Small bowel malrotation is a congenital anomaly due to either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development. More specifically, the duodenum courses down the right side of the abdomen, the ligament of Treitz fails to cross the midline to the left side, and the cecum remains attached to the right side of abdominal wall through peritoneal fibrous bands known as Ladd’s bands (Figure 1). Ladd’s bands can form and extend from the right colon, across the duodenum, to the right lateral abdominal wall and entrap the descending duodenum causing intermittent obstruction.\textsuperscript{1,2} Other congenital rotational disorders of the small intestine include nonrotation and reversed rotation. In nonrotation, there is complete failure of the midgut to rotate around the superior mesenteric artery leading to the duodenojejunal junction being displaced to the right side of the abdomen and the ileum entering the cecum from the right side, with the large bowel located in the left abdomen\textsuperscript{1} (Figure 2).
Nonrotation is generally discovered in adulthood and is more commonly an incidental finding on imaging or during surgery.\textsuperscript{3} Reversed rotation is an abnormal clockwise rotation of the midgut by approximately 90 degrees wherein the transverse colon lies to the right of the superior mesenteric artery and passes through a retroduodenal tunnel dorsal to the artery in the small intestinal mesentery (Figure 3). It is associated with the cecum and the colon being poorly fixed which can lead to torsion.\textsuperscript{4} Malfixation can also occur where there is only a small vertical attachment of the small bowel mesentery resulting in limited fixation to the retroperitoneum. This makes the small bowel highly mobile and prone to midgut volvulus.\textsuperscript{2}

Roughly 64-80% of patients with malrotation present with bilious vomiting in the first month of life due to duodenal obstruction or a volvulus.\textsuperscript{6,7} This is reviewed in the companion article addressing the pediatric perspective. In adults, malrotation can manifest with symptoms ranging from acute abdomen, intestinal obstruction, unexplained abdominal pain, nausea and vomiting and more likely as an incidental finding in otherwise asymptomatic patients. Due to the variable presentation and rarity of this condition in adulthood, the proper management and role for operative intervention in these patients remain controversial.\textsuperscript{1} For instance, Ladd’s procedure, which involves counterclockwise reduction of volvulus, division of coloduodenal bands, widening of mesenteric base to prevent repeated volvulus and prophylactic appendectomy, has been shown to provide similar benefits in adults as it does in children. However, in adults this experience is not extensive and clinical presentation does not exactly parallel that observed in children.\textsuperscript{10,11}

**Epidemiology**

In adults, there is a slight female predominance.\textsuperscript{1} In the pediatric literature, the impression is that up to 80% of patients with malrotation are diagnosed in the first month and 90% of these are diagnosed within the first year of life.\textsuperscript{8} However, in a study of 170 patients of both pediatric and adult ages who did receive the diagnosis of malrotation, 48% were > 18 years old demonstrating that adults could be affected as well.\textsuperscript{1}

**Clinical Presentation**

Most patients present with poorly characterized abdominal pain, unexplained nausea and vomiting.\textsuperscript{1} They can also present with diarrhea, early satiety, bloating, dyspepsia, abdominal swelling, palpable abdominal mass or melena.\textsuperscript{13,14} Many of them have these symptoms for months to years and are diagnosed with small bowel malrotation when a CT scan was performed as part of the work up.\textsuperscript{1} However, up to half (continued on page 26)
of adult patients with malrotation can be asymptomatic. Complications of malrotation in adults include intestinal obstruction, acute abdomen, and volvulus of the midgut or ileocecum. There have also been reports of malabsorption, peritonitis and septic shock. Due to the rarity of this disease in adults, it can be misdiagnosed as irritable bowel syndrome, peptic ulcer disease, acute appendicitis, cholecystitis, enteritis, left colonic diverticulitis, biliary and pancreatic disease and psychiatric disorders before the correct diagnosis is finally made.

**Diagnosis**

Historically, patients with suspected midgut volvulus would undergo barium enema to evaluate cecal position. In the 1960s, upper gastrointestinal (UGI) and small bowel contrast series became the gold standard for evaluation of suspected malrotation in a child since it allows for visualization of the duodenojejunal junction and has an accuracy of approximately 80%. Malrotation can manifest as a right sided duodenojejunal junction or proximal jejunal loops on the right side on UGI series. Conventional plain radiography is neither sensitive nor specific for malrotation although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive. Abdominal doppler ultrasound may show malposition of the SMA with or without abnormal location of the hollow viscus. Recently CT has become more popular due to its ability to better illustrate the findings predictive of malrotation, such as inversion of the SMA and SMV, as well as bowel position, viability and volvulus. In malrotation, the SMV is often located to the left of the SMA or rotates around it. Indeed, in a study of 170 patients with malrotation of whom 82 were adults, 61% of them were diagnosed by CT. It is important to note, however, that some patients with malrotation will have a normal SMA/SMV relationship and an inverted relationship can also be seen in patients without malrotation. Therefore, isolated detection of such an abnormality is not sufficient for diagnosis but should warrant closer examination of the bowel. On angiography, the appearance of the “barber pole sign” is suggestive of malrotation if there is volvulus of the entire small bowel with twisting of the SMA and SMV (Figure 4).

