Intestinal malrotation is a rare congenital abnormality that occurs when the normal rotation and fixation of the intestines does not occur. About 90% of the cases of malrotation are diagnosed before the first birthday with the majority of the diagnosis being made in the first month of life. The most common complication of intestinal malrotation is midgut volvulus which could lead to intestinal necrosis and be potentially fatal if not promptly diagnosed and surgically treated. The reported incidence for malrotation is estimated at 1/6,000 live births while the absolute incidence is unknown. Autopsy studies have reported an incidence as high as 1% of the total population.

The spectrum of intestinal malrotation includes non-rotation, incomplete rotation, reversed rotation, and mesocolic hernia. Non-rotation is characterized by incomplete counterclockwise rotation of the midgut around the superior mesenteric artery (SMA). Essentially the small bowel is on the left of the abdomen, the colon on the right, and the cecum displaced anteriorly and midline. This type of malrotation has an increased risk for midgut volvulus. Incomplete rotation occurs when the intestine only rotates 180 degrees rather than the total 270 degrees. The duodenojejunal junction is on the right and the cecum is in the upper abdomen just left of the SMA; the same risk of midgut volvulus is present. Reversed rotation and mesocolic hernia are rare and difficult to diagnose. Both conditions can present with chronic abdominal symptoms or acute bowel obstruction in the absence of previous abdominal surgeries.

Most of the patients have an asymptomatic malrotation which sometimes is incidentally found while testing, specifically imaging for other clinical conditions. Symptomatic malrotation usually present...
early in life with bilious emesis and abdominal pain as the most common symptoms. Other symptoms may include hematemesis and rectal bleeding. Older children with symptomatic malrotation have more vague symptoms that include: anorexia or nausea, intermittent apnea, failure to thrive, constipation, and diarrhea. It is important to suspect intestinal malrotation in older children that present with recurrent episodes of abdominal pain associated with symptoms of intestinal obstruction pain, irritability, vomiting, or failure to thrive.

For patients who are symptomatic as a result of malrotation, surgery is the treatment of choice. When it comes to asymptomatic malrotation, there is a lack of consensus. There are no prospective studies and only retrospective chart reviews and computer modeling. The questions that often arise are:

1. Are there asymptomatic patients that should be screened for malrotation?
2. If malrotation is found, what should the next step be? Should an elective Ladd’s procedure be performed or should observation be instituted? Surgery is a consideration in these asymptomatic patients because volvulus is a theoretical risk.

Graziano et al. summarized the available literature and concluded that there was a lack of evidence to support screening asymptomatic patients for malrotation. Also, there was minimal evidence to screen those patients with heterotaxy syndrome (HS). Patients with HS have cardiac and visceral malpositioning. In these patients, volvulus most commonly occurs in the first year of life and most cases occur in the first month of life. A recent literature review performed by Landisch, et al. found that 47% of patients with HS have intestinal rotation abnormalities on upper gastrointestinal series, but only 1.2% have an increased risk of developing volvulus. Pockett et al. found that 57% of patients with HS have serious complications and argued against prophylactic Ladd procedure.

What is true is that prospective data is needed and in the interim, we recommended that with each malrotation, the physician carefully weigh their experience and how it coincides with patient outcomes. From a pediatric gastroenterology perspective, we follow our field’s common adage—once a malrotation, always a malrotation. We therefore observe until clinical symptoms tell us otherwise.

References