Malrotation is a part of the spectrum of anatomic anomalies comprising malrotation. Most cases of nonrotation are symptomatic and managed surgically with a Ladd’s procedure. However, incidental discovery of asymptomatic patients has also been reported. In these cases, the role of surgery is debated. The authors describe a case of nonrotation in a young woman with constipation-predominant irritable bowel syndrome that caused symptoms associated with nonrotation. Medical management of her irritable bowel syndrome resulted in the resolution of all abdominal complaints and a Ladd’s procedure was not recommended. The authors include a literature review of nonrotation with an emphasis on the treatment of asymptomatic disease.


Malrotation is classically thought of as a pediatric disease, as it is a product of altered embryologic development and most commonly presents in the first 12 months of life. Surgical literature, however, is replete with case reports of malrotation in older children and adults, highlighting the fact that the disease is treated by both pediatric and adult general surgeons alike.

Malrotation is a collection of several distinct anatomic anomalies, which can be described based on the location of the duodenal-jejunal junction and the cecum. The most common of the anomalies is nonrotation of the duodenum and cecum, in which both the pre-arterial and postarterial midgut are malpositioned. However, additional entities including isolated duodenal malrotation (aberrant pre-arterial midgut positioning with normal postarterial positioning), mixed rotation (aberrant pre-arterial midgut positioning with partial postarterial rotation), hyperrotation (normal pre-arterial midgut positioning with overrotation of the postarterial midgut), reversed rotation (normal pre-arterial midgut positioning with inverted postarterial midgut positioning), and others have been described.

Despite the heterogeneity of anomalies grouped under the heading of malrotation, the treatment is rather homogeneous as patients with all types are treated with Ladd’s procedures. While the role of surgery is clear in the setting of symptomatic disease, its role in asymptomatic malrotation has been challenged by a series of publications.

Report of Case
A 26-year-old woman presented to the Tripler Army Medical Center’s emergency department in Honolulu, Hawaii, with severe, cramping abdominal pain of 10 hours duration. The pain was centered over the periumbilical and suprapubic regions and did not radiate into other areas. She denied having fevers or chills but stated that it had been more than 24 hours since her last bowel movement.

The patient reported a history of constipation since childhood and autoimmune alopecia. Three months before presentation, the patient was diagnosed as having acute colitis. At that time, she was experiencing abdominal pain with up to six foul-smelling bowel movements per day. A colonoscopy was performed, and the biopsy was remarkable for colonic inflammation with prominent lymphocytic aggregates. Her symptoms resolved after 1 month of mesalamine. Her family history was notable only for a paternal uncle with ulcerative colitis.

At presentation, the patient’s vital signs were within normal limits. Physical examination revealed a distended abdomen with normal bowel sounds. The patient reported tenderness to palpation in the left lower quadrant, but we found no peritoneal signs. Results of her digital rectal examination were normal, without stool in the rectal vault. Laboratory tests revealed a clinically significant high white blood cell count of $10.6 \times 10^9$/mL (87.9% granulocytes).

A computed tomography scan of the abdomen and pelvis showed that the duodenal-jejunal junction was in the right side of the abdomen, as was the entire small bowel (Figure 1).
The terminal ileum emptied into the cecum in the left lower quadrant. The entire large bowel was in the left side of the abdomen. The duodenum did not pass posterior to the superior mesenteric artery, and the superior mesenteric artery was to the right lateral aspect of the superior mesenteric vein. There was no evidence of volvulus, ischemia, perforation, or appendicitis.

The patient was initially admitted to the hospital for pain control, which fully resolved after a bowel movement. She was discharged to home the same day with outpatient follow-up.

The radiologic findings suggestive of nonrotation were further studied with a fluoroscopic contrast examination of the midgut, which confirmed the presence of nonrotation (Figure 2). Given the diagnosis of nonrotation and recent abdominal pain, we discussed the option of a Ladd’s procedure with the patient. However, in light of her long history of constipation and the fact that she had no pain after a bowel movement, we referred her to a gastroenterologist for management of presumptive constipation-predominant irritable bowel syndrome before undergoing a surgical procedure.

Given the patient’s prior episode of colitis, the gastroenterologist repeated a colonoscopy that demonstrated no evidence of colitis, and he confirmed the diagnosis of constipation-predominant irritable bowel syndrome. The patient was initially treated with polyethylene glycol 3350, which failed to resolve her constipation. She was switched to lubiprostone, which resulted in daily soft bowel movements. The patient has been without abdominal complaints for more than 1 year, and a Ladd’s procedure has not been recommended.

