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Intestinal Obstruction in Newborns

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Morbidity and Mortality Conference

Case Presentation

- SG -10 day old baby girl, born at 30 weeks gestation
- Associated with an in utero twin fetal demise
- Apgar score 2, 7 (1, 5 min); Birth weight 1.3 kg
- PMH: patent PDA
- Increasing abdominal distention and obstipation for first 4 days of life

Physical Exam

- VS: T 97.8 F BP 68/34 HR 164 RR 16 O_2 sat 91%
- CV: RRR, S1S2 normal
- Pulm: CTA bilaterally ; intubated
- Abdomen: very distended with visible bowel loops, soft, nontender
- Rectal: unable to go beyond 1.5 cm

- Labs:

CBC : 3.1/12/36/261

BMP: 138/3.8 108/24 8.4/0.7
<158

PT/INR/PTT: 13/1.33/29.8

Radiologic Studies

- AXR-dilated small bowel loops with no air in pelvis
- Barium enema: microcolon with only left colon visualized
- Upper GI: no progression of contrast beyond the stomach consistent with marked ileus

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Abdominal Exam

Intraoperatively

- 6/9: Exploratory laparotomy
  - Reduction of jejunal volvulus
  - Intestinal resection of atretic segments
  - Tapering enteroplasty of the dilated reduced bowel with primary anastamosis


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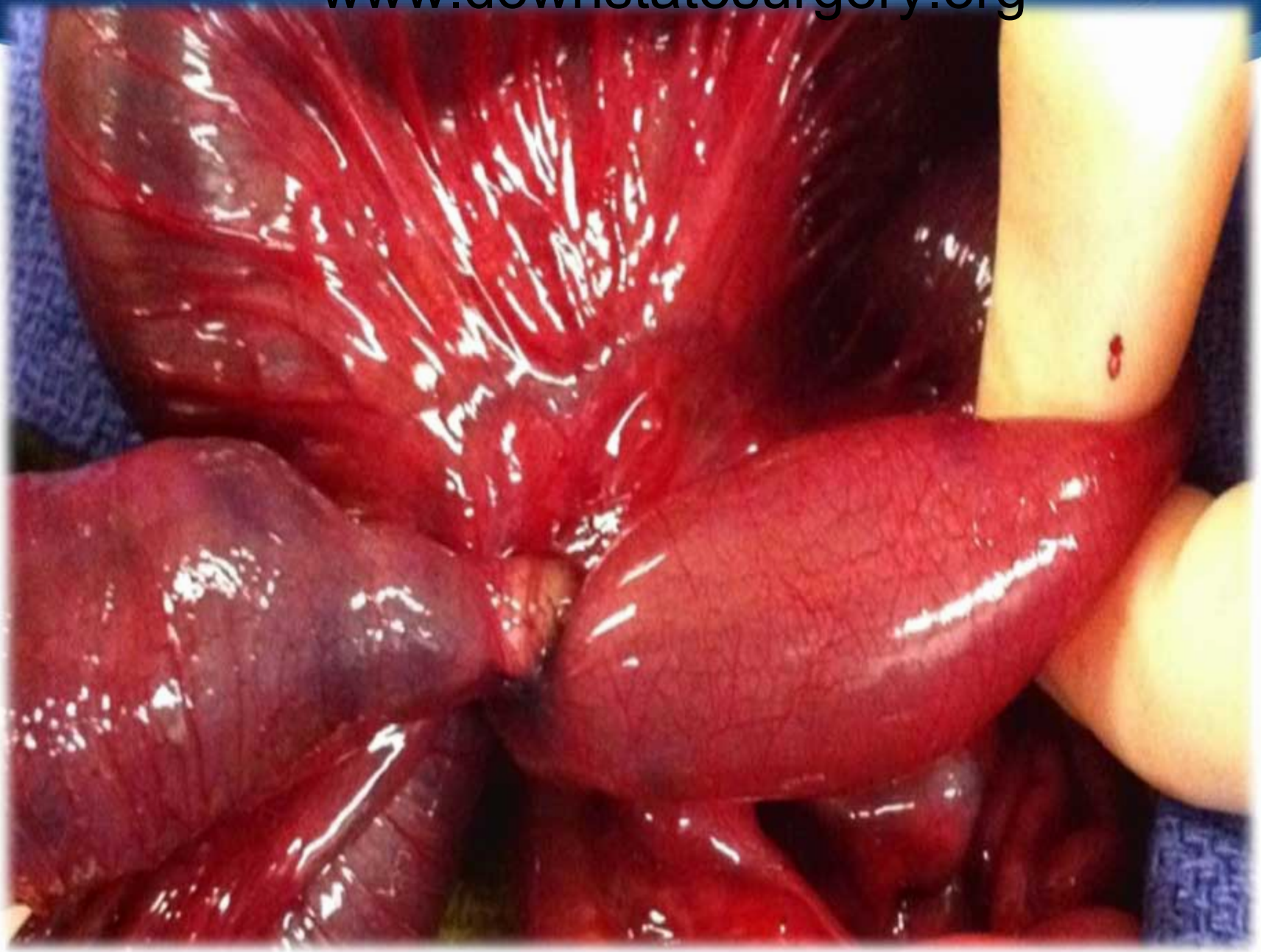

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Meconium

Hospital course

- Abdomen remained nondistended and soft
- No bowel movement for 3 ½ weeks except for mucus plugs
- Upper GI series: dilated proximal loops of bowel
- 7/5: Exploratory laparotomy, adhesiolysis, small bowel resection, enterorrhaphy (from staple line) and intestinal decompression with removal of meconium plugs

Hospital Course

- Post operatively no bowel movement for 2 weeks
- 7/19: exploratory laparotomy, adhesiolysis, intestinal decompression through enterotomy, jejunal resection of previously volvulated segment
- Meconium passed on POD #3.
- Extubated currently with normal bowel function.

Intestinal Obstruction in the Newborn

- Think of…
  - Atresia
  - Malrotation with volvulus
  - Meconium ileus
  - Hirschsprung's disease

Differential Diagnosis in neonate with abdominal distention and obstipation

- Meconium Ileus
- Distal jejuno-ileal atresia
- Hirschsprung's Disease

Intestinal Atresia

- Congenital obstruction caused by complete occlusion of intestinal lumen
- Mesenteric vascular accidents in utero
- Multifactorial
- Incidence:
  - Jejunoileal: 1 in 330 (US)- 1 in 1500
- Increased in maternal use of pseudoephedrine and ergotamine + caffeine

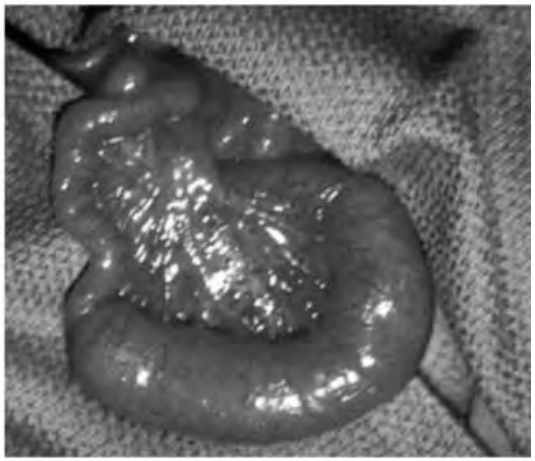
Clinical Presentation

- Symptoms:
  - Bilious emesis- proximal
  - Abdominal distention- distal
  - Maternal polyhydramnios (24%)
  - Failure to pass meconium
- Prenatal Ultrasound
  - More detectable in duodenal atresia
  - Multiple distended loops of bowel with vigorous peristalsis
  - Echogenic bowel
  - <1/3 cases recognized

Radiographic findings

A

B



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- High Jejunal Atresia
  - Few air fluid levels

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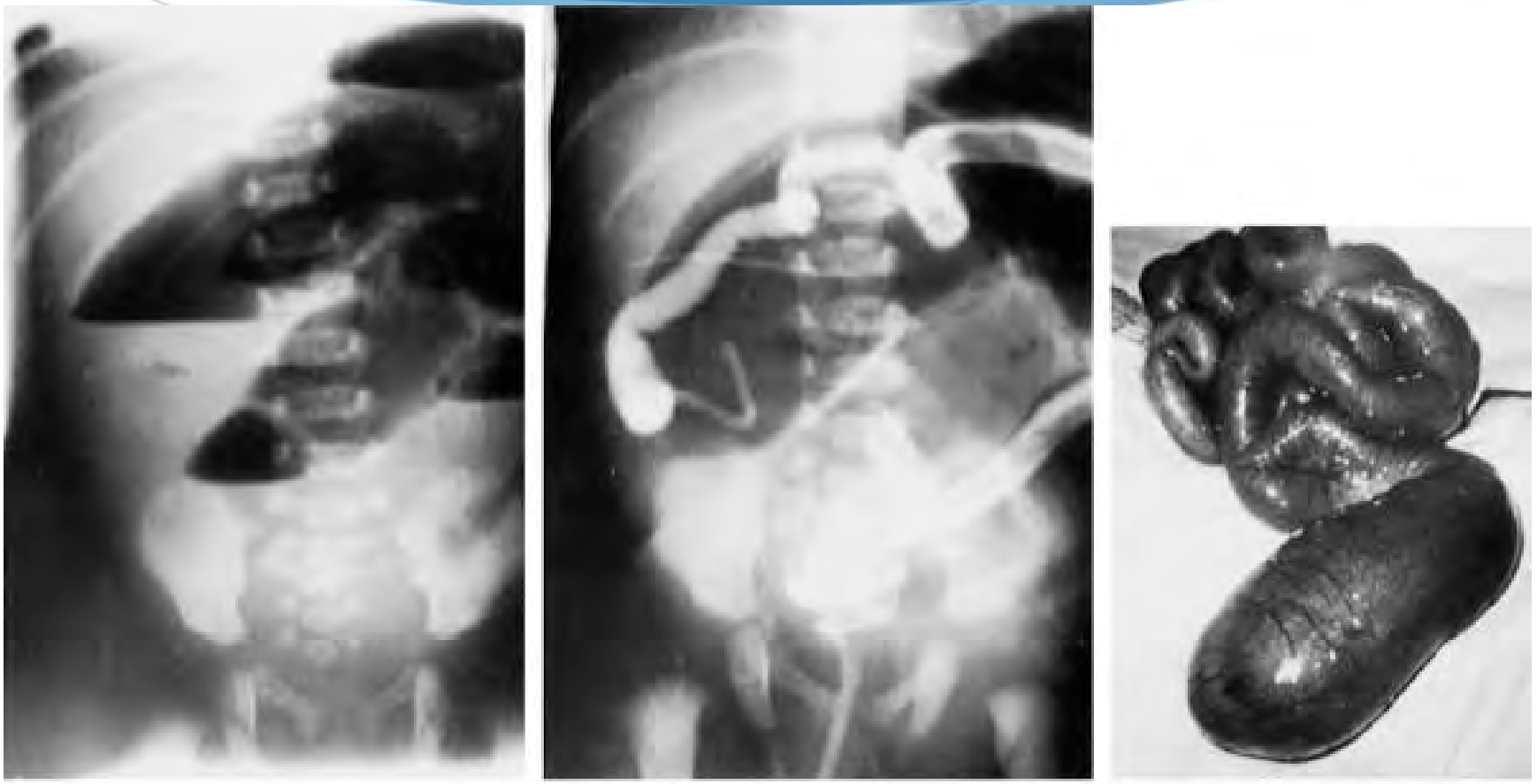
Ileal Atresia



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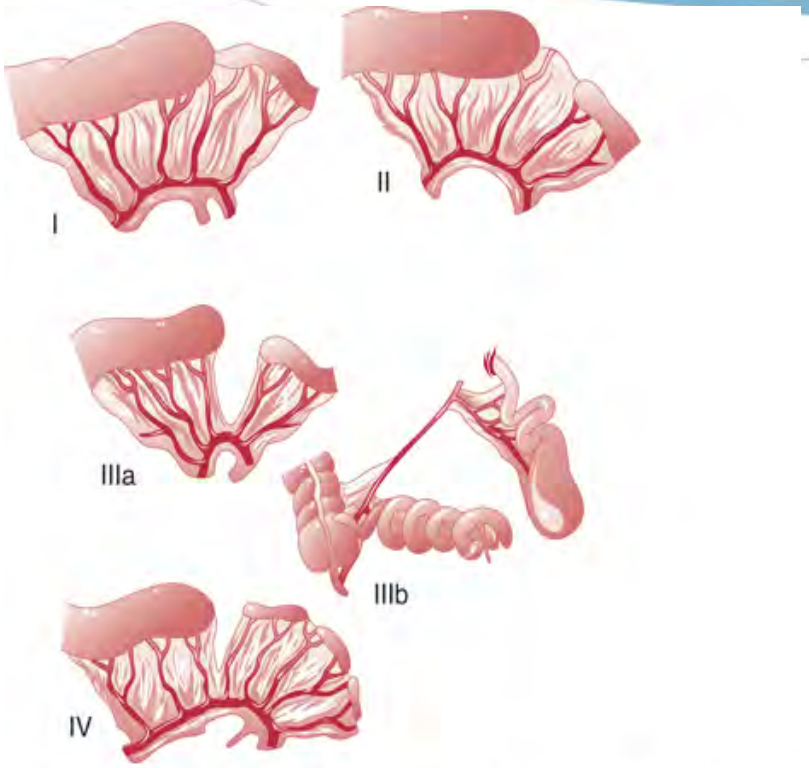
Intestinal Atresia Classifications



