Intestinal atresia

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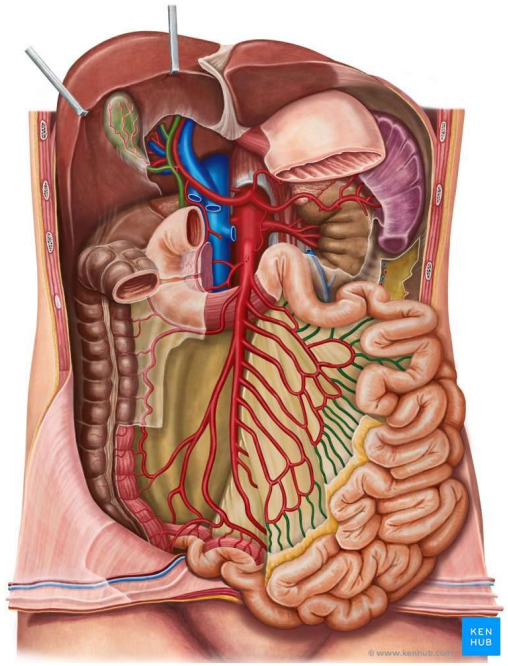
Outline

1. Epidemiology
2. Etiology
3. Classification
4. Presentation
5. Management
6. Outcome



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Introduction

- **Defect in continuity of the bowel** (closed or absent)
- **Named based on** part of intestine affected
  - Duodenal
  - Jejunoileal
  - colonic
- A Difficult problem in the management is the **poor intestinal motility**.
- Local literatures show **higher mortality.**

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Ethiopian Literature

NEONATAL GASTROINTESTINAL SURGICAL EMERGENCIES: A 5- YEAR REVIEW IN A TEACHING HOSPITAL ADDIS ABABA, ETHIOPIA

Endale Tefera,MD[1], Telahun Teka, MD[1], Milliard Derbew,MD[2]

- **12 jejunoilal atesias**
  - 30% post op death (2/6)
  - **60% death** overall (8/12)

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]

- **6 jejunoilal atesias**
  - **30% death** (2/6)… all operated
- **1 duodenal atresia, 1 colonic**
  - No mortality….all operated

*25% death overall

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Part 1-
Duodenal atresia

Differential diagnosis: duodenal obstruction

- **Duodenal atresia/stenosis**
- **Malrotation**
- **Annular pancreas** (associated atresia/stenosis)
- **Preduodenal portal vein** (associated malrotation or duodenal atresia)

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1. Epidemiology

- 1 per 5000–10,000 live births
- Duodenal atresia is 2x more common than duodena stenosis
- M>F
- Associated problems in 50%
  - **Prematurity** 50%
  - **Trisomy 21** in 30%
  - **Cardiac anomaly** in 30%
  - **Annular pancreas**: ~20%
  - **Malrotation** in 30%
  - **EA** 8%, **ARM** 4%

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2. Etiology

- **Pathophysiology** – early gestation (ass. With other anomalies)
- **Embryologic theories**
  - failure of recanalization (Tandler 1990)
  - * normal esophagus is solid at 6 wks, becomes hollow again around 9 wks (vacuolation)

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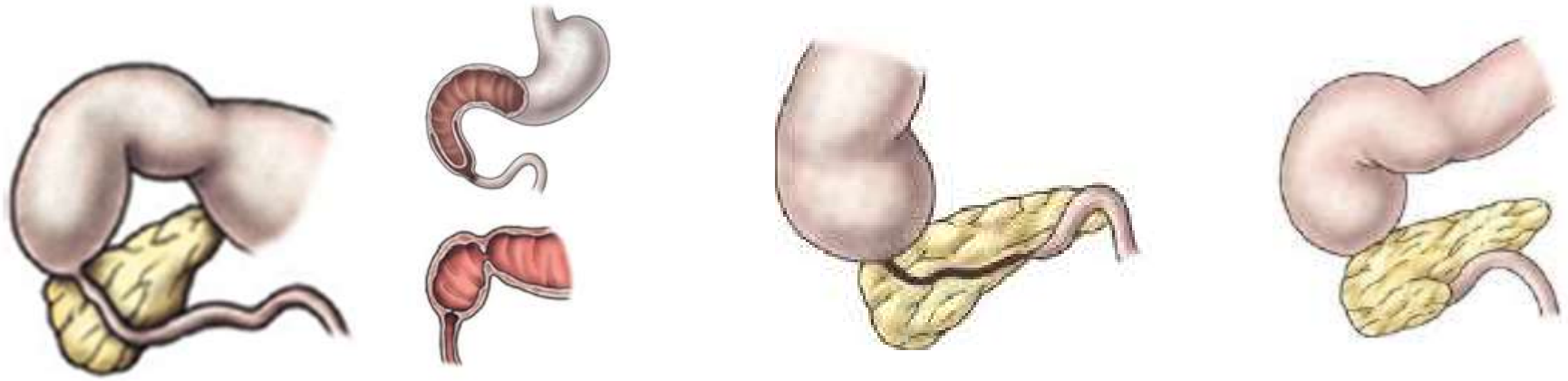
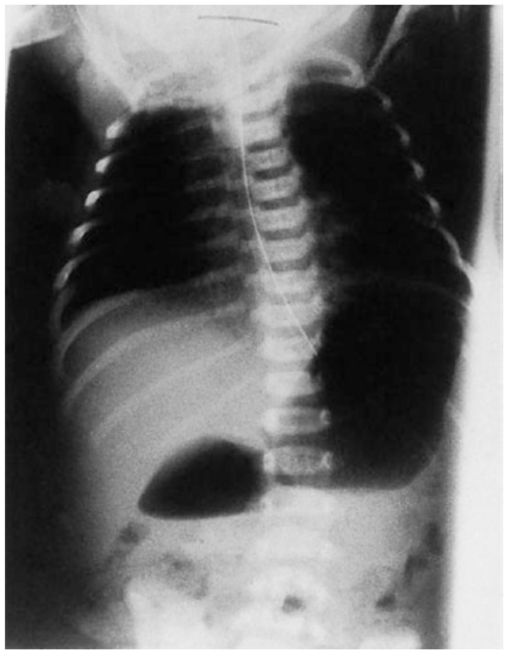
3. Classification

