Edited by: Tasnim Ahmed

61 page

NEONATAL INTESTINAL OBSTRUCTION

ABEER ALDIAB, MBBS ,JBPS,EBPS,FRCS ENG (PAED SURG)

- Age : neonatal , infantile , children
- Organ : duodenal , small bowel , colon
- Cause :ischemic , inflammatory , mechanical (adhesions ,congenital bands)

Topics

1. **Intestinal atresia**
2. **Malrotation**
3. **Meconium ileus**
   - Meconium plug
4. **Hirschsprung disease**
5. **Anorectal malformations**
6. **Necrotizing enterocolitis**
   - Inguinal hernia

1- INTESTINAL ATRESIA

- Abnormal formation of the bowel resulting in discontinuation of the bowel length … complete or partial obstruction
- Can occur any where

Duodenal atresia

happens embryonically due to failure of canalization

- 1 per 5000– 10,000 live births
- Affecting boys more commonly than girls. M > F
- Associated anomalies in 45–65% of cases. 2/3
- Most commonly trisomy 21 (half of the cases) , cardiac malformations (25–30%) ,GI anomalies (25-30%).
- Approximately 45% of babies are premature, and about one-third exhibit growth retardation.

TYPES

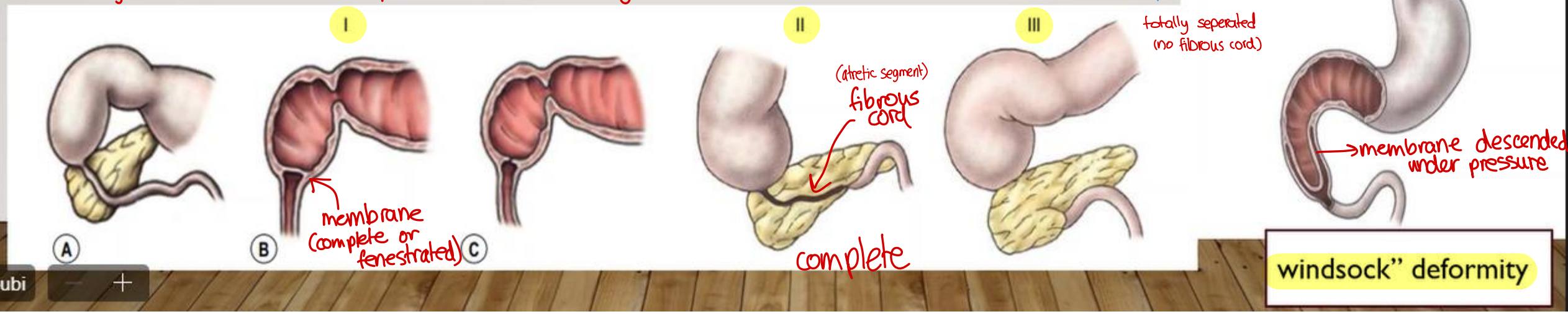
- Congenital Duodenal obstructions are classified into complete or incomplete obstruction

Incomplete obstruction (type I) as fenestrated web or diaphragm and stenosis and most involve the third and/or fourth part of the duodenum.

complete obstruction (type II : 2 ends connected with fibrous cord , type III :complete separation)

- ¬85% of obstructions located distal to the ampulla.
  any obstruction below ampulla of vatar → biliary vomit

II + III → ± annular pancreas
↳ ring of pancreatic tissue encircles the duodenum & causes complete or partial obstruction



DIAGNOSIS

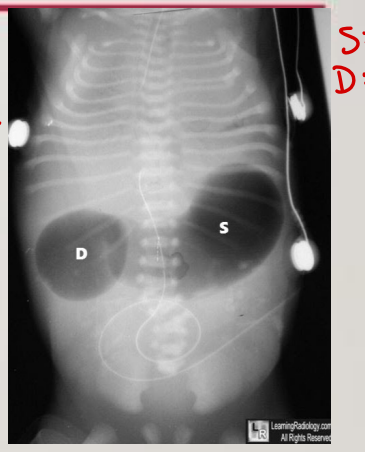
- **Antenatally :**

polyhydramnios (32–81% with complete obstruction)

Double bubble sign (up to 44%)

Karyotyping : find other anomalies

Double bubble



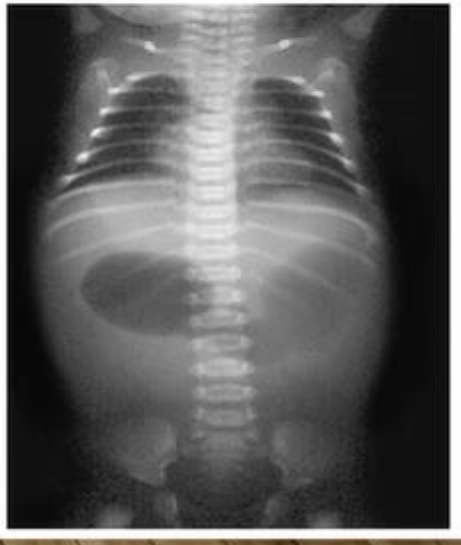
S: stomach
D: duodenum

- **Postnatally:**

→ obstruction under ampulla

Classic presentation is that of bilious emesis within the first hours of life in an otherw
stable neonate +/- upper abdominal distension .

On exam : abdomen is scaphoid. → no gas in small + large bowel



MANAGEMENT

- Resuscitation with correction of fluid balance and electrolyte abnormalities
- NPO + IV access
- Gastric decompression with NG
- **Echocardiography prior to any operation.** → they could have cardiac anomalies
- Not an urgent operation unless malrotation with concurrent volvulus cannot be excluded.

