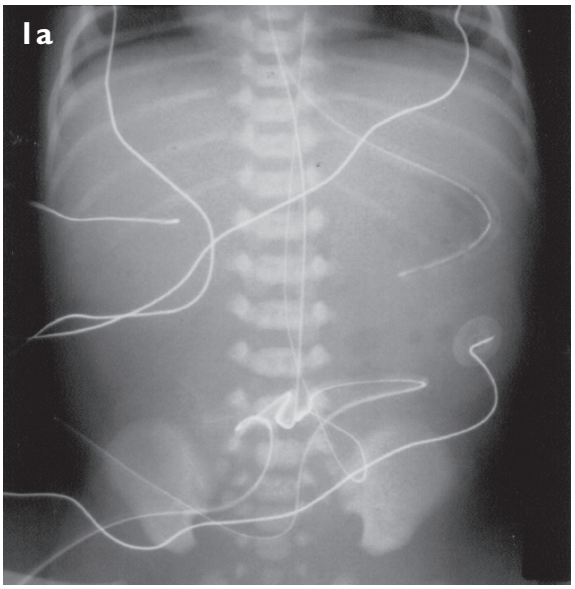
Clinics in Diagnostic Imaging (74)

P J Strouse



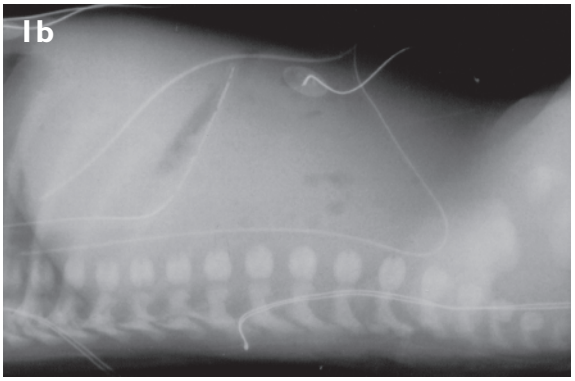
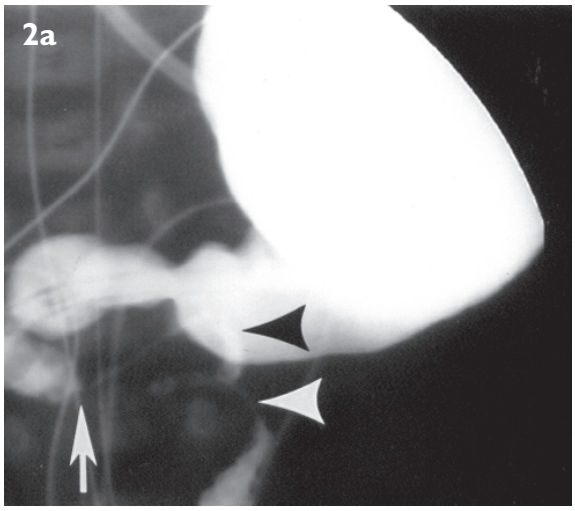


Fig. 1 (a) Anteroposterior and **(b)** cross-table lateral (b) radiographs of the abdomen.



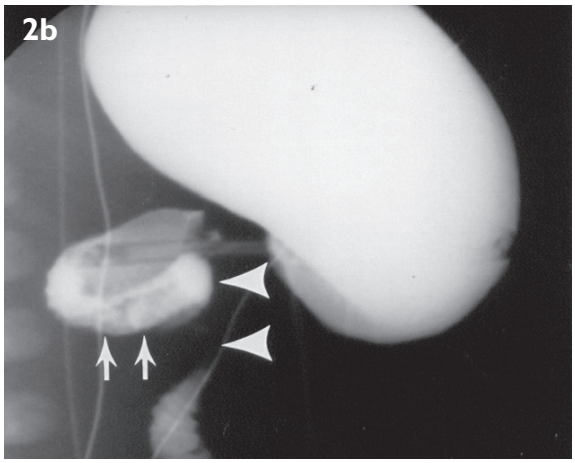


Fig. 2 (a) Anteroposterior and **(b)** lateral views obtained during an upper GI examinaion.