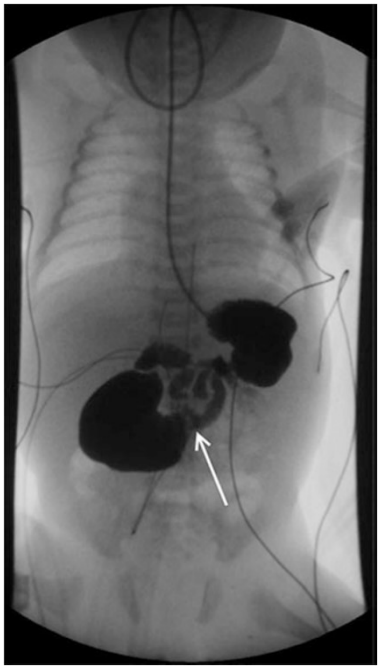


- **type I:** (92%) bowel continuity (variants include perforated diaphragm, “windosck web”, stenosis)
- **type II:** (1%) bowel discontinuity with a connecting bridge of tissue
- **type III:** (7%) complete separation with a mesenteric defect

4. Presentation

- **Prenatal ultrasound** – polyhydramnios, double bubble (detedted in late gestation)
- **Bilious vomiting on first day** of life (Obstruction is post-ampullary in 85%)
- **May pass meconium** the first day (can have bifid termination of CBD above and below atresia)
- **NG tube aspirate >20 ml** suggests obstruction (normal <5ml)
- **Delayed diagnosis** leads to **Dehydration** with weight loss and **electrolyte imbalance** (hypokalemic/hypochloremic metabolic alkalosis)
- Presentation of **Incomplete** duodenal obstruction may be **delayed months-years**





- **Double bubble** sign is seen on x-ray. If decompressed by NGT/vomiting 40-60ml air instilled will reproduce double bubble. There may be some **gas in distal intestine if** there is CBD bifurcation or obstruction is partial. Contrast study in perforated diaphragm shows dilated duodeunum terminating abruptly.

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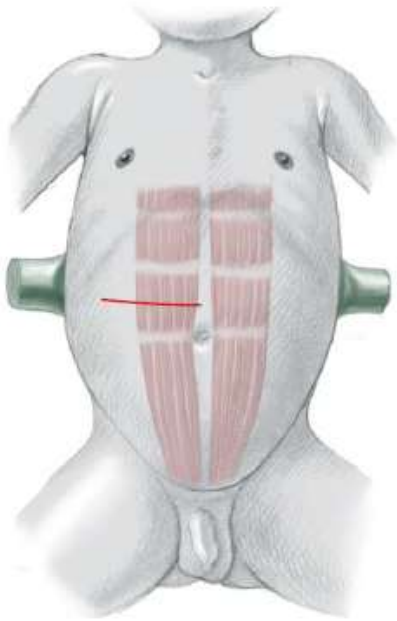
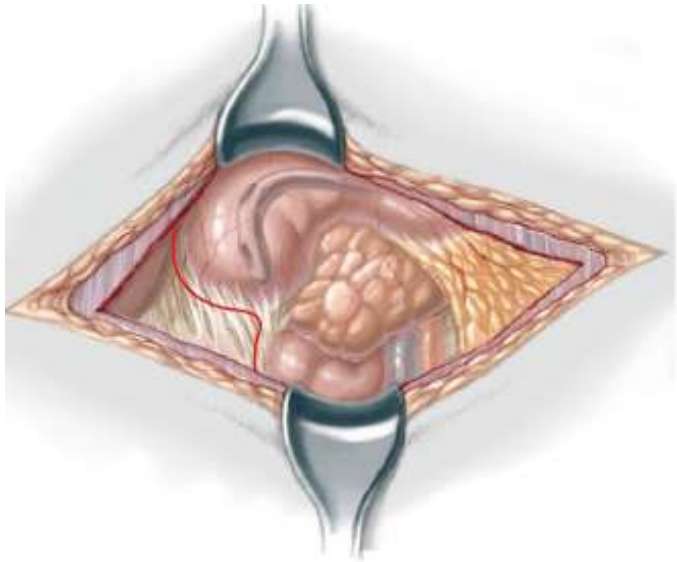
5. Surgery

- **Duodenoduodenostomy**
  - Diamond shaped (proximal transverse to distal longitudinal)
  - Side to side
  - partial web resection with Heineke–Mikulicz-type duodenoplasty

**<u>laparoscopy</u> is an option in b/c bowel is decompressed unlike other causes of NBO*

- **transanastomotic feeding** tube is started within 24–48 h postoperatively.
- **Oral feeding** delayed several days depending on gastric aspirates (occasionally 1-2wk)





- **Duodenoduodenostomy exposure:** RUQ transverse or umbilical incision. Evaluate for **malrotation**. Liver retracted superiorly. The ascending colon and the hepatic flexure of the **colon are mobilized** medially and downwards to expose duodenum. Duodenum mobilized from retroperitoneum (**kocherized**). Avoid dissecting medially (CBD). Distal duodenum mobilized behind SMV as needed to allow a tension-free anastomosis. If necessary, the ligament of Treitz is divided.

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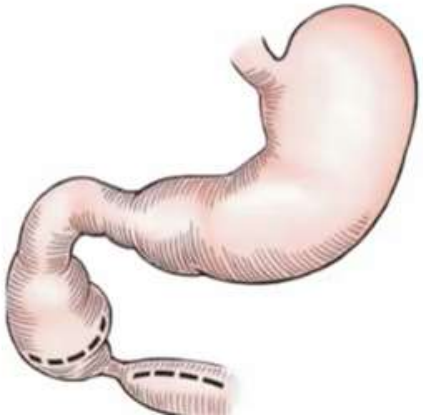

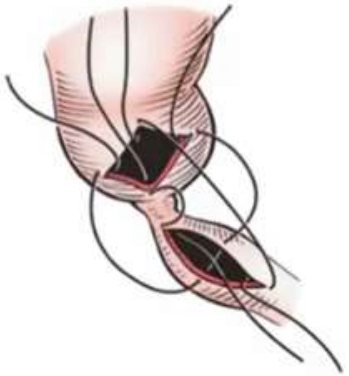

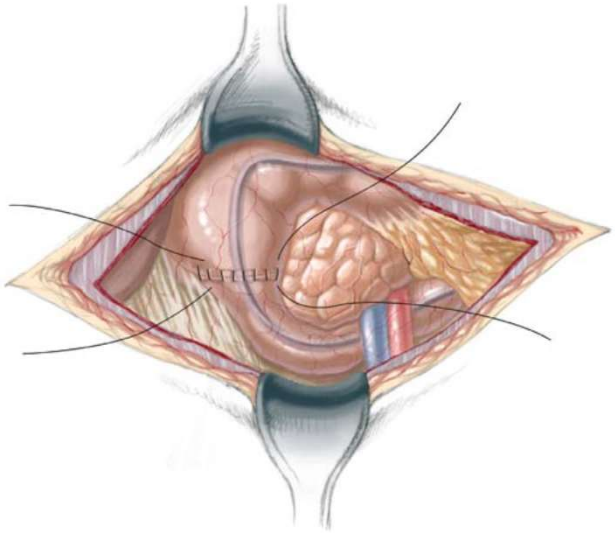

- **Dimond-shaped anastomosis as described by Kimura is preferred:** Transverse incision on proximal segment with longitudinal incision on distal atretic. The papilla of Vater is located by observing bile flow. This is performed by gentle compression of the gallbladder Careful not to miss membrane (windsock) in type 1. Tapering duodenoplasty usually not needed because dilatation resolves after obstruction is relived. Transanastomotic tube may be passed.

