, damp laparotomy pad or towel to help prevent evaporative losses and vasoconstriction. Following this, a Ladd procedure is performed. This operation consists of 4 discreet steps, the goal of which is to place the bowel into a position of nonrotation, with the small bowel on the right side of the abdomen and the colon on the left: (1) division of any abnormal bands (Ladd bands) fixing the bowel to the right upper quadrant retroperitoneal or intra-abdominal structures; (2) mobilization and rotation of the colon toward the left, taking care not to injure the colonic mesentery in the process, so that the entire colon sits on the left side of the abdomen; (3) mobilization and straightening of the duodenum so that it heads inferiorly and all the small bowel sits on the right side of the abdomen; (4) broadening of the base of the mesentery by dividing the congenital bands along the SMA and SMV. If the child has presented with duodenal obstruction, duodenal patency should be tested by milking gastric contents from the proximal to the distal duodenum to rule out associated duodenal atresia or web. Most surgeons also perform an appendectomy, either by excision or using an inversion technique (**Fig. 4**).

Before closing the abdomen, small bowel viability should be reassessed. Any frankly necrotic sections should be resected. If the rest of the bowel is completely viable, a primary anastomosis can be performed, and, if not, stomas should be created. If there are large sections of bowel in which viability is still unclear, resection should not be done, and a second-look laparotomy should be planned for 24 to 48 hours later. More ethically problematic is the situation in which the entire midgut is clearly necrotic, and resection of the bowel will result in short bowel syndrome. Options include closing the abdomen without resection and offering palliative care, or performing a massive resection and creating intestinal failure, with long-term need for total parenteral nutrition. Although historically the prognosis for neonatal intestinal failure was dismal because of the extremely high incidence of fatal cholestatic liver failure, improvements in intestinal rehabilitation and small bowel transplantation have resulted in new management paradigms for children with extreme short bowel and, in selected cases, massive intestinal resection may be a reasonable option.[7]

A laparoscopic approach to the Ladd procedure can also be used.[8] In most cases, laparoscopy should only be used for malrotation not associated with midgut volvulus, because the bowel in patients with volvulus can be friable and subject to perforation.[9] In addition, surgery must be done as quickly as possible in patients with volvulus to maximize the chance of survival, and laparoscopy may waste precious time. The operation begins with the placement of an umbilical trocar and abdominal insufflation, followed by the placement of 2 additional trocars in the right lower quadrant and left midabdomen (depending on the size of the child). A fourth port may be placed in the right upper quadrant to assist with retraction. Careful exploration of the abdomen is then performed and the specific anatomy of the patient delineated. If midgut volvulus is present, the bowel must be detorsed, which can be difficult, especially if the bowel is distended or fragile. If this is the case, the operation should be converted to an open approach. If there is no volvulus, the next step is determination of the length of the small bowel mesentery; that is, the distance between the DJJ and the cecum. If this distance is long (in our center this is defined as greater than half the diameter of the abdomen, although this is an arbitrary threshold that so far is not evidence based), as is seen in both near-normal rotation and in nonrotation, the patient is not considered to be at risk for midgut volvulus, and a Ladd procedure is not considered to be necessary. In this scenario, obstructing bands around the duodenum should be identified and divided (especially if the child is having symptoms that might be caused by partial duodenal obstruction), any internal hernias should be identified and repaired, and an