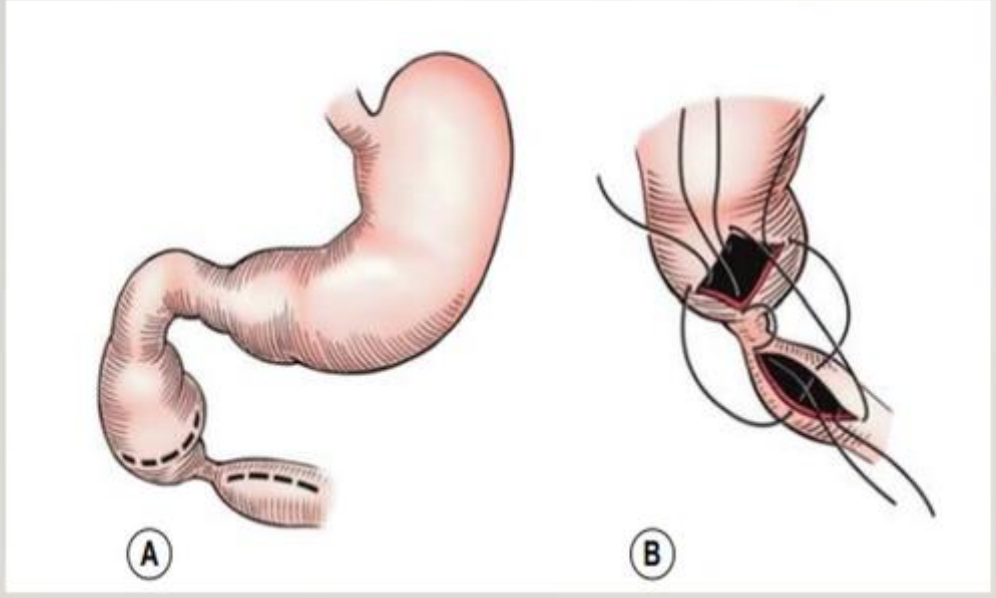
Duodenoduodenostomy

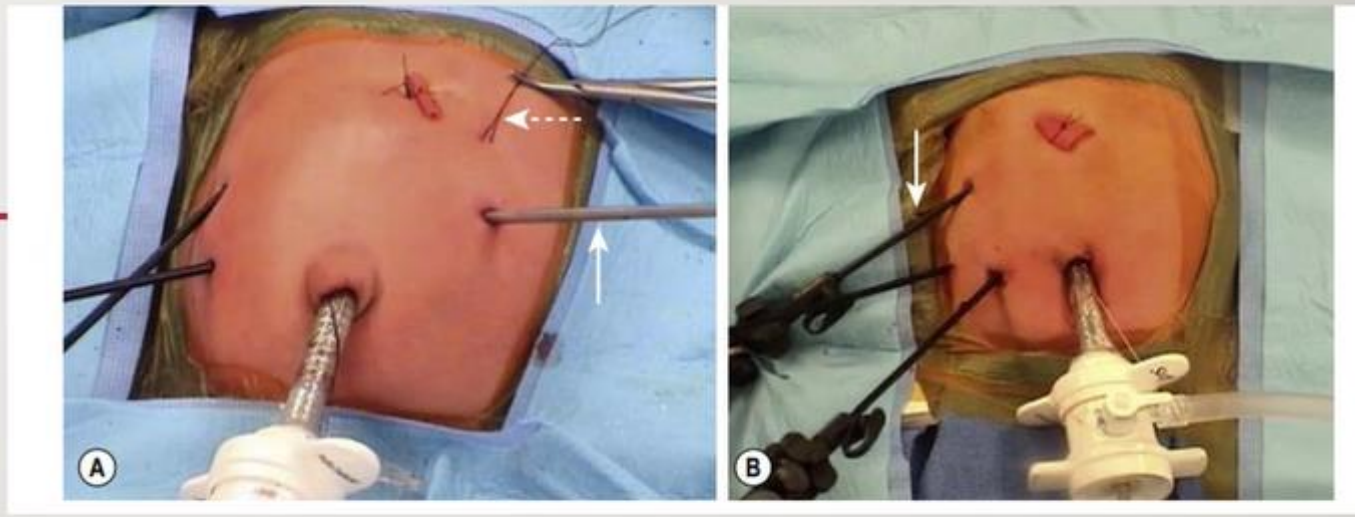
- **Surgery :**

minimally invasive surgery

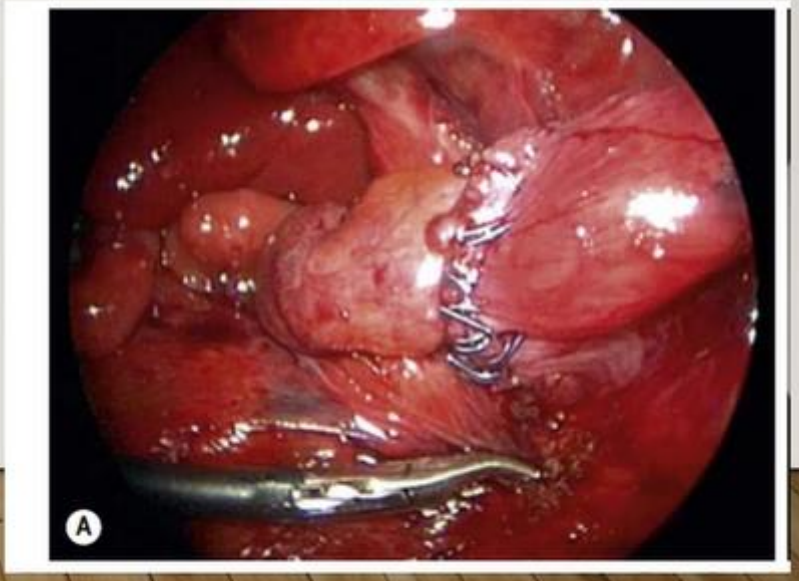
MIS (laparoscopic) or open

(diamond-shaped) anastomosis





duodeno-duodenostomy



- Early postoperative mortality is low (3–5%) and its related to associated congenital abnormalities.

