A 14-year-old boy presented to the University of Maryland emergency department due to constant abdominal pain for 3 days and vomiting, which had been intermittent over the prior 2 weeks. He described having episodic abdominal pain with vomiting 2 or 3 times per year for the past 3 years. Each episode lasted approximately 2 days before resolving spontaneously.

Physical examination revealed a 115-kg male with a heart rate of 101 beats per minute in moderate distress. Abdominal examination revealed an obese but soft abdomen with normally active bowel sounds and tenderness localized to the epigastrium. He exhibited voluntary abdominal guarding, but no rebound tenderness. Rectal examination was negative.

The white blood cell count was $9.0 \times 10^9/L$, bicarbonate 30 mEq/L, anion gap 17, and chloride 95 mEq/L. All other electrolytes were within normal ranges. Urinalysis revealed specific gravity greater than 1.030 with 3+ ketones. Computerized tomography (CT) revealed a mid-abdominal transition area, beyond which there was no evidence of contrast-filled small bowel. An area of concentric small bowel was noted. There was no evidence of free fluid or free air (Figure 1). The CT scan was suggestive of intussusception but also as consistent with volvulus, prompting a fluoroscopic small bowel evaluation. This study revealed mild dilation of the first 2 sections of duodenum, a tapered, highly obstructive appearance of the third portion of the duodenum; and a corkscrew configuration in the duodenal junction region—highly suggestive of volvulus (Figure 2). The duodenal-jejunal junction was also abnormally low and rightward in location, consistent with malrotation. Exploratory laparotomy revealed a midgut volvulus of 540 degrees, in addition to malrotation with no evidence of bowel ischemia or necrosis. A Ladd procedure was performed with an appendectomy, and the patient recovered uneventfully.

Intestinal malrotation is thought to occur in 1 in 200 to 500 newborns, but is symptomatic in only 1 in 6000. Up to 90% of malrotations with volvulus require surgical repair during the first year of life. Thirty percent to 60% of infants diagnosed with malrotation have an associated gastrointestinal anomaly, ranging from diaphragmatic hernia to omphalocele. These rotational anomalies form during the 4th to 10th week of gestation. Malrotation typically results in the cecum resting in the mid-upper abdomen, with attachments to the posterior wall via peritoneal bands (Ladd bands).
Newborns or infants with intestinal malrotation classically and commonly, 97% of the time, present with bilious emesis and constipation. Other possible findings include hematochezia and abdominal distention, although these symptoms may indicate intestinal ischemia. Symptoms are more varied and nonspecific with increasing age and can include episodic attacks of vomiting, failure to thrive, and recurrent colicky abdominal pain. The intermittent symptoms of older children often result in diagnostic delays as the patients are diagnosed with more common ailments: irritable bowel, peptic ulcer disease, pancreatic disease, and psychiatric disorders. These delays are important because the most serious consequence of volvulus is bowel necrosis and resultant shock, sepsis, and death.

The treatment for volvulus is surgical. The Ladd procedure, the most common surgical technique, involves untwisting the bowel, removing any nonviable bowel, and placing the bowel in a position of nonrotation. A prophylactic appendectomy is usually also done.

References