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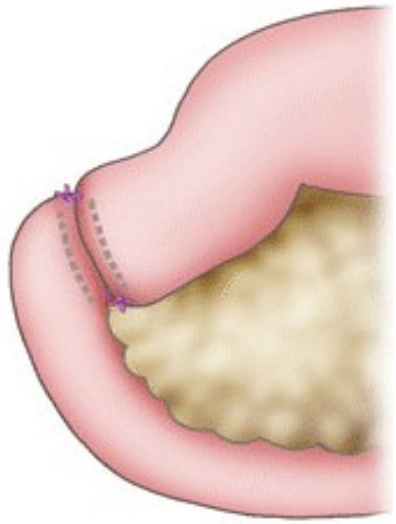
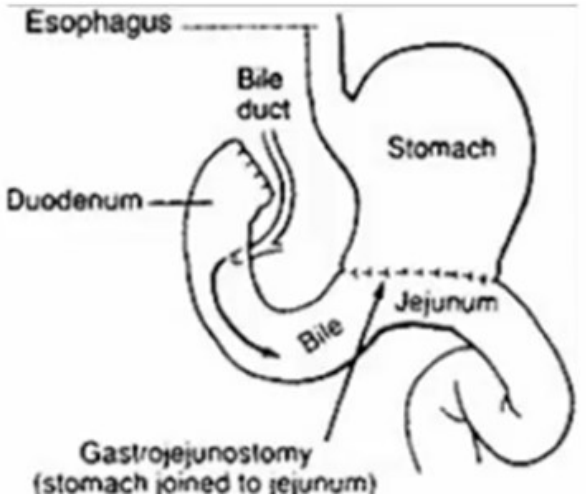
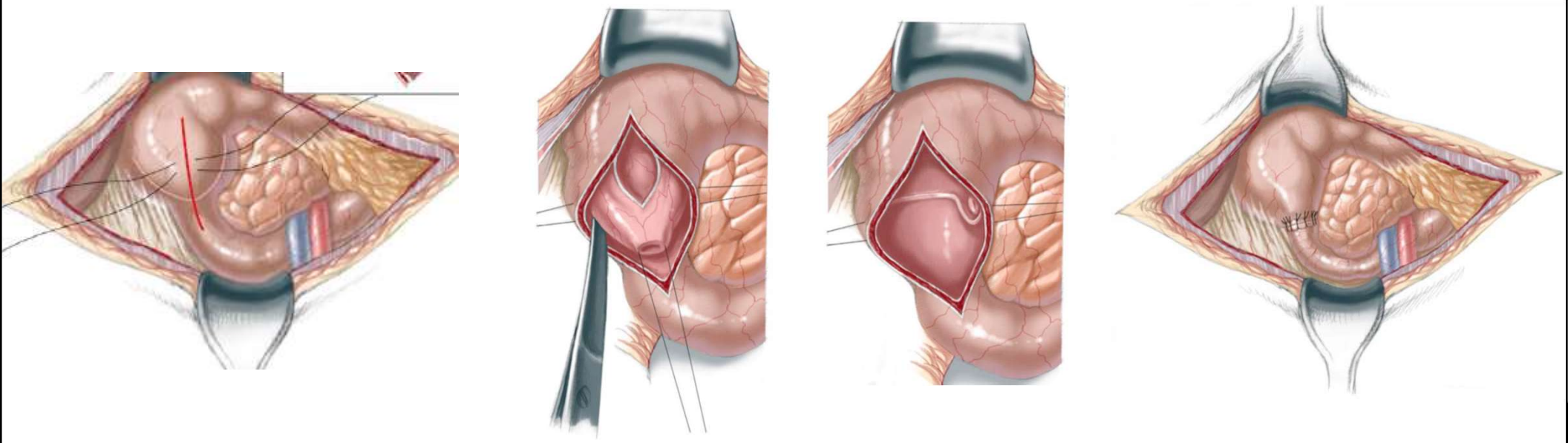




- **long side-to-side anastomosis** was the original duodenoduodenostomy, and although effective, is associated with a high incidence of anastomotic dysfunction and prolonged obstruction. **Gastrojejunostomy** is not standard but may be performed by some surgeons. Complications include Blind loop syndrome, ulceration and bleeding which usually needs revision of anastomosis or conversion to duoenouduodenostomy



- **Partial web resection with Heineke–Mikulicz-type duodenoplasty:** Longitudinal incision made over transition. Traction applied at the apex of the web deforms the duodenum at its point of attachment and allows excision at the base. The web is opened along the lateral side of the membrane and excision from the duodenal wall takes place, leaving a rim of tissue of 2–3 mm. The medial portion of the membrane should remain intact, to avoid damage to the ampulla of Vater.

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6. Outcome

- > 95% long term survival
- Long term complications (duodeno-duodenostomy)
  - **Megaduodenum,** dysmotility
    - poor wt gain, vomiting, pain
    - tapering/plication duodenoplasty ± revision of anastomosis
  - Delayed gastric emptying, GERD,
  - Duodenogastric reflux, PUD
  - Adhesions

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Part 2 - Jejunoileal atresia

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Differential diagnosis for Ileal atresia

- **meconium ileus**
- **Hirschsprung disease (total colonic)**
- **colonic atresia**
- **midgut volvulus, internal hernias**
- **duplication cysts**
- **Medical** - ileus due to sepsis, maternal medications, prematurity, and hypothyroidism

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1. Epidemiology

- **1:100 – 1: 10,000** (higher incidence in Africa than Europe/USA)
- **Most common atresia**, 1/3 of NBO
- **M=F**
- Associated anomalies are rare
  - **Prematurity** (3x more common in preterm, 30% are preterm)
  - Hirschsprung disease
  - cystic fibrosis
  - malrotation
  - Down syndrome
  - other GI atresias
  - ARM, NTD, CHD