CASE PRESENTATION

A two-day-old, 2.5 kg male infant from a 35-week gestation pregnancy had initially done well. When the baby was given feedings, he had several episodes of bilious emesis. The child had passed meconium promptly; however, frank bloody stools were noted approximately 48 hours after birth. On physical examination, the child appeared distressed and was lethargic to stimulation. The abdomen was slightly distended, with slight apparent tenderness elicited during palpation. Bowel sounds were hypoactive. Rectal examination revealed a small amount of meconium with a normally positioned, patent anus. The child's heart rate was 145 beats per minute, blood pressure was 59/40 mm Hg, and respiratory rate was 30-35 breaths per minute (ventilated). Capillary refill was delayed. Laboratory investigation showed sodium of 139 meq/l, potassium of 3.4 meq/l, chloride of 105 meq/l, pH of 7.47, pCO_2 of 32 mm Hg, pO_2 of 151 mm Hg, haematocrit of 28.9% and white blood cell count of 5.9 x $10^3/mm^3$. Supine anteroposterior and cross-table lateral radiographs (Fig. 1) were obtained, followed by an upper gastrointestinal (GI) examination using barium (Fig. 2).

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IMAGE INTERPRETION

The radiographs of the abdomen (Fig. 1) were essentially normal. There was a paucity of bowel gas, a non-specific finding. A small amount of gas was seen in non-dilated bowel. A nasogastric tube was present in the stomach, and umbilical artery and vein catheters were present. No pneumatosis intestinalis, portal venous gas or free intraperitoneal gas was seen to suggest necrotising enterocolitis. Although there were no findings to specifically suggest malrotation, the diagnosis was not excluded as radiographs are usually normal in the setting of malrotation.

The upper Gl films showed barium in the stomach and duodenum (Fig. 2). The barium was instilled via the nasogastric tube, the tip of which was in the duodenal bulb. Little barium coursed through the duodenum and most had filled the stomach, indicative of a duodenal obstruction. Although the duodenum extended across the midline in this patient, it tapered at the point of partial obstruction (arrow) and then made a 720° "corkscrew" (arrowheads) throughout which the calibre was narrowed. The lateral view (Fig. 2b) showed an abnormal anterior course of the distal duodenum (arrows) entering the "corkscrew" (arrowheads).

DIAGNOSIS

Midgut malrotation with volvulus.

CLINICAL COURSE

The baby was urgently taken to the operating room and an exploratory laparotomy was performed. A midgut malrotation was noted with approximately a 360° torsion and Ladd bands coursing from caecum across the duodenum. The midgut was dusky initially, but seemed to reperfuse slowly. Peristalsis was noted throughout the small bowel. A Ladd's procedure was performed. The child received fluid resuscitation during and after surgery, as well as post operative antibiotics. Vital signs and fluid balances were stable post-operatively. Due to the questionable status of bowel at the time of the initial surgery, a planned re-exploration was performed 24 hours later. At the second surgery, the bowel appeared fully viable. The child did well after the second surgery. Oral feedings were initiated six days later without difficulty. The child was discharged in good condition after an additional week. The child has not developed new symptoms during follow-up two years post-surgery.

DISCUSSION

Midgut malrotation with volvulus is a true surgical emergency. An expedient diagnosis and treatment are paramount to maintaining viability of the gut. If the gut is infarcted and requiring resection, the child may need long term parenteral nutrition for short gut syndrome, or worse still, may even die. Midgut malrotation is a congenital anomaly. Under normal circumstances, the midgut herniates out of the abdomen into the umbilical cord at about eight weeks gestation. By twelve weeks, the midgut has returned into the abdomen, undergoing a 270° counter clockwise rotation as it returns, resulting in the familiar C-shaped course of the duodenum and the caecum in the right lower quadrant of the abdomen[1]. The normal bowel is fixed in position by a long mesentery extending from the upper left to the lower right abdomen. This broad mesenteric attachment prevents the bowel from twisting on itself. In malrotation, the returning bowel fails to undergo the 270° counter-clockwise rotation and fails to normally fixate. There are several variants of malrotation with varying susceptibility to volvulus[2,3]. Those forms predisposed to volvulus have a narrow mesenteric root which fails to prevent the bowel from twisting on itself.

Midgut malrotation is usually an isolated anomaly. It is invariably present in children with congenital diaphragmatic hernia, omphalocoele and gastroschisis. However, these children are not prone to volvulus as a result of the corrective surgery performed early in life. Malrotation is also seen in most, if not all, children with heterotaxy – asplenia and polysplenia. This is problematic as these infants may have severe congenital heart disease requiring treatment, thus temporising semi-elective surgery for malrotation. Midgut malrotation, in itself, is often asymptomatic. Occasionally, children may have persistent vomiting, abdominal pain or malabsorption. The diagnosis is frequently incidentally made. Classically, malrotation with volvulus presents with bilious vomiting. Children with severe vomiting or ischaemic bowel may be profoundly ill, as was the illustrated patient. Most patients present in the first month of life. However, a patient with malrotation may develop volvulus at any time in life.