- Long-term survival 90%.

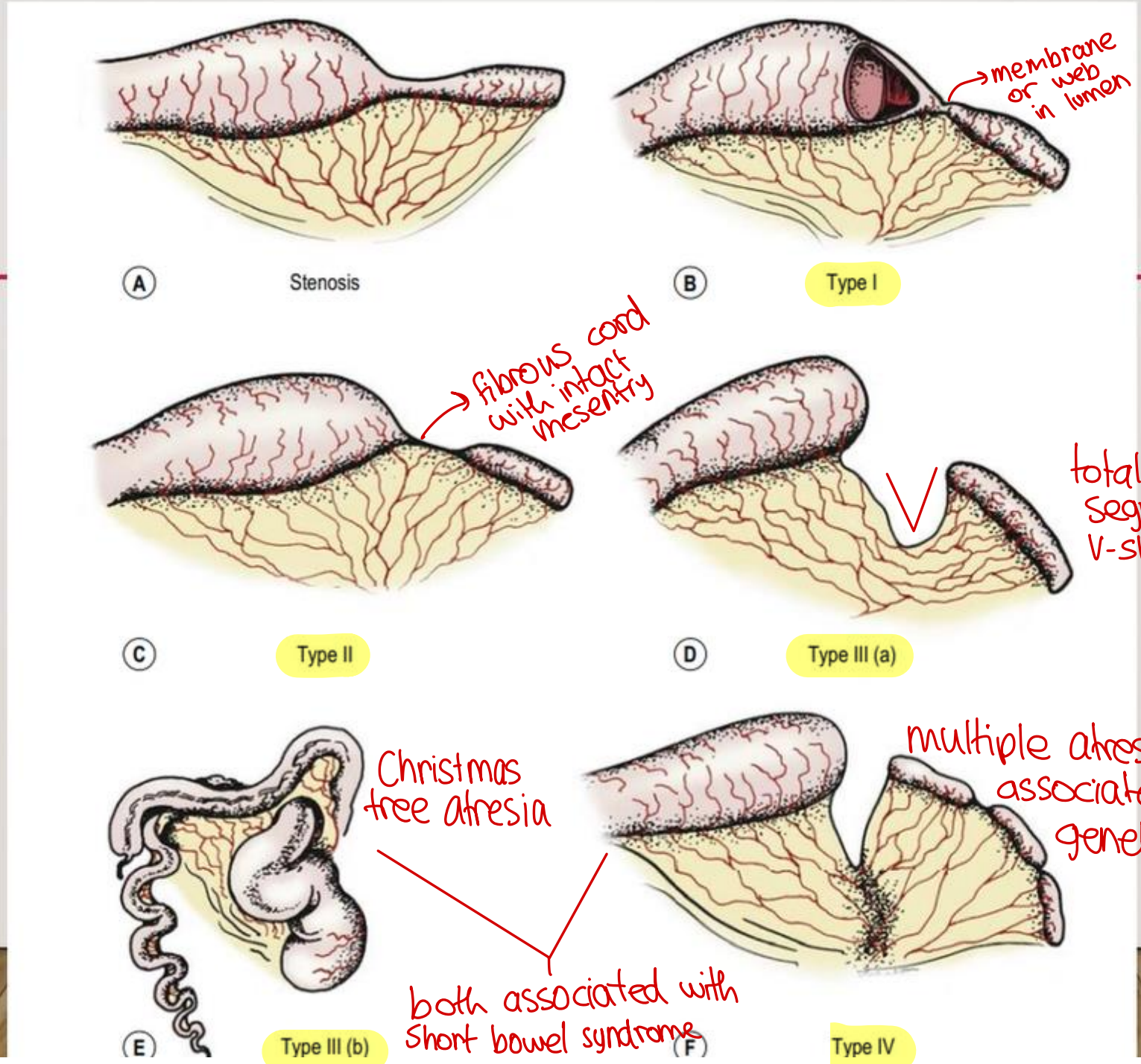
- Long-term complications have been noted following repair and include delayed gastric emptying, severe gastroesophageal reflux, bleeding peptic ulcer, megaduodenum, duodenogastric reflux, gastritis, blind-loop syndrome, and intestinal obstruction related to adhesions.

