BMJ Best Practice

Intestinal malrotation

Straight to the point of care

Last updated: Jan 27, 2022

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Summary

Intestinal malrotation is an entire spectrum of rotational and fixation disturbances that can occur during embryonic development.

The anatomical variant that poses the highest risk of volvulus is a narrow midgut mesenteric base accompanied by lack of retro-peritoneal midgut fixation. This variant cannot be reliably determined from any radiological studies.

Malrotation predisposes patients to a risk of midgut volvulus.

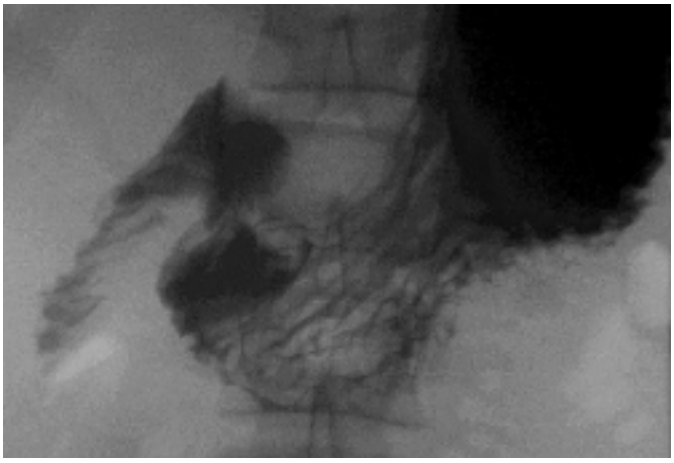
Emergency surgical consultation is appropriate before obtaining any diagnostic studies if midgut volvulus is suspected. Upper gastrointestinal contrast studies should be ordered in patients who are stable.

Treatment is surgical correction, the procedure of choice being the Ladd's procedure.

Definition

Intestinal malrotation is a term used to describe an entire spectrum of rotational and fixation disturbances that can occur during embryonic development.

The most significant pathological concerns in malrotation are a lack of intestinal fixation to the retro-peritoneum and a narrow midgut mesenteric base that predisposes to a twisting of the small bowel in the form of midgut volvulus. This condition creates an abrupt obstruction of the duodenum, resulting in bilious vomiting.



Malrotation with volvulus causing complete obstruction of the duodenum that does not sweep to the left, as seen on upper GI contrast

From the collection of Dr S.D. St Peter

If the twisting at the base also obstructs flow in the superior mesenteric artery, the entire small intestine and proximal colon may become acutely ischaemic and subsequently necrotic within a few hours. Abnormal caecal attachments to the right upper peritoneal cavity (i.e., the Ladd’s bands) can cross the second portion of the duodenum creating an extraluminal low-grade obstruction of the duodenum, which may present with signs of partial obstruction.

Patients with malrotation are at risk for volvulus and should be identified whenever possible to allow for proper therapy.

Epidemiology

All reported epidemiological statistics on rotational abnormalities are likely to dramatically underestimate the true prevalence, given that many people with incomplete rotation remain asymptomatic and are never diagnosed. Rotational abnormalities have been noted as an incidental finding in 0.2% of upper gastrointestinal contrast studies.[1] This denominator is also misleading, as it selects patients with some illness given that they required such a study. Autopsy series have documented non-rotation in 0.5%.[2] Those who come to clinically evident presentation have been estimated as 1 in 6000 live births.[3]

Aetiology

The primitive midgut becomes identifiable in the fourth week of gestation.[4] In the 5-week embryo, the midgut is a linear tube from duodenum to rectum suspended from the dorsal abdominal wall by a continuous mesentery.[5]At this stage, the duodenum begins extending in a straight anterior direction into the cephalic limb (jejunum and ileum) to the vitelline duct, which communicates with the extra-corporeal yolk sac. The duct exits at the terminal point of the superior mesenteric artery (SMA). This cephalic, or duodeno-jejunal, limb runs parallel and superior to the SMA. The caudal, or caeco-colic, limb courses posteriorly, running parallel and inferior to the SMA to the cloaca.

At 6 weeks, the midgut herniates out of the abdomen into the umbilical cord until the 10th week, when it returns. There is a 90° anti-clockwise rotation during herniation and an additional 180° rotation during return to a complete 270° anti-clockwise rotation.[5]

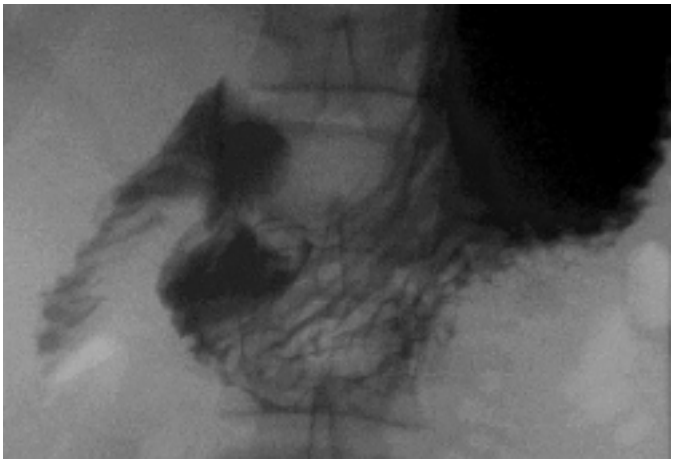
During the return, while the caecum is completing a separate 270° anti-clockwise rotation anterior to the small intestine, the colonic attachments to the posterior abdominal wall develop.

Rotation may be arrested at any point in this process, creating subsequent abnormalities in intestinal fixation points, and the myriad conditions described by the term malrotation.

Pathophysiology

In the scenario of little or no rotation, the duodenum does not completely rotate and cross posterior to the SMA to allow creation of a normal ligament of Treitz on the left side of the abdomen. The duodenum continues out into the proximal jejunum on the right side of the SMA continuing to the terminus of the SMA within the ileum. The remainder of the bowel courses back so that the colon is on the left side of the SMA. If lateral attachments (Ladd's bands) between the caecum and the right upper abdominal wall are created in direct proximity to the duodenum, as in the circumstance of classic incomplete rotation, the caecum will lie anterior to the duodenum and the proximal and distal portions of the small bowel mesentery are folded, allowing the caecum to be located back to the right side of the SMA.

If there is less rotation initially, as in non-rotation, the duodenum then lies at the base of the mesentery on the right and the colon at the base on the left of the SMA with a variable distance between these two points. The relative narrowness between these two fixation points in either rotational anomaly is what creates the risk of midgut volvulus. A shorter distance between these fixation points allows the narrow pedicle of the small bowel to easily twist around into a volvulus. An abrupt obstruction of the duodenum results, creating the bilious vomiting seen with volvulus.



Malrotation with volvulus causing complete obstruction of the duodenum that does not sweep to the left, as seen on upper GI contrast

From the collection of Dr S.D. St Peter

If the twisting at the base also obstructs flow in the SMA, the entire small intestine and a portion of the colon become acutely ischaemic, and necrosis may soon follow. As such, midgut volvulus causes significant morbidity and mortality unless emergency surgical intervention is performed to prevent irreversible loss of most of the intestine.[6]

In addition to volvulus, Ladd's bands may contribute to more subtle and likely chronic symptoms from a partial duodenal obstruction. Without concomitant volvulus, these symptoms rarely cause significant morbidity.

