

CHAPTER 65

INTESTINAL MALROTATION AND MIDGUT VOLVULUS

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Introduction

Malrotation is a spectrum of anatomic abnormalities of incomplete rotation and fixation of the intestinal tract during foetal development. Disorders of intestinal rotation and fixation are of paramount importance to the paediatric surgeon because they are most commonly seen in infancy and childhood and can have catastrophic consequences when midgut volvulus occurs. Early diagnosis and surgical treatment of this disorder can be life saving.

Demographics

Malrotation is thought to occur in approximately 1 in 500 live births.^{1,2} The exact incidence is not known because many patients may live their entire lives without experiencing problems or consequences from their malrotation. Approximately 80% of patients with malrotation will present within the first month of life, and of those, most will present within the first week of life.¹⁻⁴

Embryology/Pathophysiology

Embryology

The adult midgut extends from the second portion of the duodenum to the proximal third of the transverse colon, and is derived from the embryologic midgut loop. The normal development of the human intestine involves two processes: rotation of the midgut and the subsequent fixation of the colon and mesentery. These processes occur in three stages.

Stage 1 consists of umbilical cord herniation, lasting from approximately weeks 5 to 10 of embryonic development. The midgut lengthens disproportionately during this period and undergoes rotation around the superior mesenteric artery (SMA) axis for a total of 270° in the counterclockwise direction. Stage 2 is the return of the midgut loop back into the abdomen; it occurs at approximately weeks 10 to 11. As the intestines re-enter the abdominal cavity, the cephalad midgut completes its 270° counterclockwise rotation as the caudad midgut also completes its rotation, resulting in the duodenum coursing inferior and posterior to the SMA and the caecum located in the right lower quadrant. When completed, this rotation ensures that the attachment of the base of the midgut loop is spread along a diagonal stretching from the ligament of Trietz on the left upper quadrant to the ileocecal junction in the right lower quadrant of the abdomen. Stage 3 is the period of fixation, and lasts from the end of stage 2 until just after birth. The descending and ascending colon mesenteries fuse with the retroperitoneum, and the small bowel is fixed by a broad mesentery from the duodenojejunal junction in the left upper quadrant to the ileocecal valve in the right lower abdomen. The broad base of the small bowel mesentery stabilises its position and prevents volvulus.^{5,6}

Malrotation can be grouped into syndromes arising from anomalies of three categories: migration, rotation, and fixation.

Anomalies of Migration

Omphalocele

Return of the midgut from the yolk sac back into the abdominal cavity is usually completed by week 12 of intrauterine life. This enables the anterior abdominal wall mesodermal folds to meet at the central umbilical ring, thereby closing the anterior abdominal wall. When the return of the midgut is delayed or arrested, the anterior abdominal wall folds fail to meet, and an omphalocele in the central umbilical area of the abdomen is the result.

Congenital diaphragmatic hernia

If return of the midgut into the abdominal cavity, which divides the celomic cavity into peritoneal and pleural compartments, occurs before the closure of the pleuroperitoneal membrane at 8 weeks gestation, part of the returning midgut loop may herniate into the pleural cavity. This occurs usually in the posterolateral position on the left side.

Subhepatic appendix

With completion of the 270° rotation of the ileocecal limb of the midgut loop, the caecum is brought to the right upper quadrant of the abdomen. The caecum with the attached appendix then further descends down to the right lower quadrant position in the right iliac fossa and becomes fixed to the posterior abdominal wall. The caecum and appendix may fail to migrate and remain in that subhepatic position. This condition may cause a serious diagnostic dilemma in acute appendicitis.

Anomalies of Rotation

Nonrotation

Nonrotation may occur when the midgut returns to the abdominal cavity en masse without rotating. Then, the first and second parts of the duodenum are situated normally but the third and fourth parts descend vertically downward along the right side of the superior mesenteric artery. The small bowel lies on the right and the colon is doubled on itself to the left of midline.⁷

Reversed rotation

Reversed rotation has the caecum and colon positioned posterior to the superior mesenteric vessels, and the duodenum subsequently crosses anterior to it.

