

# Volvulus and midgut malrotations

## What is a volvulus?

A volvulus is a complete twisting of a loop of intestine around its mesenteric attachment site. This can occur at various locations of the gastrointestinal (GI) tract, including the stomach, small intestine, caecum, transverse colon and sigmoid colon.

## What is midgut malrotation?

Midgut malrotation refers to twisting of the entire midgut about the axis of the superior mesenteric artery (SMA).<sup>[1]</sup>

A knowledge of embryology is necessary to understand the mechanisms that lead to the development of volvulus and malrotation.<sup>[2]</sup> At the fourth week of gestation, the GI system is a straight tube centrally located in the abdomen. During the ensuing eight weeks, the midgut rotates and becomes fixed to the posterior abdominal wall. Arrest of development at any stage narrows the mesenteric base and impairs fixation, leaving the bowel at high risk for volvulus.

## Malrotation epidemiology

- The true prevalence of malrotation is unknown, since many asymptomatic patients go undiagnosed. Estimates vary, from 1 in 500 to 1 in 2500 live births.<sup>[3]</sup> Either sex can be affected with the anomaly.
- Approximately 90% of patients with malrotation are diagnosed within the first year of life, 80% of whom are diagnosed within the first month of life.<sup>[4]</sup>
- Thereafter, it can present at any age and may present in adults of any age.<sup>[5] [6] [7]</sup>

- Recurrent bowel obstruction in patients with previous abdominal operation for midgut malrotation is mostly due to adhesions but very few reported cases have been due to recurrent volvulus.<sup>[8]</sup>

## Volvulus epidemiology

Most cases of volvulus associated with malrotation occur in infants and young children. The incidence of malrotation-associated volvulus decreases with age.<sup>[9]</sup>

## Presentation

It should be remembered that malrotation and volvulus are two distinct entities.

### Malrotation symptoms

Non-rotation may be asymptomatic and be detected as an incidental finding during GI imaging for some other purpose. Malrotation may cause intermittent symptoms of intestinal obstruction but if a volvulus develops, the obstruction is typically complete. The presenting features will also vary, depending on age.

Bilious vomiting is the key presenting symptom. Neonates with bilious (green) vomiting should be considered to have intestinal obstruction until proved otherwise.<sup>[10]</sup> . Infants presenting in the first 24 hours after birth through the first week of life tend to have more severe obstruction and present with bilious vomiting and feeding intolerance. After the age of 2 months, bilious and non-bilious vomiting occur with equal frequency. Other symptoms may include failure to thrive, anorexia, constipation, bloody stools and intermittent apnoea.<sup>[11]</sup>

Older children may present more insidiously with cyclical vomiting, recurrent abdominal pain, protein-calorie malnutrition and immunodeficiency. Once intestinal ischaemia develops, pain becomes the most pronounced symptom.

Intestinal malrotation can be a rare cause of chronic abdominal pain in adults. The diagnosis is often missed initially, leading to increased morbidity.<sup>[12]</sup>

### Volvulus symptoms

Rarely, volvulus can occur in utero and even more rarely can lead to intrauterine death.<sup>[13]</sup> Abdominal examination may be normal in the early stages or may show distension.<sup>[14]</sup>

In midgut volvulus associated with malrotation, the entire midgut from the proximal jejunum to the proximal colon may twist around the mesenteric base. Volvulus may lead to intestinal necrosis in several hours without surgical treatment. Rapid-onset and bilious vomiting occurs, leading to metabolic acidosis, lactataemia, oliguria, hypotension and shock with advancing ischaemia.<sup>[9]</sup>

Careful examination may reveal a palpable abdominal mass in some patients. If ischaemia develops, the presentation is of acute abdomen, with significant abdominal distension, and signs of peritonitis. Blood or sloughed tissue may pass per rectum. Tachycardia, hypovolaemia and septic shock herald the onset of gangrene.<sup>[15]</sup>

## Differential diagnosis

In the acute phase, the differential diagnosis is of acute abdomen/obstruction. Chronic vague abdominal symptoms in older children or adults raise the possibility of a wide range of other symptoms, which should include:

- Annular pancreas.
- Appendicitis.
- Cholecystitis.
- Colic.
- Constipation.
- Duodenal atresia.
- Duodenal web.
- Gastroenteritis.
- Gastro-oesophageal reflux.
- Henoch-Schönlein purpura.
- Hepatitis B.

- Hirschsprung's disease.
- Hypertrophic pyloric stenosis.
- Incarcerated hernia.
- Intussusception.
- Meckel's diverticulum.
- Necrotising enterocolitis.
- Ovarian torsion.
- Pancreatitis.
- Peptic ulcer.
- Perforated viscus.
- Renal stones.
- Sickle cell crisis.
- Urinary tract infection.

## Investigations<sup>[16]</sup>

### Laboratory investigations

The diagnosis is usually made clinically and management should not be delayed in order to obtain the results of laboratory tests.