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2. Etiology

- **Risk factors**
  - Maternal smoking, vasoconstrictive meds
  - Chromosomal anomaly <1%
- **Pathophysiology** = late gestation
  - **isolated anomaly** (extra-abdominal anomaly is <10%)
  - finding of meconium (bile) distal to the atresia indicate patency at some point (>12wk)
  - ***Autosomal recessive pattern** (hereditary, rare, no gene identified) = Multiple atresias, No rotation abnormality or mesenteric defects, No lanugo or squamus cells distally

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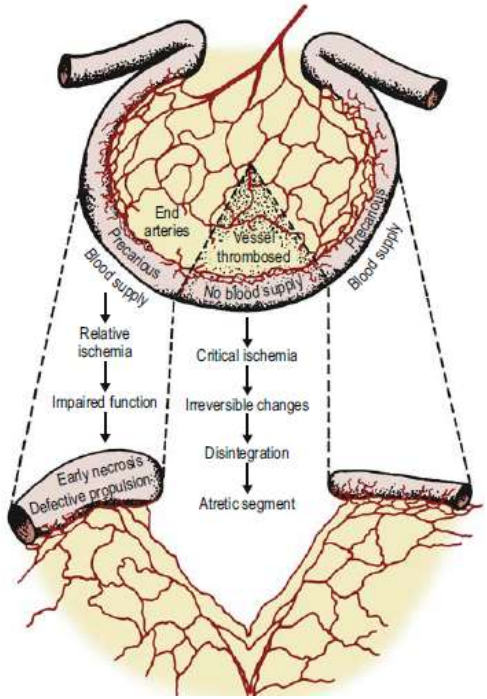

- **Theory of vascular disturbance** (Lauw/Barnard 1955): focal resorption of the sterile gut occurs after ischemic necrosis. Supported by Association with other intrauterine vascular insults (intussusception, volvulus, incarceration in an omphalocele or gastroschisis). Theory also proven in fetal dogs.

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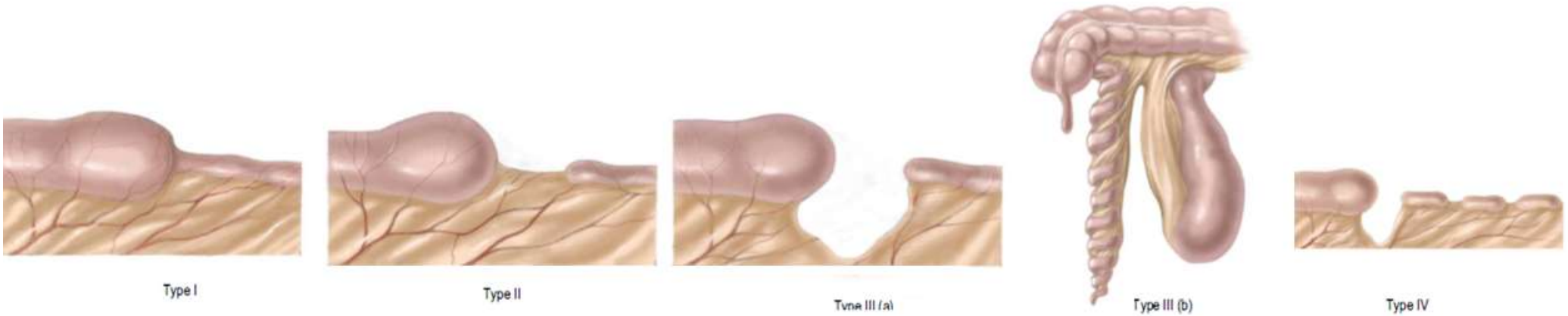
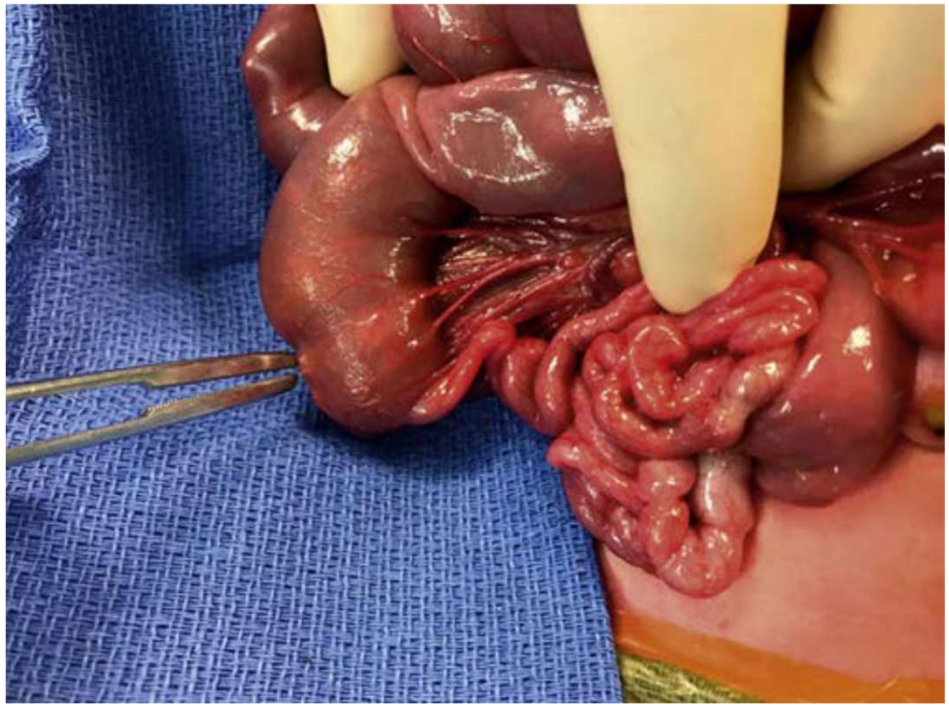
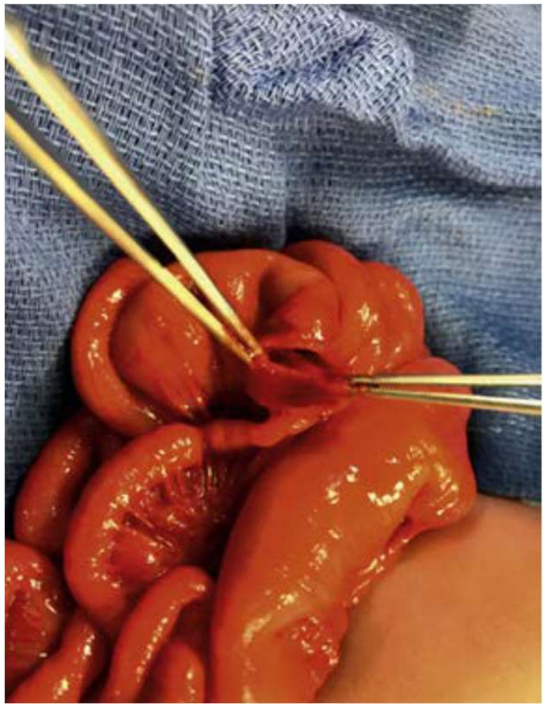
3. Classification



- **distal ileum and proximal jejunum** are the most frequent sites. **30% are type 4** (Multiple atresias) appearing as string of sausages. Named jejunal/ileal based on the most proximal atresia.