Malrotation

Malrotation is a spectrum of abnormalities that occurs when the normal process of rotation is arrested at various stages. Most frequently, the duodenojejunal flexure is located inferiorly and to the right of the midline. In addition, the caecum has failed to reach its normal position in the right iliac fossa and lies in a subhepatic or central position.

Anomalies of Fixation

Volvulus neonatorum

A normal fixation of the midgut loop results in a broad diagonal attach-

ment of the loop to the posterior abdominal wall, extending from the ligament of Trietz to the ileocecal junction. With malfixation, the distance between these two points of attachment may become shortened, leaving the midgut loop hanging on a narrow and unstable pedicle that easily predisposes to twisting (volvulus) and strangulation.

Ladd's bands

When the caecum has failed to descend from the right upper quadrant to the right iliac fossa, anomalous fixation may occur, whereby dense fibrous bands (Ladd's bands) extend from the caecum and right colon across the duodenum to the retroperitoneum of the right upper quadrant. These bands may cause duodenal obstruction via extrinsic compression; however, the obstruction of the duodenum is most commonly caused by torsion at the base of the midgut mesentery. Bands may also form between the colon and the duodenum, drawing them closer together and predisposing the midgut towards volvulus.

Mobile caecum

Failure of fixation of the caecum to the posterior abdominal wall results in a floating caecum that may predispose to cecal volvulus.

Internal hernias

Failure of fixation of the mesentery of the duodenum, right colon, or left colon may result in the formation of potential spaces for internal or mesocolic hernias. Internal hernias are associated with partial bowel obstructions, as there may be recurrent entrapment of bowel, which may eventually lead to obstruction and strangulation.⁵

Other Associated Anomalies

Malrotation may be present in patients with heterotaxy syndrome (asplenia or right isomerism and polysplenia or left isomerism). Patients presenting with this syndrome should be investigated for the possibility of malrotation. Malrotation may also be seen in conjunction with intestinal atresias and may be the cause for developing atresias in some of these patients. Vecchia et al., in a large series, found that 28% of infants with duodenal atresias had malrotation and 19% of infants with jejunoileal atresia had malrotation.⁸

Clinical Presentation

The classic presentation of malrotation with acute midgut volvulus is a neonate with bilious emesis. The point of obstruction is typically beyond the ampulla of Vater, as demonstrated by the bilious emesis. However, this symptom is not synonymous with the diagnosis of malrotation. A majority (around 60%) of infants with bilious emesis will prove to have no anatomic obstruction, but imaging is necessary to exclude the potentially catastrophic event of midgut volvulus as a consequence of malrotation. Most patients with malrotation and many with volvulus have a normal history and have a normal physical exam. Other acute symptoms that may occur with malrotation are intermittent abdominal pain, diarrhoea, constipation, and haematochezia. The latter involves 10–15% of patients and is associated with a poorer prognosis because it is indicative of bowel ischemia.⁹ Patients presenting with peritonitis, abdominal distention, bloody stools, and haemodynamic instability (signs and symptoms of shock) have a much worse prognosis; the clinician may be misled from the diagnosis of malrotation with volvulus due to the other symptoms related to sepsis.

Malrotation may present in an insidious manner with chronic symptoms that develop over days, months, and even years. In one series by Spigland et al., when malrotation presented beyond the neonatal period, the delay in diagnosis was a mean of 1.7 years.¹⁰ Chronic symptoms include intermittent pain, intermittent vomiting, malabsorption, and failure to thrive. Patients may be chronically misdiagnosed with other abdominal pain syndromes, "cyclic vomiting," or even psychological disorders.¹¹ Howell et al. noted that 70% of children

presenting with malrotation had clinical evidence of malnutrition.¹²

Physical Examination

There are often very few, if any, diagnostic physical exam findings with malrotation and midgut volvulus. Late presentations may have abdominal distention and abdominal tenderness, and some patients may have haemodynamic instability if bowel necrosis and sepsis have occurred. The herald sign is bilious emesis and requires prompt diagnostic studies in order to prevent bowel ischaemia and necrosis.

