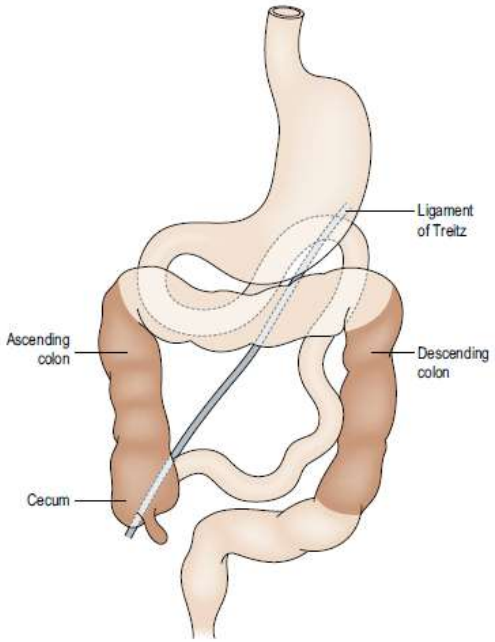
Malrotation

Samuel Negash MD
Pediatric Surgery Resident
Addis Ababa University

Outline

1. Epidemiology
2. Embryology
3. Presentation
4. Imaging
5. Treatment



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1. Epidemiology

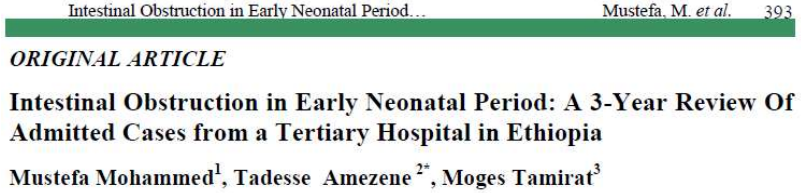
- 1: 6000 LB (*1:500 on imaging, 1:100 in autopsies)
- 2:1 male predominance in neonates but equal in older children

ORIGINAL ARTICLE

PATTERN AND OUTCOME OF NEONATAL SURGICAL CASES AT TIKUR ANBESSA UNIVERSITY TEACHING HOSPITAL, ADDIS ABABA, ETHIOPIA

Nebiyu Shitaye, MD[1]*, Belachew Dejene, MD[1]

- **13 cases** (2% of neonatal surgery)
  - **46% death**



ORIGINAL ARTICLE

Intestinal Obstruction in Early Neonatal Period: A 3-Year Review Of Admitted Cases from a Tertiary Hospital in Ethiopia

Mustefa Mohammed[1], Tadesse Amezene[2*], Moges Tamirat[3]

- **3 cases** (6%of NBO)
  - **1 death**

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2. Embryology

- **Traditional theory**: Arrest of normal rotation

- **Applied embryology**: there is no rotation around axis of mesentery. All processes are the result of longitudinal lengthening .

- May be a component of gastroschisis, omphalocele, CDH or hetrotaxy syndrome.

