

COLONIC VOLVULUS: AN UNUSUAL PRESENTATION OF MALROTATION

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Midgut volvulus secondary to midgut malrotation usually occurs in the neonatal period.¹⁻⁴ When malrotation presents beyond the neonatal period, it is associated with a spectrum of symptoms which are non-specific. Midgut volvulus in older patients has been reported to be seen in as few as 14% patients.^{1,5,6}

Whereas midgut volvulus secondary to midgut malrotation is the most common and feared complication, there are instances where the defect is secondary to lack of fixation of the large bowel.^{7,8} The purpose of this report is to describe an unusual and rare cause of colonic volvulus secondary to lack of fixation of the right side of the colon, and to alert pediatricians and surgeons to this unusual presentation of malrotation.

Case Report

A 2½-year-old male patient was admitted to our hospital because of abdominal distension, constipation and vomiting of three days' duration. He was a product of a full-term normal delivery at home, who had been referred to our hospital immediately after delivery because of multiple anomalies, which included low set ears, widely spaced eyes, wide nipples, short humeri on both sides, penoscrotal hypospadias with chordee, bilateral impalpable testis, congenital cataract and exomphalos minor (Figure 1). The exomphalos was 3 cm at the neck and 8 cm in diameter. This was repaired without difficulty. The patient's birthweight was 2.2 kg. He was readmitted at the age of three months with sepsis, treated with antibiotics and recovered. At the age of five months, he was readmitted with left axillary lymphadenopathy, following BCG vaccination. This was biopsied, and the histology revealed granulomatous lymphadenitis. At the age of 2½-years, his weight is now only 3.9 kg, and he has features of arthrogryphosis, mental and growth retardation. On examination, he was found to have abdominal distension with generalized tenderness. His bowel sound

was hypoactive, and rectal examination revealed small discrete hard pieces of feces. His abdominal x-ray (Figure 2) revealed a hugely dilated colon. He was diagnosed as a case of volvulus. During surgery, he was found to have volvulus of the right side of the colon and transverse colon with the site of the volvulus at the splenic flexure (Figure 3). The cecum was located at the upper left quadrant. The right side of the colon and transverse colon was found to be ischemic, with areas of necrosis and perforation at one site. After reduction of the volvulus, the right side of the colon was found to be totally mobile, and together with the transverse colon twisted at the point of fixation at the splenic flexure. There were no adhesions. About 10-15 cm of terminal ileum was also involved and this, together with the ascending and transverse colon, was resected, and end-to-end anastomosis of terminal ileum and descending colon done. Except for a wound infection, the patient did well after operation and was discharged after two weeks.

Discussion

Embryologically,⁹⁻¹¹ rotation of the gut occurs between 10 and 12 weeks of intrauterine life, coinciding with the return of the midgut from its ventral extracelomic herniation. The duodenal loop rotates 270 degrees counterclockwise from the pylorus, forming a "C" loop that passes behind the superior mesenteric artery and vein. As a result of this, the duodenojejunal junction will be

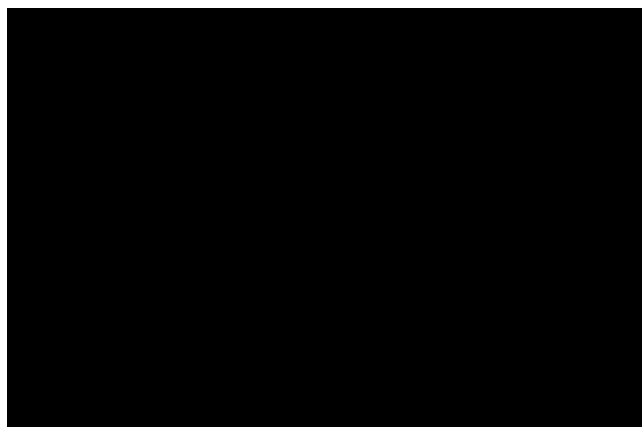


FIGURE 1. A newborn with exomphalos minor.