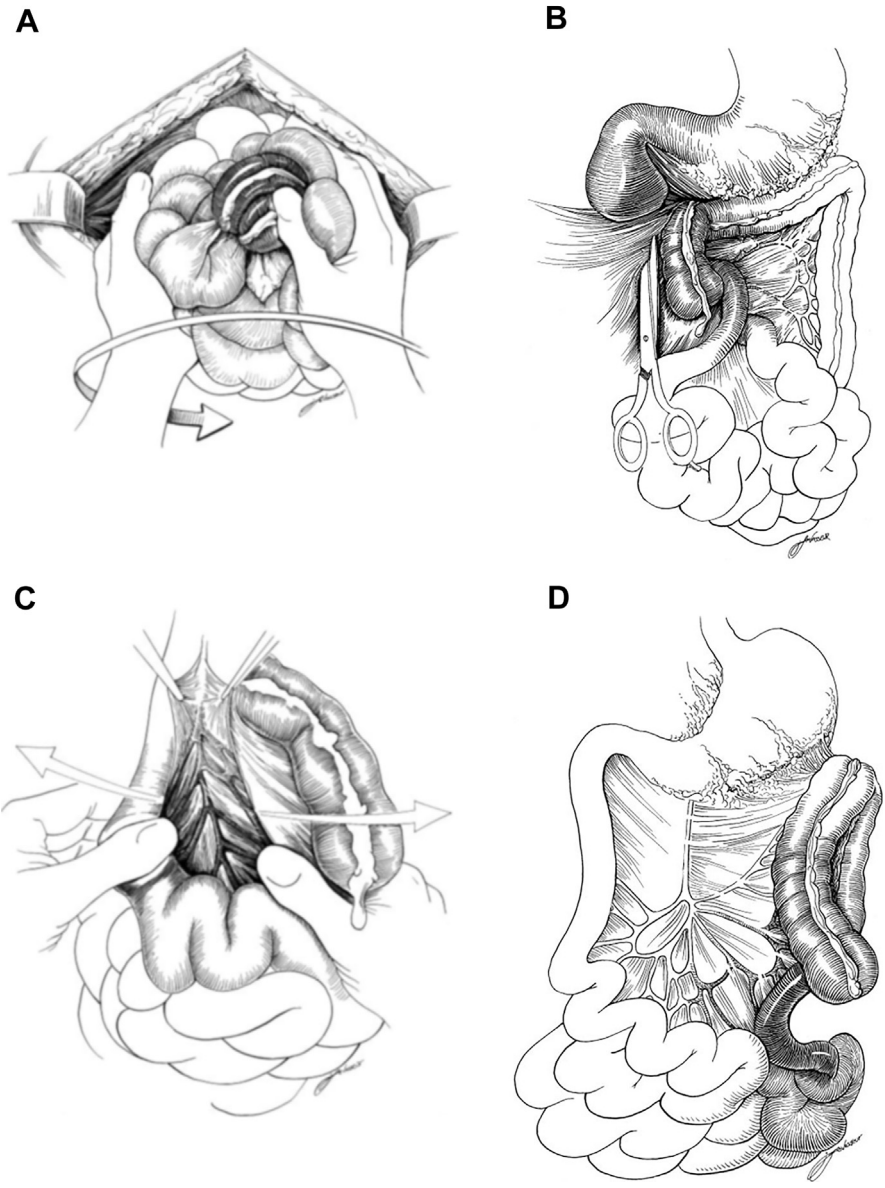

Fig. 4. Operative steps of Ladd procedure. (*A*) Bowel is assessed and, if volvulus is present, gently detorsed in a counterclockwise direction (*arrow*). (*B*) Ladd bands attaching the colon to the liver, gallbladder, or retroperitoneum are divided sharply or with electrocautery. (*C*) Adhesions to the mesentery are divided (*arrows*) and the mesenteric pedicle widened, allowing the colon to be placed on the left side of the patient and the small bowel with a straightened duodenum on the right. (*D*) Final position of the bowel contents at the completion of the Ladd procedure. An appendectomy has been performed to avoid future confusion with the presentation of atypical appendicitis in the left abdomen.

appendectomy should be considered if the appendix is in an abnormal location. If the base of the small bowel mesentery is short, the patient should be considered to be at risk for midgut volvulus, and a full Ladd procedure should be done. The steps are the same as for the open procedure, and dissection can be done using hook electrocautery, sharp scissors, or scissors attached to cautery. In larger children, a sealing device may also be used. If an appendectomy is performed, it can be done extracorporeally through the umbilical port site or intra-abdominally with an Endoloop or stapler.

Advocates of a laparoscopic approach cite decreased postoperative pain and more rapid return of bowel function (and thus shorter hospital stay) as well as an obvious cosmetic advantage. Detractors suggest that intraoperative visualization of the mesenteric pedicle is inadequate, especially in children less than 1 year of age, who are most commonly affected by malrotation. It has also been suggested that open correction of malrotation may be more effective in preventing recurrent volvulus by facilitating the formation of intra-abdominal adhesions and that the laparoscopic approach may not achieve this ancillary benefit to the same extent. Advocates of the laparoscopic approach argue that prevention of recurrent volvulus is accomplished by adequate broadening of the mesenteric base rather than by adhesions, and that adhesion formation results in a long-term risk of intestinal obstruction requiring further surgical correction. To date there have been no large-scale studies with enough longitudinal follow-up to demonstrate this theoretic benefit.

SPECIAL CONSIDERATIONS

Asymptomatic Rotation Abnormalities

Although there is general consensus that symptomatic malrotation should be addressed surgically, the role of prophylactic surgery in children with incidentally diagnosed, asymptomatic rotation abnormalities is less clear. Advocates of routine operative intervention cite reports of midgut volvulus secondary to malrotation throughout adult life and further argue that a careful history often elicits subtle symptoms of malrotation that may have been dismissed or attributed to other causes. However, population-based evidence suggests that the incidence of midgut volvulus secondary to malrotation decreases significantly after infancy and that many patients with rotation abnormalities remain asymptomatic throughout life.

Ultimately, the most important decision in asymptomatic patients is whether there is a risk of midgut volvulus or not; that is, what is the width of the small bowel mesentery? Sometimes this can be well seen on contrast imaging, and a reasonable decision can be made regarding surgical intervention. However, contrast imaging has clearly delineated false-positive and false-negative rates, and laparoscopy may be a safer and more definitive way of determining the need for a Ladd procedure. If, at the time of laparoscopy, the mesenteric base is found to be wide, the operation can be concluded, with minimal morbidity. If the mesenteric base is found to be narrow, a Ladd procedure can be done either laparoscopically or open, at the discretion of the surgeon.