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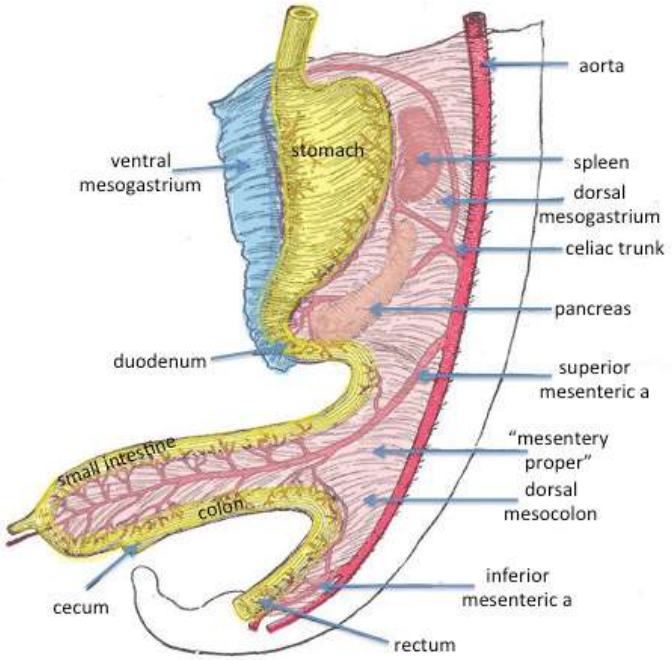
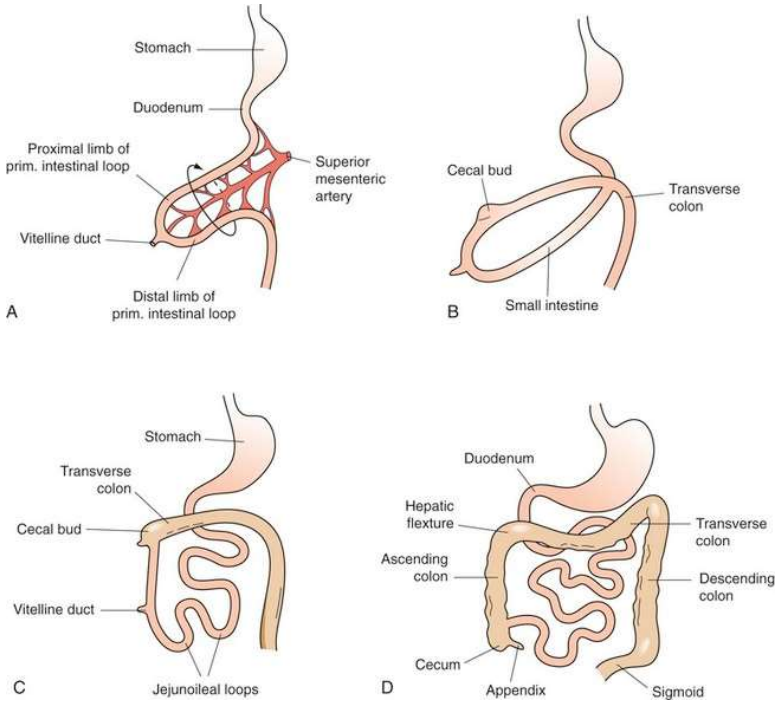
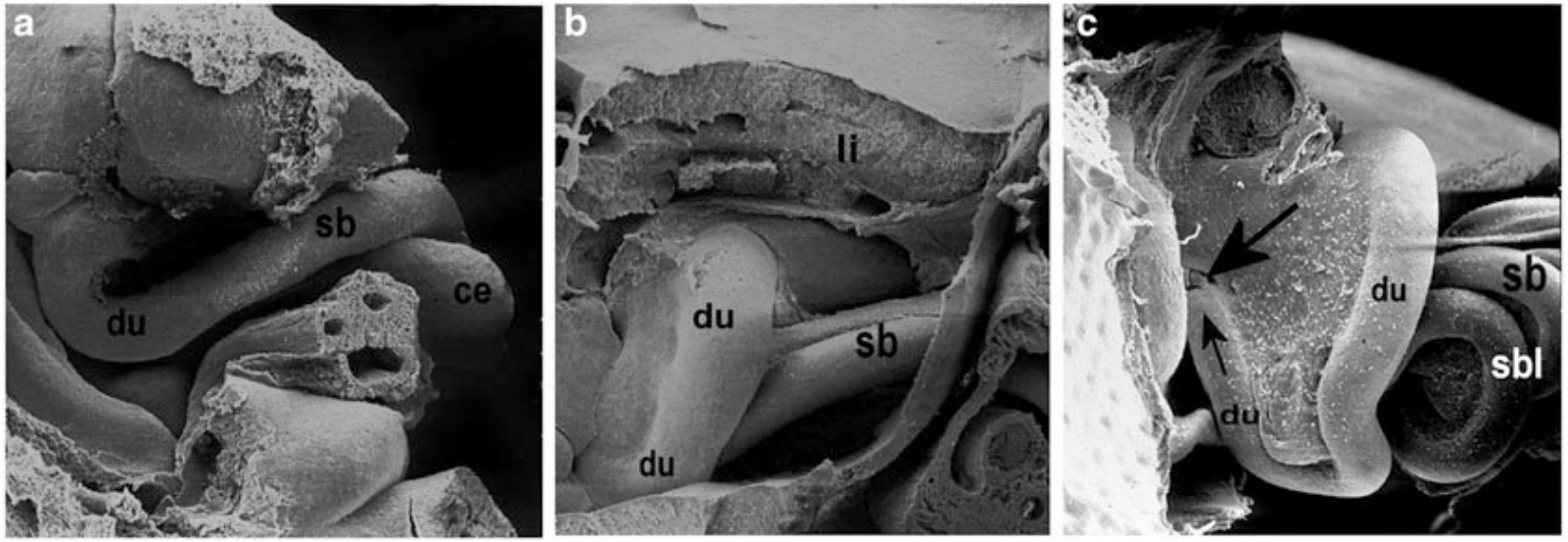




- **Traditional theory** (6-10 wk): Intestinal initially a U shaped loop around SMA with omphalomesentric duct at apex. **Herniate** due to **elongation**. Make **three 90 degree turns** (1 outside, 2 inside abdomen). **Fixation** then occurs



- **Observation in rat embryo**: three parts with a duodenal loop (du), a loop in the extraembryonic sac of the umbilicus (ce), and a straight part in between (sb). With longitudinal growth of duodenal loop, the duodenojejunal junction is pushed beneath the root of the mesentery

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3. Presentation

- **Age**: 75% during the first month of life, another 15% present within the first year
- **Variety of presentations**
  - subtle manner (chronic volvulus) **:** bilious vomiting, distention, Pain, FTT, reflux, constipation
    - *high index of suspicion in a previously healthy baby who presents with bilious emesis
    - *rule out associated atresia if the condition develops immediately after birth.
  - catastrophic (acute midgut volvulus): infarction (bloody stool, erythema and edema of abd wall)
  - Other: ladds band with duodenal obst, reverse rotation with colonic obst, internal hernia, cecal volvulus
- **Associated anomalies**
  - Duodenal atresia (1/3 have malrotation), ARM, Cardiac anomalies, Down's syndrome
  - Intestinal Atresia, Meckel's diverticulum