Midgut volvulus is a complication of malrotation in which clockwise twisting of the bowel around the SMA axis occurs because of the narrowed mesenteric attachment. It can manifest as a corkscrew appearance on UGI series. CT can show a whirlpool sign in which the bowel and mesentery twist around the axis of the SMA creating a swirling appearance. Other radiographic findings include duodenal obstruction, congestion of mesenteric vasculature, and malpositioned duodenum and cecum from the underlying malrotation. However, there are no reliable means of predicting which group of patients with intestinal malrotation will develop complications such as midgut volvulus.

**Internal hernia due to abnormal peritoneal bands** is another complication which may be life-threatening because of possible bowel obstruction and strangulation. It can manifest as malrotation with small bowel obstruction (without volvulus) on CT.

**Treatment**

In patients with symptomatic malrotation, the established treatment is Ladd’s procedure to improve symptoms and reduce the risk of future complications such as volvulus and bowel ischemia. The urgency of the intervention depends on symptoms at presentation. In a study of 170 patients with malrotation of whom 82 were adults, approximately 60% of these patients underwent surgery and 50% of them had resolution of their symptoms post-operation. Of the patients who underwent surgery, only 35% had Ladd’s procedure. However, the management of the asymptomatic patients with incidentally discovered malrotation is controversial. While some authors recommended patient education and avoidance of operation unless symptoms...
Small Bowel Malrotation: A Perspective for the Adult Gastroenterologist

Discussion

Intestinal malrotation is any deviation from the normal 270 counterclockwise rotation of the midgut during embryologic development. It was first described through embryology by Dott in 1923 and then later in 1936 Dr. William Ladd, an American pediatric surgeon from Massachusetts, wrote an article detailing the surgical treatment of intestinal malrotation that now bears his name. This procedure, also known as “Ladd’s procedure”, involves counterclockwise detorsion of the bowel, division of bands crossing from the cecum to the lateral peritoneal gutter, and widening of the mesenteric base with positioning of the colon in the left abdomen and the small bowel in the right abdomen. The appendix is removed to prevent a future diagnostic dilemma because the cecum is not in its usual location in the right lower quadrant. According to Nehra et al, the majority of patients under the age of 18 with malrotation underwent Ladd’s procedure but only 35% of adults had a Ladd’s procedure. The reasons for this difference include increased operative risk, less familiarity in management of malrotation among adult general surgeons compared with pediatric surgeons, and other adult surgical issues including adhesions and anatomical changes from previous surgeries. Others have argued that the risk of midgut volvulus warrants surgical intervention regardless of age in all operative candidates. Surgical intervention in asymptomatic patients in the form of the Ladd’s procedure as described can also eliminate the possibility of ongoing morphological progression thereby decreasing the future risk of volvulus.

or complications arise, others have argued that the risk of midgut volvulus warrants surgical intervention regardless of age in all operative candidates. Surgical intervention in asymptomatic patients in the form of the Ladd’s procedure as described can also eliminate the possibility of ongoing morphological progression thereby decreasing the future risk of volvulus.

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Surgical intervention in asymptomatic patients in the form of the Ladd’s procedure as described can also eliminate the possibility of ongoing morphological progression thereby decreasing the future risk of volvulus.

This emphasizes the importance of considering small bowel malrotation in the differential diagnosis of unexplained abdominal pain in any age groups and perhaps with a low threshold to perform earlier surgical intervention in patients who are otherwise asymptomatic or with...
mild symptoms due to potential life-threatening complications later on in the disease course. However, in patients with a working diagnosis of IBS, peptic ulcer disease, functional dyspepsia, intestinal adhesions, the possibility that malrotation has been missed and could explain their symptoms needs to be considered.

On the other hand, in patients with an established diagnosis of irritable bowel syndrome (IBS), or nausea and vomiting attributed to gastroparesis, or abdominal pain attributed to adhesions from prior surgeries, or peptic ulcer disease, or even severe constipation there can be the incidental finding of small bowel malrotation during imaging. The risk of surgery may outweigh the benefits of prophylactic Ladd’s procedure given the fact that these patients have lived into adulthood without complications from small bowel malrotation. This is an ongoing dilemma for the adult gastroenterologist, namely when to even consider intervening if the malrotation was found incidentally on a CT imaging performed for other indications. For instance, in a study of 177 patients over a 35 year period it was found that asymptomatic patients had a low risk of intestinal volvulus, and as such elective surgery was not recommended. It is also impossible to predict which patients will develop life-threatening complications. Therefore, further long term observational studies are needed to determine which subset of patients with otherwise asymptomatic small bowel malrotation are at high risk for complications and warrant prophylactic Ladd’s procedure.

**CLINICAL PEARLS**

When does the adult gastroenterologist even consider ruling out malrotation in his patients; 1) If the patient gives a history of pediatric surgery(ies) of unclear or forgotten indications. 2) Surgery for an acute abdominal problem at a young age never fully clarified for the patient, such as suspected internal hernias, volvulus, or aberrantly located appendix; 3) Where abdominal pain, nausea, or vomiting are out of proportion to objective data by standard or routine testing and where treatment approaches have been suboptimal.

We hope that our review has planted a seed for your appreciation of the spectrum of small bowel malrotation in adults. We have attempted to provide a rational approach for the diagnosis and summarized the treatment options available for these patients. ■

**References**