- **Type 1** (No gap) membrane/web formed from mucosa and submucosa.

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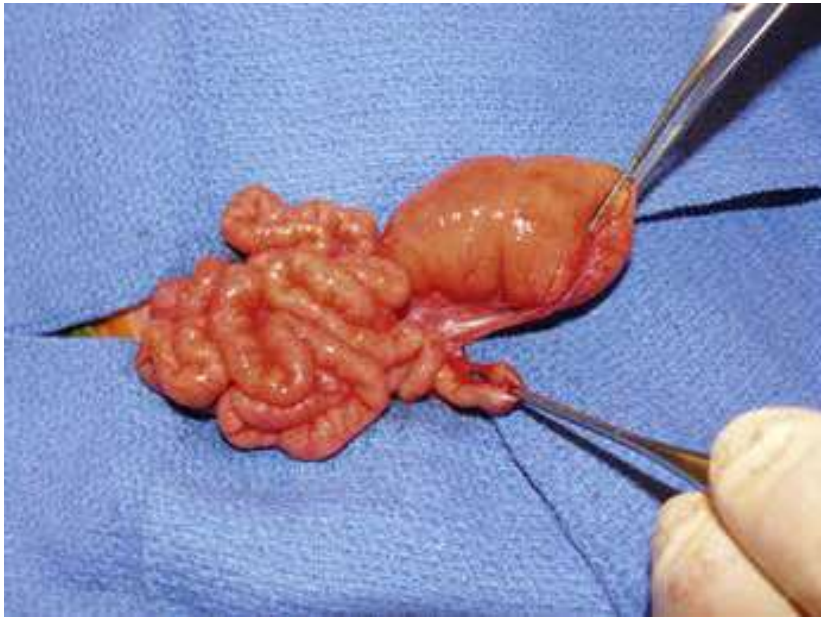

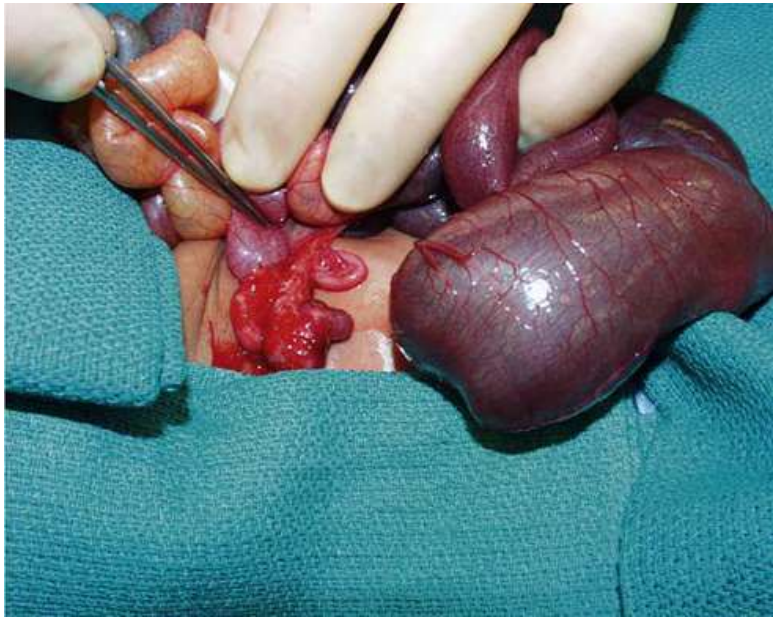

- **Type III A (**V Shaped gap in mesentry). **Bowel length is less than normal**. The dilated, blind-ending proximal bowel is often **aperistaltic** and may appear cyanotic. Pressure induced dilation may cause **ischemia** and frequently undergoes torsion (**vovulus**)

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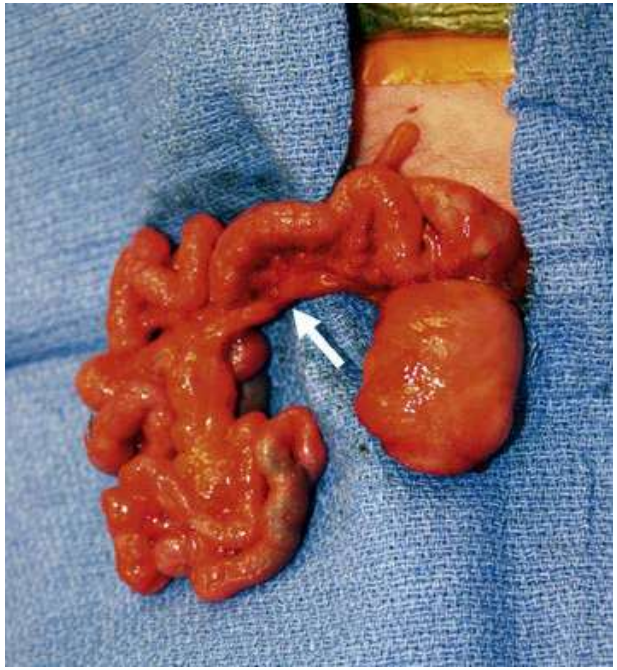

- **Type III b** (apple-peel, Christmas tree, or Maypole deformity) proximal jejunal atresia, **absence of distal SMA** beyond the origin of the middle colic, agenesis of the dorsal mesentery, **significant loss of bowel length**, and a **large mesenteric defect**. Distal bowel assumes a **helical configuration** around a single perfusing vessel with **retrograde perfusion** via right/middle colic arcades. More likely to present with **volvulus** and are at risk for impaired vascularity of the distal bowel. **Genetic transmition** in ~20%.