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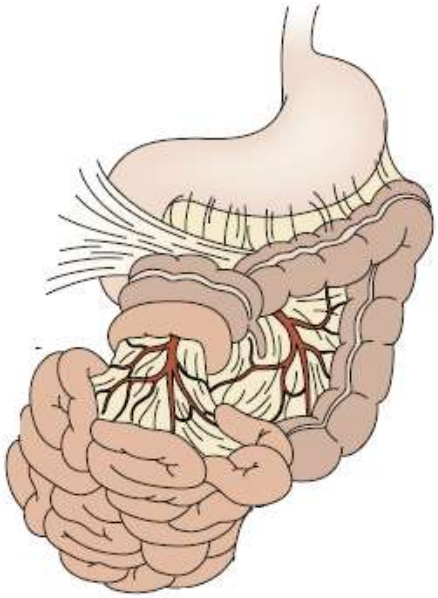
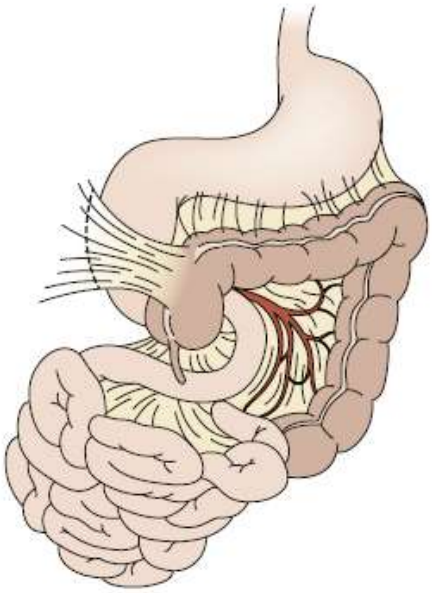
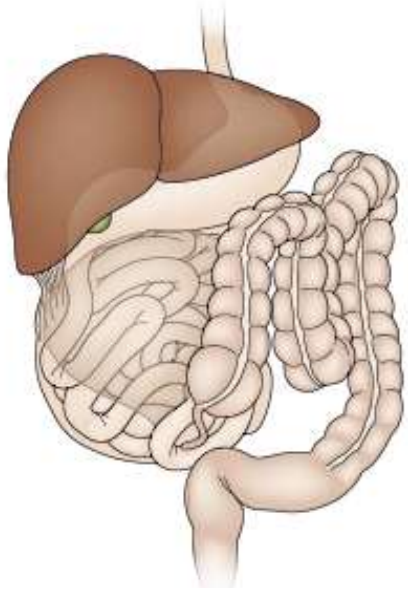
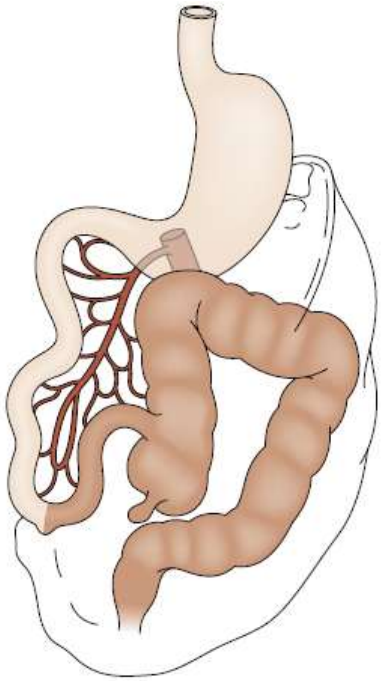
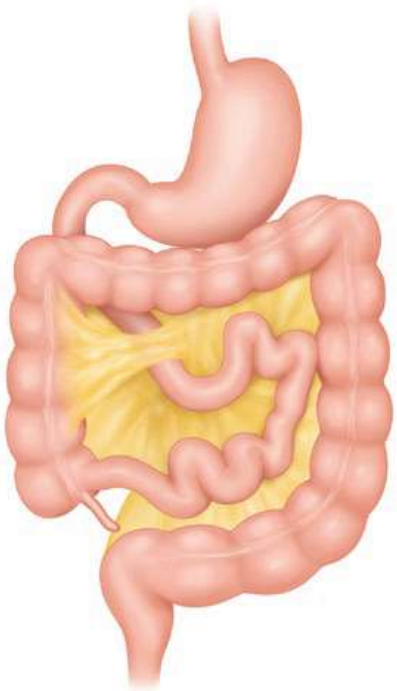




- **Incomplete rotation** *(arrest around 180 degree, DJ & cecum in RUQ).* Is the most common type of malrotation. Highest predisposition to **Midgut Volvulus** (clockwise, around narrow mesentery) because very narrow mesentry. May also present with duodenal obstruction due to **Ladd bands (**from cecum to lat abd wall). **Right mesocolic/paraduodenal hernia** may occur due to trapping of small bowel behind mesocolon.





- **Non rotation** *(arrest at 90 degree, DJ in RUQ & cecum in LLQ),* Less predisposed to volvulus because relatively wider mesenteric base.
- **Variant:** *Isolated non rotation of the duodenojejnal limb.* May present with duodenal obstruction due to **Ladds band** running across duodenum

