SUNY Downstate Medical Center

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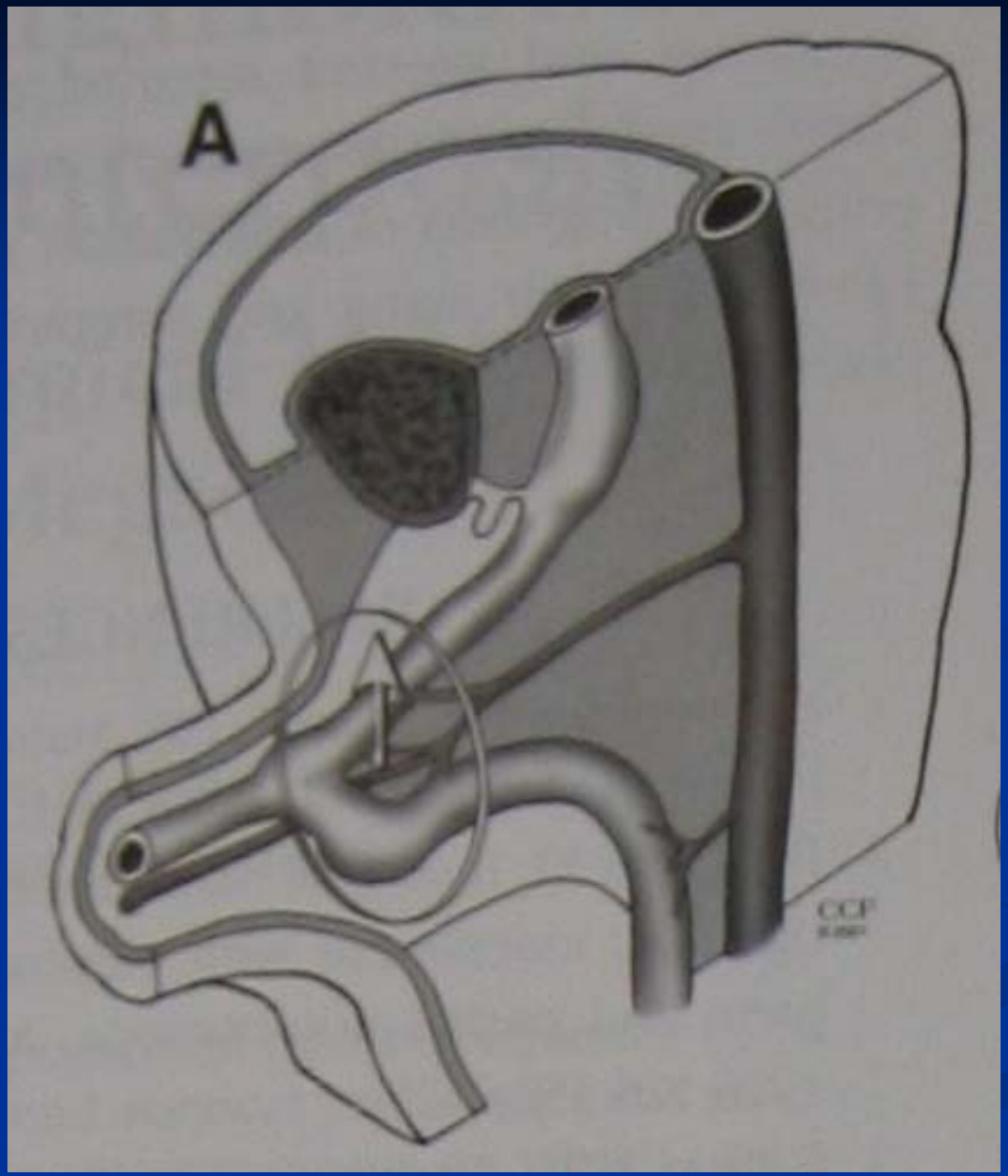
Intestinal Malrotation

Historical Perspective

- 1700's first reported cases in literature
- 1898 Mall - embryology of malrotation described
- 1923 Dott – relationship between anatomy and clinical outcome described
- 1936 William E. Ladd – described the treatment of malrotation