Source: Gerard M. Doherty: *CURRENT Diagnosis & Treatment: Surgery, 13th Edition*: http://www.accessmedicine.com

- Type I: mucosal atresia with intact bowel wall and mesentary
- Type 2: 2 atretic blind ends joined by fibrous cord + intact mesentary

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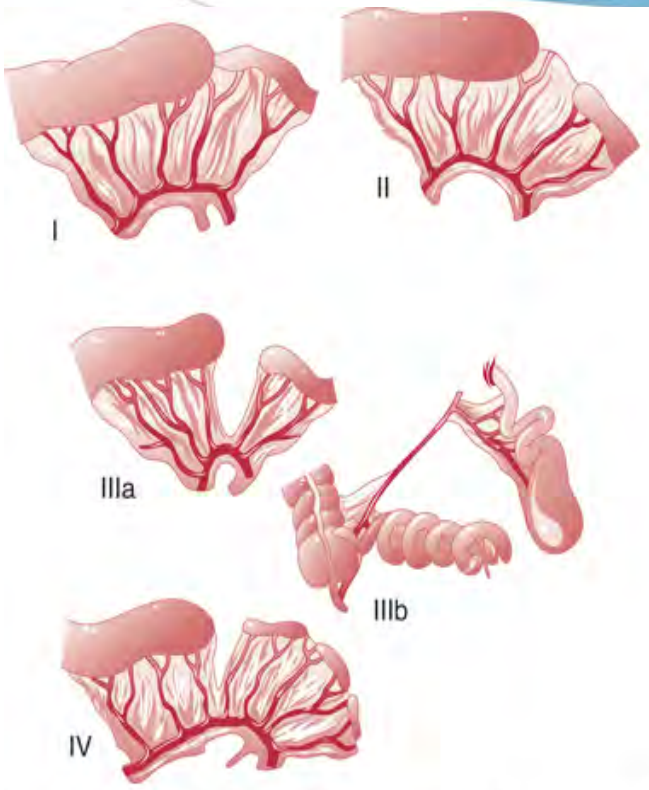
Intestinal Atresia Classifications



Source: Gerard M. Doherty: *CURRENT Diagnosis & Treatment: Surgery, 13th Edition*: http://www.accessmedicine.com

- Type 3:
  - A- two ends of atresia separated by V shaped mesenteric defect
  - B- “Christmas tree” deformity
    - Bowel distal to the atresia receives its blood supply in a retrograde fashion from ileocolic or right colic artery
- Type 4- mutliple atresias
  - “String of sausage” or “string of beads”

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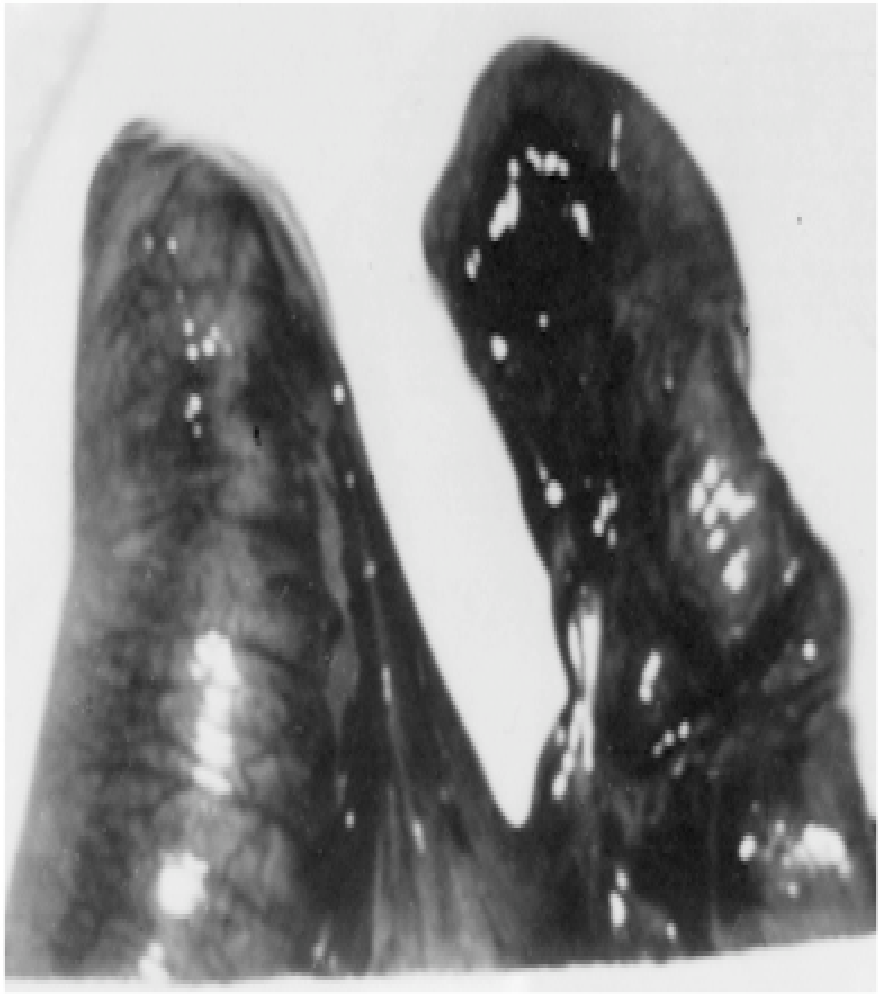
Type 3 Intestinal Atresia

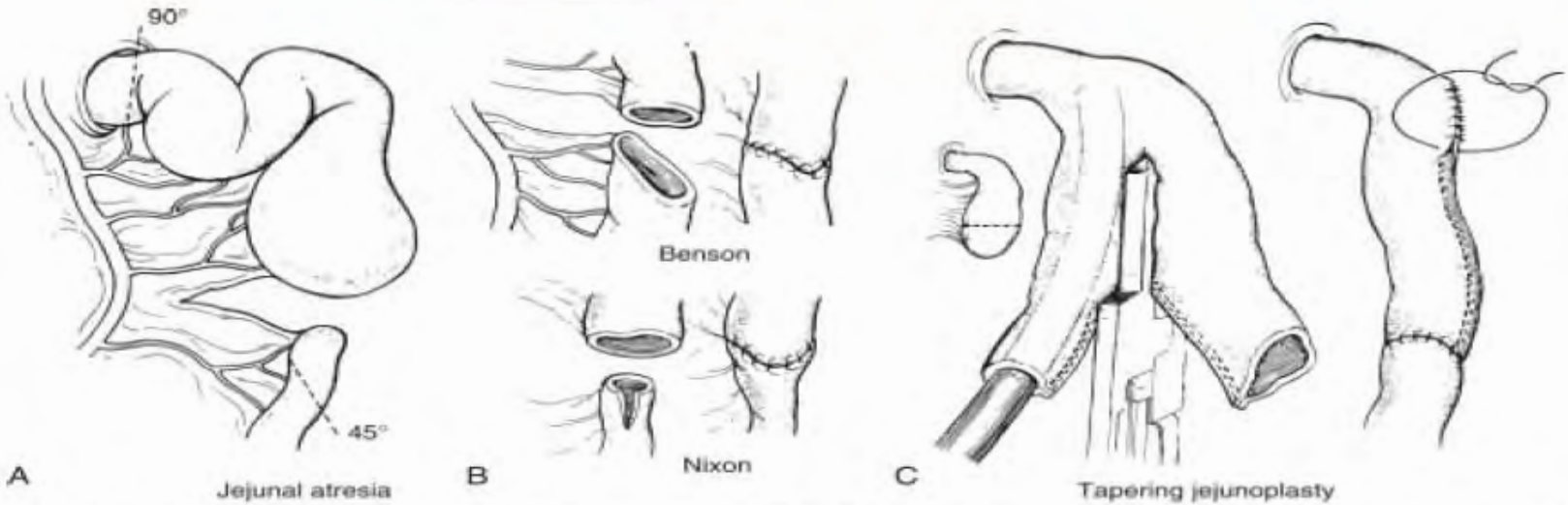


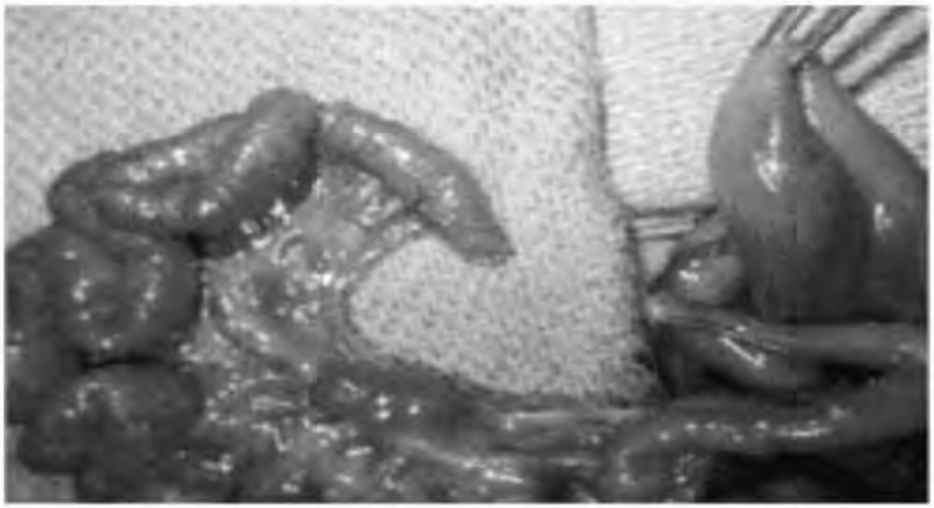
3A

3B

Operative Interventions

- In proximal jejunal atresia- resection at ligament of Treitz followed by end – to- oblique anastamosis
- If limited length- tapering enteroplasty
  - Retention of dilated blind proximal segment- functional obstruction
  - Smooth muscle hypertrophy and enlargement of bowel diameter
  - Ineffective peristalsis


90°
45°
A
Jejunal atresia
Benson
Nixon
B
C
Tapering jejunoplasty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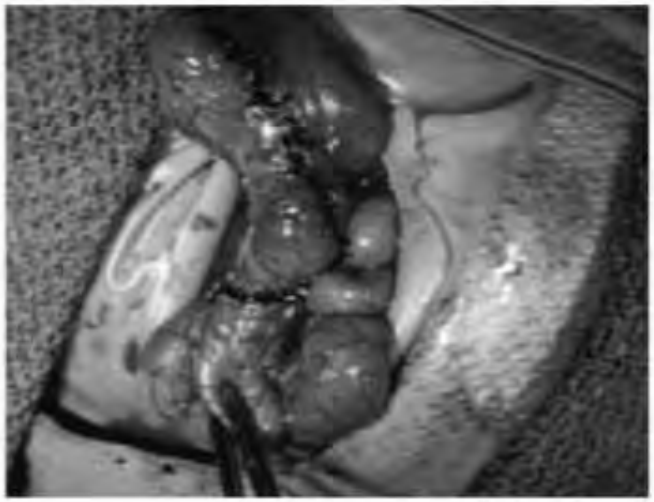
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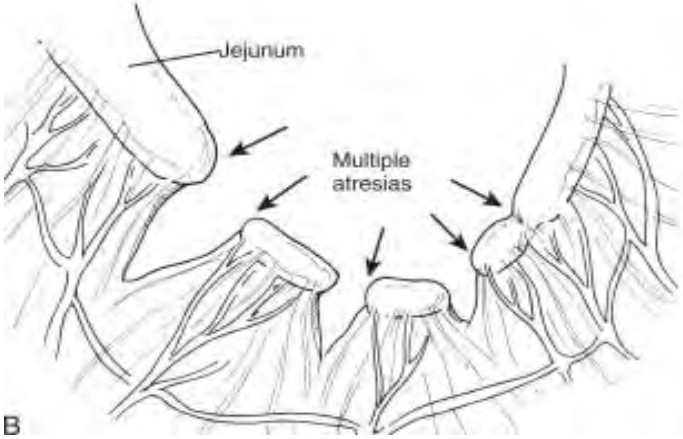
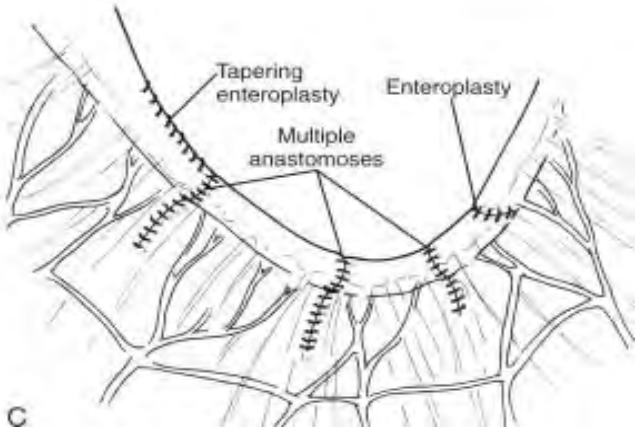
Multiple Atresias