Classification

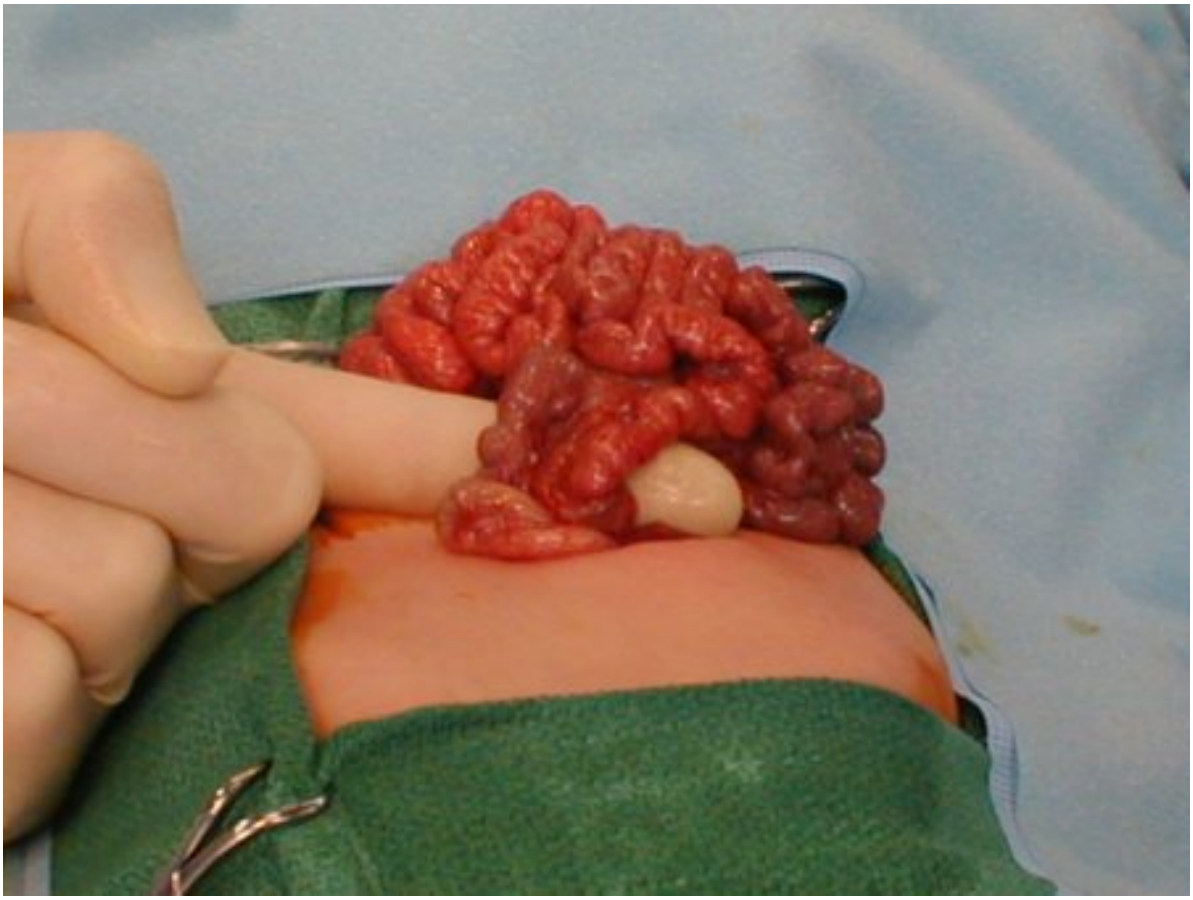
Clinical classification schema

Normal rotation

- Two separate, but connected, 270° anti-clockwise turns occur in the foregut-midgut junction and midgut-hindgut junction resulting in a retro-peritoneal duodenum, ligament of Treitz on the left of the spine, and caecum fixed to the right lower lateral abdominal wall. Hence, a broad-based mesentery is created, fixed at opposing sites in the left upper abdomen and right lower abdomen.
- Occurrence of midgut volvulus is difficult in this setting without secondary adhesions present anteriorly.

Incomplete rotation (or classic malrotation) is limited (90° to 180°) counterclockwise rotation in the foregut-midgut and midgut-hindgut junctions resulting in:

- Failure of the duodeno-jejunal limb to rotate posterior and to the left of the superior mesenteric artery, resulting in the lack of a ligament of Treitz, or one that lies to the right of the spine
- Incomplete rotation of the caeco-colic limb, which normally leads to Ladd's bands attaching the caecum across the duodenum in the right upper abdomen
- Close proximity of the fixation points for the proximal and distal midgut mesentery.



Narrow base of mesentery in malrotation

From the collection of Dr S. Shew

Non-rotation

- Minimal (<90°) rotations occurring in the primitive gut junctions around the superior mesenteric artery resulting in no ligament of Treitz, lack of lateral caecal fixation, and variable proximity between the proximal and distal midgut mesentery.

Reverse rotation

- Partial clockwise rotation of the foregut-midgut junction anterior to the superior mesenteric artery; this is rare.

Case history

Case history #1

A 7-day-old female infant is brought to the emergency department with acute-onset bilious vomiting of 6 hours' duration. The patient has previously tolerated breastfeeding without emesis and has stooled normally. Physical exam is benign without significant tenderness or distention.

THEORY

Case history #2

A 1-month-old male infant is brought to the emergency department with acute onset of inconsolable discomfort and bilious vomiting of 6 hours' duration. Physical exam reveals a tight, distended abdomen with some dark blood in the nappy. He is tachycardic and tachypnoeic.

Approach

The diagnostic approach follows the pattern of history, examination, and radiological evaluation. Emergency surgical consultation is appropriate before obtaining any diagnostic studies if midgut volvulus is suspected, as emergency surgical exploration without pre-operative studies may be warranted if peritonitis is present or a gas-less abdomen is seen on abdominal x-ray. If the patient appears stable, an urgent upper gastrointestinal (GI) contrast study should be performed to identify the patient who will need an operation for malrotation and/or midgut volvulus. If the history and physical findings are more suggestive of bowel ischaemia/necrosis (e.g., peritonitis), which may occur with midgut volvulus, then immediate surgical intervention is performed without radiological investigation in order to expedite diagnosis and reversal of ischaemia.

History

Of patients with malrotation who become symptomatic, 50% to 75% do so in the first month and about 90% within the first year of life.[6]

The presence of bilious (dark green) emesis is the key factor in immediately alerting the carer to consider the possibility of midgut volvulus. Any patient without previous abdominal surgery and/or an upper GI contrast study showing normal rotation, who has a history of green vomiting, should undergo emergency investigation to rule out midgut volvulus.

In the presence of malrotation with midgut volvulus, the vomiting is often sudden in onset and can be associated with severe abdominal pain. Carers of infants may give a history of the child becoming inconsolable for a period of time. Abdominal pain may be out of proportion to physical findings. However, history or duration of abdominal pain can otherwise be challenging to elicit in infants and young children, who account for most of the patients with symptomatic malrotation.

More subtle and chronic symptoms are frequently found in malrotation patients without volvulus. Moreover, patients may be asymptomatic and are discovered to have malrotation incidentally on work-up of other unrelated conditions.

Some less common congenital anomalies can be found with various degrees of malrotation (i.e., gastroschisis, omphalocoele, congenital diaphragmatic hernia, and heterotaxy). The presence of heterotaxy adds another layer of complexity to the variation of malrotation patterns. Although a high percentage of these patients have some element of malrotation, the risk of volvulus is still evident, albeit lower than with classic incomplete rotation.

The carer may also see dark blood in the nappy.