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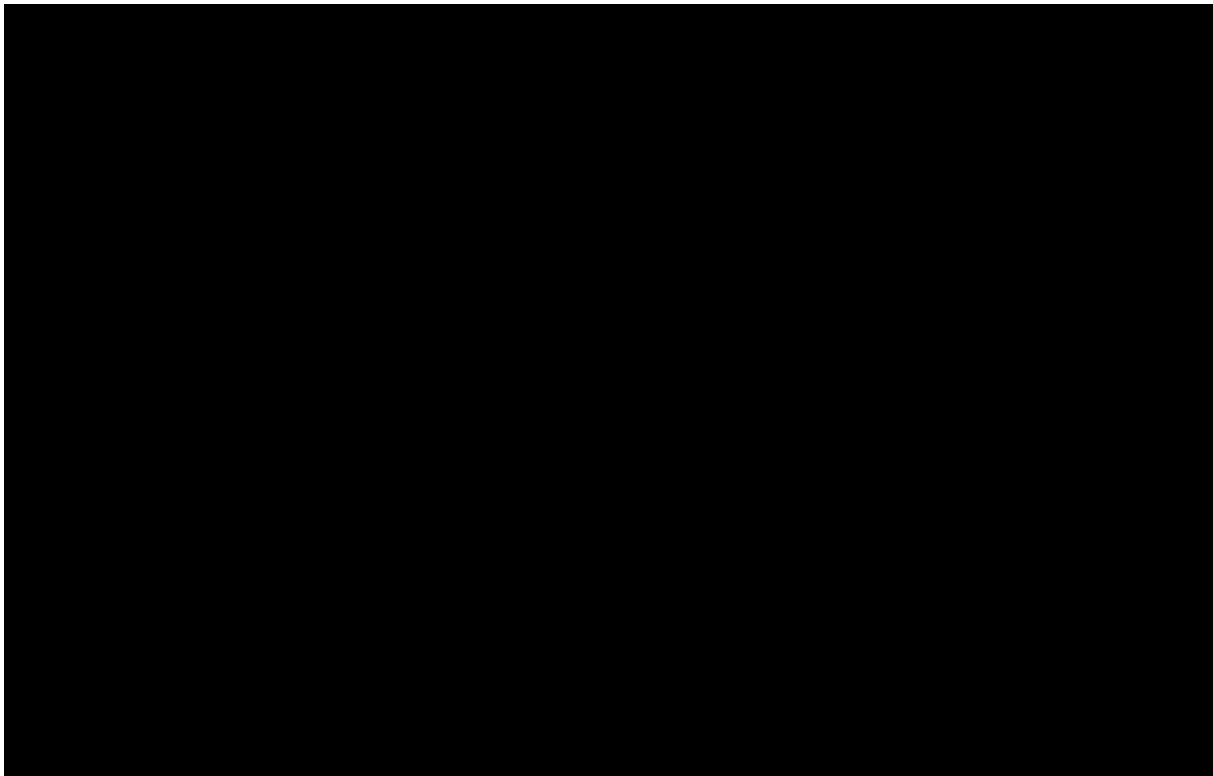


FIGURE 2. Supine and erect abdominal x-rays showing massively dilated colon.

located to the left of the spine and attached to the retroperitoneum by the ligament of Treitz. The cecocolic loop, on the other hand, rotates 270 degrees counterclockwise, but passes in front of the superior mesenteric vessels. This places the ileocecal junction in the right lower quadrant of the peritoneal cavity. It is then fixed by peritonealization bands.

In malrotation, the site of obstruction is usually the second or third portions of the duodenum. Although duodenal obstruction is a common presentation of malrotation, midgut volvulus with intestinal gangrene is the most feared complication of intestinal obstruction. To obviate this, Powell et al.⁶ questioned labelling patients as asymptomatic or incidental malrotation, and recommended prompt elective Ladd's procedure in all patients found to have abnormalities of malrotation and fixation. Malrotation is easily diagnosed when it presents in the neonatal period. Biliary vomiting is the most frequent symptom in the first month of life. The frequency of midgut volvulus among infants presenting in the neonatal period ranges from 45% to 80%. By contrast, the frequency in older patients has been reported to be as low as 14%.¹ Malrotation presenting beyond the neonatal period is associated with a multiplicity of symptoms which are often nonspecific. These include diarrhea, constipation, failure to thrive, malabsorption and protein

deficiency.^{5,11,12}

Although the most common site of obstruction in malrotation is at the second or third portion of the duodenum, there are reports of unusual types of malrotation. In the Spigland et al.⁵ series of 82 patients, six unusual types of malrotation were documented. Of these, four had right paramesocolic hernia, one had malrotation of the duodenum of the small bowel without malrotation of the colon, and the other had reversed rotation with the colon passing beyond the superior mesenteric artery and duodenum. None of these cases resembled that of our patient. Attention has been focused mostly on midgut volvulus, which frequently occurs in the immediate neonatal period and is usually associated with midgut malrotation. However, there are instances such as our case where the defect is related to lack of fixation of the large bowel rather than malrotation. Pochaczewsky et al.⁷ reported two cases of volvulus in older children. One of them was the result of lack of fixation of the right colon in its normal retroperitoneal position, which resulted in massive necrosis of the small bowel and the right side of the colon. Colonic volvulus is unusual in children, and has been reported to affect the sigmoid or cecum.^{5,13} Our patient had volvulus of the right and transverse colon up to the splenic flexure with necrosis, as a result of lack of fixation of the right colon. The presence of omphaloceles,



FIGURE 3. Intraoperative photograph showing massively dilated colon with volvulus.

which is sometimes associated with malrotation, was diagnosed in the neonatal period in our patient, and so hinted at the possibility of malrotation with volvulus. Awareness of the possibility of these rare types of malrotation in patients who, like ours, present beyond the neonatal period, may help reduce the delay in diagnosis and treatment, as well as minimize associated morbidity and mortality.

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