A



B



C

Attempting to preserve bowel length

Morbidity and Mortality

- Most common cause of early death: infection related to pneumonia, peritonitis or sepsis
- Postoperative complications:
  - Functional intestinal obstuction at anastamosis
  - Anastamotic leak (15%)
- De Lorimier and associates concluded that resection improved survival in jejunal atresia from 39% to 66%.
  - Little effect on overall survival in ileal atresia

Hirschsprung's Disease

- Incidence: 1 in 5000 live births
- M:F ratio 4:1
- Among families of children with HD incidence ↑ to 6%
- Associated with trisomy 21 (4.5-16%)
- atresias

HD- Clinical Presentation

- Any child with constipation dating back to newborn period
  - 90% diagnosed as newborns
- History of delayed passage of meconium **w/in 48 hours of life**
- Other si/sx:
  - Abdominal distention absent at birth and tight anus
  - Poor feeding
  - Emesis

Radiographic Studies

- AXR- distended loops of intestine with paucity of air in rectum
- Contrast enema (76-92% accuracy)
  - Narrow spastic distal intestinal segement with a dilated proximal segment
  - **the point of caliber change is the key radiographic finding
  - Most commonly the transition point at rectosigmoid

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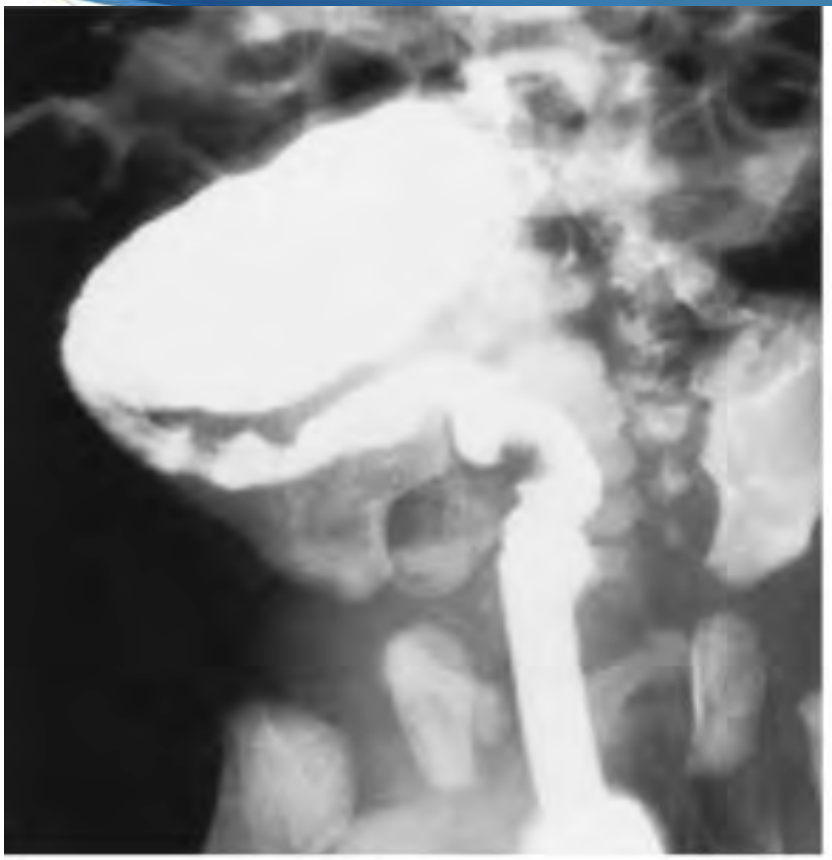
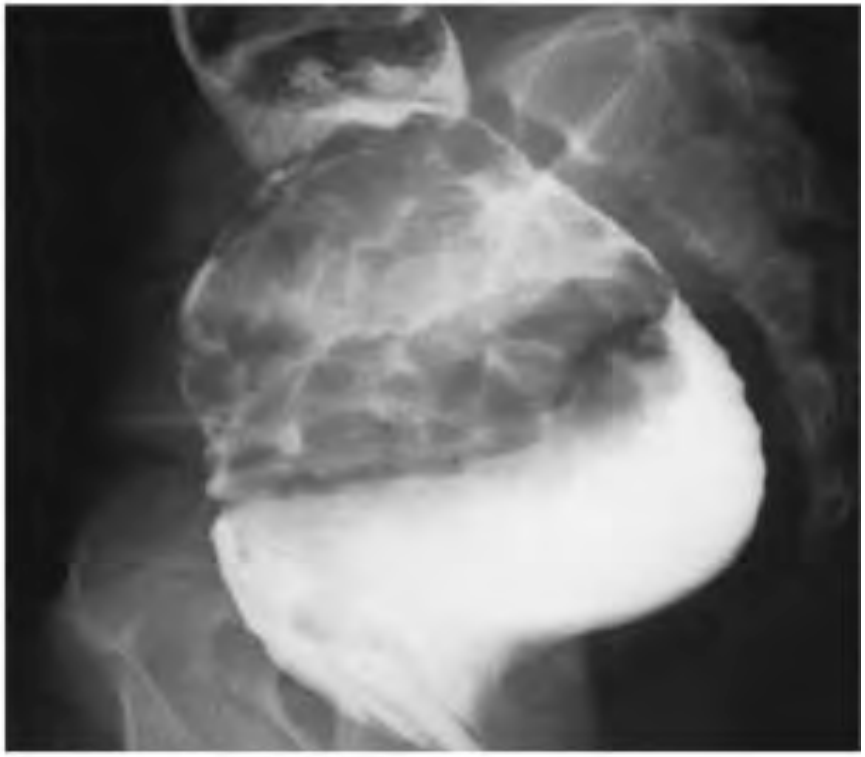
Hirschsprung's Disease



A

Hirschsprung's Disease



B

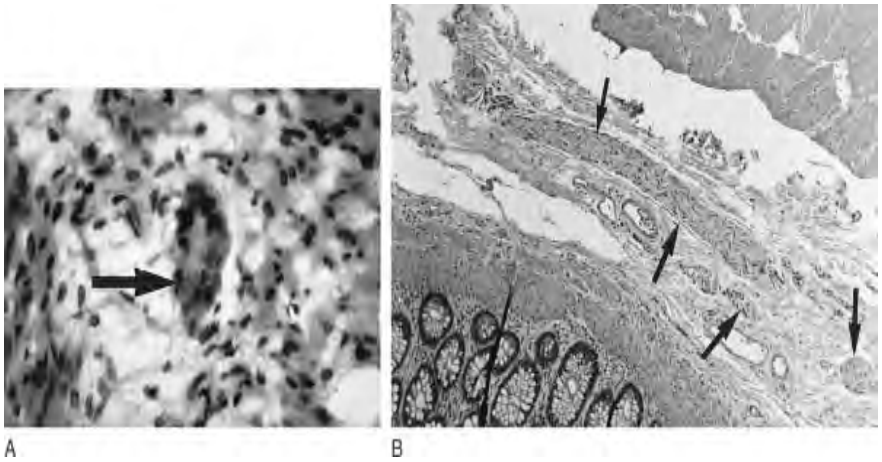
Constipation

Anorectal Manometry

- Measures absence of a relaxation reflex after a distending bolus is created in the rectal lumen
- Elevated resting anal sphincter pressure

Rectal Biopsy

- GOLD STANDARD for diagnosis of Hirschsrung's disease
- Diagnostic accuracy 99.7%
- Alternate method: full thickness posterior rectal wall biopsy



Cause of Hirschsprung's Disease

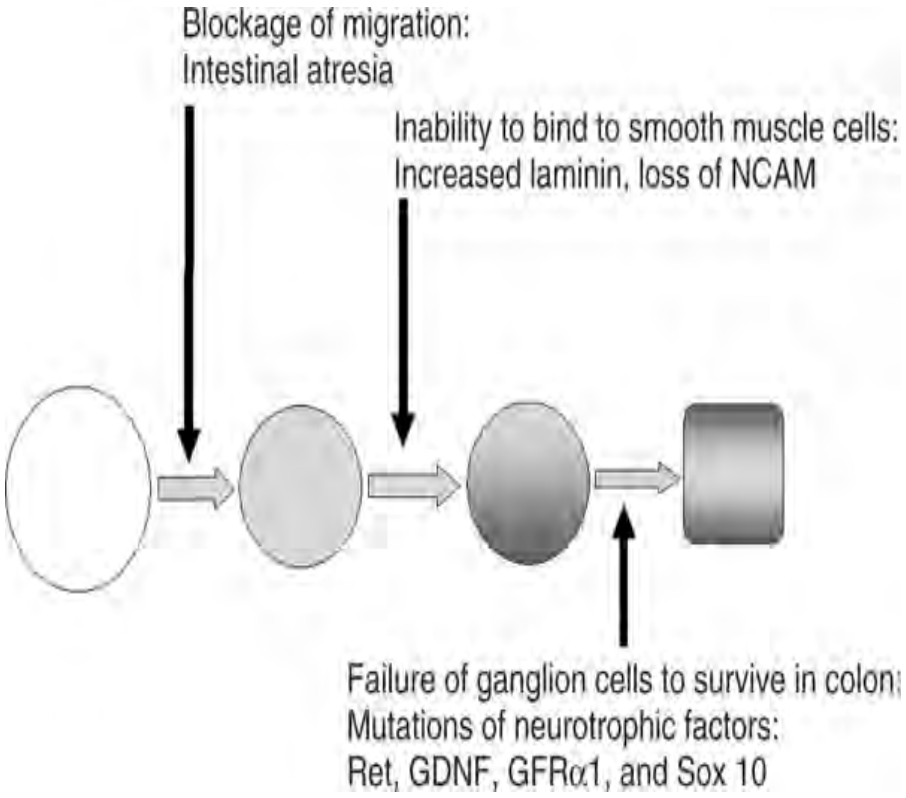
- Sporadic occurrence accounts for 80-90% of cases
- RET gene mutations in 35% of sporadic cases and 49% of familial cases
- 5-10% show mutations in other genes

Embryology

- 3 phases in development of enteric nervous system:
  - **Induction phase**
  - **Neural crest cell migration**
  - **Differentiation of the neural crest cell precursos**
- 5 wks gestation- NC cells in esophagus
- 7 weeks- midgut
- 12 weeks- distal colon

Theories of embyologic defect:

- "Failure of migration"
- "Hostile environment"



Pathology

- In infancy the intestine may appear fairly normal
- As infant ages- proximal ganglionic bowel hypertrophies
- Rectosigmoid – 80%
- Histology:
  - Absence of ganglion cells in the distal intestine

Endorectal Pull-Through

- Single stage operation; Initially described by Soave in 1964
- LLQ incision
- At least 5 cm proximal to the first area of normal ganglion cells
- Endorectal dissection approximatly 2 cm below peritoneal reflection by incising the seromuscular layer
  - Carried down 0.5cm of the dentate line in newborns

Endorectal Pull through -2

- Evert mucosal-submucosal tube and incise on anterior half
- Normal ganglionic intestine brought down to this point
- Anastamosis with absorbable sutures

Source: Brunicardi FC, Andersen DK, Billiar TR, Dunn DL, Hun
Pollock RE: *Schwartz's Principles of Surgery, 9th Edition:* http:

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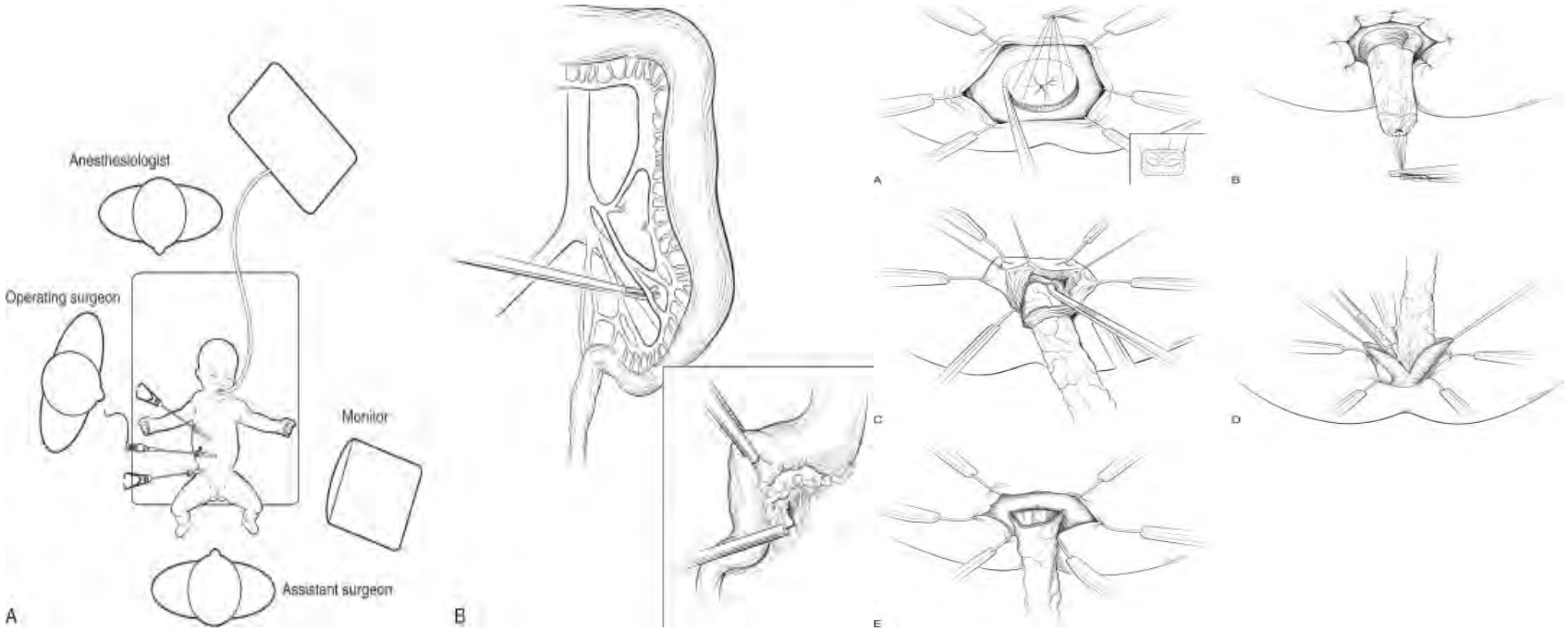
Different surgical techniques



Laparoscopi
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Transana
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Complications

- Early Post-Pull-Through
  - Intestinal obstruction (8-13%)
  - Early anorectal stenosis (10-20%)
- Late
  - Incontinence (3-8%)
  - Constipation (6-30%)

Meconium Plug Syndrome

- Confirmed by contrast enema radiograph to find “plugs” in sigmoid or descending colon
- Spontaneously pass after withdrawal of the enema catheter
- Pathogenesis may relate to bowel hypomotility
- Associated with: prematurity, hypotonia, hypermagnesemia, sepsis, hypothyroidism, and Hirshsprung’s disease in 5%
- Sweat test and rectal biopsy

Meconium Ileus

- 1905- Landsteiner
- Seen in 20-35% patients with cystic fibrosis
  - Mutation in CF transmembrane regulator (CFTR) gene
- Patients with meconium ileus represent a distinct phenotype
  - Earlier presentation and worse pulmonary function
- Survival 95-100% with aggressive management, nutrition and close monitoring of pulmonary function

Pathophysiology of Meconium Ileus

- Intestinal obstruction secondary to intraluminal accumulation of inspissated and dessicated meconium
- EARLIEST clinical manifestation of CF- 20.8%
- CFTR- chromosome 7, band q31 (Δ F508 mutation)
  - Cyclic adenosine monophosphate-induced chloride channel
- Reduced clearance of secretions
  - Respiratory, GI, biliary, pancreatic and reproductive systems

Pathogenesis of Meconium Ileus

- Intestinal glandular disease plays a dominant role
  - Pancreatic disease plays a secondary role
- Meconium in CF patients is twice as high in concentrations of sodium, potassium and magnesium; ↑ protein nitrogen

− Degradation enzymes

- Protein + mucopolysaccharides → highly viscid rubbery meconium
- Complications : volvulus of heavy loop with perforation, peritonitis, atresia
  - microcolon

Meconium Ileus

- Uncomplicated
  - Presents immediately at birth
  - Abdominal distention, bilious emesis, failure to stool
- Complicated
  - Presents in utero or postnatally
  - Bowel obstruction with evidence of perforation and/or necrosis

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Clinical Features

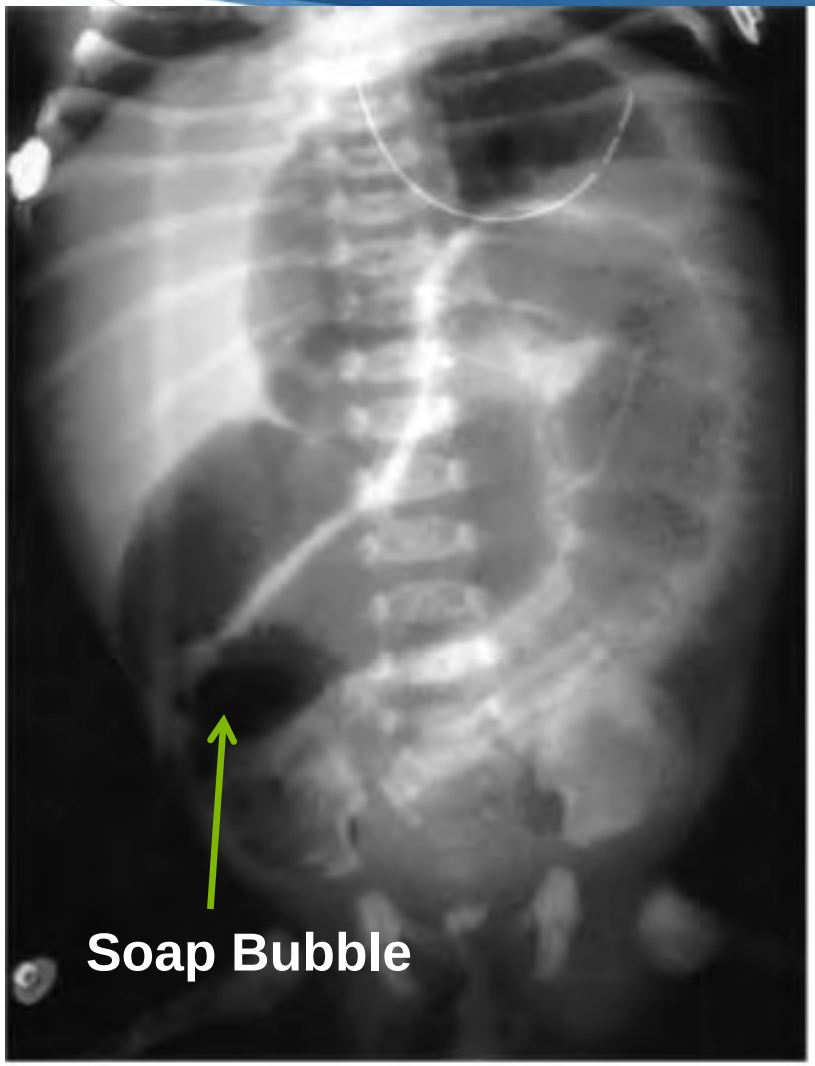
- Family history CF 10-33%
- Polyhydramnios
- Physical Exam:
  - Abdominal distention
  - Visible peristaltic waves
  - Palpable doughy bowel loops
  - **Putty sign**

Radiologic Studies

- **In utero:**
  - echogenic bowel in 3rd trimester;
  - distended bowel
- **Abdominal XR:**
  - Great disparity in size of bowel loops
  - NO or few air fluid levels
  - "**soap bubble" or "ground glass"** in meconium peritonitis
- **Contrast Enema-** microcolon

Lab studies

- **Labs:**
  - Sweat test- 100mg
    - Sweat Cl >60 mEq/L
- **Stool Studies**
  - Meconium albumin>80mg/g
  - Stool trypsin >80 mg/g



A

B

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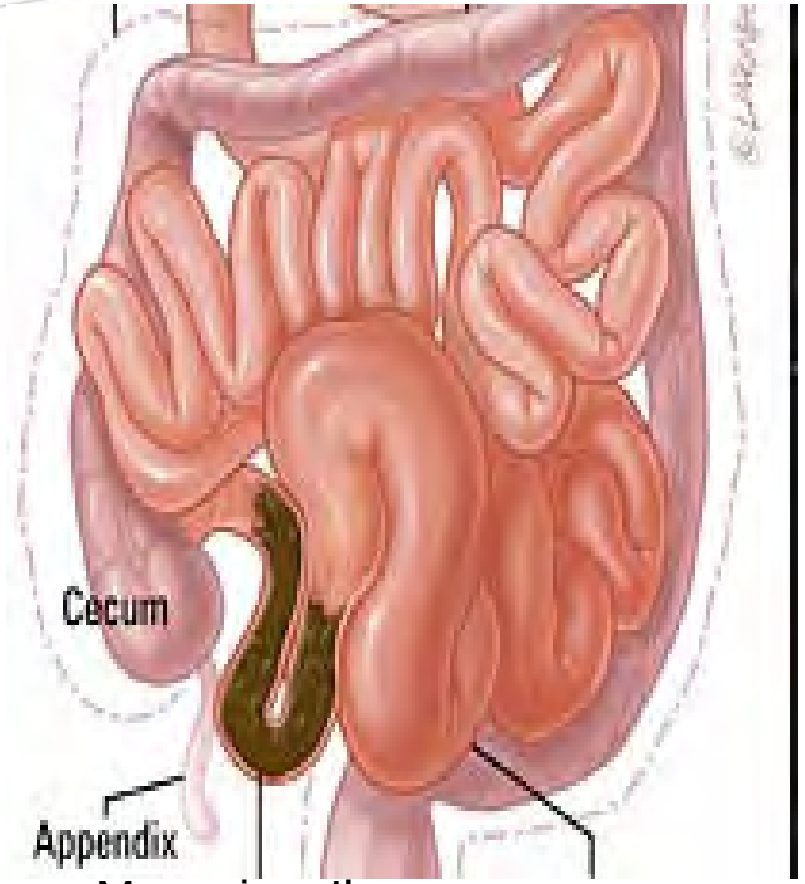
Meconium Ileus



Meconium Ileus

Non-operative Management

- Gastrograffin enema
  - Hyperosmolar, water-soluble, radioopaque solution
  - 0.1% polysorbate 80 (solubilizing agent) and 37% organically bound iodine
  - Transient osmotic diarrhea and a putative osmotic diuresis
- Advantages: reduction in pulmonary morbidity, decreased hospital length of stay
- Disadvantages: delay, intestinal injury, hypovolemia

Operative Management

- Enterotomies with irrigation
  - Warmed saline, 50% diatrizoate solution, H_2O_2, Mucomyst
  - Meconium milked distally into the colon or enterotomy
  - Enterostomy + T- tube
    - Continued irrigation until POD #14 where catheter removed
- Mikulicz double-barreled enterostomy
- Santulli – proximal chimney enterostomy

Operative Management

- Bishop-Koop procedure
  - Limit intraoperative bowel trauma in neonatal period
  - Resect disparately enlarged ileal loop filled with meconium
  - Create an approximately sized end of prox to side of distal ileum
  - Access to the insertion of a catheter into the distal bowel containing the meconium pellets
  - Permit eventual enterostomy closure by bedside ligation of “chimney” stoma