→due to hypomotility bc of distention prenatally

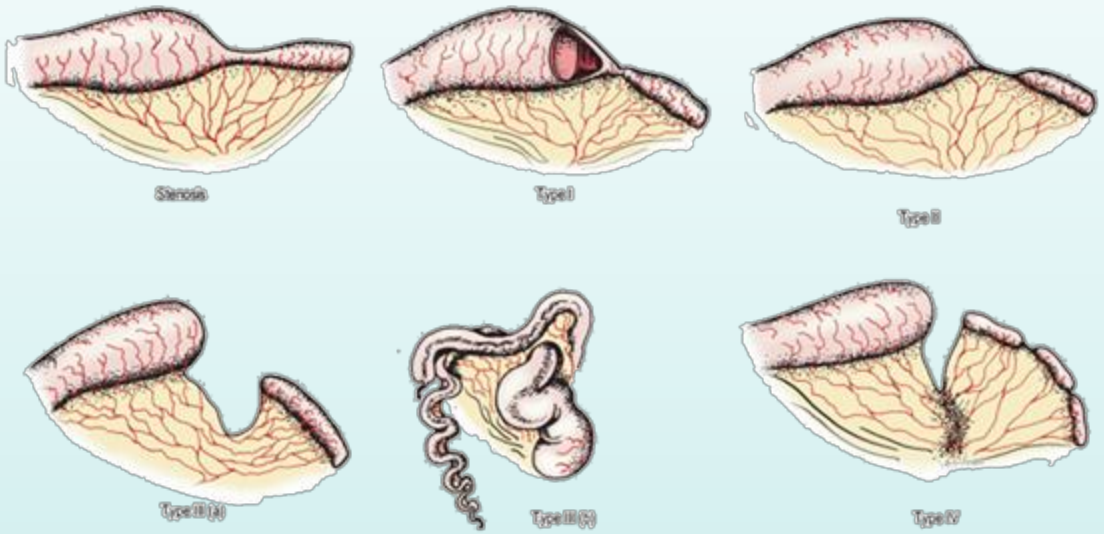
Small bowel atresia

- 1 in 3000-5000 live births.
- It occurs equally in males and females M=F
- 1/3 of infants are premature
- Majority of cases are sporadically
- Intrauterine ischemic insult to the midgut
- Affecting single or multiple segments of the already developed intestine

TYPES

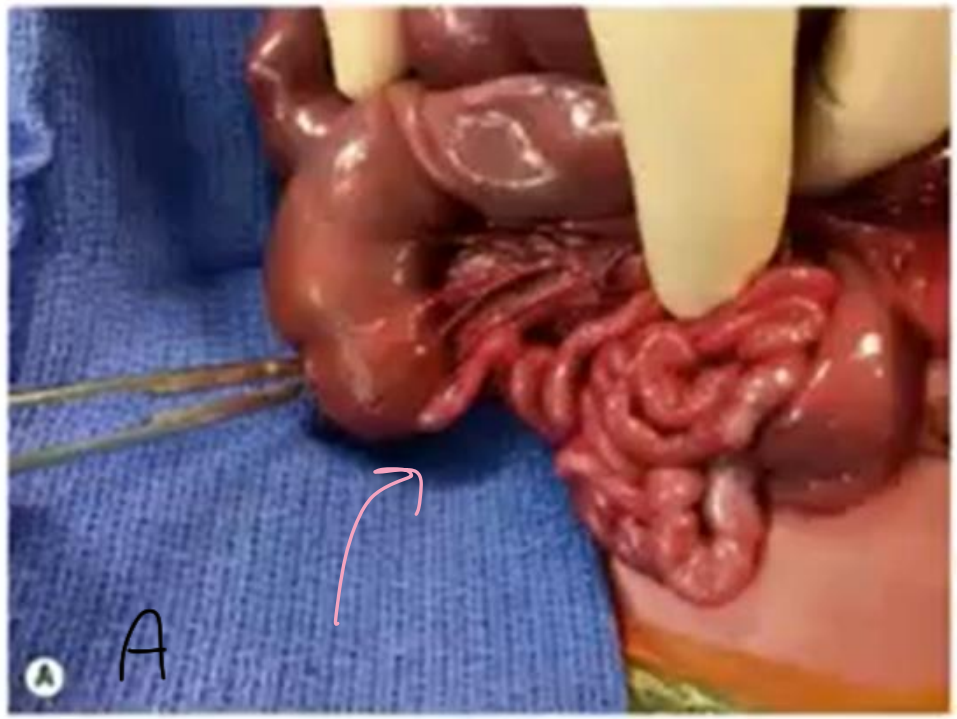

membrane or web in lumen
A
Stenosis
B
Type I
fibrous cord with intact mesentry
C
Type II
D
Type III (a)
totally seperated segments with V-shaped mesentric defect
Christmas tree atresia
multiple atresia associated with familial or genetic association
both associated with short bowel syndrome
E
Type III (b)
F
Type IV

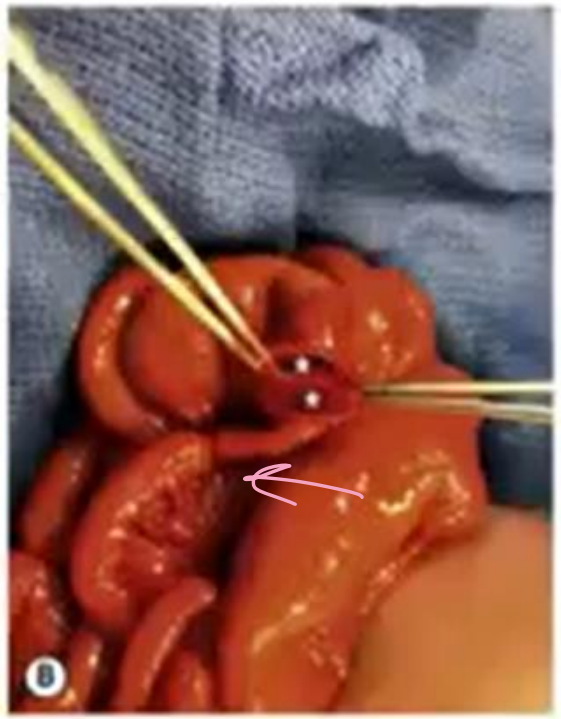
Intestinal Atresia



Classification (applicable to all parts of the intestine)