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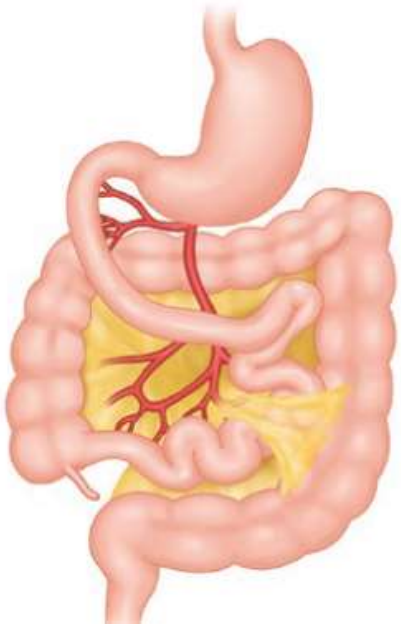
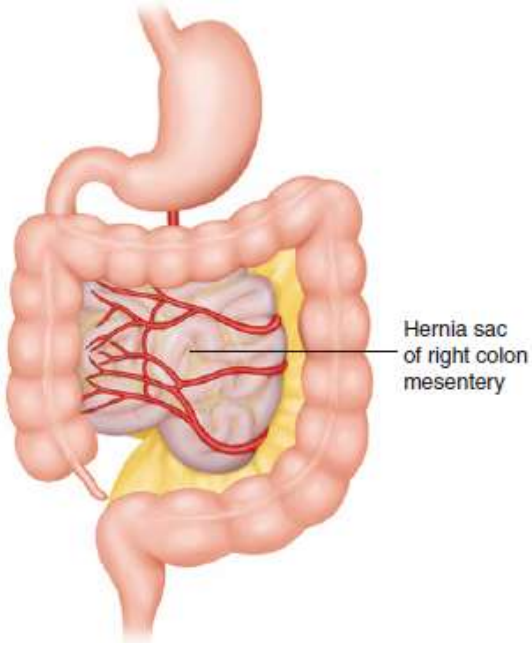
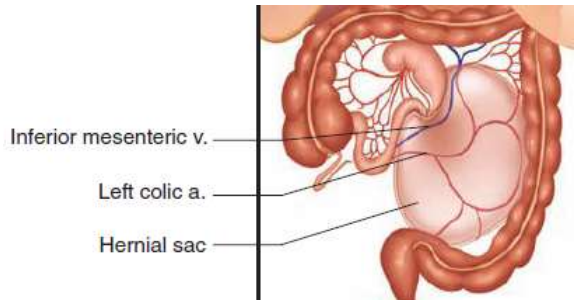
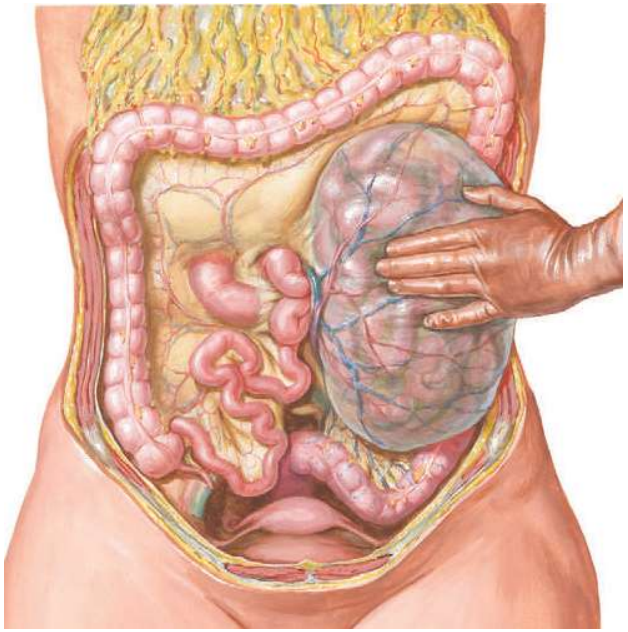


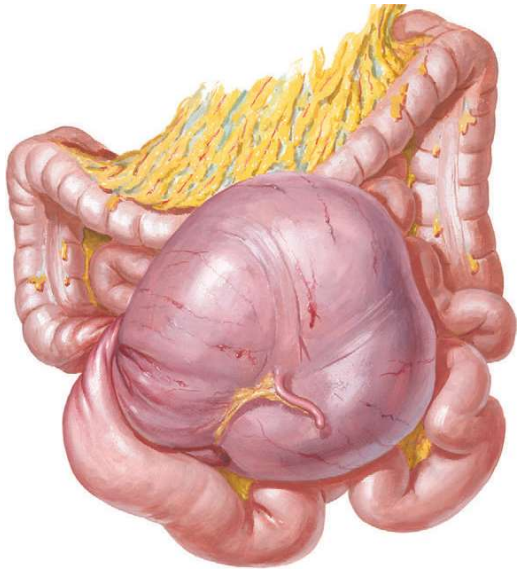


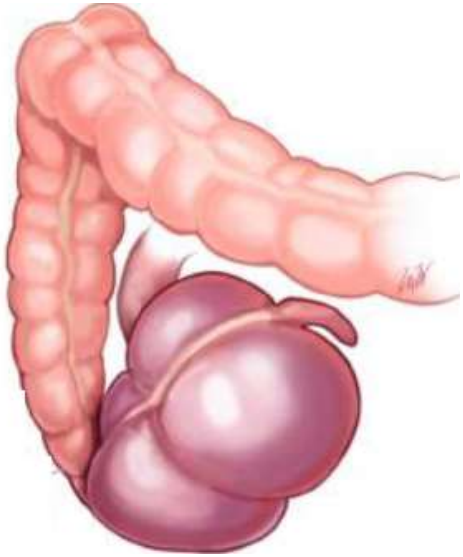
- **Reverse Rotation** *(90 degree clockwise rotation, DJ in RUQ & cecum in LLQ but duodenum is anterior to SMA and Transverse colon):* Transverse colon obstruction may occur due to overlying SMA.
- **Variant:** *Isolated reverse rotation of the duodenojejnal limb* may form **Right mesocolic hernia** due to failure of mesocolon to fuse to the posterior body wall creating a potential space .