Various Techniques

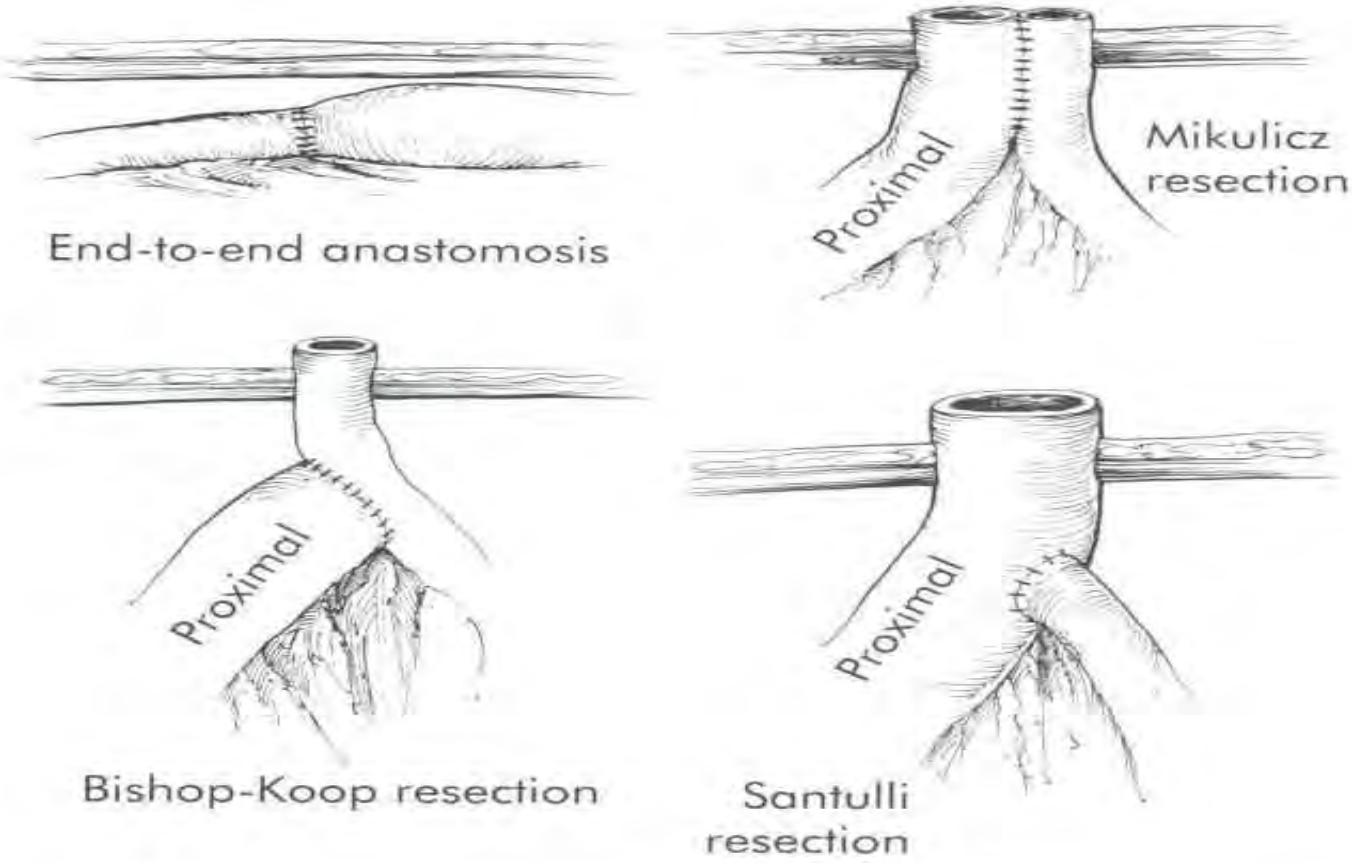
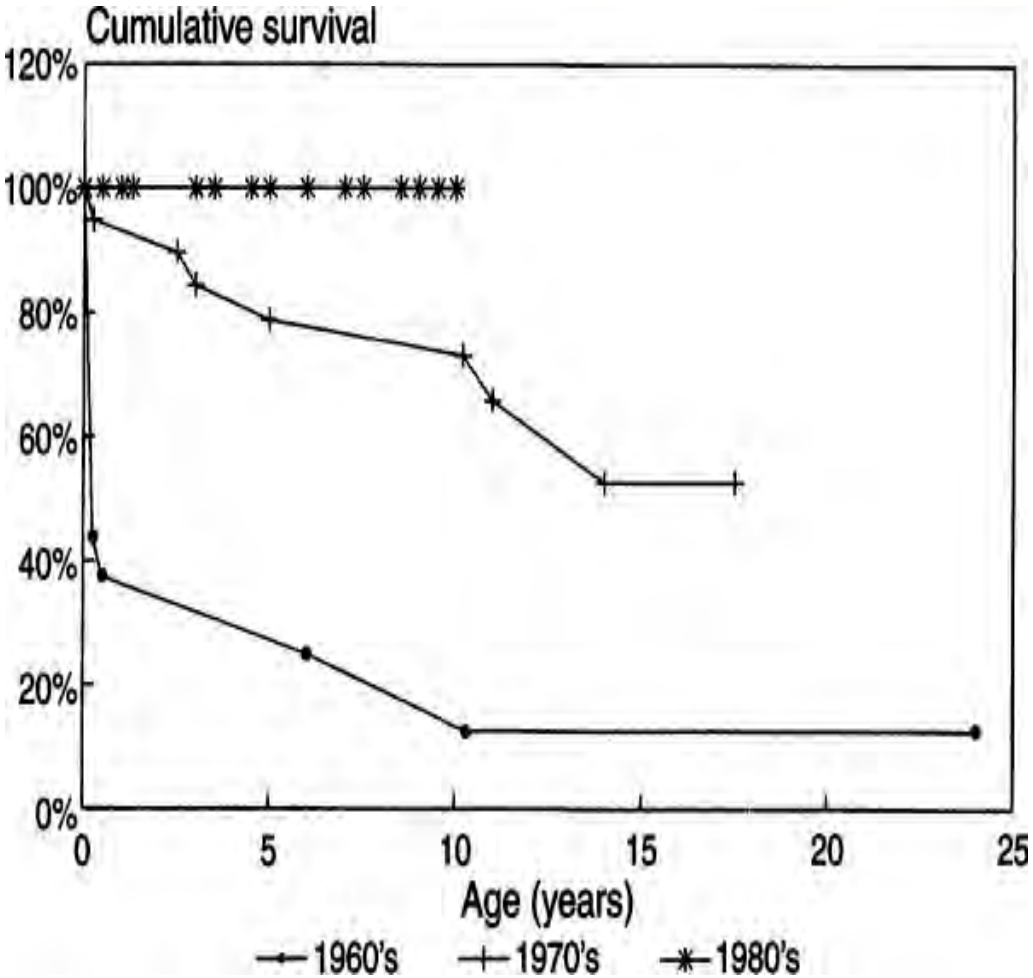


Fig. 69-2 Operative options for the surgical treatment of meconium ileus.

Postoperative Management

- Instillation of 2%-4% acetylcysteine delivered through a nasogastric tube to solubilize the residual meconium
- When gut patency verified – elemental formula
- Supplemental pancreatic enzymes also begun with formula
- Short term post operative antibiotics
- Total parenteral nutrition

Outcomes



- Survival rates approaching 100%
- No overall significant differences in outcome was present with regard to patient gender, complication of meconium ileus or type of operation performed

Diminishing role of contrast enema in simple meconium ileus

- Hypothesis: the role of contrast enema in simple meconium ileus is not as effective as previously reported (30-50% reduction)
- 37 cases of simple MI over a 12 year period (16M:21F)
  - 22/37 patients had CF (89%)
- 8/37 cases (22%) had successful relief of obstruction
  - 3/8 required one attempt only

Diminishing role of contrast enema in simple meconium ileus. Copeland DRS, J Pediatr Surg. 2009 Nov;44(11):2130-2.

Demographics and Results

Table 1 Demographics of simple MI patients

	Contemporary group	Historic group	*P*
Gestational age (wk)	36 ± 3.8	37 ± 3.8	.32
Birth weight (kg)	3 ± 0.9	2.9 ± 0.7	.83
Male (n)	10	6	.13
Age at diagnosis (d)	4.7 ± 6	2.6 ± 1.8	.16

No differences between groups in demographic variables

Significant decrease in successful resolution of obstruction by enema

Table 2 Results of contrast enema in simple MI patients

	Contemporary group (n = 18)	Historic group (n = 19)	*P*
Successful reduction	1 (5.5%)	7 (39%)	.05
No. of attempts	1.4 ± 0.7	1.9 ± 1	.12

Discussion

- Low perforation rate with a high failure rate of enema
- Enema substrate used in this study (cysto-conray II- 400 mOsm/kg water) less osmotically active than Gastrografin (1940 mOsm/kg water).
- Small sample size- need a prospective multicenter evaluation

References

- Diminishing role of contrast enema in simple **meconium ileus**.Copeland DR, St Peter SD, Sharp SW, Islam S, Cuenca A, Tolleson JS, Dassinger MS, Little DC, Jackson RJ, Kokoska ER, Smith SD. J Pediatr Surg. 2009 Nov;44(11):2130-2.
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- Dalla V, Grosfeld JL, West KWl. Intestinal atresia and stenosis: a 25 year experience with 277 cases. *Arch Surg*. 1998 May; 133 (5) 490-6.
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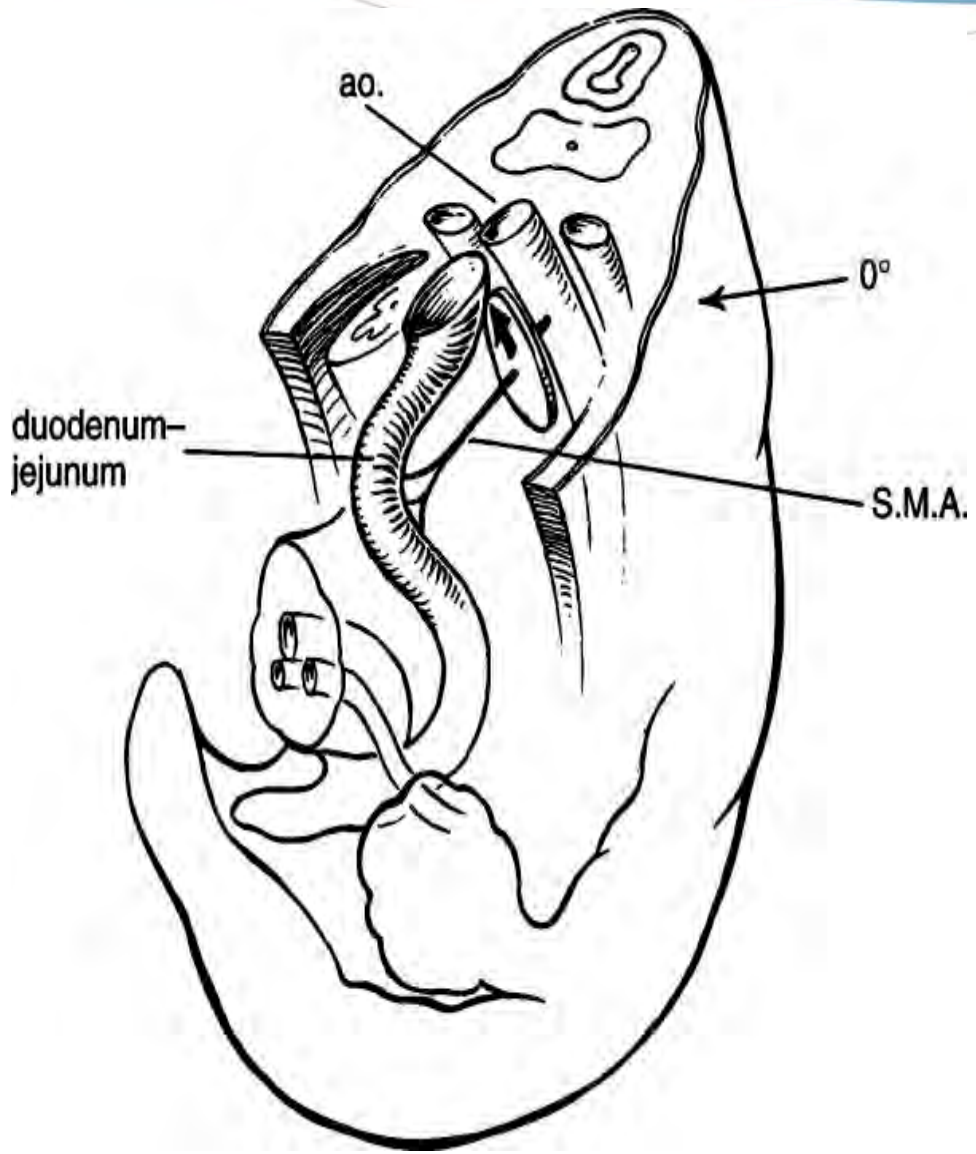
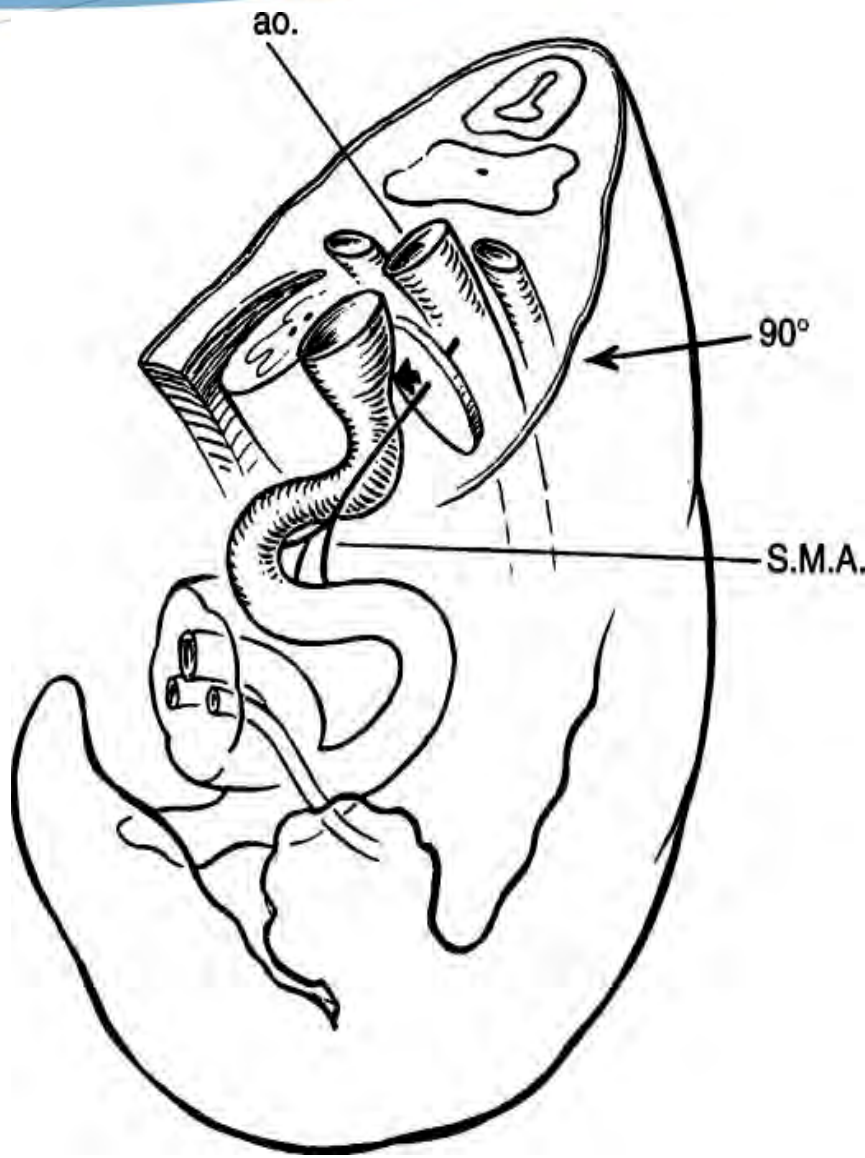
Normal Rotation and Fixation of Duodenojejunal loop