Physical examination

Abdominal exam findings can vary substantially depending on the presence and duration of midgut volvulus. Without volvulus, the exam is frequently benign and history is more elaborative. Early in the setting of midgut volvulus, the exam may demonstrate only minimal or no tenderness and no distention, as the initial point of bowel obstruction is in the duodenum. Later during midgut volvulus, examination can reveal a more worrying and obvious presentation of abdominal distention, peritoneal signs, and septic shock with tachycardia and tachypnoea. These are ominous signs of ongoing intestinal ischaemia/infarction.

Summary of clinical findings

Features of obstruction with ischaemia (midgut volvulus with vascular compromise) include:

- Acutely ill patient with severe abdominal pain
- Bilious vomiting
- Tachycardia
- Tachypnoea
- Abdominal tenderness
- Acidosis
- Signs of peritoneal catastrophe (re-bound and guarding).

Features of obstruction without ischaemia (midgut volvulus without vascular compromise) include:

- Bilious vomiting
- Crampy abdominal pain in waves
- Non-tender abdomen
- Non-distended abdomen
- No severe physiological perturbation.

Features of intermittent or partial volvulus or obstructing Ladd's bands include:

- Intermittent vomiting
- No signs of acute illness
- Intermittent abdominal pain (typically post-prandial)
- Weight loss.

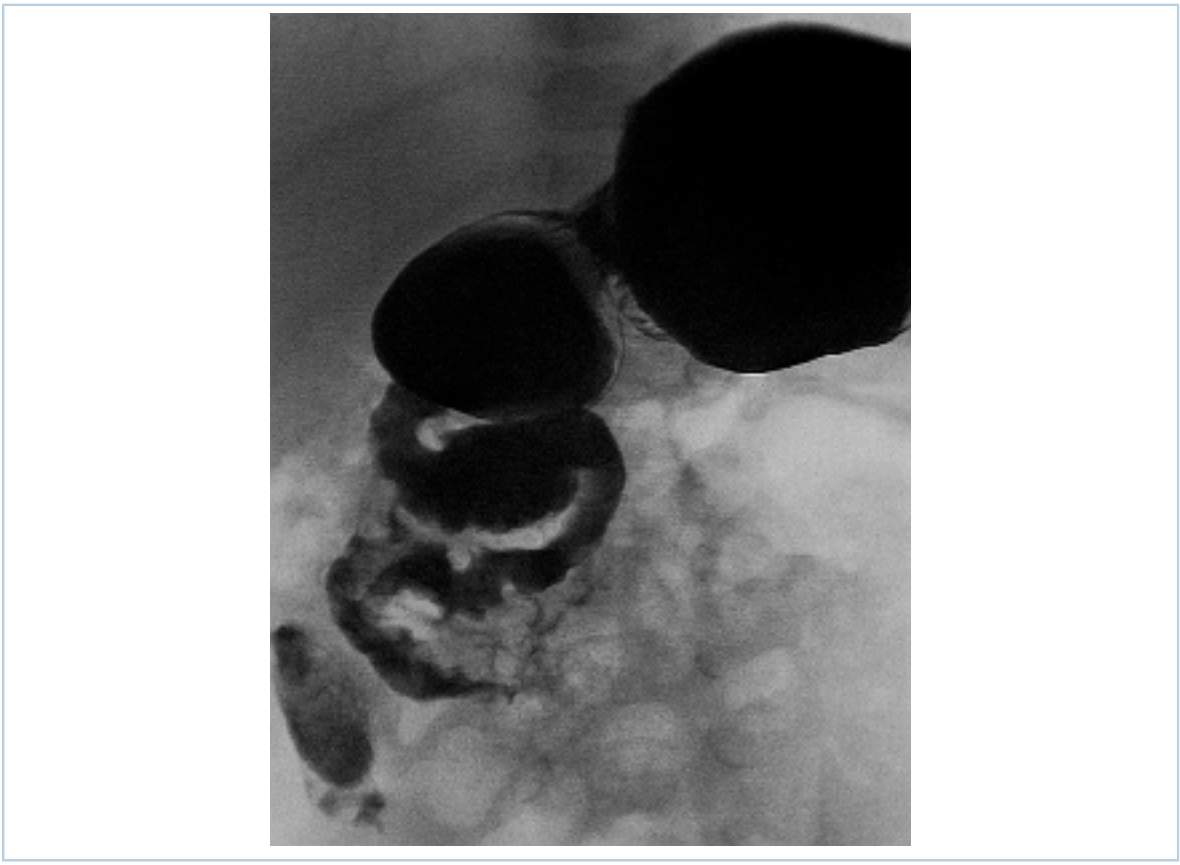
Features of questionable malrotation or asymptomatic/incidental findings in investigating for reflux (fussiness, arching, apnoeic events, reactive airways, pneumonia) include:

- Intermittent vomiting
- No signs of acute illness.

Radiological evaluation

Upper GI contrast series

- This is the diagnostic standard for malrotation with or without midgut volvulus and must include anteroposterior and lateral projections.
- In normal rotation, the duodeno-jejunal junction at the ligament of Treitz is located to the left of the left vertebral pedicle and is posterior to the approximate height of the pylorus.
- In malrotation, the duodeno-jejunal junction is located to the right of the spine and inferior to the level of the pylorus, with a more anterior projection than normal.
- In midgut volvulus there may be a bird's beak cut-off of contrast in the second portion of the duodenum, or the duodenum may have a corkscrew appearance if contrast can get into the loops of bowel that have twisted.



Malrotation with volvulus without obstruction as seen on upper GI contrast

From the collection of Dr S.D. St Peter

- The typical sensitivity is approximately 95%, but it may have a false-positive rate up to 15% and false-negative rate of up to 6%.[8]

CT (with oral and IV contrast)

- CT scan is not the preferred study to assess for malrotation and/or midgut volvulus; however, when this study is performed to assess abdominal pain without bilious emesis, malrotation and/or midgut volvulus may be seen.
- In the setting of volvulus, CT scan of the abdomen could potentially show oral contrast cutting off at the duodenum, and intravenous contrast may show a twirling of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). The image of the vessels may depict whether there is good arterial flow beyond the point of twisting.
- In the setting of malrotation without volvulus, findings would be similar to those of the upper GI series. Furthermore, an abnormal orientation of the SMA and SMV may be noted, with the SMA located to the right and anterior to the SMV.

Abdominal plain x-rays

- This study is usually ordered as a preliminary investigation for patients presenting with abdominal complaints. It is infrequently helpful in preventing other more definitive tests to rule out malrotation unless other conditions are highly suspected (e.g., necrotising enterocolitis or congenital duodenal obstruction).
- Stomach and proximal duodenum distended with gas and little distal gas is the most consistent sign of volvulus on plain x-rays.[9]

- The build-up of air in the stomach and proximal duodenum during volvulus gives the double-bubble appearance that is pathognomonic for duodenal atresia. However, if any air is present distally, malrotation with volvulus is suggested and an emergency contrast study should be performed.

Lower GI contrast series

- This study may be ordered in the work-up of newborn patients with stooling difficulty or distal intestinal obstruction.
- Malrotation may be recognised by abnormal position of the caecum (present in 85%).[10]
- If there is any concern of GI obstruction, the abnormal caecal position should prompt contrast being placed from above.
- Rarely, lower GI series may augment the investigation for malrotation if the upper GI series is inconclusive.[10]

Ultrasound

- This study can be done quickly without contrast or radiation, but if midgut volvulus is a concern, it may be clinically misguided to order an ultrasound. An ultrasound is more commonly ordered to assess for intussusception if malrotation or midgut volvulus is not the primary consideration. However, when it is ordered to rule out intussusception, or in circumstances where the upper GI series is not conclusive, images of the mesenteric vessels may be useful, demonstrating a reversed SMA-SMV relationship or even a "whirlpool" pattern of volvulus.
- Sensitivity (75%) and specificity (80%) are less than those of upper GI series.[8]

Laboratory evaluation

FBC

- In cases of malrotation without volvulus, WBC count is normal. It is also typically normal for several hours in cases of patients with volvulus.
- If the volvulus exists for a period of time, WBC count may be elevated. If the presentation is delayed, the loss of plasma in the intravascular space leads to haemoconcentration.

