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EDUCATION COMMITTEE PUBLICATION: APP Resources

Malrotation/Volvulus

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Malrotation is a term used to represent congenital anomalies that result from an arrest or deviation of normal rotation and fixation of the embryonic gut. This process occurs within the first 3 months of gestation. Volvulus is a complication of malrotation where the intestine rotates around itself, causing compromised blood flow to that segment of intestine which leads to bowel ischemia and necrosis.

Bilious emesis in an infant less than one year old is a surgical emergency until malrotation is ruled out.

Epidemiology:

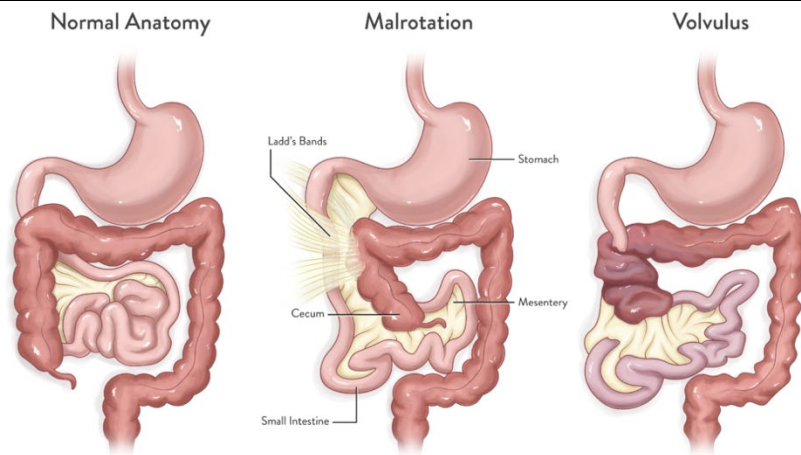
- Symptomatic malrotation occurs in approximately 1 in 6000 live births
- The true incidence of malrotation is unknown as patients may remain asymptomatic throughout their lifetime.

Embryology:

Normal Rotation and Fixation	A wide based mesentery that extends from the Ligament of Treitz in the LUQ to the ileocecal valve in the RLQ
Non-rotation	• Small bowel is located on the right of the abdomen and the colon on the left • Base of mesentery is wider than with malrotation leading to decreased risk of volvulus
Malrotation (Incomplete Rotation)	Cecum is in the mid-upper abdomen and is fixated to the lateral abdominal wall by bands of peritoneum called Ladd bands (can cross the duodenum and cause extrinsic compression and obstruction of the duodenum)

The risk of volvulus occurs because rotation anomalies result in an abnormal narrow mesenteric base.

<https://malrotationresearch.org/what-is-intestinal-malrotation/>



It must be assumed that patients with the following conditions have malrotation by definition:

- Abdominal wall defects (gastroschisis, omphalocele, prune belly)
- Congenital diaphragmatic hernia

Associated Congenital Defects

- Intestinal atresia
- VACTERL anomalies
- Meckel diverticulum
- Abnormalities of the biliary tree
- Major congenital cardiac anomalies (especially heterotaxy)

Clinical Manifestations

Presentation can vary depending on the age of the patient.

In pediatric patients malrotation may be the cause of a critically ill newborn or a newborn/infant with acute intestinal obstruction. The presentations for newborns and infants are described below.

Bilious emesis is the principal feature of neonatal intestinal obstruction and requires urgent evaluation to rule out malrotation with volvulus.

- Abdominal distension
- Abdominal tenderness
- Occult or gross blood in the stool (from mucosal injury and ischemia)
- Lethargy
- Shock

Adults or older children may have chronic or intermittent symptoms (abdominal pain, vomiting, weight loss) or may have incidentally found malrotation on radiographic studies.