- **Abnormal small bowel migration** *:* may cause **left mesocolic hernia** due to trapping of small bowel behind left mesocolon & IMV during its migration to left upper quadrant. IMV delineates the right margin of the sac and is an integral part of the neck of the sac.





- **Inadequate fixation of cecum & ascending colon***:* predisposes to **cecal volvulus.** This is most often seen in adults but relatively younger age than sigmoid volvulus (20-40yr). A variant called **cecal bascule** can occur when the ascending colon fills up without twisting. It can present with similar symptoms.

4. Imaging

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X-ray

Initial test

- **Variety of findings, usually non specific**
  - Normal
  - gasless abdomen
  - Dilatation
  - double-bubble (duodenal obstruction)

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Upper GI Contrast

if patient is clinically stable (no suspicion volvulus)

- **malposition** of duodenum (to right of vertebra)
  - In patients with an enlarged stomach, may be displaced inferiorly, so gastric decompression may be needed prior
- **volvulus** (corckscrew)
- duodenal **obstruction** (beak)

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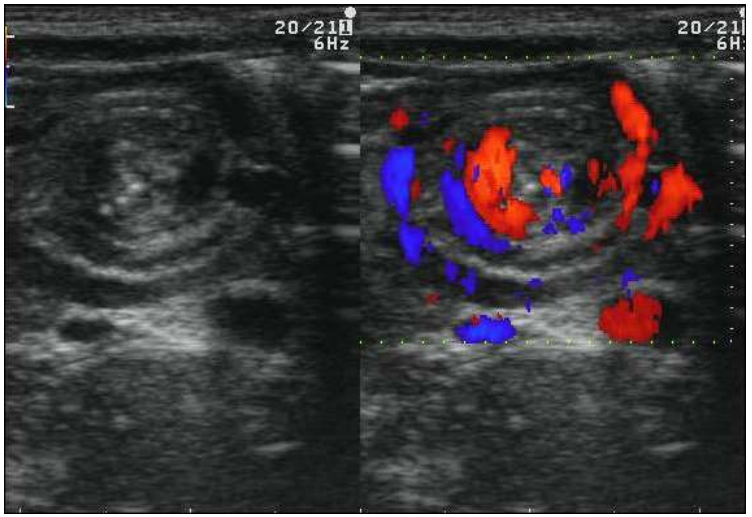
Doppler Ultrasound

Can't rule out malrotation. (upto 30% false negative)

- **reverse position** of SMV (normally SMV to right of SMA)
- **abn position** of 3rd part duodenum (normal b/n aorta & SMA)
- **volvulus** (whirlpool)



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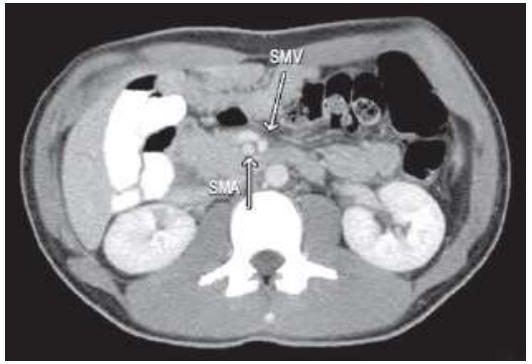
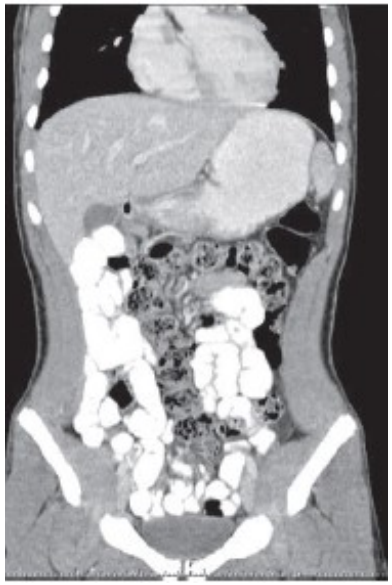
CT scan

In older patients

- dilated duodenum
- malposition of the DJ flexure
- whirlpool sign
- internal hernia,
- reverse position of SMV





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5. Treatment

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Management options

- Clinically Suspect Volvulus (45-65%)
  - **Resuscitate and immediate Surgery** (no confirmatory test or prolonged resuscitation)
- Symptomatic malrotation
  - **Surgery**
- Mild symptoms, Equivocal imaging (DJ junction in equivocal position)
  - **Close observation and repeat contrast study**
  - **Diagnostic laparoscopy**
- Incidentally diagnosed (10%)
  - **Controversial**