Blood gas analysis

- When bowel perfusion is compromised from twisting of the mesenteric vessels, lactic acidosis ensues dropping the serum pH with increased anion gap.
- If the diagnosis of volvulus is confirmed with radiological studies, blood gas analysis is important to determine the physiological status of the patient and to guide resuscitation.

DIAGNOSIS

History and exam

Key diagnostic factors

bilious vomiting (common)

- Green vomit is often of sudden onset and vomiting of some form is present in >90% of cases of midgut volvulus.

abdominal pain (common)

- Pain is usually severe with an onset so sudden the parents may recall the exact time it started. As many of the patients are infants, the pain manifests as a notable transition to an inconsolable state.

Other diagnostic factors

infant age <1 year (common)

- Of patients with malrotation who become symptomatic, 50% to 75% do so in the first month and about 90% within the first year of life.[6]

normal abdominal examination (common)

- A distended abdomen is more commonly a bowel obstruction further downstream. Vomiting and abdominal pain with a flat abdomen should alert physicians to the presence of volvulus.

abdominal distension (common)

- Progressive distension usually follows bowel ischaemia and swelling, which foreshadows a poor outcome.

abdominal tenderness (common)

- May be a presenting feature of obstruction with ischaemia (midgut volvulus with vascular compromise).

tachycardia with hypertension (common)

- Initially the severe abdominal pain causes tachycardia with hypertension. An elevated heart rate from pain and bowel ischaemia may reach >200 bpm in an infant, which should precipitate an early surgical consultation.

tachycardia with hypotension (common)

- If ischaemia exists, the toxic products of tissue infarction create a picture of early systemic inflammatory response syndrome. The resultant loss of endothelial integrity creates loss of intravascular volume and tone, causing severe hypotension and tachycardia.

tachypnoea (uncommon)

- In the physiological response to acidosis, the patient may be breathing quickly and if the abdomen is becoming tender from infarction, the breaths are shallow and short.

weight loss (uncommon)

- May be a feature of intermittent or partial volvulus or obstructing Ladd's bands.

dark blood in nappy (uncommon)

- Carer may notice this.

rebound tenderness and guarding (uncommon)

- Peritoneal signs may occur with midgut volvulus.

Risk factors

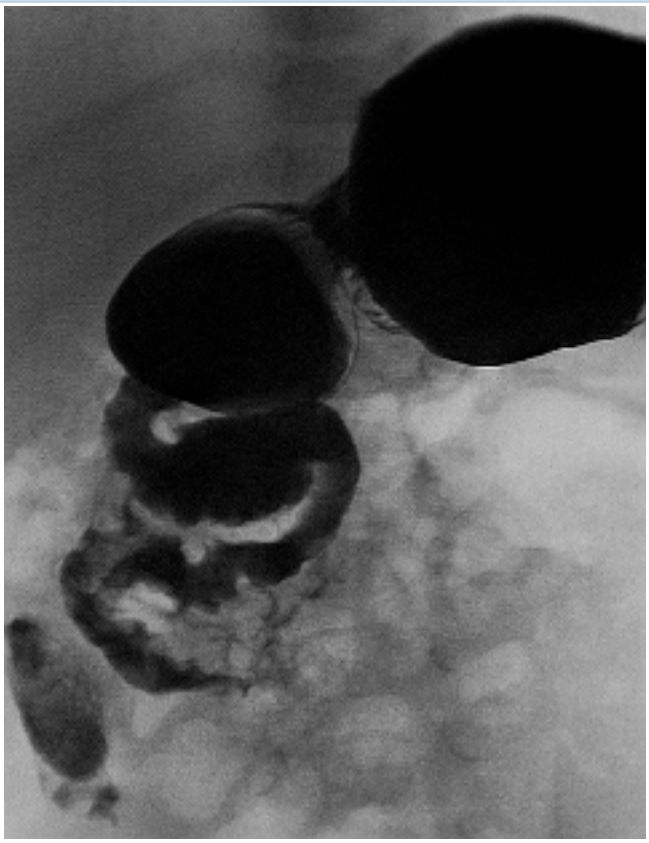
Strong

embryological abnormality

- Rotation may be arrested at any point in midgut development creating the myriad conditions described by the term malrotation.
- Furthermore, other embryological abnormalities that disrupt the intestinal rotation and fixation process may lead to the development of various forms of malrotation.[7] These include abdominal wall defects (i.e., gastroschisis, omphalocele); congenital diaphragmatic hernia; or heterotaxy syndromes.

Investigations

1st test to order

Test	Result
upper gastrointestinal contrast series • Diagnostic standard test for malrotation, and should be performed with AP and lateral projections if there is any history of bilious vomiting without previous abdominal surgery. • Normal pattern of rotation: the transition of duodenum to jejunum (ligament of Treitz) lying left of the left vertebral pedicles in AP view. The position of this transition should also be at the level of the pylorus or superior to this.  *Malrotation with volvulus without obstruction as seen on upper GI contrast* *From the collection of Dr S.D. St Peter* • Normal anatomy on this examination rules out malrotation, midgut volvulus, and obstruction of the upper small intestine. • Delayed images in 1 or 2 hours can depict or rule out obstruction of the distal small bowel.	**right-sided duodenum (malrotation); duodenum courses inferior or medial to normal (malrotation); bird-beak cut-off of duodenum (volvulus); corkscrew of duodenum (volvulus); a web in the duodenum (duodenal atresia)**
CT abdomen (with oral and intravenous [IV] contrast) • Should be ordered when the presentation is abdominal pain without witnessed vomiting, and the concern for malrotation is low. If midgut	**no oral contrast beyond duodenum (volvulus); no contrast in the distal superior mesenteric artery (volvulus with**

Test	Result
volvulus is being considered, then CT scan should not be performed before an upper gastrointestinal (GI) series.[10] • Performed using oral and IV contrast to delineate intestinal pathologies. • Coronal reconstructions are particularly useful in following the course of the duodenum to assure the transition from duodenum to jejunum lies left of the left side of the spine and is appropriately high enough. • Additional information that can be obtained from the scan, which is not offered by the upper GI contrast series, is immediate assessment of caecal position/colonic rotation and 3-dimensional evaluation of the entire extra-intestinal space.	**ischaemia); twirling of the superior mesenteric artery and vein (volvulus); transposition of superior mesenteric artery and vein (malrotation); a transition point in bowel calibre, right-sided duodenum; duodenum courses anterior or to right of superior mesenteric artery**
abdominal plain x-rays • Usually ordered in the emergency department for patients presenting with abdominal complaints, but is rarely helpful in changing management or preventing other tests. • In the newborn nursery, a patient with bilious vomiting should be studied first with emergency plain films, as they provide a better yield for demonstrating other congenital aetiologies for symptoms. • Stomach and proximal duodenum distended with gas and some distal gas is the most consistent but non-specific sign of volvulus on plain films.[9] • The build-up of air in the stomach and proximal duodenum during volvulus may give the double-bubble appearance that is otherwise pathognomonic for duodenal atresia, but if any air is present distally, malrotation with volvulus is suggested and an emergency upper gastrointestinal (GI) series should be performed. • If the amount of distended loops of bowel is large, suggesting more of a distal bowel obstruction, but a midgut volvulus cannot be ruled out, a carefully combined upper and lower GI series may need to be performed in co-ordination with the paediatric radiologist.	**distended stomach and proximal duodenum with paucity of bowel gas seen distally (volvulus or duodenal web); normal x-ray (malrotation without volvulus or volvulus without complete obstruction)**
FBC • In cases of malrotation without volvulus, WBC count is normal. It is also typically normal for several hours in cases of patients with volvulus. • If the volvulus exists for a period of time, WBC count may be elevated. If the presentation is delayed the loss of plasma in the intravascular space leads to haemoconcentration.	**elevated WBC count; polycythaemia**