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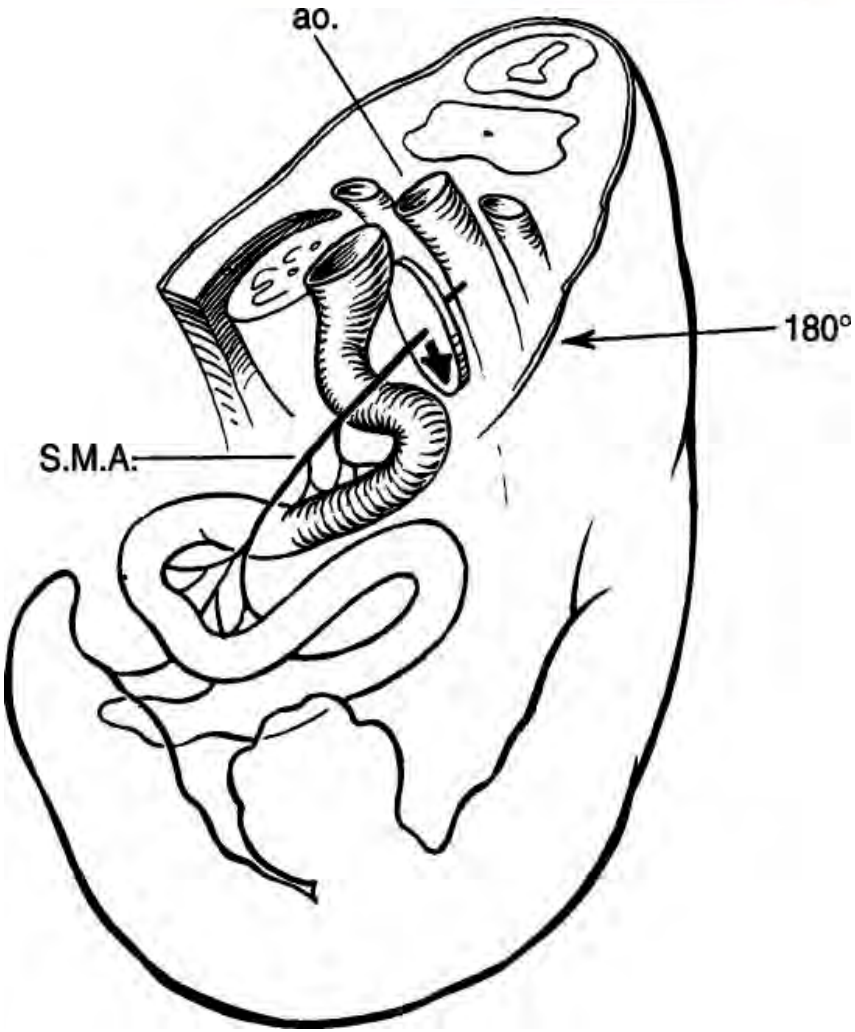
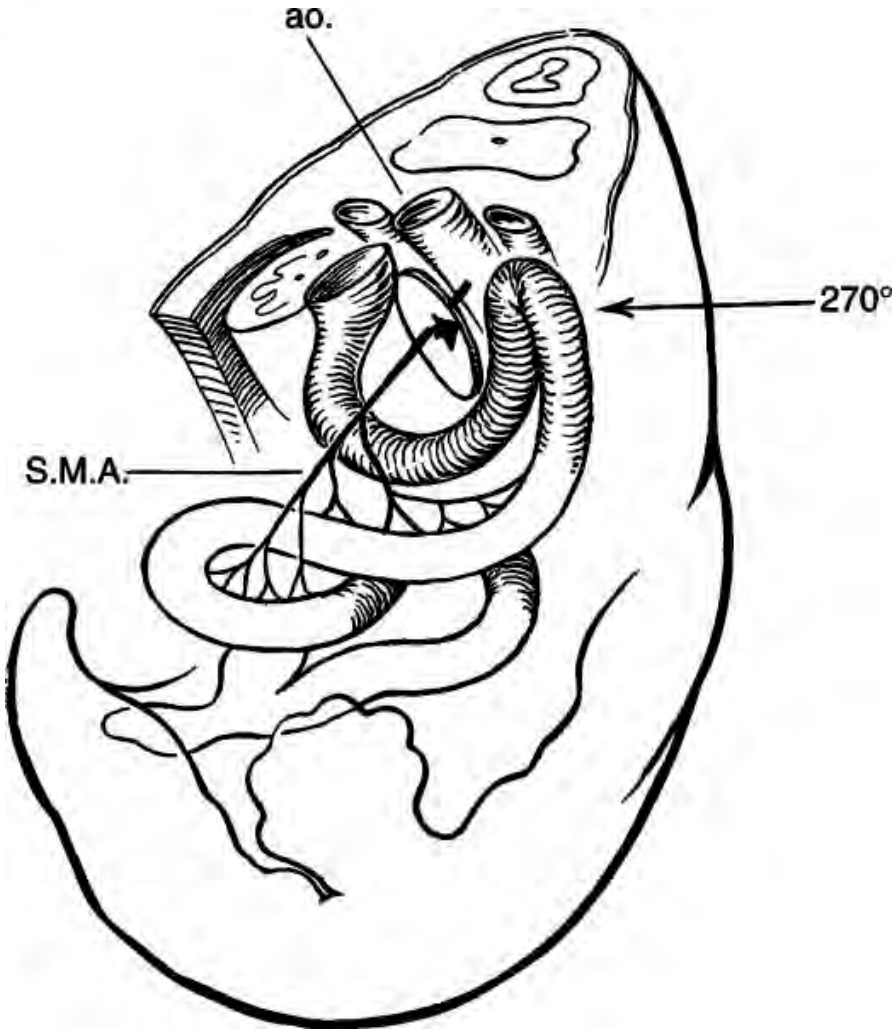
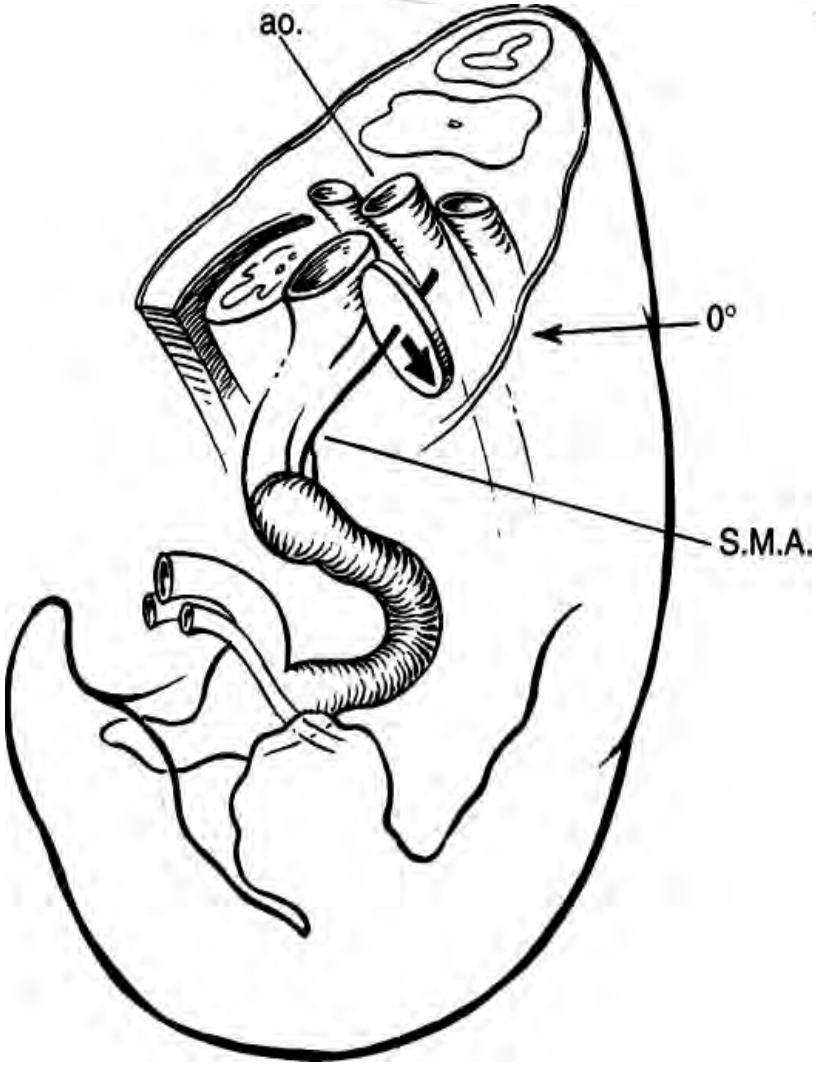
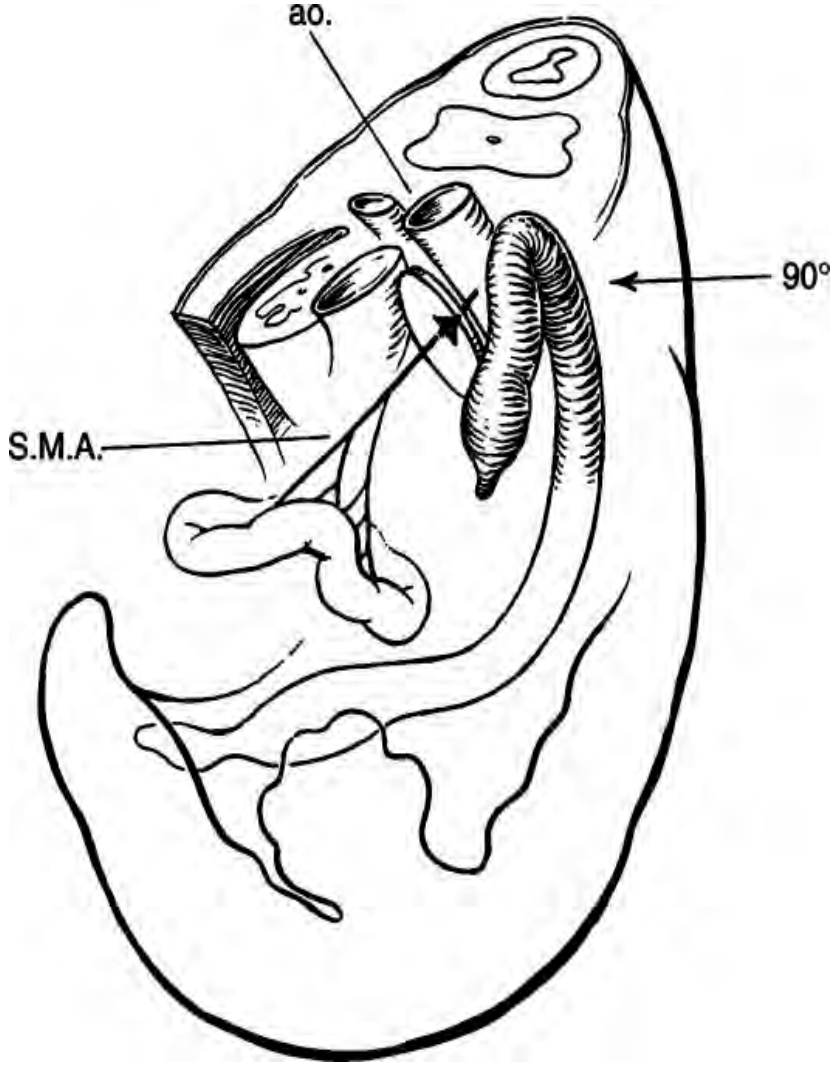