Heterotaxia Syndromes

Patients with HS (defined as any arrangement of organs along the left-right body axis that is neither situs solitus nor situs inversus) are known to have a high rate of rotation anomalies, which cover the spectrum from nonrotation to classic malrotation to near-normal rotation, as well as the more uncommon rotation abnormalities such as reverse rotation. The coexistence in many cases of congenital heart disease places these children at an increased risk of operative intervention, which has resulted in controversy

around the role of generalized screening for rotation abnormalities in patients with heterotaxia, and the role of intervention in asymptomatic patients with documented rotation abnormalities.[10] Although several centers have found that the morbidity and mortality associated with a Ladd procedure in patients with HS is not increased compared with a control population, others have documented a higher anesthetic and surgical risk in children with HS who have more complex cardiac disease. In addition, the Ladd procedure is associated with at least a 10% risk of postoperative bowel obstruction, and overall longer term childhood mortality in patients with HS is 23%, mainly caused by cardiac disease. In our own study following 152 asymptomatic neonates with HS, only 4 developed gastrointestinal symptoms over a median follow-up of 18 months (range, 4–216 months), and only 1 of these 4 was found to have malrotation on UGI. Of the remaining asymptomatic patients, 43% died of cardiac disease and none developed intestinal symptoms or complications. The authors have therefore adopted a more conservative approach in which asymptomatic patients with HS are not screened for rotation abnormalities unless they develop symptoms.[11] Those with documented rotation abnormalities and either mild symptoms or no symptoms are evaluated laparoscopically.

CLINICAL OUTCOMES

Outcomes for children with malrotation and midgut volvulus depend on the degree of intestinal ischemia and the need for intestinal resection. If intestinal ischemia is extensive and/or the child presents with overwhelming sepsis, death is the usual result. If massive resection is done and the child survives, outcome depends on the management of the resulting intestinal failure; however, many advances have been made in this area, including the development of intestinal rehabilitation teams and strategies for the prevention of sepsis, venous thrombosis, and cholestatic liver failure.

Surviving children with midgut volvulus who have an adequate length of small bowel, and those children without midgut volvulus, have an excellent outcome after the Ladd procedure. There is a very low rate of recurrence in those who presented with volvulus. The reported rate of adhesive intestinal obstruction is 10% to 15%, which may be lower if a laparoscopic approach is used. Some children who presented with vague symptoms may remain symptomatic, and families should be warned about that possibility before the Ladd procedure. Some persistently symptomatic children ultimately are diagnosed with an intestinal motility disorder that may not have been suspected before the Ladd procedure, and which may be difficult to differentiate from an adhesive bowel obstruction.

SUMMARY

Rotation abnormalities represent a spectrum of anomalies that may be asymptomatic or may be associated with obstruction caused by bands, midgut volvulus, or associated atresia or web. The most important goal of clinicians is to determine whether the patient has midgut volvulus with intestinal ischemia, in which case an emergency laparotomy should be done. If the patient is not acutely ill, the next goal is to determine whether the patient has a narrow-based small bowel mesentery, which may be causing nonischemic midgut volvulus, or may predispose to midgut volvulus in the future. This decision based on imaging studies or laparoscopy, and should be followed by Ladd procedure if the mesenteric base is thought to be narrow. There is still controversy around the role of laparoscopy, the management of atypical and asymptomatic rotation abnormalities, and the management of rotation abnormalities in children

with HS. In general, the outcomes for children with a rotation abnormality are excellent, unless there has been midgut volvulus with significant intestinal ischemia.

Pearls and pitfalls

The gold standard for the diagnosis of a rotation abnormality is an upper gastrointestinal contrast study, specifically looking for the location of the DJJ. Ultrasonography may be useful as a screening tool.

The 3 potential causes of duodenal obstruction in children with a rotation abnormality are midgut volvulus, Ladd bands, and an intrinsic duodenal obstruction. Surgeons should look for all 3 in children undergoing surgery for a rotation abnormality with duodenal obstruction.

The distance between the DJJ and the ileocecal junction represents the length of the base of the small bowel mesentery, and can be estimated by contrast study or more accurately by laparoscopy. If this distance is less than half the width of the abdomen, the patient may be at risk for midgut volvulus and should have a Ladd procedure.

A laparoscopic approach is advantageous for older children without clinical or radiological evidence of midgut volvulus. Infants and children with midgut volvulus should be approached by laparotomy.

Controversies

Use of laparoscopy for children with a rotation abnormality

Pros

Ability to determine the length of the small bowel mesentery and potentially avoid the need for a Ladd procedure if the mesenteric base is long enough to prevent midgut volvulus

Theoretic decrease in risk of adhesive small bowel obstruction

Less pain, faster recovery, better cosmetic result

Cons

Lack of adhesions may increase the risk of recurrent rotation abnormality (assuming that adhesions are important in preventing recurrence, which is controversial)

Technically challenging in some cases, particularly infants and in children with midgut volvulus

Routine investigation for a rotation abnormality in children with heterotaxia

Pros

Many of these children have rotation abnormalities

Some of these may predispose to midgut volvulus

Cons

Risk of midgut volvulus is extremely low in asymptomatic children

Ladd procedure has a high risk in children with significant cardiac lesions

Long-term risk of adhesive bowel obstruction in children undergoing Ladd procedure

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