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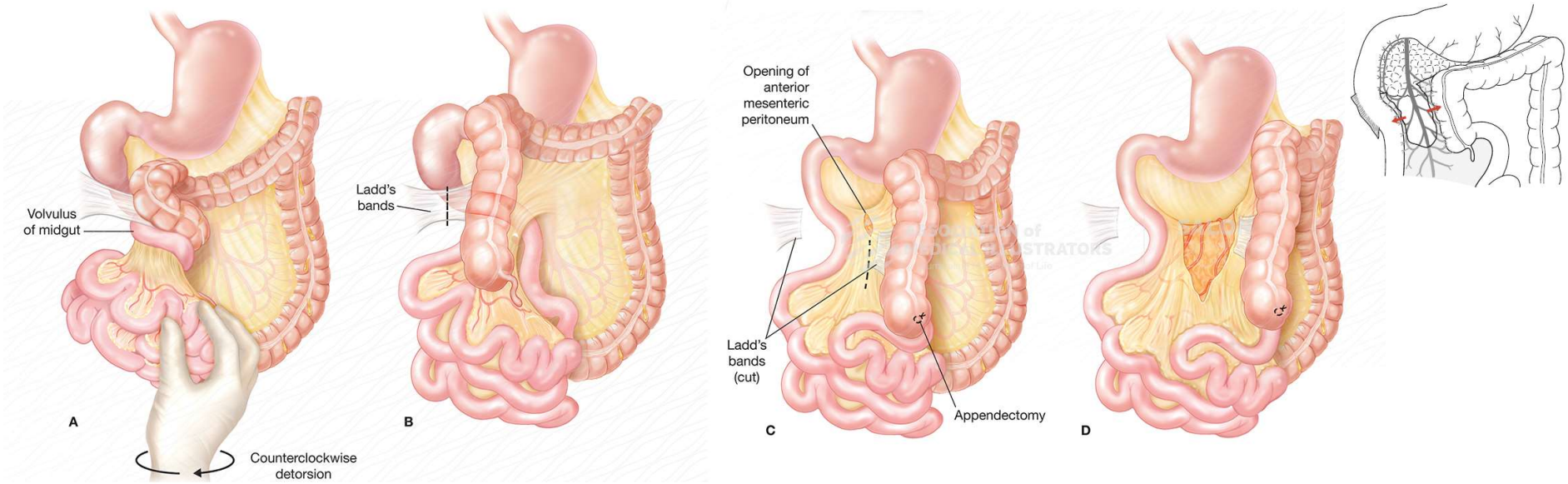
CONGENITAL OBSTRUCTION OF THE DUODENUM IN CHILDREN*

BY WILLIAM E. LADD, M.D.†



- **Ladd's procedure**: first described in 1932, the original technique remains almost unchanged. Eviscerate bowel. Counter-clockwise detorsion. division of ladd bands. broaden mesentery. Appendectomy. place in position of non rotation .

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- **Bowel ischemia**: Once detorsion has been accomplished, warm soaked lap pads are placed on the bowel, and the surgeon should patiently observe for reperfusion. If bowel perfusion is uncertain, a temporary closure/silo should be placed and second look operation done in 24-48 hrs. Unfortunately, on occasion, complete infarction of the midgut may be found. Decision depend on surgeon and parents (need transplant)

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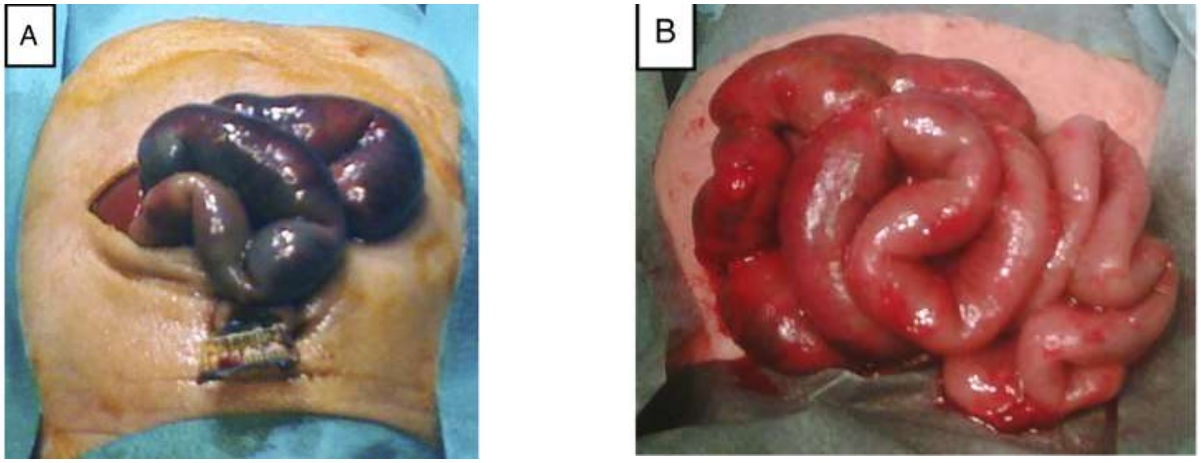
CASE REPORT

Clot Dissolution: A Novel Treatment of Midgut Volvulus



- **Novel technique for ischemia**: it has been proposed that after deterosion, we need to deal with mesenteric thrombosis, which causes continuing ischemia of the intestine. This includes digital massage of the superior mesenteric vessels after derotation to restore intestinal perfusion and postoperative systemic infusion of t-PA (tissue-type plasminogen activator). A second look operation has shown success in the 2 neonates reported.

