A 16-year-old Female With Menstrual Cramps and a History of Anorexia

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Presentation
A 16-year-old African American girl presents to the emergency department with severe cramping and pain in all quadrants of her abdomen, with the most severe pain located in her right upper quadrant. The pain had started 24 hours previously and initially was dull and diffuse. The pain has worsened progressively with sudden, more severe pain developing in the right upper quadrant 5 to 6 hours before she came to the emergency department.

She has had three episodes of nonbilious brown emesis over the past 24 hours. Her last bowel movement was 7 days ago, and she denies any blood or mucus in the stool. She reports having mild abdominal cramps over the past week that she attributes to her recent menstrual period; her last spotting was 8 days ago.

There is no history of recent fever, diarrhea, or genitourinary problems. She is not sexually active, and there is no history of alcohol or illicit drug use. The patient has occasionally taken a combined medication of acetaminophen, aspirin, and caffeine for her pain. She has no allergies. She is an excellent student, and there are no known psychosocial stressors in the family.

The girl has a chronic history of vague abdominal pain while eating, followed by nonbilious, nonbloody emesis and subsequent relief of pain. These episodes of pain started in early childhood and continued until age 12 years. However, her abdominal pain returned at age 14 years; although that pain was severe, there was no emesis. The pain was unrelated to meals but would present sometimes 2 weeks before menses and sometimes during menses. The pain also was associated with occasional inability to pass a stool for a few days.

Menarche occurred at age 13 years. For the past 2 years, she thought her abdominal pain was due to her menstrual periods; yet her parents, concerned about a possible eating disorder, brought her to her pediatrician.
for evaluation. At that time, her physical examination was unremarkable, and no abdominal imaging was performed.

Today, the patient’s hemodynamic, respiratory, neurologic, and hydration statuses are normal. Her abdomen is distended and tender upon palpation of the left upper, left lower, and right lower quadrants. The tenderness is particularly severe over the right lower quadrant. There is no guarding or rebound tenderness. Bowel sounds are present. The rest of her physical examination is unremarkable. She vomits while she is being examined.

Complete blood count and metabolic panel are within normal limits. A urine pregnancy test is negative. Urinalysis reveals a specific gravity of 1.030, 1+ ketones, and 2+ protein. Blood cultures and urine cultures are obtained.

An abdominal axial computed tomography (CT) scan reveals the diagnosis.

**Diagnosis: Intestinal Malrotation With Volvulus**

The CT scan revealed a variant malrotation of the small and large bowel, with a low lying duodenal-jejunal junction in the left upper quadrant and the cecum in the left lower quadrant (Fig 2). Also present was a midgut volvulus revealing characteristic twisting of mesenteric vessels around the superior mesenteric artery axis (Fig 3). The proximal small bowel loops were distented up to 7.0 cm in the greatest transverse dimension, with the loops returning to normal caliber in the region of the volvulus.

**Discussion**

Although intestinal malrotation and mid gut volvulus have been reported in the adolescent and adult literature, this patient’s presentation was atypical given the combination of abdominal pain and nonbilious vomiting during her childhood; the temporal relationship of symptoms to monthly period cycles once puberty was achieved; and the acute presentation of 1 week of constipation and 24 hours of abdominal pain and intense nonbilious vomiting. In general, malrotation must be considered when faced with a patient who has a history of chronic vomiting and abdominal pain.

Intestinal malrotation is a congenital abnormality consisting of nonrotation or incomplete rotation of the bowel around the superior mesenteric artery during embryonic development. Most anomalies of rotation result in an abnormally narrow mesenteric base. Because the mid gut is suspended on a narrow vascular pedicle instead of the usual wide base of the mesentery, there is a risk for the bowel to twist itself, thereby developing a volvulus. Malrotation is accompanied by abnormal bowel fixation by mesenteric bands. Ladd bands are
abnormal coloduodenal peritoneal bands that tether the midgut and can compress the midgut or duodenum extrinsically.