Other tests to consider

Test	Result
ultrasound • Can be done quickly without contrast or radiation, but if midgut volvulus is a concern, it may be clinically misguided to order an ultrasound. • An ultrasound is more commonly ordered to assess for intussusception if malrotation or midgut volvulus is not the primary consideration. However, when it is ordered to rule out intussusception, or in circumstances where the upper gastrointestinal series is not conclusive, images of the mesenteric vessels may be useful, demonstrating a reversed SMA-SMV relationship or even a "whirlpool" pattern of volvulus.	**no flow beyond the proximal superior mesenteric artery ([SMA] volvulus with ischaemia); twirling of the superior mesenteric artery and vein ([SMV] volvulus)**
lower gastrointestinal (GI) contrast series • May be ordered in the work-up of newborn patients with stooling difficulty or distal intestinal obstruction. • Malrotation may be recognised by abnormal position of the caecum (present in 85%).[10] • If there is any concern of GI obstruction, the abnormal caecal position should prompt contrast being placed from above. • Rarely, lower GI series may augment the investigation for malrotation if the upper GI series is inconclusive.[10]	**abnormal position of the caecum**
ABG • When bowel perfusion is compromised from twisting of the mesenteric vessels, lactic acidosis ensues dropping the serum pH with increased anion gap. • Acidosis is non-specific and non-diagnostic. If the diagnosis of volvulus is confirmed with radiological studies, blood gas analysis is important to know the physiological status of the patient and to guide resuscitation.	**metabolic acidosis with respiratory alkalosis resulting in profound decrease in partial pressure of carbon dioxide**

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Duodenal atresia/web	• Duodenal atresia is usually not associated with abdominal pain, physiological perturbation, or metabolic acidosis. Any of these signs should press urgent action towards diagnosis and treatment of volvulus.	• On plain x-ray in a newborn with bilious vomiting, the presence of bowel gas beyond the duodenum suggests either midgut volvulus or type 1 duodenal atresia; therefore, an upper gastrointestinal series should be done to differentiate between them.
Intussusception	• The incidence of intussusception increases sharply at 5 months when volvulus is beginning to decrease in incidence. • Bilious vomiting is delayed or secondary if present at all in the presentation of intussusception as opposed to volvulus. • Intussusception is typically distinguished by periods of sudden abdominal pain followed by complete cessation. These continue to return as equally sharp in waves, until the intussusception is reduced.	• Ultrasound identifies intussusception in most cases. • Contrast enema (with air or barium) is thereafter used to confirm the diagnosis and reduce the intussusceptum.
Gastro-oesophageal reflux	• Malrotation without volvulus may cause intermittent non-bilious vomiting that appears identical to reflux, and the two cannot be distinguished by history or physical examination.	• Upper gastrointestinal contrast series definitively clarifies malrotation.
Non-specific symptoms	• It is important to consider that malrotation without volvulus is diagnosed for a wide variety of complaints that lead to a contrast study, including diarrhoea, constipation, anorexia, irritability, apnoea, lethargy, failure to thrive, blood in stool, and fever.[11]	• Upper gastrointestinal contrast series definitively clarifies malrotation.

Criteria

Summary of clinical findings in malrotation resulting in midgut volvulus

Features of obstruction with ischaemia (midgut volvulus with vascular compromise) include:

- Acutely ill patient with severe acute abdominal pain
- Bilious vomiting
- Tachycardia
- Tachypnoea
- Abdominal tenderness
- Acidosis
- Signs of peritoneal catastrophe (re-bound and guarding).

Features of obstruction without ischaemia (midgut volvulus without vascular compromise) include:

- Bilious vomiting
- Crampy abdominal pain in waves
- Non-tender abdomen
- Non-distended abdomen
- No severe physiological perturbation.

Features of intermittent or partial volvulus or obstructing Ladd's bands include:

- Intermittent vomiting
- No signs of acute illness
- Intermittent abdominal pain (typically post-prandial)
- Weight loss.

Features of questionable malrotation or asymptomatic/incidental finding in investigating for reflux (fussiness, arching, apnoeic events, reactive airways, pneumonia) include:

- Intermittent vomiting
- No signs of acute illness.

Screening

Asymptomatic diagnosis

While the infant population is not screened for malrotation, which may be present in an asymptomatic manner, it is important to consider that malrotation without volvulus is diagnosed for a wide variety of complaints that lead to a contrast study, including diarrhoea, constipation, anorexia, irritability, apnoea, lethargy, failure to thrive, blood in stool, and fever.[11] Patients with malrotation remain at risk for volvulus and should be identified whenever possible to allow for proper therapy; thus, infants with persistent demonstration of any of the aforementioned symptoms should have a prompt upper gastrointestinal contrast study.

Approach

If malrotation is suspected or the diagnosis is confirmed, the timing of surgery (i.e., elective, urgent, or emergency) is predicated on the level of concern for bowel ischaemia, the type of malrotation, and the underlying condition of the patient. Most patients with intestinal malrotation without volvulus require urgent surgery in the form of the Ladd's procedure. Patients with midgut volvulus require an emergency Ladd's procedure to potentially reverse intestinal ischaemia before necrosis occurs.[6] Less frequently, the Ladd's procedure may be performed electively if there are truly incidental findings without symptoms, or when the risk of surgery and anaesthesia for the patient outweighs the risk of developing midgut volvulus (e.g., patients with congenital heart disease).[12]

Ladd's procedure

Once bowel viability has been assessed at exploration, the Ladd's procedure is completed by detorsion of volvulus when present, lysis of Ladd's bands, and separation of the duodenum and caecum by broadening the mesenteric base. Once the duodenum and caecum are separated, the small bowel is placed in the right peritoneal cavity and the colon on the left.

Controversy exists as to whether it is still beneficial to perform an incidental appendectomy for the concern of future atypical appendicitis from abnormal appendiceal location, and this decision is left to surgeon preference along with parental discussion.

The Ladd's procedure can be challenging to perform for those surgeons inexperienced with the technique, particularly in the infant with volvulus. The base of the volved bowel can be disorienting, and the volvulus may inadvertently be tightened with erratic exploration.

One needs to remember that in almost all patients the twist is in a clockwise direction. Therefore, detorsion consists of "turning back the hands of time" with anti-clockwise rotation 1 to 3 complete turns until the duodenum and caecum are aligned in parallel.[7] Furthermore, meticulous technique in mesenteric separation is necessary to avoid catastrophic iatrogenic injury to the vessels and full thickness opening in the mesentery that may predispose to an internal hernia.