- Type 0 – Stenosis (no atresia)
- Type I – Membrane or web
- Type II – Fibrous cord joins two blind ends of bowel
- Type III
  - IIIa – Gap between ends with a V-shaped mesenteric defect
  - IIIb – Large defect in the mesentery, significant intestinal loss and distal intestine winds round a single, fragile vascular pedicle ("apple-peel" or "Christmas tree" atresia)
    - ↳ jejunum
    - ↳ duodenum
- Type IV – Multiple atresia ("string of sausages" appearance)


A
A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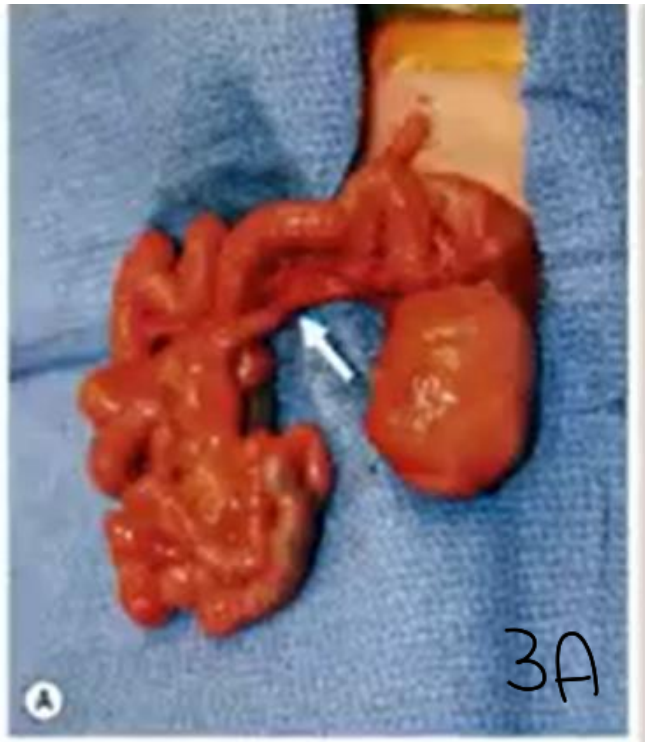
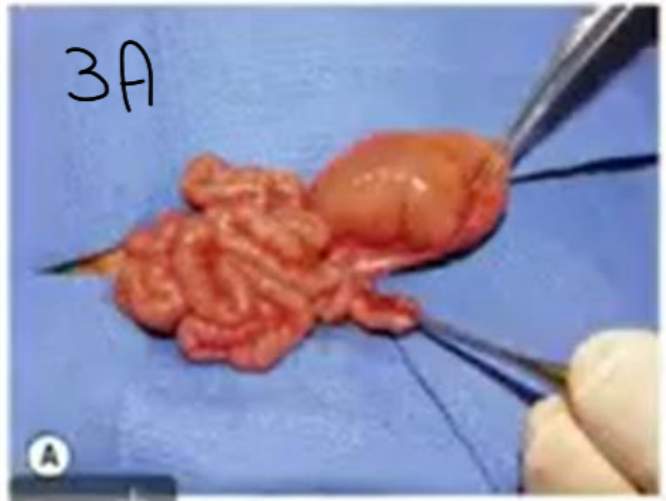
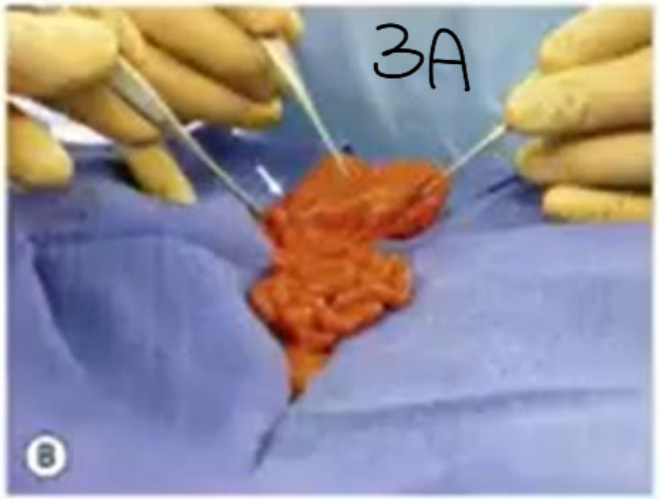