FBC helps to assess the severity of the illness; a raised white cell count may be found in sepsis or gangrene and a low haemoglobin may suggest venous oozing. Inflammatory markers (eg, CRP) may be normal in the very early stages of volvulus.<sup>[9]</sup>

Renal function and venous blood gas monitoring may help to assess the patient's general condition and detect dehydration, sepsis and acidosis:

- Large amounts of fluid can migrate into the bowel lumen and interstitial space and, in such patients, dehydration can occur without diarrhoea and vomiting.

- Hyponatraemia, hyperkalaemia, metabolic acidosis, increased urea and creatinine, hypochloraemia and lactic acidosis can occur in such cases.

## **Plain radiography**

### **Malrotation**

In simple malrotation, plain radiographs are frequently normal. Upright, supine and lateral radiographs may be helpful in diagnosing bowel obstruction but may be more contributory in large rather than small bowel obstruction. Radiographs taken several hours apart may be helpful. Dilated small-bowel loops, marked gastric or proximal duodenal dilatation - with or without intestinal gas - and air-fluid levels may be seen. Normal or equivocal results should not delay progress to other tests if the clinical situation is deteriorating.<sup>[17]</sup>

### **Volvulus**

In midgut volvulus, the classic radiographic finding is a partial duodenal obstruction (dilation of both stomach and proximal duodenum, with a small amount of distal bowel gas). This is known as the 'double bubble' sign. Complete obstruction of the duodenum may also be found; less frequently, a gasless abdomen, ileus or distal small bowel obstruction with multiple dilated loops and air-fluid levels. These are ominous signs.<sup>[18]</sup>

## **Contrast studies**

### **Malrotation**

In malrotation, the duodenojejunal (DJ) junction is misplaced, either at, or to the right of, the midline. Various displacements of lower bowel structures may also be seen.

### **Volvulus**

An upper GI (UGI) contrast series is the investigation par excellence if volvulus is suspected. In a child, the UGI series is performed with a small amount of barium being administered either by bottle or through a nasogastric tube. Various patterns may be observed, including dilation of the proximal duodenum with a 'bird-beak' obstruction and a spiral or corkscrew duodenal configuration. Selected patients may need studies of the lower bowel, using a barium enema.

## **Other imaging studies**

Volvulus:

- Ultrasound may reveal a midline abdominal mass in suspected volvulus. Ultrasound and CT scanning can help to confirm malrotation by identifying the position of the mesenteric vessels. CT scan may reveal 'the coffee bean' in sigmoid volvulus.
- The 'whirlpool sign' on colour Doppler may show a whirlpool pattern of flow within the superior mesenteric vein, indicating malrotation with volvulus.<sup>[19]</sup>
- In malrotation, abnormal relationships between the superior mesenteric artery and superior mesenteric vein may be visualised, as may an abnormal position of the third part of the duodenum.<sup>[20]</sup>
- Some authors have suggested that ultrasound may be a suitable first-line imaging modality. However, there are technical and training challenges in its implementation;<sup>[21]</sup> it may also be insufficient to completely rule-out the possibility of malrotation and volvulus if suspected.<sup>[21] [20]</sup>

## Treatment and management

### Non-surgical treatment for malrotation

- This may be appropriate for older patients with intestinal malrotation who are asymptomatic but they should be warned that volvulus can occur at any time.
- Observation and GI decompression with a nasogastric or orogastric tube should be commenced and a close watch kept for the development of symptoms or signs suggestive of intestinal obstruction.<sup>[22]</sup>

### Surgery

#### Malrotation

Because volvulus is such a devastating complication in children, evidence supports operative treatment of asymptomatic malrotation, using Ladd's procedure. This is particularly appropriate for young children with no existing comorbidity, as they recover quickly. There is no similar evidence base to support this approach in adults.<sup>[23]</sup>

#### Volvulus

- Ladd's procedure is the treatment of choice in most cases.

- Volvulus is corrected by rotating the small intestine in an anticlockwise direction, the caecum being placed in the left abdomen and the duodenum directed down the right paravertebral gutter. A second-look laparotomy may be used 36 hours later to ensure viability of the remaining bowel.
- A laparoscopic variation of Ladd's procedure has been used in some centres, with the general advantage of decreased adhesions and scarring; however, good visualisation of the entire bowel is necessary. [24]
- Laparoscopy is a particularly suitable investigative procedure for children presenting with acute abdomen. One study found that it produces better cosmesis, less postoperative pain and earlier return of bowel function than laparotomy. [25]

## Complications

### Volvulus

- Complete and persisting midgut volvulus leads to intestinal ischaemia, mucosal necrosis and sepsis. Untreated, perforation, peritonitis and death soon follow.
- Chronic intermittent volvulus may cause malabsorption with constipation interspersed with diarrhoea. [26]
- Postoperatively, the main complication is short-gut syndrome with the consequent problems surrounding parenteral nutrition, ie line sepsis, hepatobiliary dysfunction and growth restriction. [27]

## Prognosis

Although malrotation can usually be successfully treated with early diagnosis, sudden catastrophic events, such as intense ischaemia and intestinal necrosis, may occur and may be fatal. [28]

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## Further reading

- [Mohan P, Ramamoorthy M, Venkataraman J](#); Clinical vistas: nonrotation of the intestine. CMAJ. 2008 Jul 1;179(1):49-50.

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