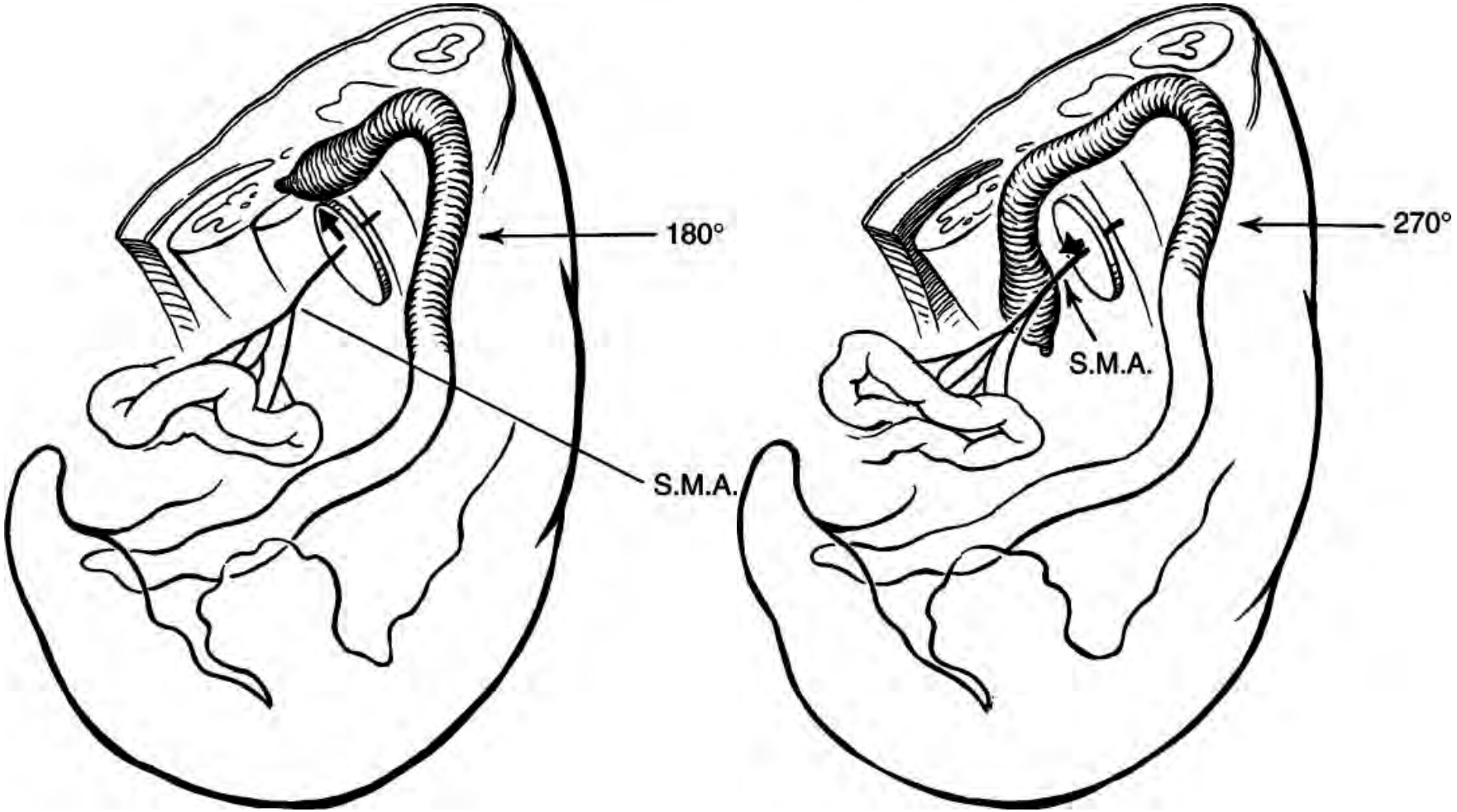


Normal Rotation and Fixation of Cecocolic loop

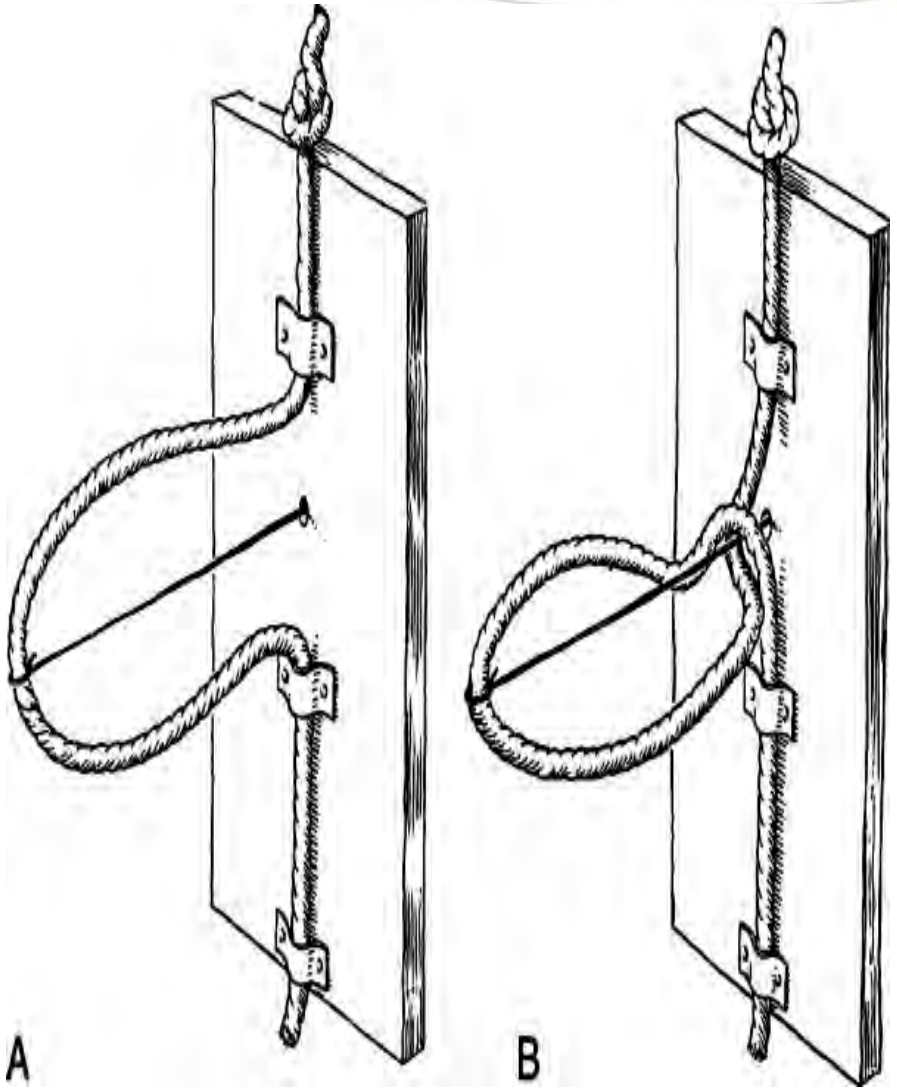




Normal Rotation and Fixation of Cecocolic loop



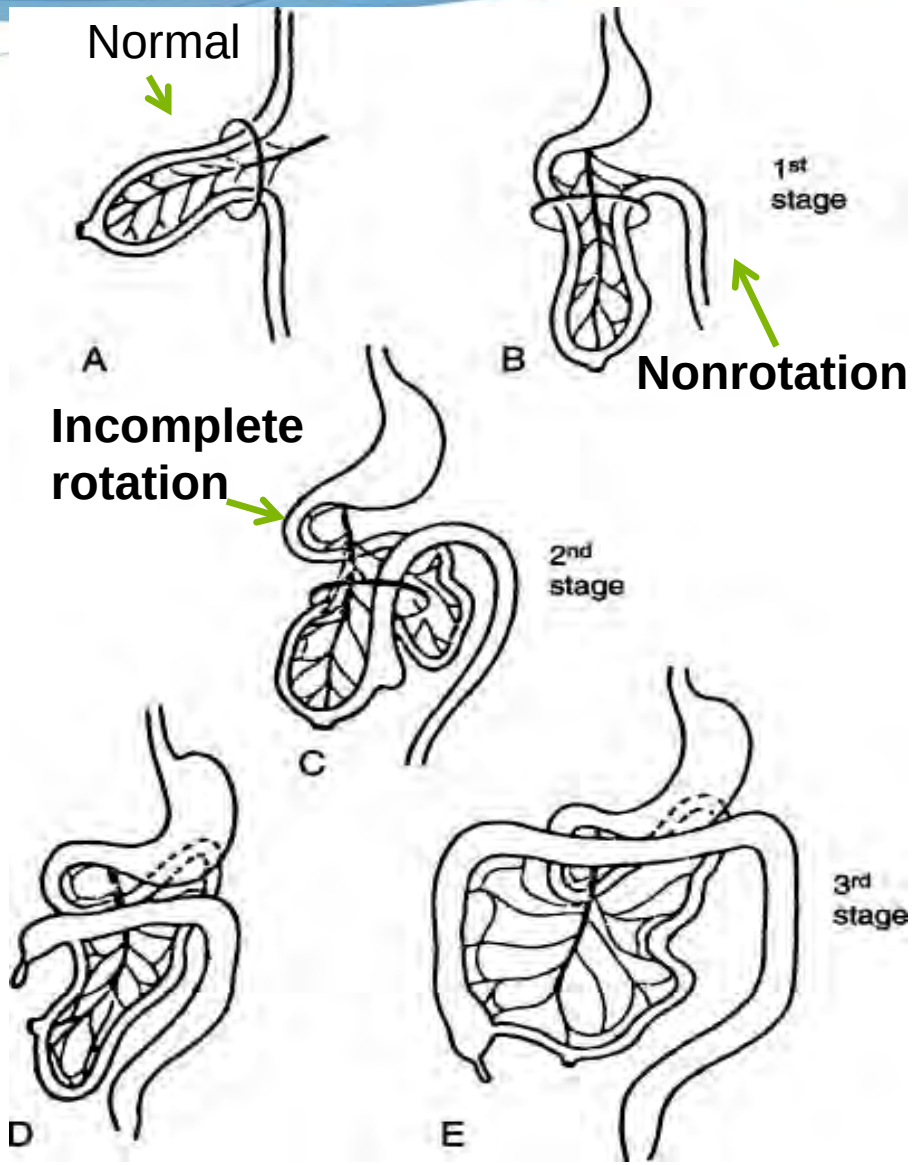
Intestinal Rotation



- 4th week of fetal life the embryo is at 5 mm stage
  - Intestinal tract is a straight tube with slight anterior bulge in central portion
- 8th week
  - Duodenojejunal rotates during extracoelomic phase to 180 degress
- 10th week
  - Intestines return to abdomen