A
3A


3A
A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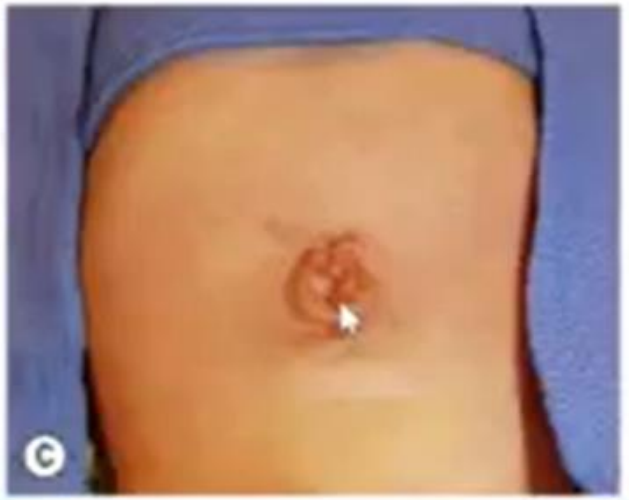
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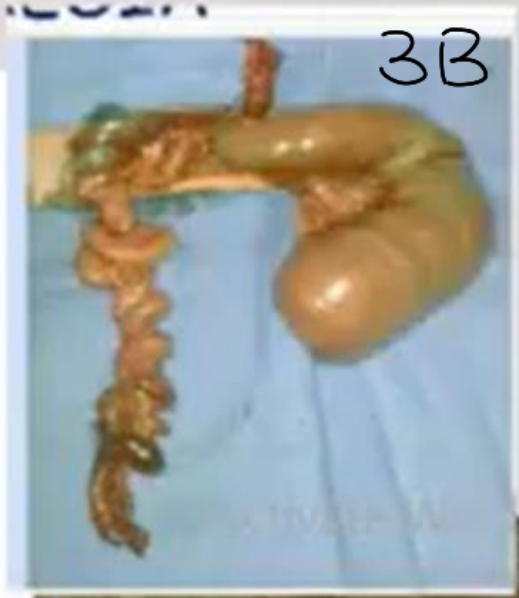

3B

PRESENTATION

- **Antenatal:**
- polyhydramnios (more prominent with proximal atresia).
- Dilated bowel loops above obstruction
- echogenic bowel

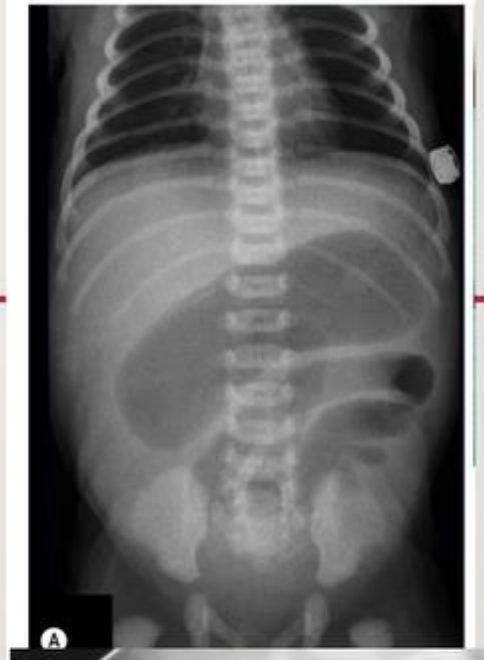
- **Post natally :**
- Bilious vomits
- Distension (depends on the level of the obstruction)
- Might pass minimal amount of meconium

DIAGNOSIS

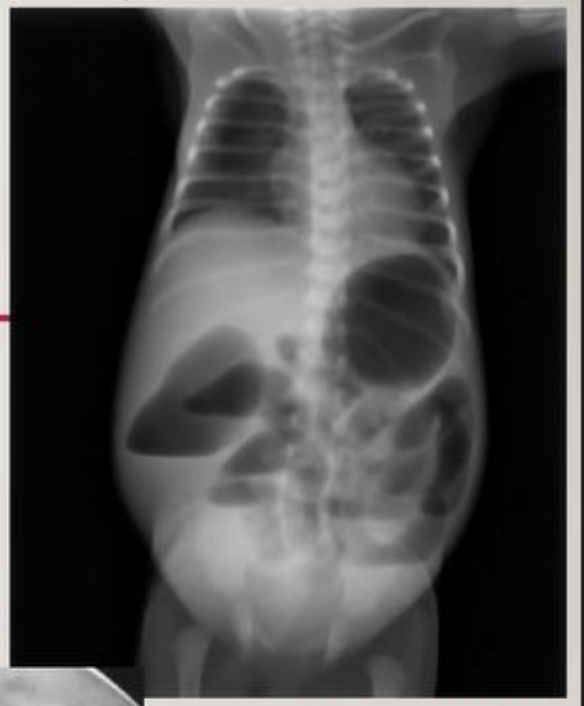
- Abd Xray

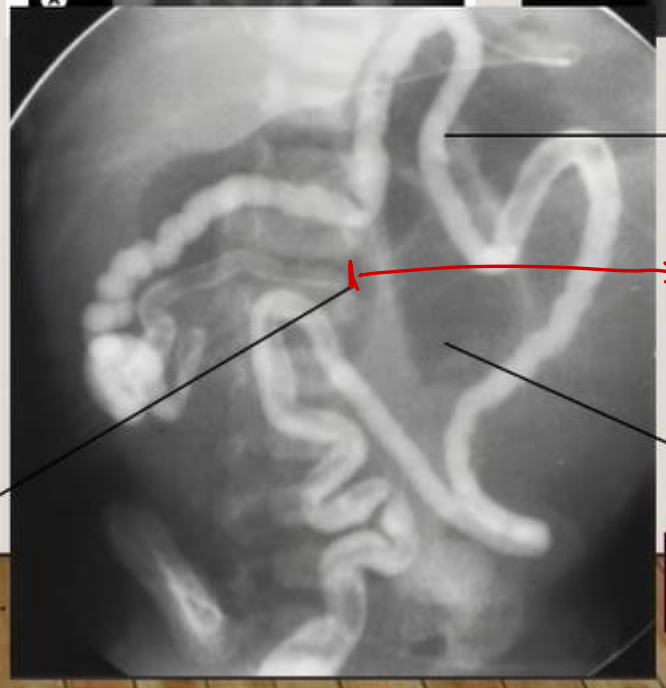
- Lower contrast study /contrast enema
  it help with the diagnosis and rule out other differential diagnosis (like HD and meconium ileus/plug)
  ↳hirshpung disease