Embryology

- **Stage I** – *Physiological umbilical herniation*
  - 5^{th} – 10^{th} week of Gestation
  - Herniation of midgut loop into base of umbilical cord
  - DJ loop begins superior to SMA & rotates 180 degrees counterclockwise to lie behind the SMA
  - CC loop begins inferior to SMA & rotates 90 degrees counterclockwise to lie to the left of SMA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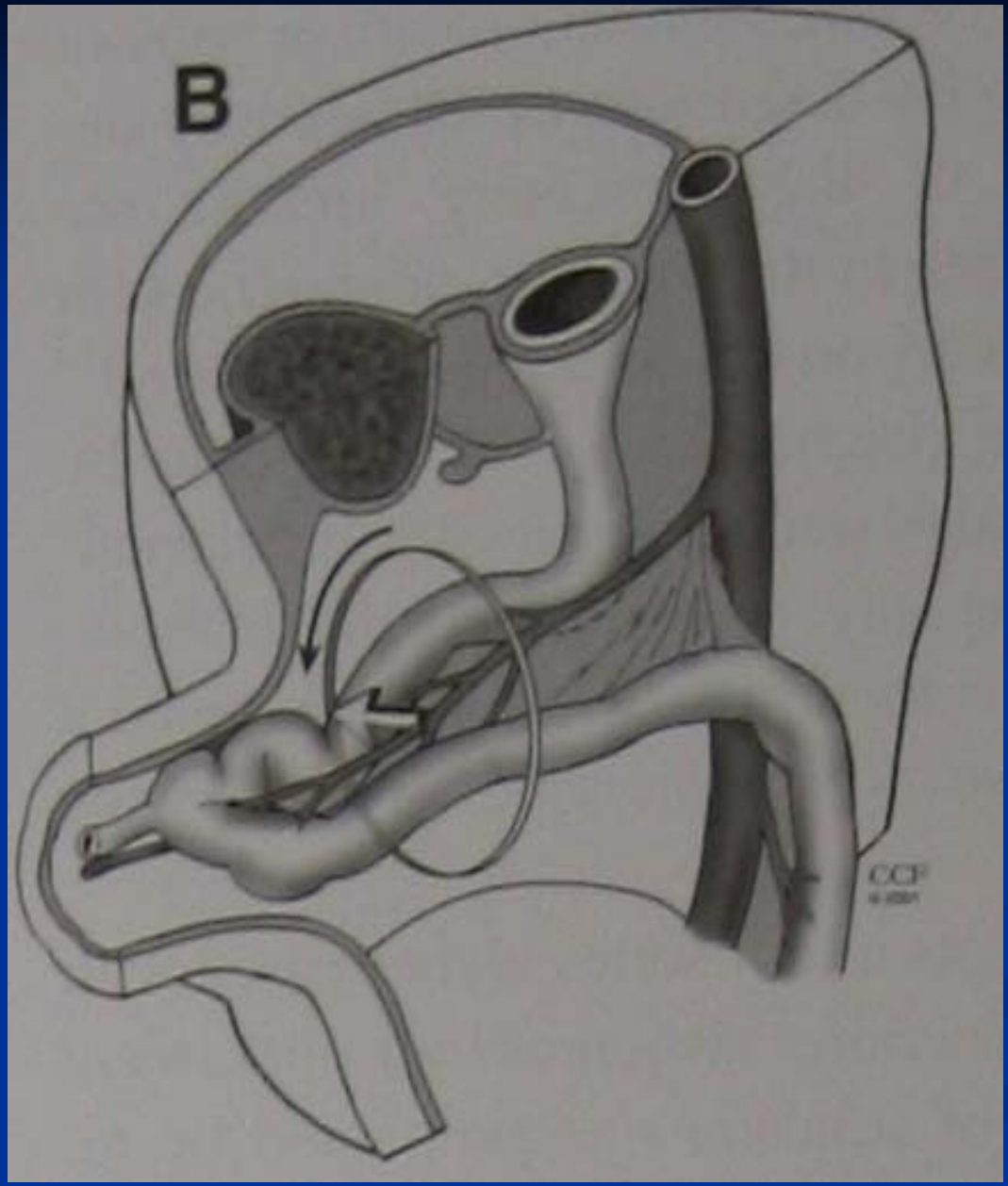
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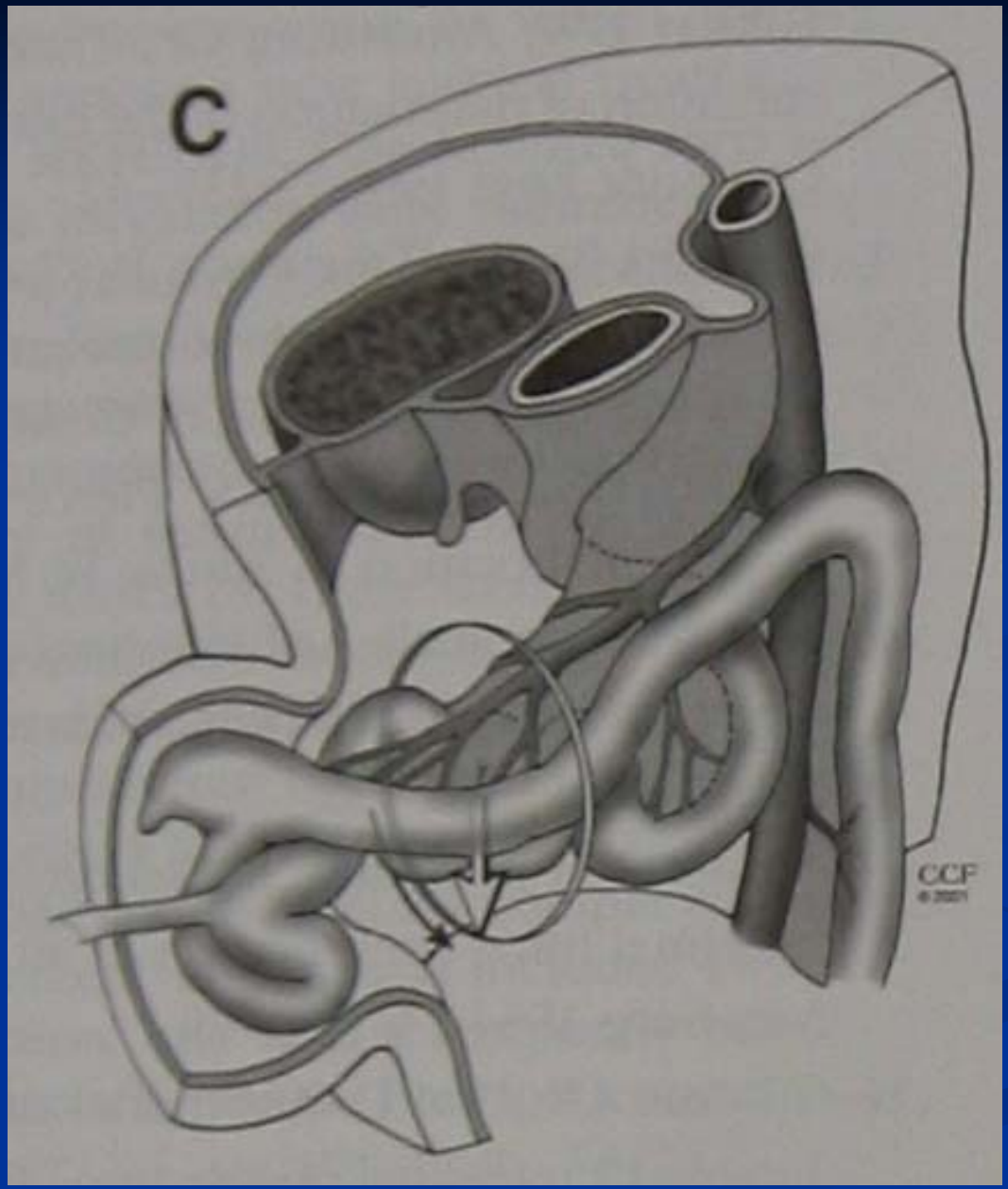
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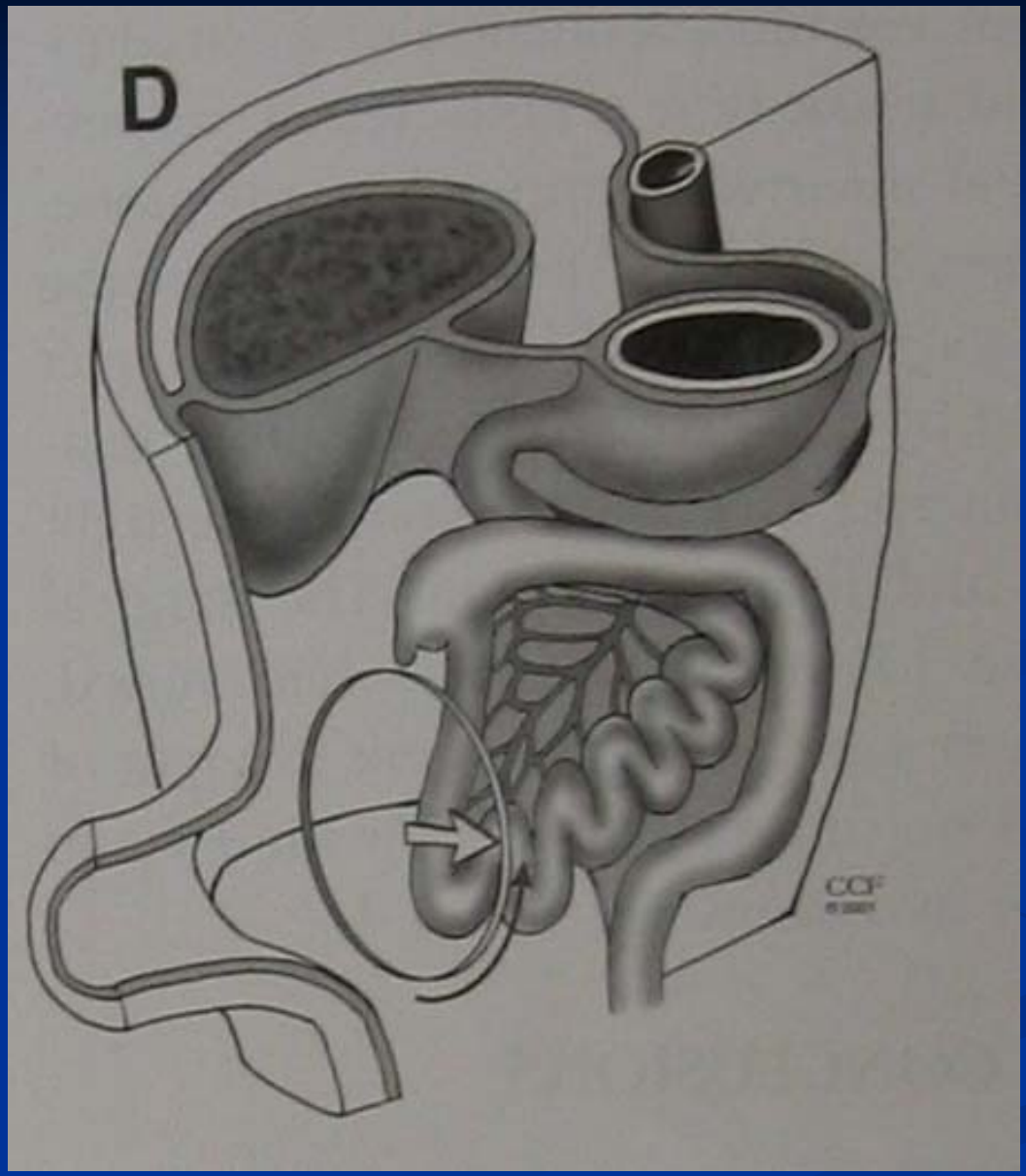
Embryology