4. Presentation

- **Prenatal** - Polyhydraminios and dilated bowel (in more proximal atresias)
- **bilious emesis** and **abdominal distention**.
- **meconium** may appear normal, or as gray plugs of mucus passed via the rectum.
- 10% present with **meconium peritonitis**
- **Delayed diagnosis** leads to ischemia (50%), perforation (10-20%), sepsis, fluid & electrolyte abn
- **Stenosis** is less common (5-10%) and present later

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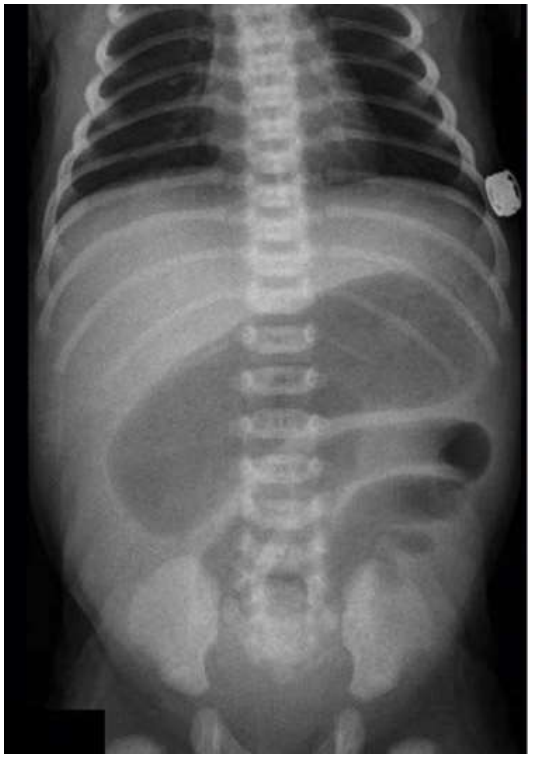

- **X-ray** usually aids in diagnosis using swallowed air as contrast. Air reaches proximal bowel in 1 hr and distal bowel in 3 hrs unless premature infants with poor sucking. In proximal atresias only few gas/fluid-filled loops seen with remainder of the abdomen being gasless

5. Surgery

- **Bowel length assessment** should be performed and documented.
- **injection of saline** into the large bowel to confirm distal bowel patency
- **Resection** of proximal (10cm) and small portion of distal atretic bowel (2cm) if adequate length remain
- **Bowel-length conservation** (limit resection if <75 cm with ileocecal valve remain)
    - **Multiple anastomosis** in multiple atreisas
    - **tapering/Imbrication** of proximal bowel
- Primary, one layer, End to back **anastomosis** (5-0/6-0, distal bowel is **obliquely resected** 4-5 cm & incised)
    - *May need to dilate distal bowel by clamping 6-8cm below and injecting saline
    - ***Stoma** may be indicated in cases of peritonitis, volvulus, meconium ileus, or type III(b) atresia

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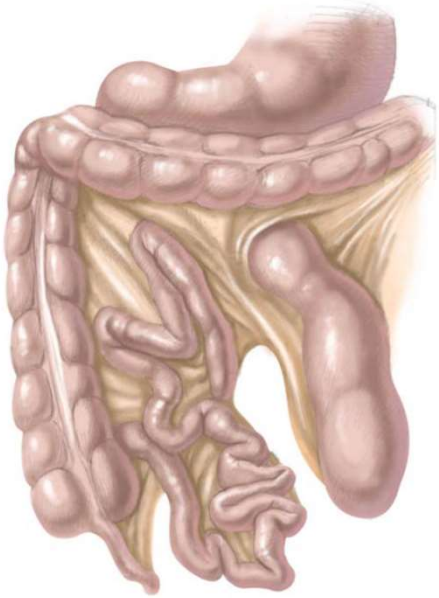

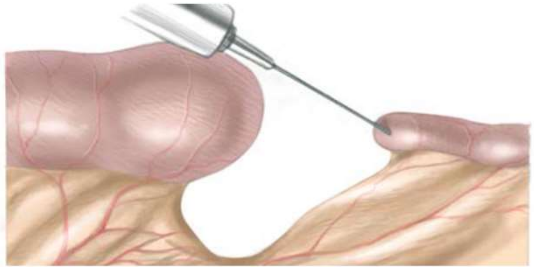

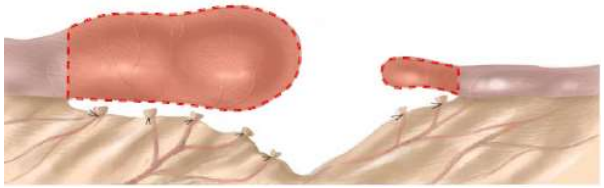

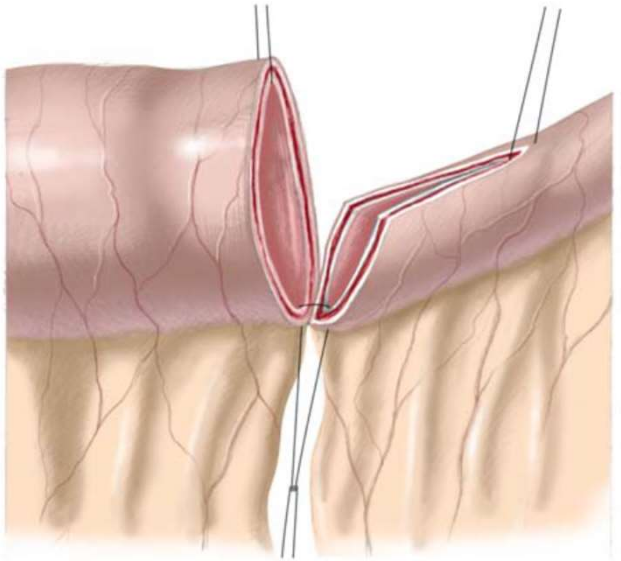

- **<u>Surgery for jejuno-ileal atresia</u>**: entails identifying anatomy, confirming distal patency, resection and end to back anastomsois. **Additional Considerations in type III B include** dividing restrictive fibrotic bands along the free edge of distal mesentry prior to anastomosis, Retaining mesentery from resected bowel (assist in closure of mesenteric defect)and Inspecting mesentery on returning bowel (prevent torsing/kinking the single marginal artery)