Proximal jejunal atresia



ileal atresia





unused colon

MICROCOLON

cut-off of contrast
↳ileal atresia
- micro-colon
- dilated bowel in background

No contrast going back into dilated loop

Dilated proximal bowel loops

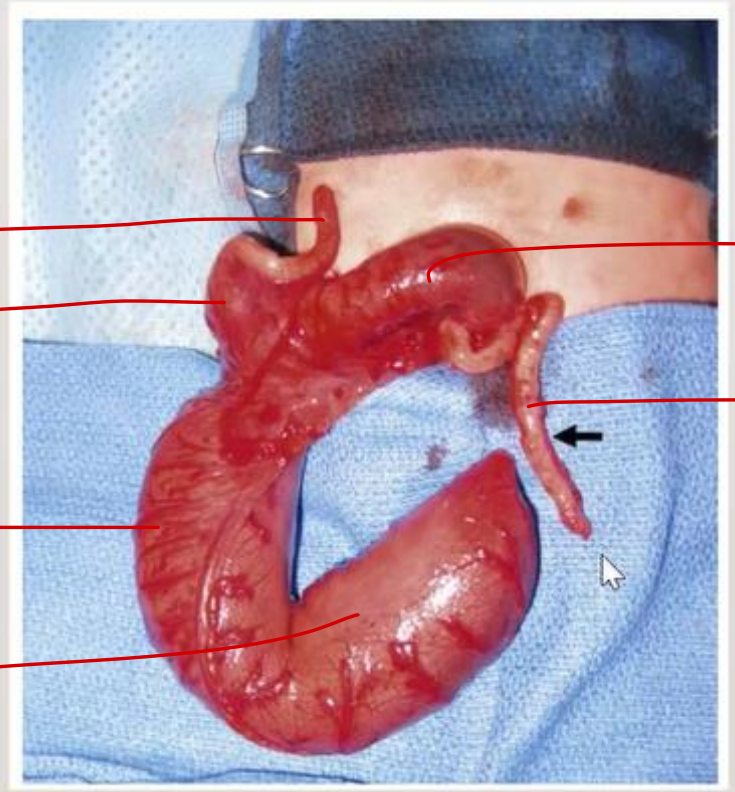
MANAGEMENT

- Resuscitation
- NPO
- IV access + fluid
- Correct electrolyte
- Gastric decompression with NG

Surgery (deliver all bowel and rule out other atresia ,assess the viability and length of remaining bowel , resection and anastomosis) MIS or lap.

↳ to see if the patient will develop short bowel syndrome or not

COLONIC ATRESIA



- Account for 2–15% of all GI atresias. rare
- 1 in 20,000 live births
- Mostly its an isolated anomaly however 1/3 of the babies have associated congenital lesions.

- Three types:

Type I : consists of mucosal atresia with an intact bowel wall and mesentery → has membrane or web

Type II: the atretic ends separated by a fibrous cord

Type III : the atretic ends are separated by a V-shaped mesenteric gap

- Type III is most common type, while types I and II are seen more commonly distal to the splenic flexure

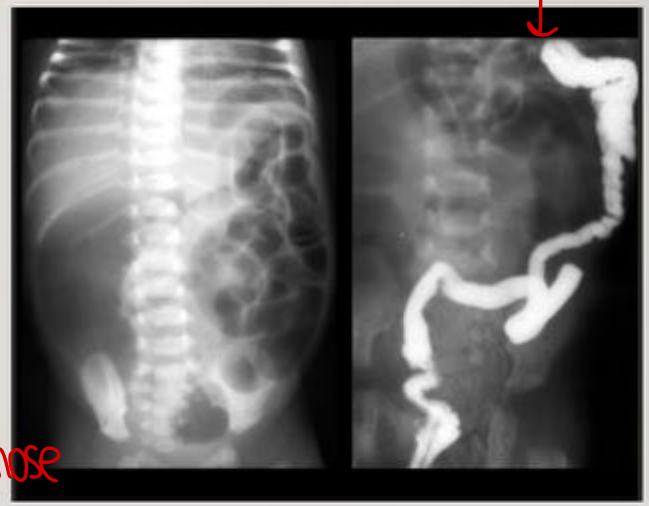
- Abdominal distention, bilious emesis, and failure to pass meconium.

- **Diagnosis :** not prominent Polyhydramnios bc small bowel is intact

Abd Xray : dilated bowel loops of large bowel often associated with a "ground-glass" appearance of meconium mixed with air

Contrast enema : small diameter distal colon that comes to an abrupt halt at the level of the obstruction