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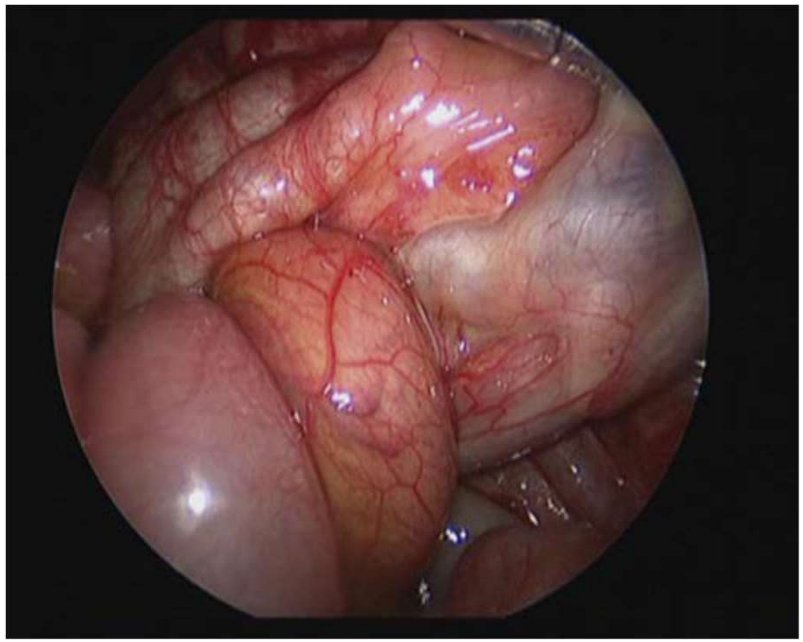

- **Laparoscopy** is useful for the diagnosis in cases of equivocal upper GI studies. It can also be therapeutic for elective cases with radiographically diagnosed malrotaiton. However it is challenging in volvulus as dilated bowel makes visualization difficult and working space isLimited. surgeon should have a low threshold to convert to laparotomy, which can be done by slightly extending the umbilical incision. Another difficult scenario is in older children with cocoon like deformity of small bowel.

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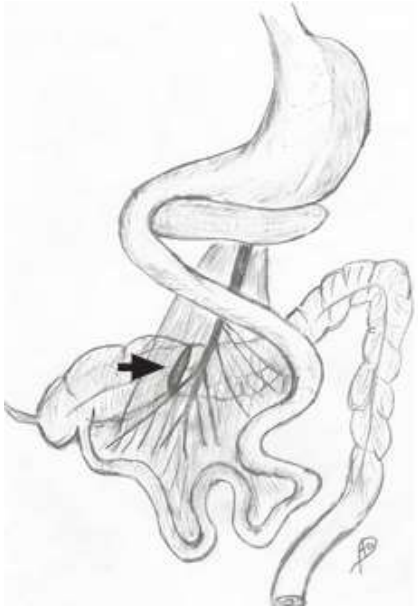

- **Surgery for colonic obstruction secondary to reverse rotation** *In the past, division of the colon with anastomosis anterior to the duodenum was recommended.* It is now recognized that this form of obstruction can be relieved by freeing mesenteric root from retroperitoneal attachment and lysing constricting bands. The liberated colon can be left in place without resection or antemesenteric transposition.

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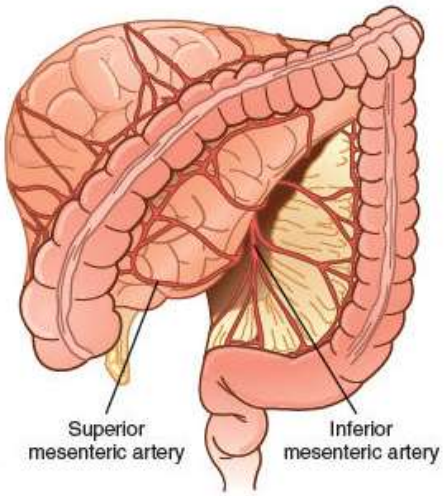

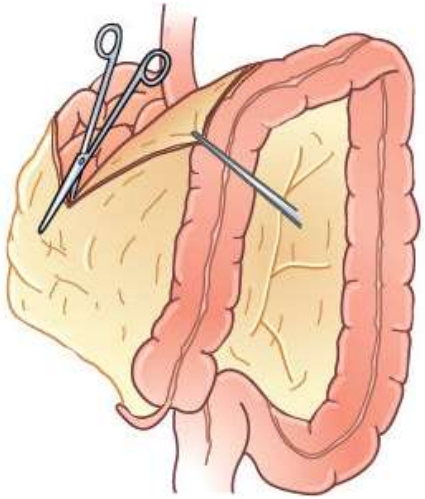

- **Right mesocolic hernia repair** is best done by incising the lateral peritoneal reflection of the right colon and rotating the colon to the left, thereby freeing the small intestine