Only tissue that is frankly necrotic or remains black after detorsion should be resected. If the viability of a long segment remains questionable it should not be resected during the initial exploration. It is more prudent to close the abdomen (or leave it open with a temporary dressing) and plan repeat exploration in 24 hours before committing to a resection that may be incompatible with life without successful enteral nutritional support.

Laparoscopic Ladd's procedure

The operation can be conducted with the exact same steps as the open procedure. Laparoscopic instruments small enough for newborn operations are becoming more widely disseminated worldwide.

There have been no trials between open and laparoscopic approaches. One of the main factors thought to contribute to the effectiveness of the Ladd's procedure is the development of intra-abdominal adhesions to help prevent volvulus.[6] The concern of the laparoscopic technique is that fewer adhesions may result that would potentially protect patients from future volvulus. The relative contribution in the prevention of future volvulus between newly formed abdominal adhesions from surgery versus the displacement and repositioning of the bowel by the Ladd's procedure has yet to be deciphered. However,

laparoscopy used in other procedures has resulted in a lowered incidence of postoperative bowel obstruction in several studies.[13]

When the diagnosis of malrotation is in question, any radiographic findings suggesting malrotation as a possibility deserve exploration that can be facilitated by the laparoscopic approach.[14] Three variables are assessed to help delineate the anatomy:

1. Presence/position of the ligament of Treitz
2. Position and attachments of the caecum and colon
3. Width of the base of the mesentery.

If malrotation is present, the Ladd's procedure can be completed laparoscopically or converted to open depending on surgeon preference.[15]

Obstruction with ischaemia (midgut volvulus with vascular compromise)

This acutely ill patient presents with bilious vomiting, severe acute abdominal pain, tachycardia, tachypnoea, abdominal tenderness, acidosis, or signs of peritoneal catastrophe (guarding and re-bound) and requires emergency abdominal exploration without a radiographically confirmed diagnosis.

Treatment is emergency surgery with open laparotomy and Ladd's procedure. Abdominal exploration is warranted without a radiographically confirmed diagnosis.[7]

Supportive care includes nasogastric (NG) tube, broad-spectrum antibiotics, and aggressive intravenous (IV) fluid resuscitation, which should be performed en route to the theatre.

Obstruction without ischaemia (midgut volvulus without vascular compromise)

The patient usually presents with bilious vomiting, crampy abdominal pain in waves, non-tender abdomen, and no severe physiological perturbation.

Treatment is urgent surgery with open laparotomy and Ladd's procedure. This group of patients allows time for a radiographically confirmed diagnosis, but little time should be wasted obtaining a contrast study, which will show the volvulus causing obstruction of the duodenum.

Immediately on radiographic confirmation, the surgeon should be called if not already involved.

Supportive care includes NG tube and aggressive IV fluid resuscitation, which should be performed en route to the theatre. Antibiotics are only prophylactic and can be used as for any bowel surgery.

Intermittent or partial volvulus or obstructing Ladd's bands

These patients may present with intermittent vomiting, but no signs of acute illness.

Timely surgery with open or laparoscopic Ladd's procedure is required. As these patients are stable, time is available for appropriate diagnostic imaging studies.

Malrotation diagnosed with this history should be operated on at the next feasible opportunity (e.g., next operating day), as this is not an emergency. Laparoscopy may have a role in treating these patients.[15] [16] [17] [18]

Regarding supportive care, no nasogastric tube is necessary before or after the operation if obstruction is not present. IV fluids before the operation are needed for maintenance only during the pre-operative period of nothing by mouth. Antibiotics are prophylactic only with the aim towards gram-positive skin flora coverage.

Questionable malrotation or asymptomatic finding

These patients may be asymptomatic or have intestinal malrotation discovered as an incidental finding in investigating for reflux (fussiness, arching, apnoeic events, reactive airways, pneumonia).

Elective surgery with open or laparoscopic Ladd's procedure is performed in these patients. This group allows time for contrast studies.

No nasogastric tube is necessary before or after the operation. IV fluids before the operation are needed for maintenance only during the pre-operative period of nothing by mouth. Antibiotics are prophylactic only and gram-positive coverage is all that is required.

Patients with heterotaxy or serious comorbidities

There are rare circumstances where malrotation is diagnosed but surgery is not recommended due to the patient's underlying medical illnesses imparting a higher risk of morbidity and mortality from surgery than the risk of malrotation itself. However, one study indicates that malrotation patients with significant comorbidities of heterotaxy or congenital heart disease benefit from a Ladd's procedure.[12]

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Initial		(summary)
obstruction with ischaemia		
	1st	**emergency surgery: open laparotomy and Ladd's procedure**
	plus	**supportive care**

MANAGEMENT

Acute (summary)		
obstruction without ischaemia		
	1st	**urgent surgery: open laparotomy and Ladd's procedure**
	plus	**supportive care**
intermittent or partial volvulus or obstructing Ladd's bands		
	1st	**timely surgery: Ladd's procedure (open or laparoscopic)**
	plus	**supportive care**
questionable malrotation or asymptomatic finding		
	1st	**elective surgery: Ladd's procedure (open or laparoscopic)**
	plus	**supportive care**

Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Initial

obstruction with ischaemia

1st **emergency surgery: open laparotomy and Ladd's procedure**

» The acutely ill patient with midgut volvulus with vascular compromise presents with bilious vomiting, acute abdominal pain, tachycardia, tachypnoea, abdominal tenderness, acidosis, and potentially with signs of peritoneal catastrophe (guarding and rebound).

» Emergency abdominal exploration, even without a radiographically confirmed diagnosis, is necessary. Therefore, the surgeon should be contacted early in the course of care when volvulus is clinically suspected in order to allow the surgeon opportunity to forgo further studies.

» If the volvulus is associated with compromised flow of superior mesenteric artery, time is of the essence to salvage the involved intestine. Immediately on opening of the abdomen, the bowel is detorsed by turning it in an anti-clockwise direction to "turn back the hands of time".[7] Warm packs are applied to allow a period of observation to see whether the bowel will be viable. The Ladd's procedure is then completed.

plus **supportive care**

Treatment recommended for ALL patients in selected patient group

Primary options

» cefoxitin: infants ≤3 months of age: consult specialist for guidance on dose; infants >3 months of age: 30-40 mg/kg intravenously as a single dose 30-60 minutes before surgery, then every 6 hours after surgery for no more than 24 hours

» Nasogastric tube, antibiotic coverage for gram-negative organisms (e.g., cefoxitin), and aggressive intravenous fluid resuscitation should be performed en route to the theatre but do not preclude an operation, as manual detorsion of the bowel is the only way to reverse an otherwise fatal course.

Initial

» Post-operative care is simply supportive. The post-operative course is dictated by bowel rest until bowel activity resumes.

» In the case of temporary closure or abdominal coverage with planned re-exploration, the patient should be sent to the intensive care unit, as there may be profound fluid shifts and haemodynamic perturbations that can progress rapidly. The course may mandate earlier re-operation sooner if the patient is unstable.

Acute

obstruction without ischaemia

1st **urgent surgery: open laparotomy and Ladd's procedure**

» The patient with midgut volvulus without vascular compromise presents with bilious vomiting, crampy abdominal pain in waves, non-tender abdomen, and no severe physiological perturbation. Therefore, a radiographically confirmed diagnosis may be sought, but little time should be wasted obtaining an upper gastrointestinal tract contrast study, which will show the volvulus obstructing the duodenum.

» Contrast may get through the torsion to give the corkscrew appearance, but this finding does not change the state of surgical urgency. Immediately on radiographic confirmation the surgeon should be called if not already involved in the care.