Investigations

Imaging

It is reasonable to start with abdominal radiographs as the initial evaluation of a patient with bilious emesis or suspected malrotation. Patients should have two views of the abdomen: an anteroposterior supine view and either an anteroposterior upright view or a cross-table lateral view. Rarely do the radiographs suggest the diagnosis of malrotation. Instead, they help to exclude other aetiologies for the patient's symptoms and serve to guide further imaging. The most common bowel gas pattern in the setting of malrotation is normal. Findings suggestive of an abnormal location of bowel include

- the presence of proximal small bowel on the right; and
- a disproportionate dilatation of the duodenum with a "double bubble"—this may be seen with severe duodenal obstruction due to volvulus or bands.¹

Upper Gastrointestinal Series

An upper gastrointestinal (UGI) series is the preferred test for radiographic diagnosis of malrotation and volvulus (Figures 65.1 and 65.2). It is usually performed with barium, except in cases of a very sick infant or child in whom the presence of infarcted bowel or perforation is possible, in which case water-soluble contrast is used. It is important to document the first bolus of contrast medium through the duodenum in the anteroposterior as well as the lateral projection. This can be done by quickly rotating the patient to the lateral position once the duodenojejunal junction is reached. The main radiographic signs of malrotation are¹

- lateral radiograph suggesting that the distal duodenum is not attached in the retroperitoneum;
- low or medial position of the duodenojejunal junction;
- spiral "corkscrew" or Z-shaped course of the duodenum and proximal jejunum; and
- location of the proximal jejunum in the right abdomen.



Figure 65.1: UGI depicting malrotation with abnormally low position of the ligament of Treitz.

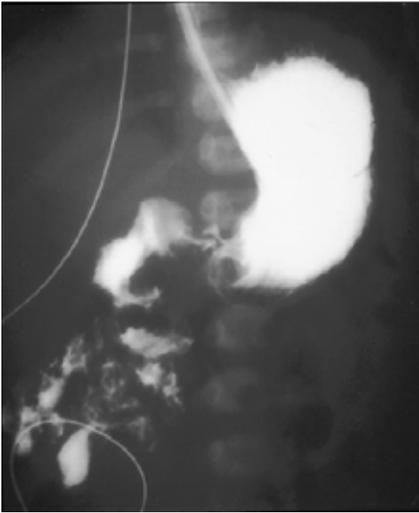


Figure 65.2: UGI depicting malrotation with all of the small bowel on the right side of the abdomen.

Ultrasound

Ultrasound is not the preferred imaging modality for malrotation, but it may be useful for some physicians with limited imaging modalities. Ultrasound can be used to evaluate other abdominal abnormalities and may be used to visualise the position of the mesenteric vessels. Normally, the superior mesenteric vein is to the right of the artery. In malrotation, the vein is frequently on the left, or it may rotate completely around the artery. These findings are neither sensitive nor specific for malrotation or volvulus, and should be further evaluated with additional diagnostic imaging studies, typically a UGI.

In resource-poor settings in most developing parts of Africa, where diagnostic facilities are limited or unavailable, it is safer to assume and handle all cases of bilious vomiting in a neonate as a potential malrotation syndrome with midgut volvulus. Such babies should be vigorously resuscitated and explored to avert the catastrophe of an entire midgut strangulation and gangrene, leading to short bowel syndrome.

Management

Preoperative Management

Preoperative management is focused on stabilising the patient and preparing for prompt surgery. The patient should be resuscitated with isotonic fluid (lactated Ringer's or normal saline) with an intravenous (IV) fluid bolus of 20 ml/kg, then kept on isotonic maintenance fluids, nothing by mouth (NPO), and nasogastric tube (NGT) decompression until surgery. The patient's urine output should be monitored; fluid resuscitation may depend on urine output or haemodynamics.

Operative Management and Technique

Ladd's procedure, first described in 1936, corrects the fundamental abnormality associated with malrotation and volvulus. The procedure consists of laparotomy with the following steps:^{13,14}

1. The bowel is eviscerated and the entire bowel and mesenteric root are inspected.
2. The midgut volvulus, if one exists, is derotated in a counterclockwise direction.
3. Ladd's bands are lysed and the duodenum is straightened.
4. An appendectomy is performed.
5. The bowel is returned into the abdominal cavity with the caecum in the left lower quadrant.^{13,14}

A laparoscopic approach may be feasible in older patients, but availability and technical comfort with this operation may be less than optimal.