Radiographs are the first line of imaging. In the newborn where any obstruction distal to the ampulla of Vater may cause bilious vomiting, radiographs may suggest an alternative diagnosis. Often, however, the radiographs are normal. Occasionally, one may appreciate malposition of bowel. Rarely, there may be mass effect seen centrally produced from the mass of volvulised bowel. If the gut is ischaemic, bowel wall thickening or pneumatosis intestinalis may be seen. Normal radiographs do not exclude the diagnosis of malrotation and thus do not preclude further work-up.

If malrotation is suspected, the study of choice is an upper GI examination[2,3] Occasionally, if an infant is severely ill and the diagnosis is suspected, the patient

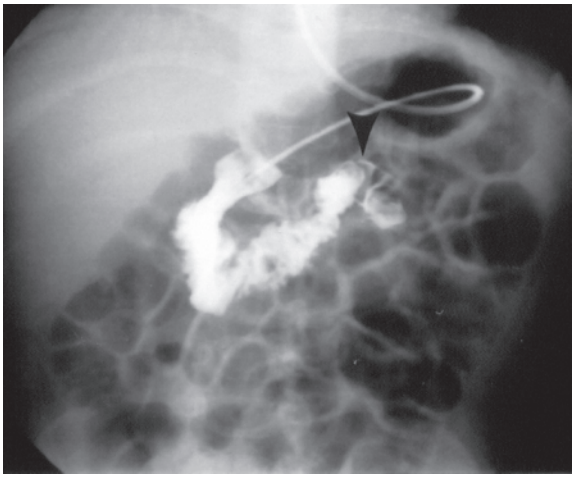

Fig. 3 Normal anteroposterior view obtained during an upper GI examination on a 10-month-old boy. The duodenum is of normal calibre throughout its course with the duodenojejunal junction (arrowhead) seen in a normal location to the left of the spine and at approximately the level of the pylorus. In this patient, the upper GI examination was facilitated by the presence of an enteric tube advanced to the duodenum. This is not usually necessary.

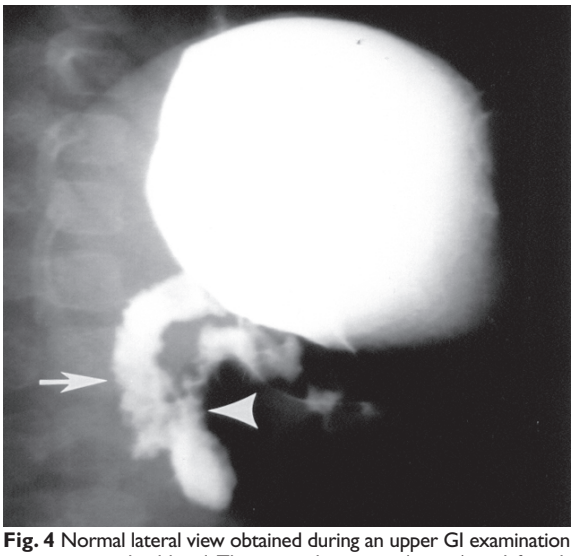

Fig. 4 Normal lateral view obtained during an upper GI examination on a six-month-old girl. The second portion (arrow) and fourth portion (arrow-head) of the duodenum are nearly superimposed in a normal, posterior location. Often, it is necessary to turn the patient slightly under fluoroscopic observation to demonstrate this anatomy.

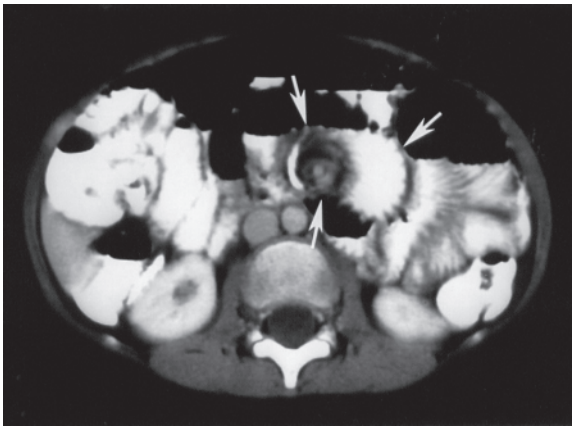

Fig. 5 Axial CT scan of a seven-year-old boy with midgut malrotation and volvulus. Note the whorled appearance of vasculature and bowel around the superior mesenteric artery (arrows). The diagnosis was confirmed by an upper gastrointestinal examination and then surgery.