- **StageII** – *Reduction of the midgut hernia*
  - 10^{th} – 12^{th} week of gestation
  - DJ loop rotates an additional 90 degrees to end at the anatomic left of the SMA.
  - CC loop rotates 180 degrees to end at the anatomic right of the SMA.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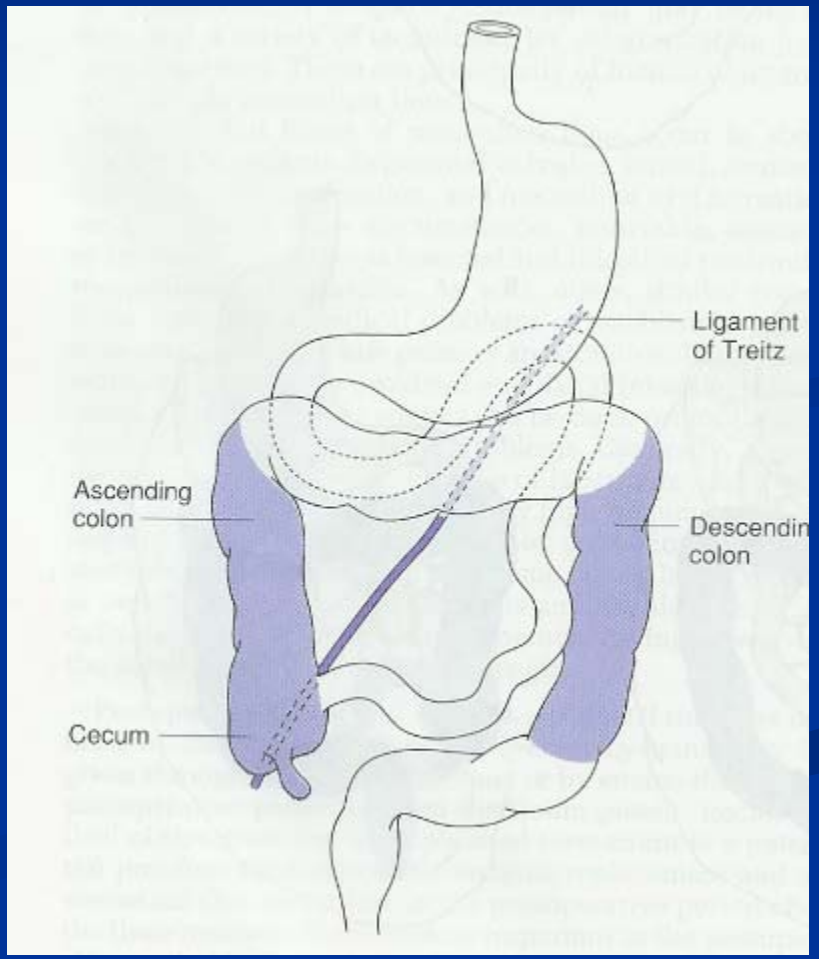
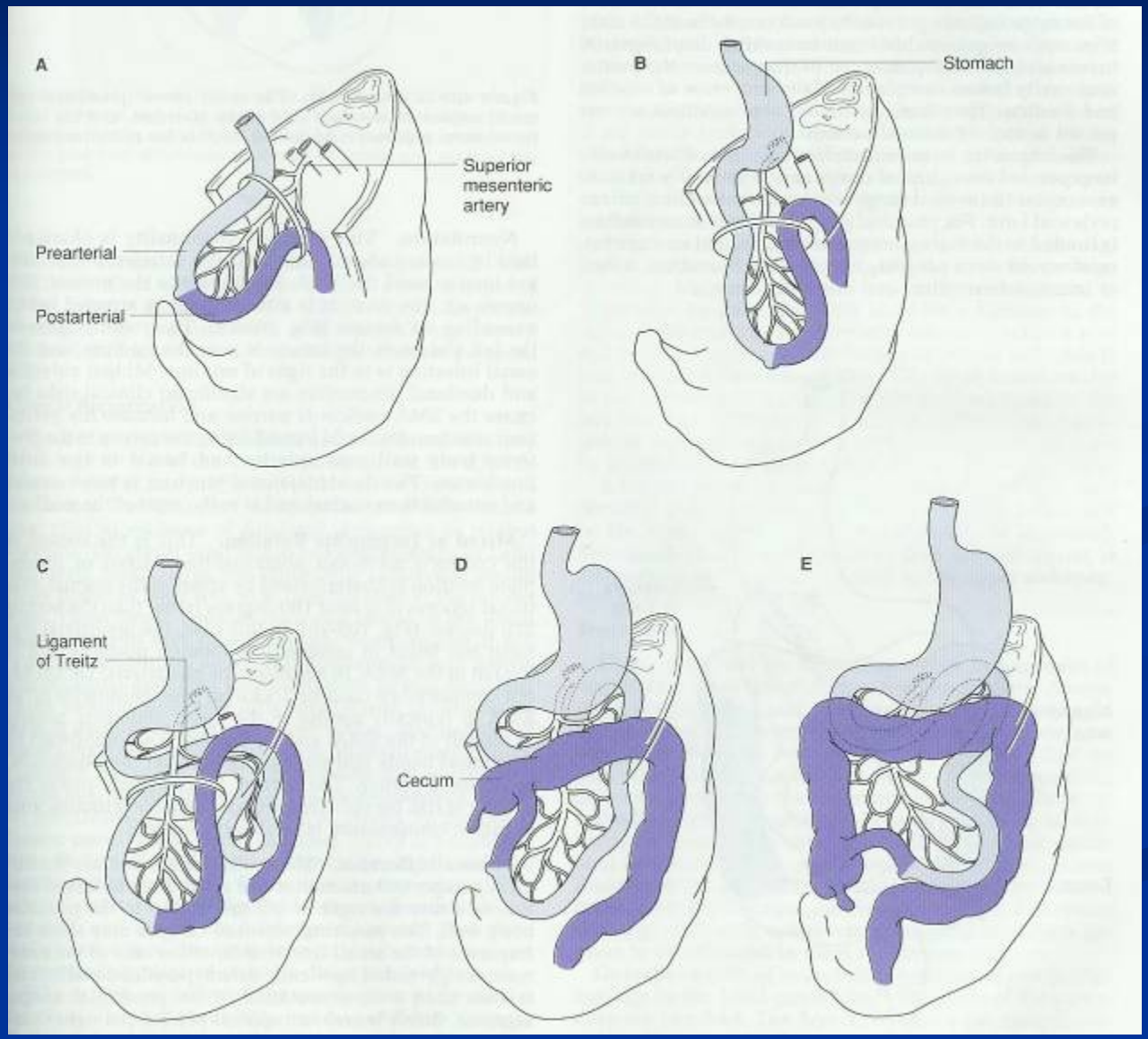
Embryology