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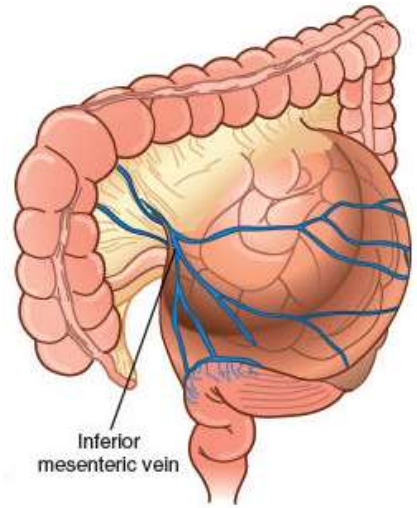
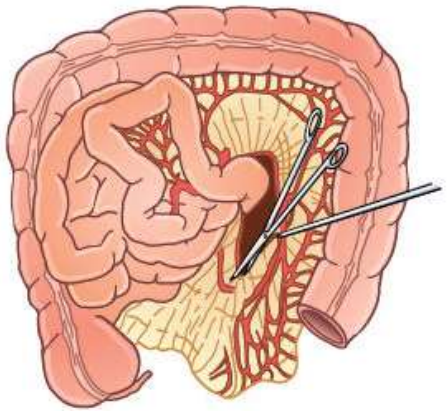
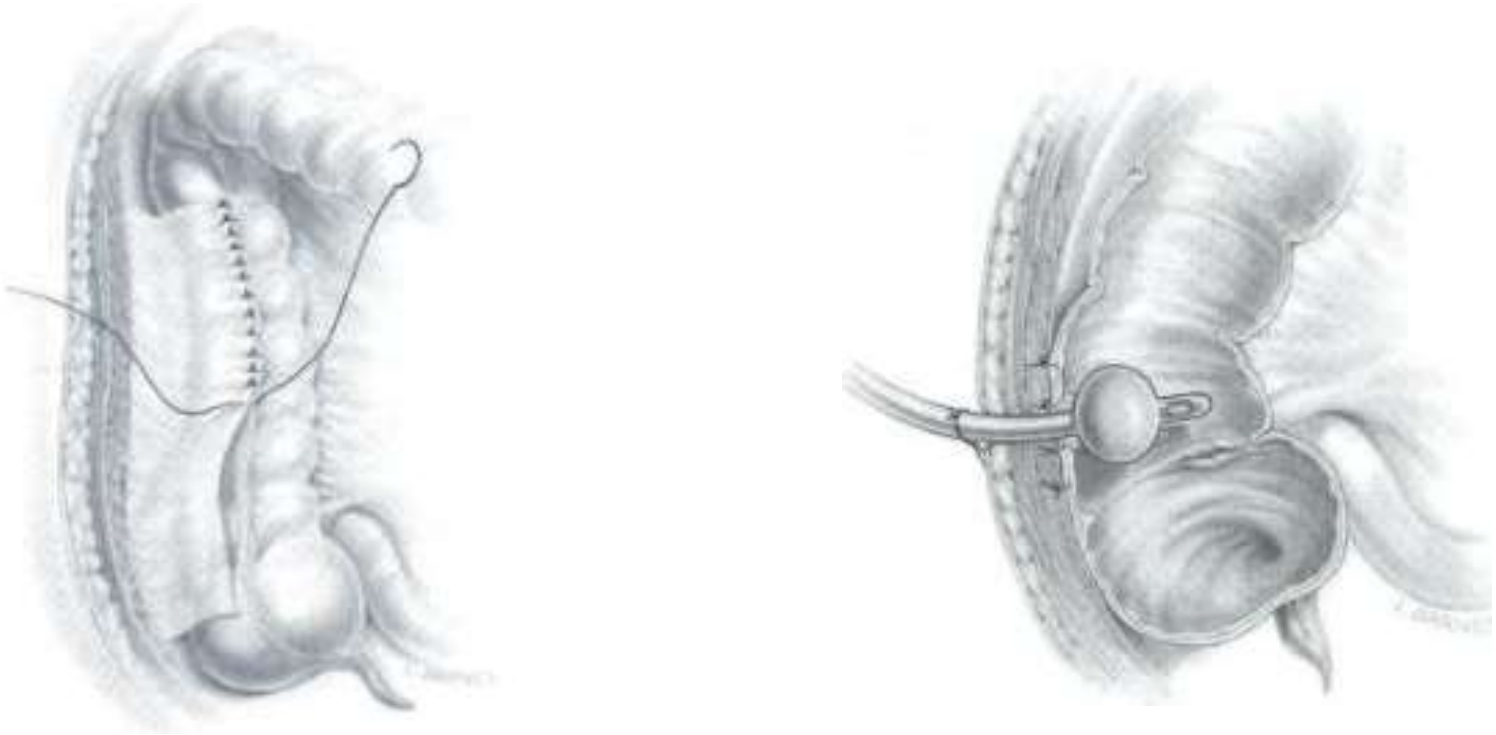




- **left mesocolic hernia repair** is more challenging. The key is mobilization of the IMV which runs along the anterior margin of the neck of the sac. The small intestine can sometimes be reduced through the neck of the sac otherwise an incision is made to the right of the vein (IMV should be spared) to reduce the bowel. The peritoneum adjacent to the vein is sutured to the posterior peritoneum to close the neck of the sac.



- **Surgery for cecal volvulus:** detorsion + cecopexy / cecostomy / resection

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Controversies (Asymptomatic malrotation)

- Incidentally diagnosed (On imaging, prior to fundoplication for gastroesophageal reflux)
  - Watchful waiting has been suggested but upto 20% adults may develop volvulus
  - *Consider age and status of cardiac disease

- Screening of asymptomatic children (for prophylactic treatment)
  - Patients with heterotaxy for intestinal foregut and rotational anomalies
  - Monozygotic twins with proven malrotation in the sibling.

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Complications

- Short bowel syndrome
  - **Extensive resection** (more than half bowel without ileocecal valve)
- volvulus (2-8%)
  - No specific guideline to predict volvulus
  - **Inadequate splaying of mesentry** (mesenteric base more than half the diameter of the abdomen is sufficient)
  - **Inadequate intra-abdominal adhesions (**higher rate of volvulus after laparoscopic approach)
- Adhesion (5%), intussusception (3%)

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