Malrotation leads to an increased risk for bowel obstruction, acute or chronic volvulus, and bowel necrosis. Congenital abnormal positioning of the duodenal-jejunal junction may lead to midgut volvulus, a potentially life-threatening complication.

Rotational anomalies, which may or may not be symptomatic, are estimated to occur between 1/200 and 1/500 live births. Symptomatic malrotation is estimated to occur in 1/6,000 live births. Approximately two thirds of children who require surgery for malrotation are younger than age 1 month; 18% to 25% present between 1 month and 1 year after birth, and 10% to 18% present after 1 year.

Between 30% and 62% of children born with intestinal malrotation also have an associated anomaly, such as congenital diaphragmatic hernia, omphalocele, gastroschisis, duodenal atresia, jejunoileal atresia, Meckel diverticulum, duodenal web or stenosis, Hirschsprung disease, esophageal atresia with tracheoesophageal fistula, biliary atresia, prune belly syndrome, cardiac anomalies, situs inversus, mesenteric cysts, and renal anomalies.

During the neonatal period, bilious emesis and bloody stools are the most common symptoms caused by malrotation with volvulus or obstruction. Beyond the neonatal period, children who have malrotation with volvulus or obstruction present commonly with bilious or nonbilious emesis, recurrent abdominal pain, and failure to thrive. In older children and adults, malrotation has been misdiagnosed as appendicitis, diverticulitis, or peritonitis when the presentation was acute, and as malabsorption syndrome or psychogenic vomiting when symptoms occurred over a prolonged period.

The diagnosis often is established by upper gastrointestinal (UGI) barium studies, which are diagnostic for most patients in whom malrotation is strongly suspected. However, UGI findings can be equivocal and can miss the diagnosis. A diagnostic UGI reveals the duodenal-jejunal junction to be located below the level of the duodenal bulb instead of horizontally crossing the midline to the left upper quadrant. The CT scan usually can reveal the diagnosis but can be falsely negative in 25% of patients if one does not use contrast to visualize the blood vessels. Other studies, such as barium enema, abdominal ultrasonography, colonoscopy, esophageogastroduodenoscopy, and plain radiographs, may be of limited value.

**Differential Diagnosis**
The differential diagnosis for chronic abdominal pain and vomiting besides malrotation depends on age. In the neonate, the differential includes necrotizing enterocolitis and duodenal atresia. In the young infant, one should consider pyloric stenosis. In older infants, one also should consider intussusception or other causes of bowel ischemia. In older children and adolescents, the differential diagnosis also includes a variety of other illnesses that can present with chronic abdominal pain and vomiting, with or without diarrhea.

**Treatment**
The treatment for intestinal malrotation is surgery. The recommended procedure is the Ladd procedure, which consists of counterclockwise detorsion of the midgut volvulus (if present), division of the abnormal coloduodenal Ladd bands that tether the midgut and extrinsically compress the bowel, and expansion of the mesenteric base (to prevent further volvulus), together with removal of the malpositioned appendix.

**Patient Course**
The patient underwent an emergent exploratory laparotomy that resulted in reduction of the midgut volvulus, resection of the Ladd bands, and an appendectomy. The small bowel proximal to the volvulus revealed signs of chronic dilation without ischemia. Her postoperative course was uneventful, and she was discharged from the hospital in 8 days.

**Summary**
- Bowel malrotation typically presents in infancy or early childhood. The condition is seen less commonly in the teenage population, in whom the diagnosis may be missed.
- Among older children born with malrotation, 10% to 15% present with volvulus. The onset of symptoms usually is acute, but some children can present with a more chronic pattern of episodic vomiting and abdominal pain suggestive of intermittent volvulus.
- The diagnosis may be missed or delayed, with resultant catastrophic midgut volvulus, together with sepsis or other complications.
- This case illustrates that clinicians must include malrotation in the differential diagnosis when evaluating an adolescent girl experiencing chronic abdominal pain that is suggestive of menstrual pain or pain of psychogenic origin.
Suggested Reading