- **StageIII** – *fixation*
  - 12th week until term
  - Fixation of intestine to posterior body wall
    - Ligament of Treitz
    - Cecum to right iliac fossa
    - Base of mesentery
    - Ascending and Descending colon
  - Cecal descent to RLQ



Normal Rotation



Epidemiology

- Incidence 1 in 6,000 to 1 in 200 live births
- Autopsy studies suggest 0.5 – 1% of the population affected
- Slight Male predilection (2:1)
- 50-75% discovered in 1st month of life, 90% in children less than 1 year old
- Unknown number go undetected some of which are discovered on coincidental GI imaging or surgery

Anomalies Associated with Intestinal Malrotation

Congenital diaphragmatic hernia

Abdominal wall defects (omphalocele and gastroschisis)

Duodenal atresia

Jejunal atresia

Meckel diverticulum

Duodenal web or stenosis

Hirschsprung disease

Imperforate anus

Esophageal atresia with tracheoesophageal fistula

Biliary atresia

Prune belly syndrome

Cardiac anomalies

Situs inversus

Mesenteric cysts

Renal anomalies

Right isomerism syndromes (polysplenia)

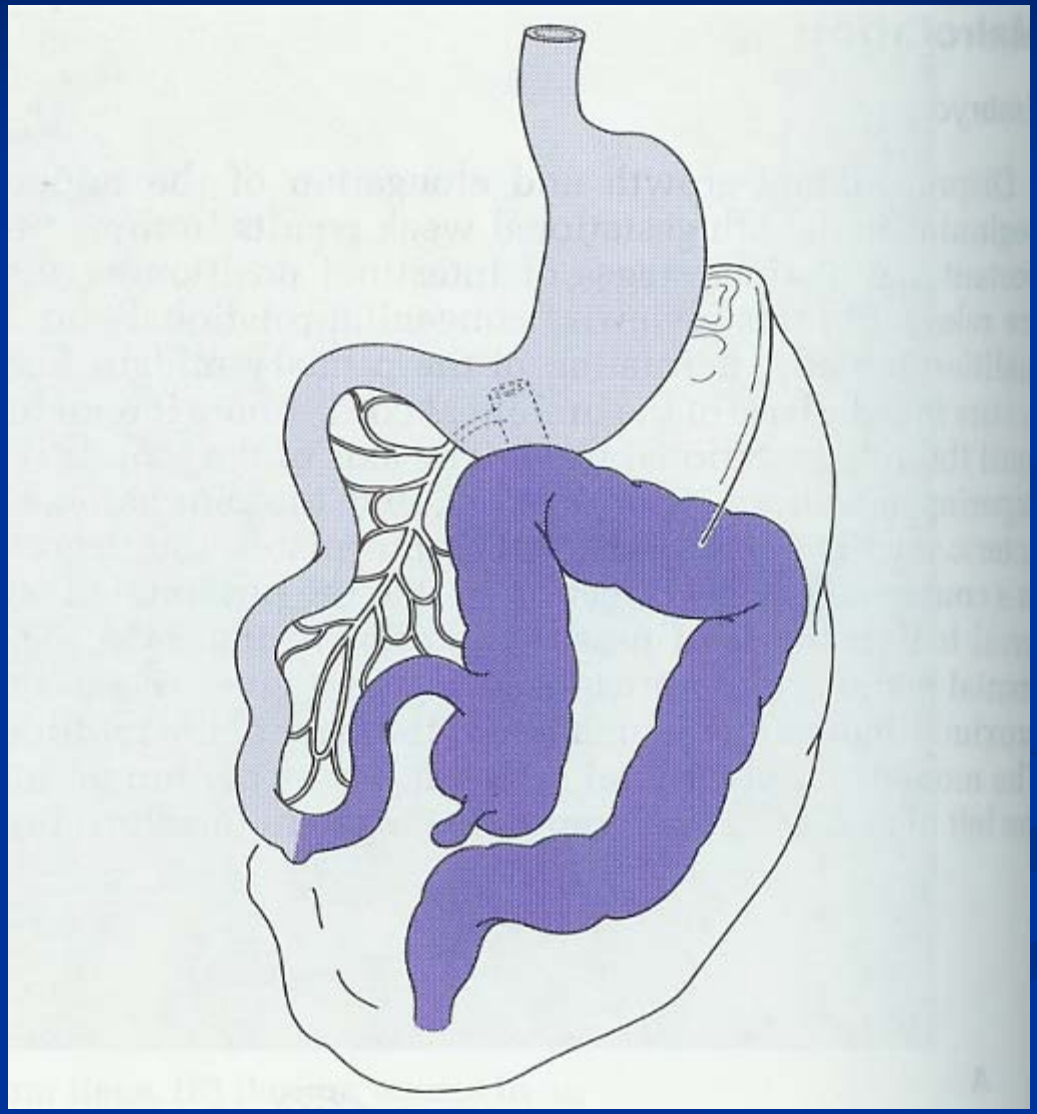
Left isomerism syndromes (asplenia)

UpToDate®: 'Intestinal malrotation'

Types of Malrotation

- **Nonrotation**
  - Lenghtening of midgut with no rotation or less than 90° CCW
  - Colon on the left and small intestine to right of midline
  - Mesentery in turn forms a narrow base as the gut lengthens on the SMA without rotation
  - Midgut volvulus and duodenal obstruction are significant risks

Nonrotation



Types of Malrotation

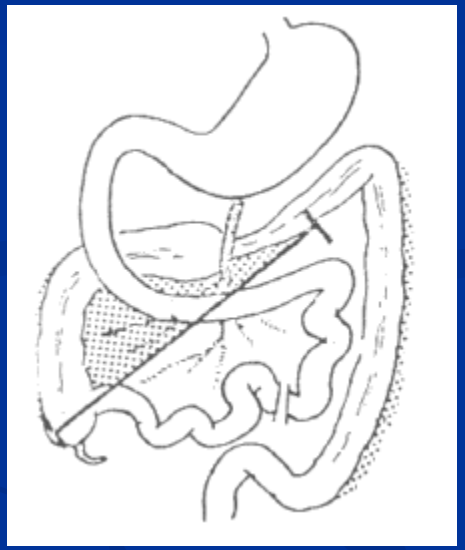
- **Mesocolic Hernias**
  - Rare anomaly
  - Failure of fixation of either the right or left mesocolon to the posterior body wall
  - Predisposed to entrapment of the small intestine on either side resulting in obstruction, incarceration , and strangulation

Types of Malrotation

- **Incomplete Rotation**
  - Arrest in Stage II.
  - Peritoneal (Ladd's) bands running from the misplaced cecum that cross the duodenum & pass to the undersurface of the liver or posterior abdominal wall to the mesentery. Can cause duodenal obstruction.
  - Ladd bands are abnormal peritoneal reflections
  - Mesentery also forms a narrow base and is prone to CLOCKWISE twisting.