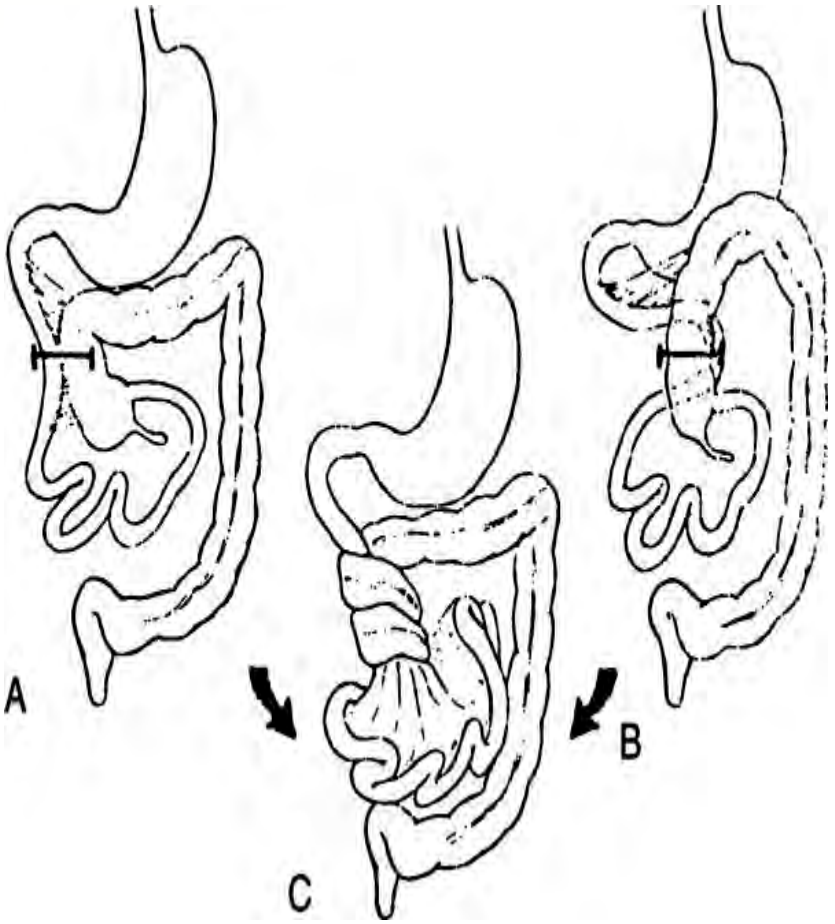
Malrotation

- All abnormalities of intestinal position and attachment
  - **Atypical malrotation**- ligament of Treitz is to the left of the midline
  - **Nonrotation**
  - **Incomplete rotation**



Acute Midgut Volvulus

- Narrow pedicle formed by the base of the mesentery in malrotation predisposes midgut to clockwise twisting from duodenum to transverse colon
- Various causes:
  - Unusual movement of torso
  - Abnormal intestinal peristalsis
  - Segmental bowel distention



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Midgut Volvulus

Acute Midgut Volvulus

- First month of life
- Bilious emesis
- As vascular compromise progresses → intraluminal bleeding
- Crampy abdominal pain
- Complete obstruction → distention, hypovolemia, shock

Chronic Midgut Volvulus

- Children > 2 years
- Lymphatic and venous obstruction with enlargement of mesenteric LN
- Sx:
  - chronic vomiting,
  - intermittent colicky abdominal pain
  - diarrhea
  - hematemesis

Duodenal Obstruction

- Ladd's bands extending across the 3rd portion of duodenum
- Most common in neonates
- Forceful bilious emesis
- "double bubble sign"
- **Upper GI contrast study**

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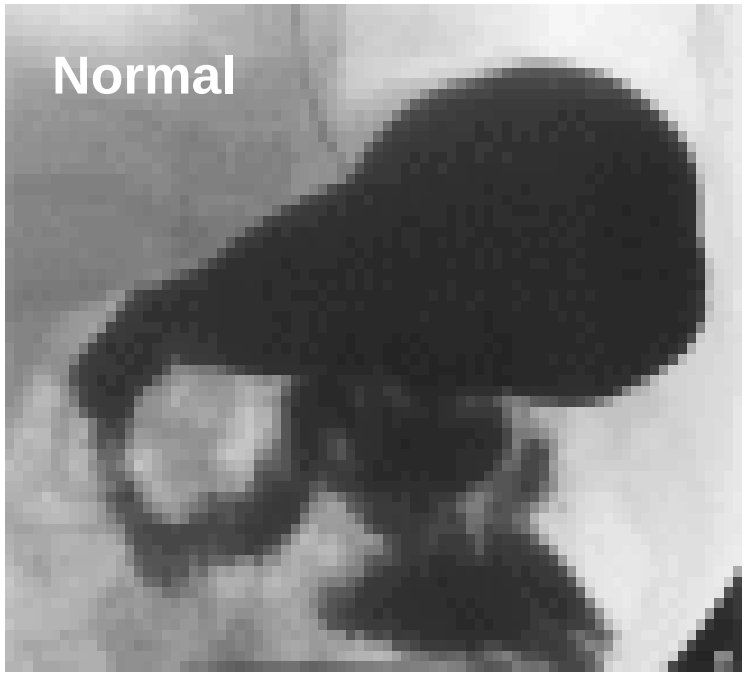
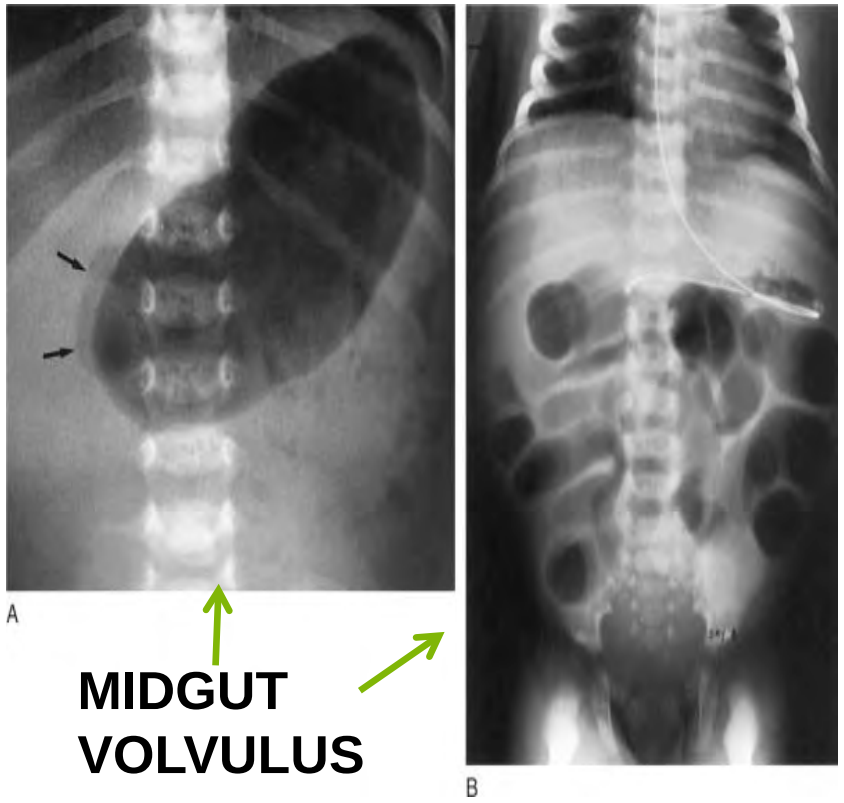
Radiologic Diagnosis

- Contrast radiography





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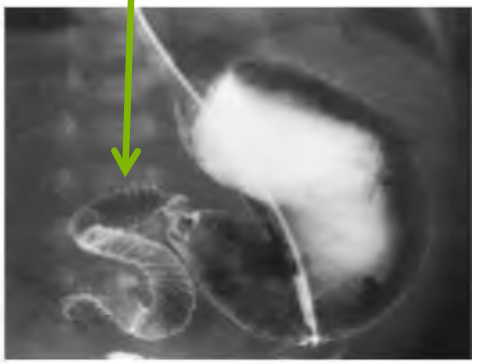
Contrast Radiography

Midgut Volvulus

A

Z sign



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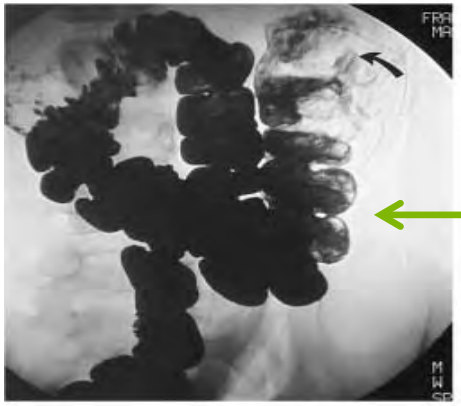

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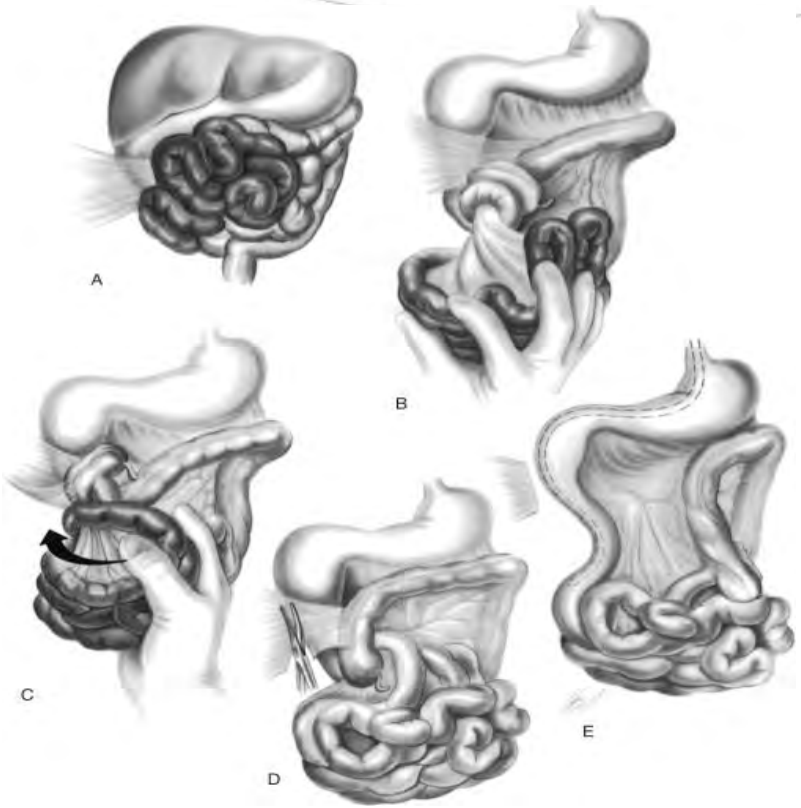
Colon Malposition

- “Z sign”- incomplete rotation and broad peritoneal bands extending across and fixing the involved small intestine without an accompanying volvulus.

Ladd Procedure

- (1) Evisceration of the bowel and inspection of mesenteric root
- (2) Counterclockwise derotation of midgut volvulus
- (3) Lysis of Ladd's peritoneal bands with straightening of the duodenum along the right abdominal gutter
- (4) Appendectomy
- (5) Placement of the cecum in the left lower quadrant

Ladd Procedure



- Supraumbilical right transverse incision
- 2 constant anatomic points: **Pylorus** and **splenic flexure**

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LADD Procedure

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Intestinal Resection and 2nd Look

- At 12-24 hours recovery of questionable bowel or demarcation is obvious
- 3 principles should be considered:
  - Preserving minimum length of intestine required for survival is of highest priority
  - Avoid anastamoses between end f intestine of ? viability
  - Resection of entire midgut will necessitate lifelong parenteral nutrition and small intestinal transplantation

Postoperative Management

- In uncomplicated duodenal obstruction- peristalsis in 1-5d
- Complications:
  - Postoperative intussusception (3.1%)
    - Abdominal distention and bilious emesis on POD # 5-8
  - Recurrent volvulus (0.5-1.25%)
  - Bowel obstruction (4%)
  - Death (associated with peritonitis)