may proceed directly to surgery. In most cases, the upper GI examination is performed with barium. If the infant is quite ill and there is a heightened concern for a perforation or ischaemic gut, non-ionic water soluble contrast material is used. The normal duodenal course is such that the distal duodenum (the fourth portion) extends to the left of the left spinous pedicle, and upwards to reach approximately the level of the pylorus (Fig. 3). This point of reference is the duodenojejunal junction, commonly termed the ligament of Treitz. The lateral view is important to corroborate normalcy[4]. The normal duodenum maintains a posterior location from its second portion to the fourth portion, reflecting its fixation to the posterior peritoneum (Fig. 4).

On the lateral view, the second and fourth portions of duodenum overlap. In malrotation, the duodenum takes an anomalous course. Rather than extending to the left and upwards, the distal duodenum stays rightwards and inferior. The duodenojejunal junction is poorly defined. Occasionally, as in the illustrated case, reliance solely on distal duodenal position on the anteroposterior film for the diagnosis may lead to error. In the lateral projection, the malrotated duodenum often takes an anterior course, reflecting its lack of fixation[4]. When no volvulus is present, the malrotated bowel maintains a normal calibre. With volvulus, the duodenum is partially or completely obstructed, often with a tapered or beaked appearance. The contrast may opacify a narrow spiral or corkscrew course of distal duodenum and proximal jejunum, reflecting the degrees of volvulus. If a volvulus is present, the child needs to be operated on emergently to save the gut and save the child.

As caecal position may be normal in the setting of malrotation and volvulus, a barium enema is not the preferred study for diagnosis. In equivocal cases however, small bowel follow-through or barium enema may serve as an adjunct to the upper GI in confirming an abnormality[2,3]. Cases in which the upper GI examination is normal and the enema shows malrotation are very rare[2,3]. Midgut malrotation and volvulus are occasionally diagnosed by ultrasonography or computed tomography (CT)[5,6]. As children with vomiting or abdominal pain are frequently imaged by these modalities, it is prudent for radiologists to know the findings of malrotation on cross-sectional imaging and look for them, particularly in the absence of another diagnosis. Inversion of the superior mesenteric artery and vein, with the vein to the left rather than the right of the artery, is strongly suggestive of malrotation, but is neither sensitive or specific[5]. When diagnosed on CT, malrotation is most commonly identified by malposition of bowel and lack of the

distal duodenum crossing the midline. In the setting of volvulus, both ultrasonography and CT may show a mass centrally located in the abdomen that is composed of a whorled appearance of bowel and blood vessels (Fig. 5).

Clinicians and radiologists involved in the care of children should be well-versed in the presentation, imaging and treatment of malrotation and malrotation with volvulus. Imaging plays a critical role in confirming the diagnosis. Expedient diagnosis and treatment of volvulus may be lifesaving.

ABSTRACT

Midgut malrotation with volvulus is a life-threatening surgical emergency. Prompt diagnosis and treatment is paramount to good outcome. Imaging plays a key role in confirming the diagnosis. A two-day-old male infant presented with bilious vomiting and was diagnosed by upper GI examination to have midgut malrotation and volvulus. The gut was ischaemic but viable at surgery. Radiological features of malrotation and volvulus are presented and discussed.

Keywords: Malrotation, volvulus, radiography, small intestine, neonate

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REFERENCES

1. Timor-Tritsh IE, Warren WP, Peisner DB, Pirrone E. First-trimester midgut herniation: a high-frequency transvaginal study. Am J Obstet Gynecol 1989; 161:831-3.
2. Jamieson D, Stringer DA. Small bowel. In: Stringer DA, Babyn PS, editors. Pediatric gastrointestinal imaging and intervention. 2nd ed. Hamilton, Canada: Decter, 2000; 315-474.
3. Long FR, Kramer SS, Markowitz Rl, Taylor GE. Radiographic patterns of intestinal malrotation in children. RadioGraphics 1996; 16:547-56.
4. Koplewitz BZ, Daneman A. The lateral view: a useful adjunct in the diagnosis of malrotation (letter). Pediatr Radiol 1999; 29:144-5.
5. Zerin JM, DiPietro MA. Superior mesenteric vascular anatomy at US in patients with surgically proved malrotation of the midgut. Radiology 1992; 183:693-4.
6. Paul AB, Dean DM. Computed tomography in volvulus of the midgut. Br J Radiol 1990; 63:893-4.