Types of Malrotation

- **Reverse Rotation**
  - Rare anomaly.
  - Bowel rotates in varying degrees in a clockwise direction.
  - DJ loop is anterior to SMA & CC loop in retroarterial leading to colonic obstruction.
  - Cecum maybe Right or Left sided.



Clinical Presentation

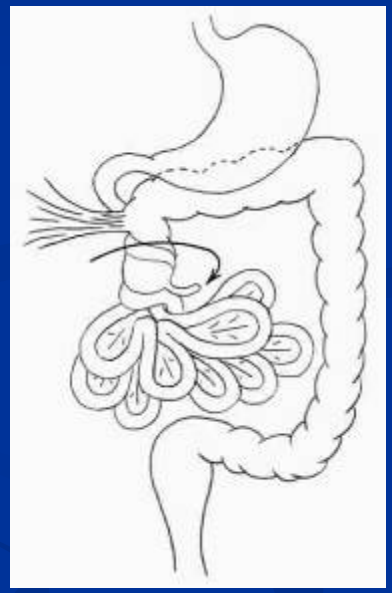
- **Newborns**
  - Bilious vomiting 95%
  - Bloody vomitus and guaiac-positive stools as a result of intestinal necrosis
  - Abdominal pain, tenderness, distention, peritonitis, shock

Clinical Presentation

- **Older Children and Adults**
  - Intermittent vomiting 30%
  - Intermittent abdominal pain 20%
  - Volvulus 10-15%
  - Less commonly enteropathy, pancreatitis, peritonitis, biliary obstruction, motility disorders, chylous ascites

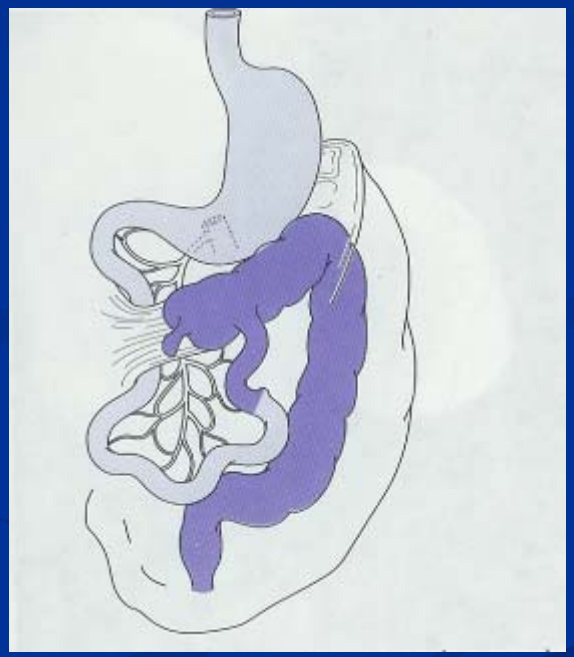
Clinical Presentation

- **Midgut Volvulus**
  - Twisting of bowel around SMA pedicle
  - Life-threatening vascular insufficiency to SMA distribution
  - Emergent surgical intervention is critical with time dependent clinical outcome -necrotic bowel: ↑ mortality by 25 times



Clinical Presentation

- **Small Bowel Obstruction**
  - Duodenal obstruction (Ladd's bands)
  - Forceful bilious vomiting