Discussion
Malrotation of the midgut occurs during embryologic development. During the fifth gestational week, rapid growth of the liver causes midgut herniation into the umbilical stalk. Simultaneously, the midgut rotates counterclockwise 90 degrees about its vascular pedicle. The herniated midgut begins to return to the abdominal cavity in the 10th week of gestation. The proximal jejunum is the first portion to return, coming to rest left of the midline. As the midgut returns into the abdominal cavity, it rotates an additional 180 degrees counterclockwise. The cecal bud is the final segment to return, bringing the postarterial midgut with it. The cecal bud initially lies immediately below the right lobe of the liver, but during the 11th and 12th weeks of gestation, descends to its final position in the right iliac fossa.

When the midgut does not undergo normal rotation or return into the abdominal space during embryologic development, a variety of anatomic configurations and anomalies...
can occur (Figure 3).4 The most common malrotation variant is nonrotation.3,10 Nonrotation results when the cecal bud is the first segment to return into the abdomen and comes to rest in the left lower quadrant.1 In this setting, the colon occupies the remainder of the left side of the abdomen, leaving the prearterial segment of the midgut to reside in the right side of the abdomen. In addition, the final 180 degrees of counterclockwise rotation does not occur. The resultant anatomic configuration can include a reversed relationship of the superior mesenteric artery and superior mesenteric vein as well as a reversed location of the duodenal-jejunal junction anterior to the superior mesenteric artery.11 All of these anatomic anomalies were noted in the patient presented herein.

Based on a literature search using the US National Library of Medicine’s PubMed database and our own familiarity with medical literature, most reports do not examine nonrotation alone, but rather discuss it within the larger context of malrotations. A range of malrotation presentations have been described, including acute surgical abdomen, vague chronic abdominal symptoms, and asymptomatic disease. The patient described in the present case represents the second group, having presented for abdominal pain originating from constipation.

In medical literature, surgery is recognized as the appropriate treatment for symptomatic disease directly related to malrotation.5,6,12 However, the role of surgery as a treatment option for patients with asymptomatic disease—especially in adult patients—is debated.

According to a PubMed database literature search using the MeSH keyword “malrotation,” operative intervention is
favored for patients with asymptomatic diseases. One reason cited for favoring surgery is the belief that if a thorough history is obtained, truly asymptomatic disease is rare. Furthermore, surgery has been found to provide complete symptom relief for two-thirds of patients and at least partial symptom relief for nearly all patients. In addition, there is an up to 20% lifetime risk of acute volvulus or ischemia in older children and adults with malrotation, and emergent surgery is associated with a 2.5-fold increased risk of complications in patients who have elective Ladd’s procedures. Given these benefits, most authors have concluded that the potential benefits of surgery appear to outweigh its risk.

A formal risk-to-benefit analysis by Malek and Burd on the management of asymptomatic malrotation with a Ladd’s procedure concludes otherwise. In their statistical model, a Ladd’s procedure was associated with improvement in quality of life only for patients younger than 20 years. A mortality benefit for surgery was also limited to pediatric patients. Additional two- and three-way analyses suggested that observation alone is beneficial for patients as young as 12 years. These results led the authors to conclude that a Ladd’s procedure is not indicated for asymptomatic disease in adults.

Additional support for observation of asymptomatic malrotation comes from a 2007 study by McVay et al. In their retrospective review, 36 patients had asymptomatic disease at initial presentation and only 6 patients eventually developed symptoms. In addition, none of the initially asymptomatic patients developed volvulus or other acute complications of malrotation. The authors concluded that nonoperative management can be an appropriate treatment strategy for asymptomatic patients, as it was rare for them to progress to symptomatic diseases or develop acute abdomens.

While the patient described in the present report did have abdominal symptoms, her symptoms completely resolved with the initiation of lubiprostone. Follow-up examination did not reveal other abdominal symptoms, suggesting that the patient’s original abdominal complaints were related to her constipation-predominant irritable bowel syndrome and not midgut nonrotation. The authors concluded that nonoperative management is an appropriate treatment option for asymptomatic patients, as it was beneficial for patients as young as 12 years. These results led the authors to conclude that a Ladd’s procedure is not indicated for asymptomatic disease in adults.

Our recommendation is based on the more recent studies demonstrating no benefit for treatment of asymptomatic disease in adults. Furthermore, the patient’s particular variant of malrotation, namely nonrotation, means her anatomy is already in a post-Ladd’s procedure configuration. While the appendix was not removed and the mesentery has not been widened, as would occur with a Ladd’s procedure, we believe that performance of these steps would not result in a measurable improvement for the patient. Finally, and most importantly, the decision to not perform a surgery was made after multiple discussions with the patient, in which the full spectrum of treatment options and their potential for benefit vs harm were discussed.

### Conclusion

Asymptomatic nonrotation of the midgut can be considered for observation alone and does not necessarily require operative intervention. Functional bowel disorders can complicate evaluation and management of patients with otherwise benign anatomy and lead to unnecessary and ultimately ineffective operations. Thoracopan patient history and physical and radiographic evaluations are critical to the appropriate treatment of the stable patient.

### References