» Immediately on opening of the abdomen, the bowel is detorsed by turning it in an anti-clockwise direction. If there is any concern about bowel viability, warm packs are applied to allow a period of observation to see whether the bowel will be viable. The Ladd's procedure is then completed.

plus **supportive care**

Treatment recommended for ALL patients in selected patient group

Primary options

» cefazolin: infants ≤1 months of age: consult specialist for guidance on dose; infants >1 months of age: 25 mg/kg intravenously as a single dose 30-60 minutes before surgery, then every 8 hours after surgery for no more than 24 hours; additional doses may be given during lengthy procedures

» Nasogastric tube and aggressive intravenous fluid resuscitation should be performed en route to the theatre but do not preclude an operation, as manual detorsion of the bowel is the only reversal of obstruction.

» Antibiotics may be used prophylactically in patients with selected risk factors for surgical-site wound infection; gram-positive coverage (e.g., cefazolin) should be used.

» Post-operative care is simply supportive and the post-operative course is dictated by bowel rest until bowel activity resumes.

Acute

intermittent or partial volvulus or obstructing Ladd's bands

1st **timely surgery: Ladd's procedure (open or laparoscopic)**

» The work-up of these patients, who present with intermittent vomiting but no signs of acute illness, consists solely of an upper gastrointestinal contrast series. If there is no malrotation, intermittent volvulus or Ladd's bands are ruled out but the contrast should be followed downstream with delayed images to rule out distal obstructing pathology. Malrotation diagnosed with this history should be operated on at the next feasible opportunity (e.g., next operating day), as this is not an emergency.

» Laparoscopy may have a role in approaching these patients.[15] [16] [17] [18] The operation remains the same with assuring no torsion around the base of the mesentery, complete duodenal mobilisation, appendectomy, possible widening of the mesentery, and placement of the small intestine on the right side of the abdomen with the colon on the left.

» Feeding resumes with resumption of bowel activity (which is immediate after the laparoscopic approach).

plus **supportive care**

Treatment recommended for ALL patients in selected patient group

Primary options

» cefazolin: infants ≤1 months of age: consult specialist for guidance on dose; infants >1 months of age: 25 mg/kg intravenously as a single dose 30-60 minutes before surgery, then every 8 hours after surgery for no more than 24 hours; additional doses may be given during lengthy procedures

» No nasogastric tube is necessary before or after the operation if obstruction is not present.

» Intravenous fluids before the operation are needed for maintenance only during the pre-operative period of nothing by mouth.

» Antibiotics may be used prophylactically in patients with selected risk factors for surgical-site wound infection; gram-positive coverage (e.g., cefazolin) should be used.

Acute

questionable malrotation or asymptomatic finding

1st **elective surgery: Ladd's procedure (open or laparoscopic)**

» These patients may be asymptomatic or have intestinal malrotation discovered as an incidental finding in investigating for reflux (fussiness, arching, apnoeic events, reactive airways, pneumonia).

» If the contrast study confirms malrotation a Ladd's procedure can be scheduled electively, either open or laparoscopic. However, the diagnosis may not be clear in this population. The duodenum may sweep normally, not be far enough to the left side, or lie low on the contrast study. The retro-peritoneal attachments may be normal (eliminating the risk of volvulus), but malrotation/non-rotation may be severe where the proximal bowel is lying to the left by circumstance.

» Therefore, any radiographic findings suggesting malrotation as a possibility deserve exploration, which is preferably done laparoscopically in this scenario.[14] If malrotation is clearly present, the Ladd's procedure can be completed laparoscopically or converted to open depending on surgeon preference.

plus **supportive care**

Treatment recommended for ALL patients in selected patient group

Primary options

» cefazolin: infants ≤1 months of age: consult specialist for guidance on dose; infants >1 months of age: 25 mg/kg intravenously as a single dose 30-60 minutes before surgery, then every 8 hours after surgery for no more than 24 hours; additional doses may be given during lengthy procedures

» No nasogastric tube is necessary before or after the operation. Intravenous fluids before the operation are needed for maintenance only during the pre-operative period of nothing by mouth.

» Antibiotics may be used prophylactically in patients with selected risk factors for surgical-site wound infection; gram-positive coverage (e.g., cefazolin) should be used.

Secondary prevention

No diet or activity has ever been documented to affect the risk of volvulus or adhesive small bowel obstruction.

Patient discussions

Patients and families need to be made well aware that any sudden onset of vomiting or abdominal pain should be immediately evaluated. The families should understand that on arrival to the emergency department, the healthcare providers must be reminded of the malrotation so they will obtain a contrast study first.

FOLLOW UP

Monitoring

Monitoring

There is no long-term follow-up that is useful after Ladd's procedure. Both adhesive bowel obstruction and volvulus are all-or-none events that will occur suddenly at an unpredictable moment. During the course of care for these patients at the initial diagnosis, providers ought to be thorough in documenting the risk of future volvulus so the medical record clearly directs future providers in the right direction.

Complications

Complications	Timeframe	Likelihood
bowel resection-related short gut syndrome	**long term**	**medium**

For survivors of midgut volvulus with ischaemic necrosis, the risk of short gut syndrome is related to the amount of bowel lost.

When a large amount of small intestine must be resected to allow for survival due to a long segment of necrotic bowel, the survivors are likely to have inadequate bowel length. This will cause inability to sustain life and growth with enteric intake and require long-term parenteral nutrition.

These patients need multi-disciplinary care ideally in an institution with a dedicated team experienced with this population.

Short bowel syndrome patients may require consideration for small bowel transplant on a case-by-case basis. It is fear of this complication that should drive a second-look operation rather than early commitment to long segment resections, unless the bowel is frankly necrotic and must be resected to establish clinical stability.

volvulus following Ladd's procedure	**variable**	**medium**

Any patient with malrotation maintains a risk of volvulus.

Rotational correction cannot be completed surgically. The retro-peritoneal attachments cannot be recreated. A pre-existing narrow base of mesentery cannot be made normal with an operation, but it can be broadened to its fullest extent.

The Ladd's procedure is intended to decrease the risk of future volvulus and decrease the complications of malrotation, such as the atypical presentation of appendicitis. However, the underlying anatomy continues to predispose to future volvulus.

The operation does allow the surgeon to have a better feel for the risk of future volvulus based on the anatomy found during the operation. Regardless of prior surgery, any patient with known malrotation who presents with bilious vomiting must have volvulus immediately ruled out.

Ladd's procedure-related adhesive small bowel obstruction	**variable**	**medium**

Any patient with a history of an abdominal operation maintains a risk of adhesive bowel obstruction.

An abdominal operation may induce adhesions, which can tether the small bowel to the abdominal wall or to other internal structures. These adhesions create spaces for internal hernias and tether points around which the bowel can twist or be pinched in an obstructive way.

A laparoscopic approach may reduce this risk.[13] [23]

FOLLOW UP

Prognosis

Prognosis after volvulus

Mortality with midgut volvulus is approximately 10% and usually depends on degree of intestinal necrosis.[21] Survivors of volvulus are at risk for recurrent volvulus of around 10%.[22] However, these estimates are likely to be low given the lack of lifelong follow-up in a large cohort. Future adhesive bowel obstruction is possible after any abdominal operation.

Prognosis after Ladd's procedure for malrotation

Mortality is entirely determined by bowel necrosis. Patients with malrotation without volvulus still have the possibility of volvulus after the Ladd's procedure, as the procedure does not create complete rotation but rather non-rotation. This risk is impossible to precisely quantify and is certainly inversely related to the width of the base of the mesentery.