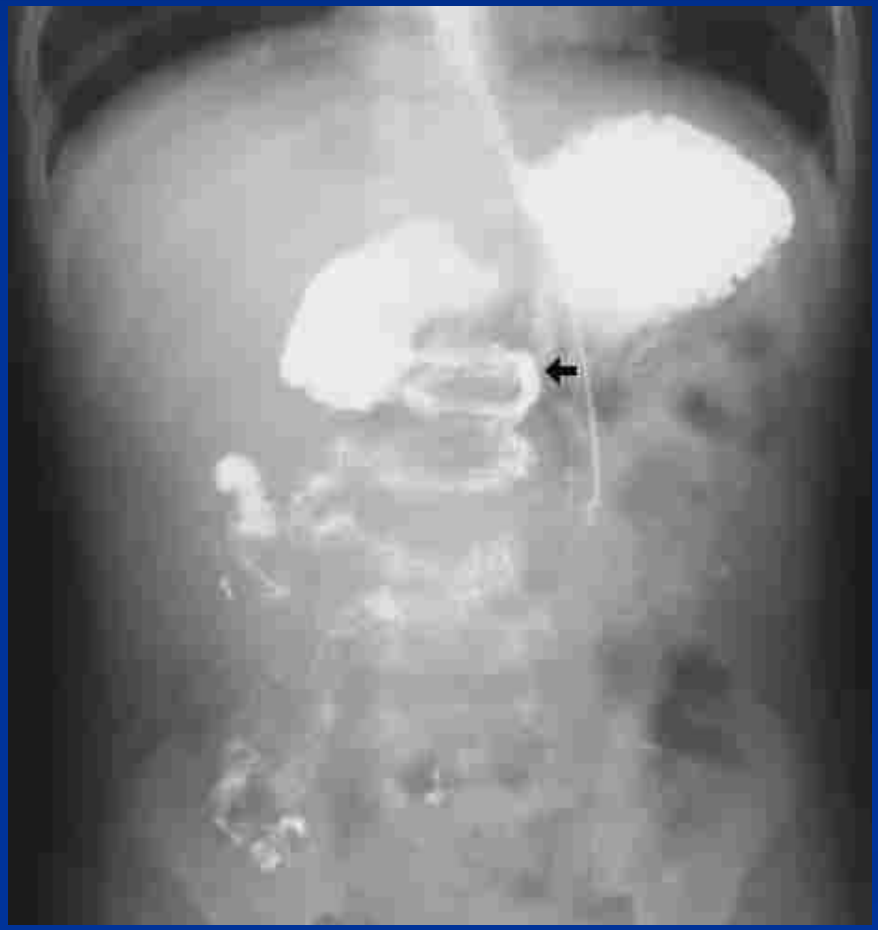
Diagnosis

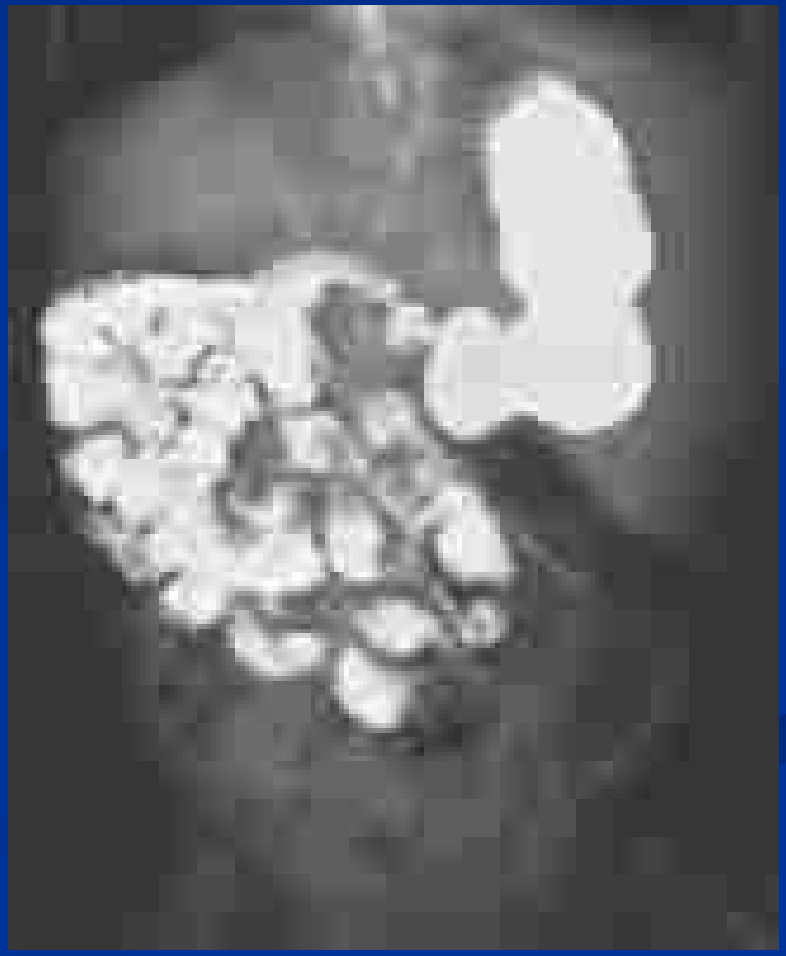
- **Plain Films**
  - Rarely helpful in diagnosing
  - Suggestive findings
    - Gasless abdomen
    - Intestinal dilatation
  - Diagnostic findings
    - NGT or OGT extending into abnormally positioned duodenum
    - "Double-bubble" sign

Diagnosing

- **UGI series**
  - "Gold Standard" best at visualizing duodenum
  - Experienced pediatric radiologist required
  - Diagnostic Findings
    - Abnormal position of duodenum (Ligament of Treitz on right)
    - Duodenal obstruction
    - Beak appearance of duodenum with volvulus
  - 6-14% false-negative, 7-15% false positive rates
  - SBFT helpful for equivocal studies

UGI Series





Diagnosis

- **Barium Enema**
  - Identifies colonic malrotation
  - Useful as adjunct to UGI series
  - High false-negative and false-positive rates
  - Diagnostic of volvulus involving transverse colon

Diagnosis

- **Ultrasound**
  - **Radiologist dependent**
  - Abnormal position of SMV (anterior or left of SMA)
  - Dilated duodenum
  - "Whirlpool" sign of volvulus