Diagnosis

Gold standard imaging study: *upper gastrointestinal study (UGI)*. The most obvious signs of malrotation include (see diagram below):

1. Incomplete duodenal obstruction with dilation of stomach and proximal duodenum
2. Ligament of Treitz on the right side of the abdomen
3. Abnormal position of proximal jejunal loops to the right of midline
4. Deformity of the duodenum with “bird’s beak,” “corkscrew,” or “coiled” configuration

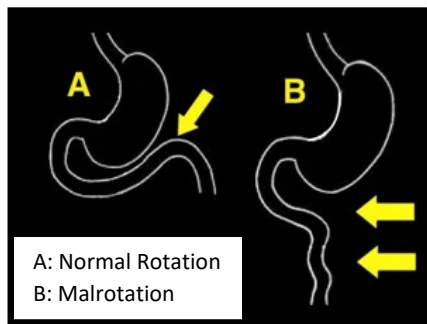
Abdominal ultrasounds are used in some centers with skilled technicians. It is more sensitive and specific when identifying volvulus compared to malrotation. Findings suggestive of malrotation include:

1. Abnormal position of the superior mesenteric vein (SMV) either anterior or to the left of the SMA (SMV is normally located to the right of the SMA)
2. “Whirlpool” sign of volvulus
3. Dilated duodenum
4. Duodenal obstruction with distal air
5. Third part of the duodenum is not in the normal retromesenteric position (normal position is between the mesenteric artery and the aorta in retroperitoneal space)

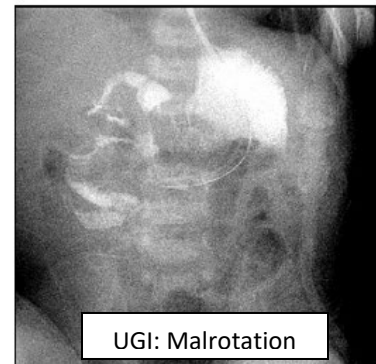
Plain radiographs: Not as **help**ful in identifying malrotation or volvulus. May demonstrate a gasless abdomen, intestinal dilation or may be completely normal.

Surgical intervention should not be delayed in a critically ill baby with an acute surgical abdomen for radiographic studies to be performed. These critical patients may need to go directly to the operating room.

UGI RADIOGRAPHIC FINDINGS: (Need to add notations at bottom of page)



Case courtesy of Dr Bruno Di Muzio, Radiopaedia.org, rID: 12548



Treatment

Malrotation with Volvulus

If there are any clinical signs or concern for volvulus, emergent surgical intervention is indicated.

Preoperative focus should include:

- Emphasis on fluid resuscitation
- Broad spectrum antibiotics
- Naso- or orogastric decompression of the stomach
- Foley catheter placement

Malrotation without Volvulus

If there are no signs of volvulus, an elective Ladd procedure (laparoscopic or open) can be performed, but should still be done in a timely manner due to the risk of volvulus.

Surgical Intervention = Ladd Procedure

- Standard approach to widen the base of the mesentery, place the bowel in a position of nonrotation and create adhesions to hold the bowel to decrease future risk of volvulus.
- Can be done laparoscopic or open
- If volvulus is present, it is untwisted in counterclockwise fashion.
- Bowel resection may or may not be indicated based on perfusion to the site of the volvulus/obstruction.

Complications

Complications arise from delayed identification: intestinal ischemia, infarction and patient death.

- Sepsis, septic shock and multi organ system failure can result from intestinal necrosis
- Short bowel syndrome can result from the extent of resection required
- Recurrent small bowel obstruction from recurrent volvulus (risk of 3.5% following laparoscopic Ladd procedure), adhesive bands or intestinal dysmotility
 - There is a continued risk of volvulus as the underlying embryologic defect is not corrected during the operation.

Results

If no intestinal necrosis, Ladd procedure has very favorable outcomes. Patients > 1 yo more likely to have persistent or intermittent symptoms, which could be related to intestinal dysmotility. The overall mortality after surgery is 3-10%. Mortality increases with the presence of midgut volvulus, intestinal necrosis, less than one month of age and associated anomalies.

References:

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