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Images

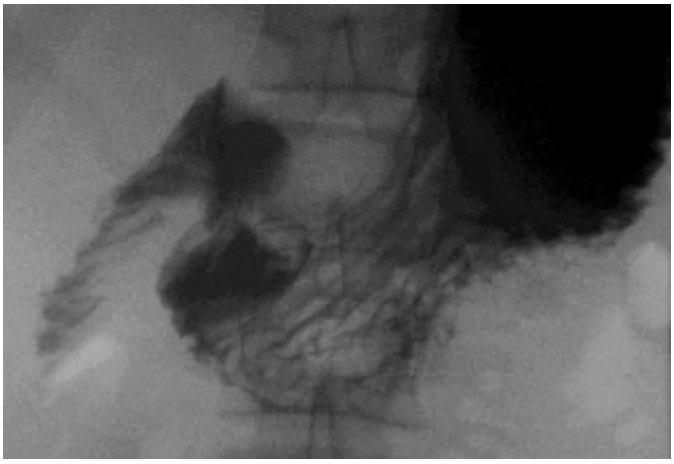

Figure 1: Malrotation with volvulus causing complete obstruction of the duodenum that does not sweep to the left, as seen on upper GI contrast

From the collection of Dr S.D. St Peter

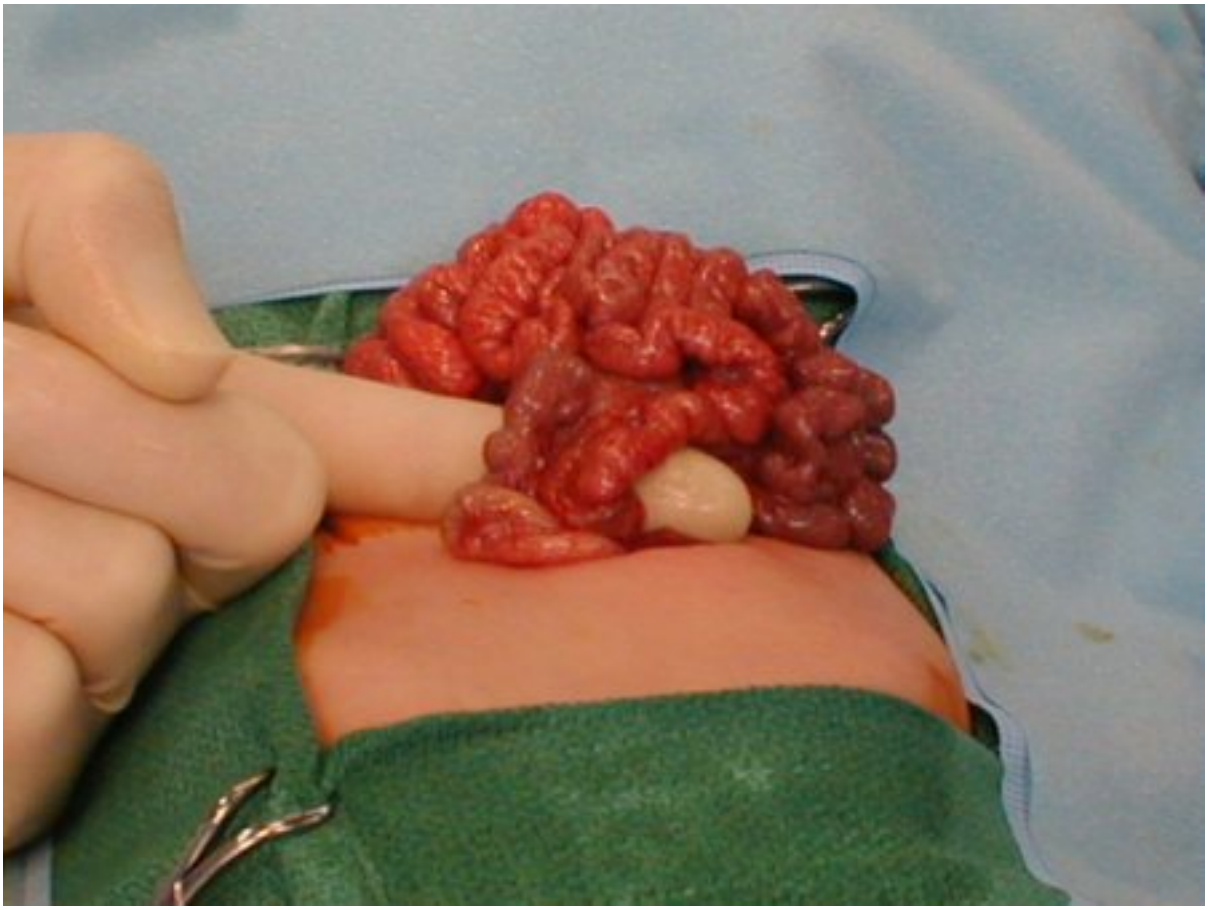

Figure 2: Narrow base of mesentery in malrotation

From the collection of Dr S. Shew

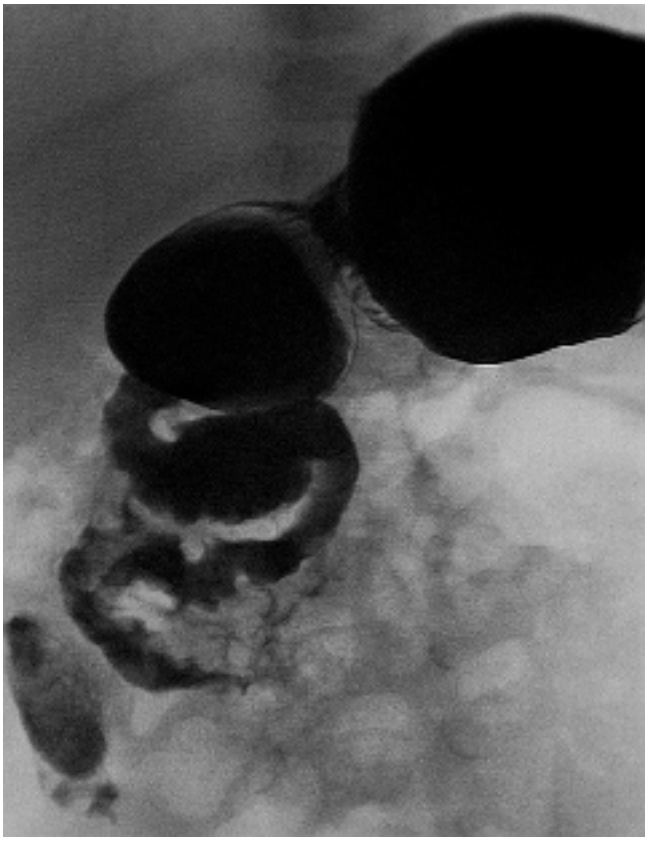

Figure 3: Malrotation with volvulus without obstruction as seen on upper GI contrast

From the collection of Dr S.D. St Peter

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This approach is in line with the guidance of the International Bureau of Weights and Measures Service.

Figure 1 – BMJ Best Practice Numeral Style

5-digit numerals: 10,000

4-digit numerals: 1000

numerals < 1: 0.25

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// Acknowledgements:

Dr Michael Stanton would like to gratefully acknowledge Dr Stephen Shew, Dr S.D. St Peter, Dr Fizan Abdullah, and Dr Omar Karim, previous contributors to this topic.
DISCLOSURES: SS is the author of two references cited in this topic. SDSP is the co-author of one reference cited in this topic. FA and OK declared that they had no competing interests. FA and OK declared that they had no competing interests.

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