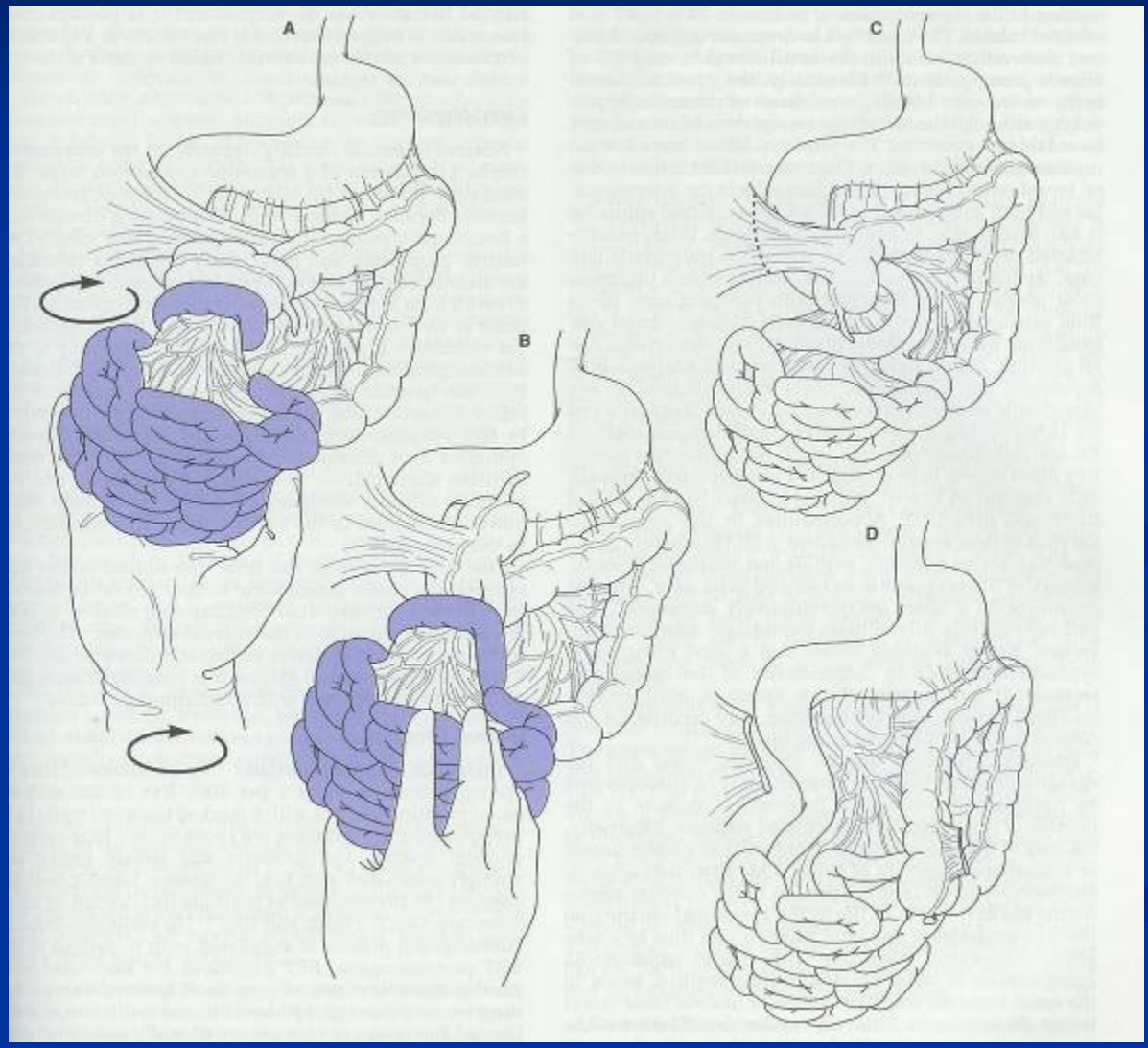
Diagnosis

- **CT scan**
  - Increasingly used in a variety of clinical settings
  - Can reveal abnormal location of small bowel and cecum
  - Inverse relationship of SMV to SMA identified
  - Whirling pattern of midgut volvulus

Treatment

- **Ladd's Procedure**
  - Detorsion of Midgut Volvulus
  - Lysis of adhesive bands
  - Placement of Small bowel in non rotated position on the right side of abdominal cavity.
  - Placement of Large bowel on the left side of abdominal cavity.
  - Inversion Appendectomy
  - Cecopexy does not alter rate of pre or postop complications

Ladd's Procedure



Laparoscopic Ladd's Procedure

- Feasibility Study 1994-1997
- 12 pts; Ages 5 days to 4 months; 3 to 7 kg
- No patients with acute volvulus or bowel ischemia included
- Standard Ladd's procedure with appendectomy
- 3 – 3.5mm trocars

Bass KD. Laparoscopic Ladd's procedure in infants with malrotation. *J Pediatric Surg* 1998.

Laparoscopic Ladd's Procedure

- Results
  - All cases completed laparoscopically
  - Avg OR time 58 min (35-120 min)
  - No operative or postoperative complications reported
- Conclusions
  - Earlier feeding
  - Decreased hospital stays

Bass KD. Laparoscopic Ladd's procedure in infants with malrotation. *J Pediatric Surg* 1998.

Malrotation in Adults

- Presentation in adults
  - Acute obstructive symptoms and signs of impending abdominal catastrophe
  - Chronic abdominal complaints
  - Atypical symptoms from a common abdominal disease (i.e. appendicitis in a subhepatic cecum)

Kapfer SA. Intestinal Malrotation – Not Just the Pediatric Surgeon's Problem. *J Am Coll Surg* 2004.

Malrotation in Adults

- Scenario 1 – Symptomatic Malrotation
  - Treatment guided by acuity of presentation
  - Resuscitation and prompt exploration of acute abdomen
  - Ladd procedure with resection of nonviable bowel
  - Second-look laparotomy for questionable viability

Kapfer SA. Intestinal Malrotation – Not Just the Pediatric Surgeon's Problem. *J Am Coll Surg* 2004.

Malrotation in Adults

- Scenario 2 – Discovery during evaluation and treatment of unrelated complaint
  - Address malrotation only if it does not add undue risk to the procedure
  - Discovery at time of operation for unrelated disease
    - Address primary disease
    - Dilemma of consent
    - Repair electively after discussion of risks and benefits

Kapfer SA. Intestinal Malrotation – Not Just the Pediatric Surgeon's Problem. *J Am Coll Surg* 2004.

Malrotation in Adults

- Scenario 3 – Asymptomatic with discovery on radiologic examination
  - Most controversial scenario
  - Intervention only for symptomatic disease vs intervention for all at risk for midgut volvulus
  - Selective intervention based on degree of malrotation

Kapfer SA. Intestinal Malrotation – Not Just the Pediatric Surgeon's Problem. *J Am Coll Surg* 2004.

Atypical Malrotation

- Records of 201 patients, undergoing operations for Malrotation retrospectively reviewed over a 5 year period.
- Malrotation classified based on location of Ligament of Trietz (LOT)
  - Typical: on the right or absent
  - Atypical:
    - High: at or left of midline (Above T12)
    - Low: at or left of midline (Below T12)
  - Cecal Position:
    - Normal: RLQ
    - Abnormal: Somewhere other than RLQ
- Atypical malrotation pts at significantly lower risk of volvulus and internal hernia

Mehall, J, MD et al. Management of Typical and Atypical Intestinal Malrotation. *J Peds Surg* 2002

Summary

- Rotational anomalies are the result of arrest of normal rotation of the embryonic gut
- Early diagnosis and surgical intervention reduces morbidity and mortality . Must have a high index of suspicion for infants with bilious vomiting.
- Malrotation, regardless of age and presence of symptoms is treated surgically with Ladd procedure.

Questions

Which study is the test of choice for diagnosing malrotation?

- Ultrasound
- CAT scan
- Endoscopy
- UGI series
- Barium enema

Questions

Which step is not part of the Ladd procedure?

A. Appendectomy
B. Cecopexy
C. Lysis of Ladd's band
D. Detorsion of volvulus

Question

Midgut volvulus is most common with which type of malrotation?

A. Reverse rotation
B. Hyperrotation
C. Nonrotation
D. Mixed rotation
E. Intraperitoneal hernia