Figure 65.3: Severe small bowel ischaemia due to volvulus.

Complicated cases with significant bowel ischaemia still demand an open approach.

The occurrence of an entire midgut strangulation and gangrene should be considered a disaster that must be prevented, especially in resource-poor settings where total parenteral nutrition (TPN) is neither available nor affordable (Figure 65.3). If widespread ischaemia of the midgut is observed at laparotomy, limited bowel resection and a second-look exploration 48 to 72 hours later to confirm viability of the remaining bowel are advised.

Postoperative Complications

Postoperative complications are similar to other surgical procedures and include infection and ileus. Patients with malrotation have been known to have postoperative intestinal dysmotility (pseudo-obstruction) that may delay return of the bowel function and contribute to their postoperative ileus. Normalisation of gut function occurs slowly in some children. Some reports in the literature suggest that there is an underlying functional abnormality of gut innervation associated with or as a consequence of malrotation.^{15,16}

If the patient had bowel necrosis and required a resection, depending the length of residual viable bowel, the patient may have short bowel syndrome. This condition can be quite difficult to handle, and typically requires parenteral nutrition for at least the short term, and potentially long term.

Patients may also have strictures, either from their resection with anastomosis, or potentially from areas of ischaemia that did not require resection. These patients may not require additional surgeries, or they may require subsequent bowel resection of the stricture and/or revision of the strictured anastomosis.

Prognosis and Outcome

Survival of children with malrotation and volvulus is high (>80%); however, despite prompt diagnosis and surgery, a significant minority of patients still die or suffer substantial morbidity due to loss of intestines.

Factors associated with an increased mortality include:

- younger age (especially less than 30 days old);¹⁷
- other clinical abnormalities; and
- bowel necrosis.^{4,17}

Prevention

There are no preventive measures to take regarding this disease process. Early detection and treatment are the only measures to help prevent a poor outcome from malrotation with volvulus.

Ethical Issues

The patient with short bowel syndrome as a result of malrotation with volvulus that occurred either in utero or in the neonatal period presents a real treatment challenge in the industrialised nations and may be even more difficult in countries where resources are more limited. These patients require TPN and significant medical care to prevent dehydration and failure to thrive. In addition, these patients require central lines for prolonged periods of time and are often plagued by complications from the central lines.

Evidence-Based Research

In the absence of comparative studies, a recent review of malrotation and volvulus is shown in Table 65.1.

Table 65.1: Evidence-based research.

Title	Malrotation and volvulus in infancy and childhood
Authors	Millar AJW, Rode H, Cywes S
Institution	University of Cape Town and Red Cross Children's Hospital, Rondebosch, Cape Town, South Africa
Reference	Sem Pediatr Surg 2003; 12:229–236
Problem	Review of malrotation and volvulus.
Intervention	Diagnostic radiology and surgical treatment.
Comparison/control (quality of evidence)	Comparison of recent cases with previously published cohort.
Historical significance/comments	A large series of patients reviewed for presenting symptoms, evaluation, and surgical management.

Key Summary Points

1. Malrotation is a spectrum of anatomic abnormalities related to fixation of the intestinal tract.
2. Bilious emesis in a newborn should be considered midgut volvulus until proven otherwise.
3. A prompt diagnostic UGI study should be done on any newborn with bilious emesis to rule out malrotation with midgut volvulus.
4. If investigative studies cannot be done, then the patient should have fluid resuscitation and prompt surgical exploration to prevent the catastrophic complications of midgut volvulus.
5. Ladd's procedure for malrotation includes detorsion of volvulus if one is present, lysis of dense fibrous bands (Ladd's bands), placement of small bowel and large bowel in abdomen in nonrotated manner, and appendectomy.
6. There is an increased mortality for malrotation in younger patients, patients with clinical abnormalities, or those with bowel necrosis.
7. Patients may have a delay in the return of bowel function after surgery, especially if volvulus was present.

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