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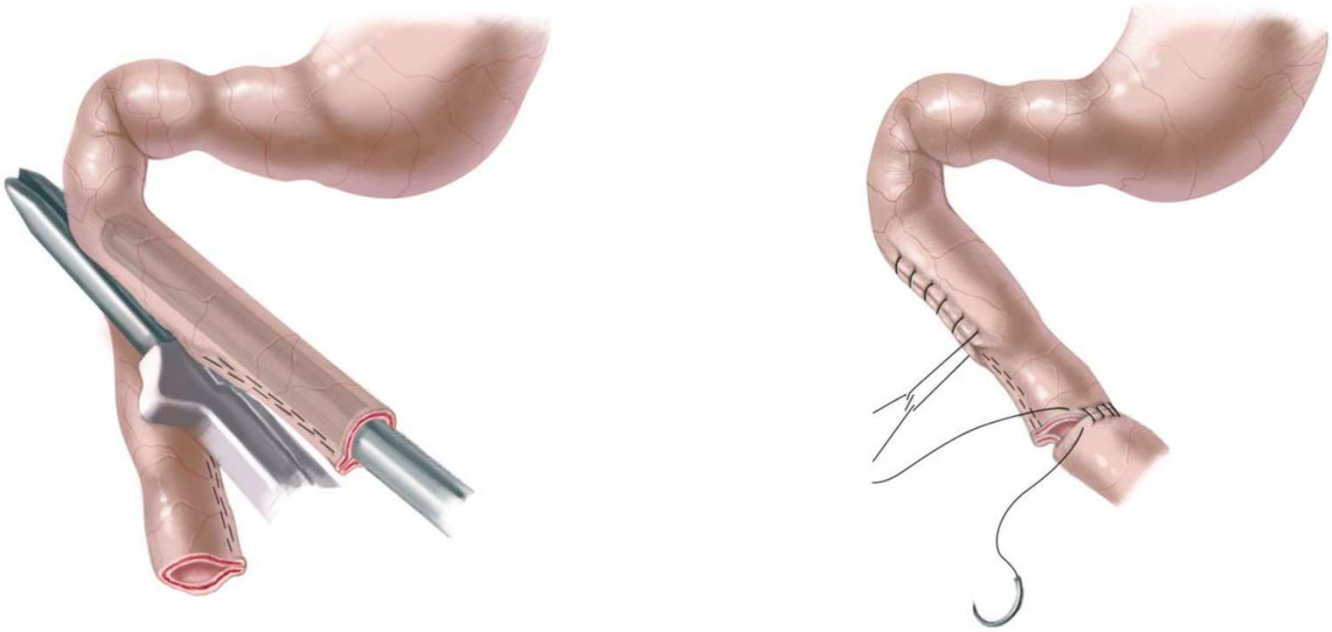

- **<u>Tapering enteroplasty</u>**: up to 35 cm is safe and Can go as far proximal as the second portion of the duodenum. The duodenum is de-rotated, allowing a direct caudal descent from stomach and cecum can be mobilized to the left, (broader mesentery and avoid kinking of anastomosis).

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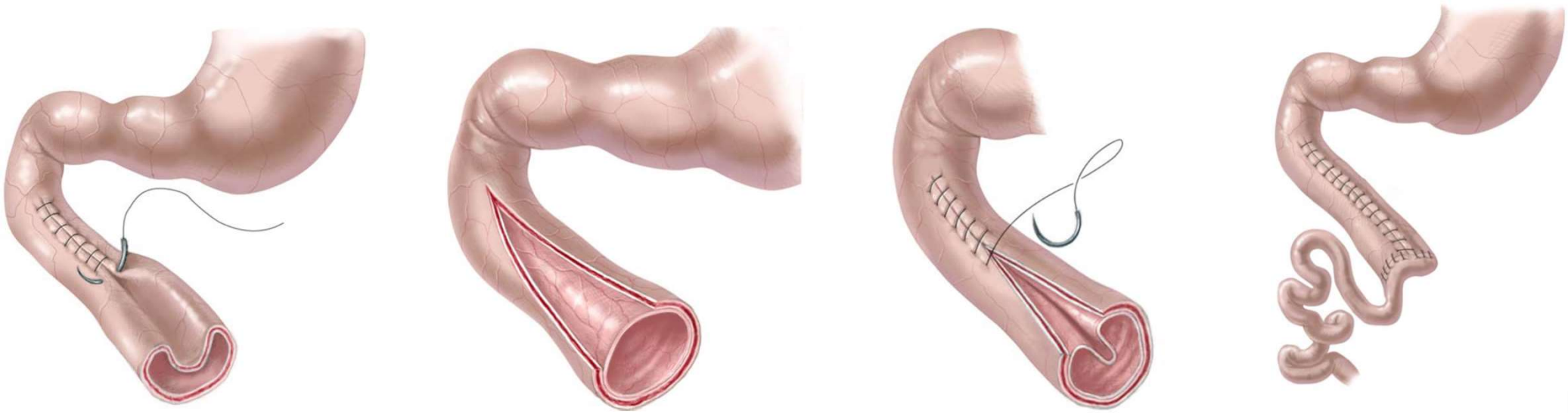

- **Antimesenteric Seromuscular Stripping and Inversion Plication:** is a technique prevents unravelling of the plication method and preserves maximal mucosal surface for absorption

5. Surgery

- **Pre-operative management**
  - Gastric decompression
  - Fluid resuscitation
  - Antibiotics
- **Postop care**
  - **Parenteral nutrition** given initially
  - Start feeding through **feeding tube** when **clear gastric output** and **stooling**. Start at a rate of **20 mL/kg/day breast milk continuous** fashion. Escalate 20-30ml/kg/d
  - **Oral feeding** when tolerate **8 ml/ hr**
  - **Monitoring intestinal overload** (increasing frequency of stooling, hematochezia)

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6. Outcome: Complications

- Transient bowel dysfunction
  - Underlying dysmotility, malabsorption (stasis and bacteria overgrowth due to aperistalisis)
  - Mucosal injury (high osmolality feeds, oral meds or infections)
- Persistent bowel dysfunction or obstruction
  - radiographic evaluation (adhesion, stenosis, dilated bowel)
  - **revision of anastomoses**, or **unkinking of bowel** or **division of adhesions**.
- Leak
  - ileus, abdominal distension, vomiting, and peritonitis
  - Pneumoperitoneum 24 hrs after surgery is indicative
  - **Immediate re-lap** with **suturing leak site** or **re-anastomosis**

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6. Outcome

- **Historically high mortality** (80-90%), currently <10% (highest mortality in type IIIB)
  - Resection of dilated proximal improved survival >80% even without TPN
- **Prognosis depends on remaining bowel length**
  - <25cm likely develop SBS with TPN
  - ?100 cm required for survival without TPN

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Part 3 -
Colonic atresia

1. Epidemiology

- Rare (1 in 20,000)
- 2-15% of atresias
- Associated anomalies
  - **abdominal wall defects** (gastroschisis, cloacal exstrophy, and more rarely omphalocele.)
  - **Hirschsprung disease** might be associated (good to send biopsy)

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2. Etiology

- **Similar to jejuno-ileal atresia**

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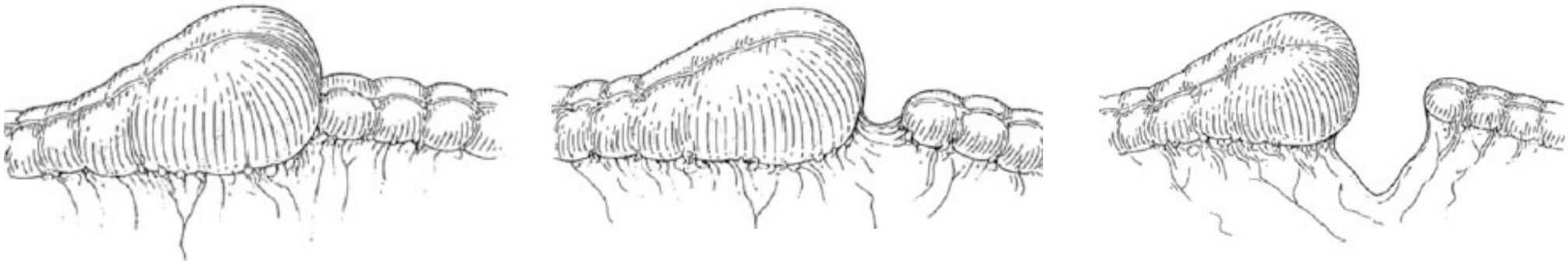
3. Classification



- **Classification** (Type 1-3): Type III atresia appears to be the most common type proximal to the splenic flexure, whereas type I and II are more common in atresias distal to the splenic flexure. Type III is the most common overall

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4. Presentation

- **Failure to pass meconium'**
- **Abdominal distention**
- **Bilious vomiting later**
- **Bowel perforation in 10%** (creation of a closed loop obstruction predisposes to higher chance of perforation than other atresias)

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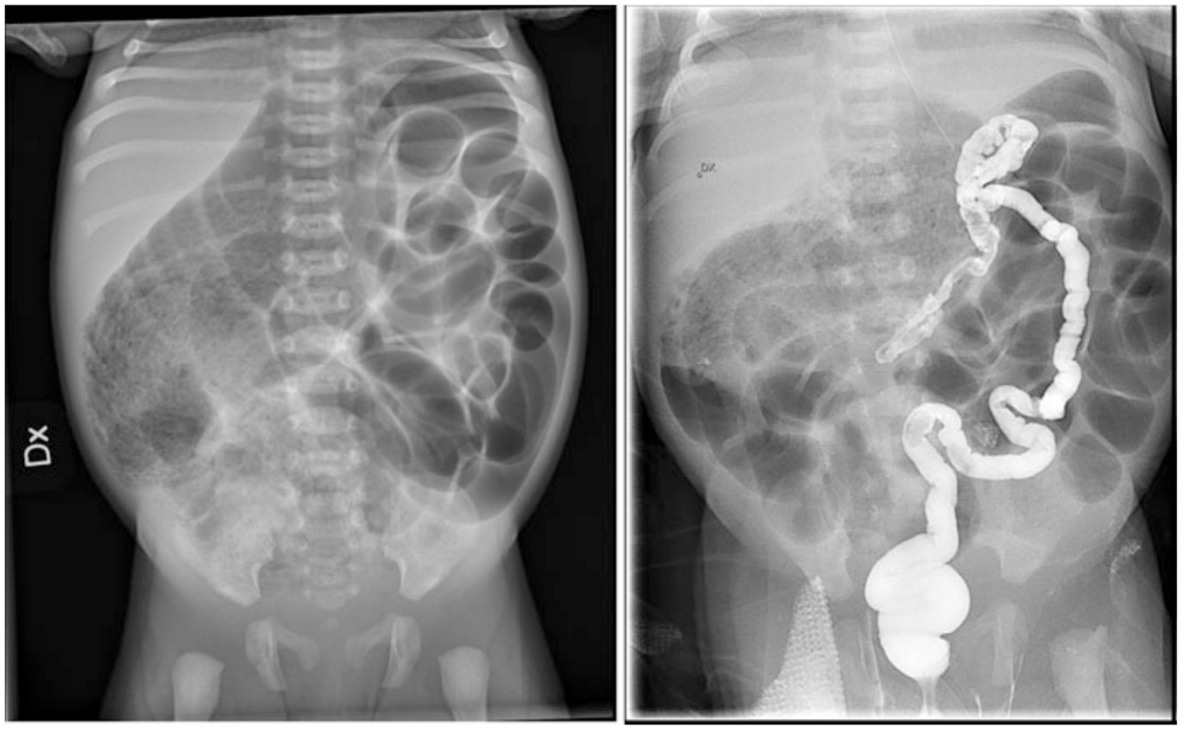

- **Large right-sided loop**, corresponding to the proximal dilated colon. Occasionally, the dilation can be so massive that it mimics pneumoperitoneum. Small colon on contrast and **failure of the contrast to move proximally** past the atresia.

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5. Surgery

- **staged repair** (colostomy)
    - complex cases (questionable bowel viability, colonic perforation, and abdominal wall defects)
- **Resection and primary anastomosis**
    - Uncomplicated atresias at all levels of the colon
- **? Transanal approach (after colostomy)**
    - Recently described for Sigmoid atresias

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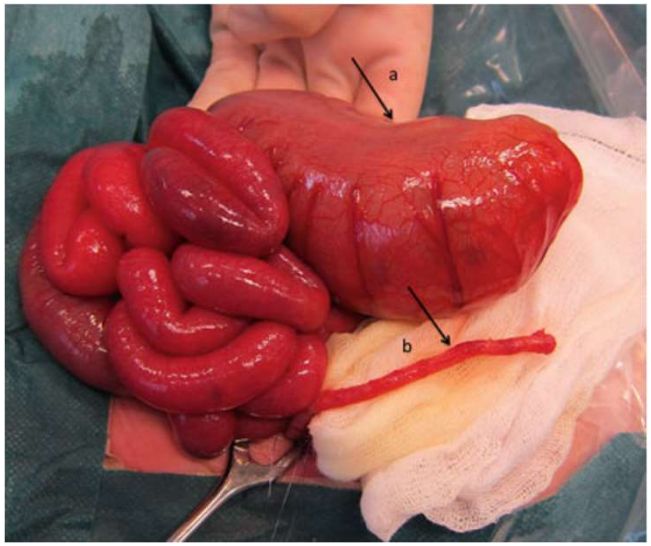

- anastomosis may be technically difficult because of the large discrepancy. resection of the bulbous proximal colon as well as a portion of the distal microcolon is suggested.

6. Outcome

- **mortality** related to the colonic atresia or its treatment is **rare**.
- However **delay in diagnosis** >72 hr may have high mortality (>60%)

References

- **Hollcomb and Aschraft Pediatric Surgery** 7th edition, 2020
- **Pediatric Surgery: General principles and newborn surgery**, 2020