round end



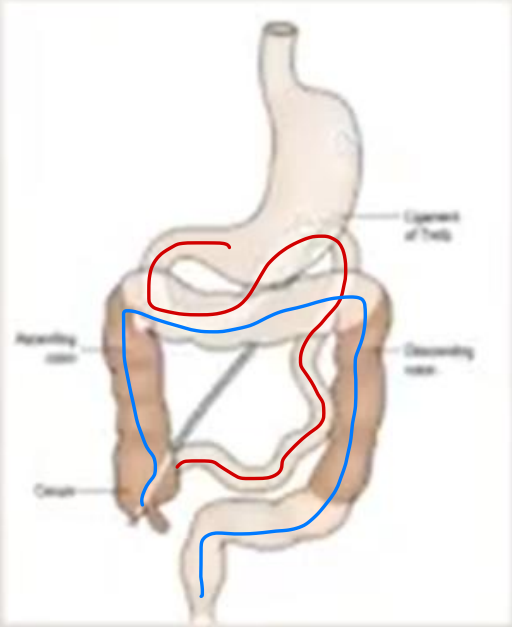
- **Management:**

1. NPO
2. fluid + electrolyte correction
3. NG Tube

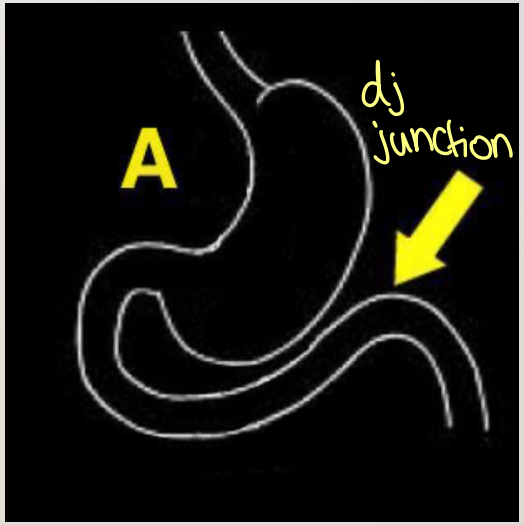
1.Resection and primary anastomaoss 1ry anastomosis → 1. to rule out Hirshpung disease 2. to reduce caliber of the proximal segment to make it easier to anastomose

2.Staged approach (colostomy with mucous fistula followed by anasatamosis 2ry anastomosis

2- MALROTATION



Normal anatomy



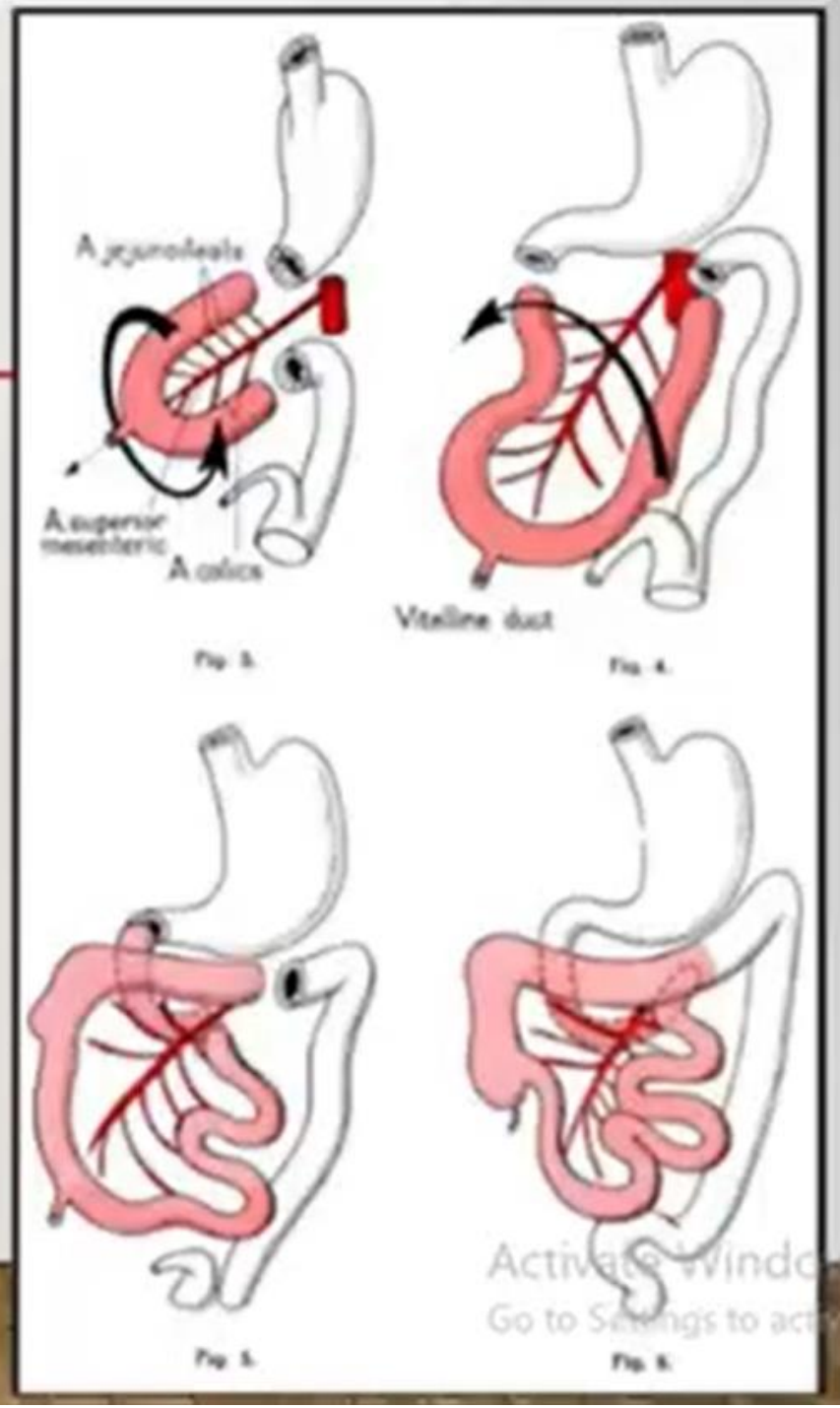
EMBRYOLOGY

- midgut maturation involves four stages: (1) herniation, (2) rotation, (3) retraction, (4) fixation

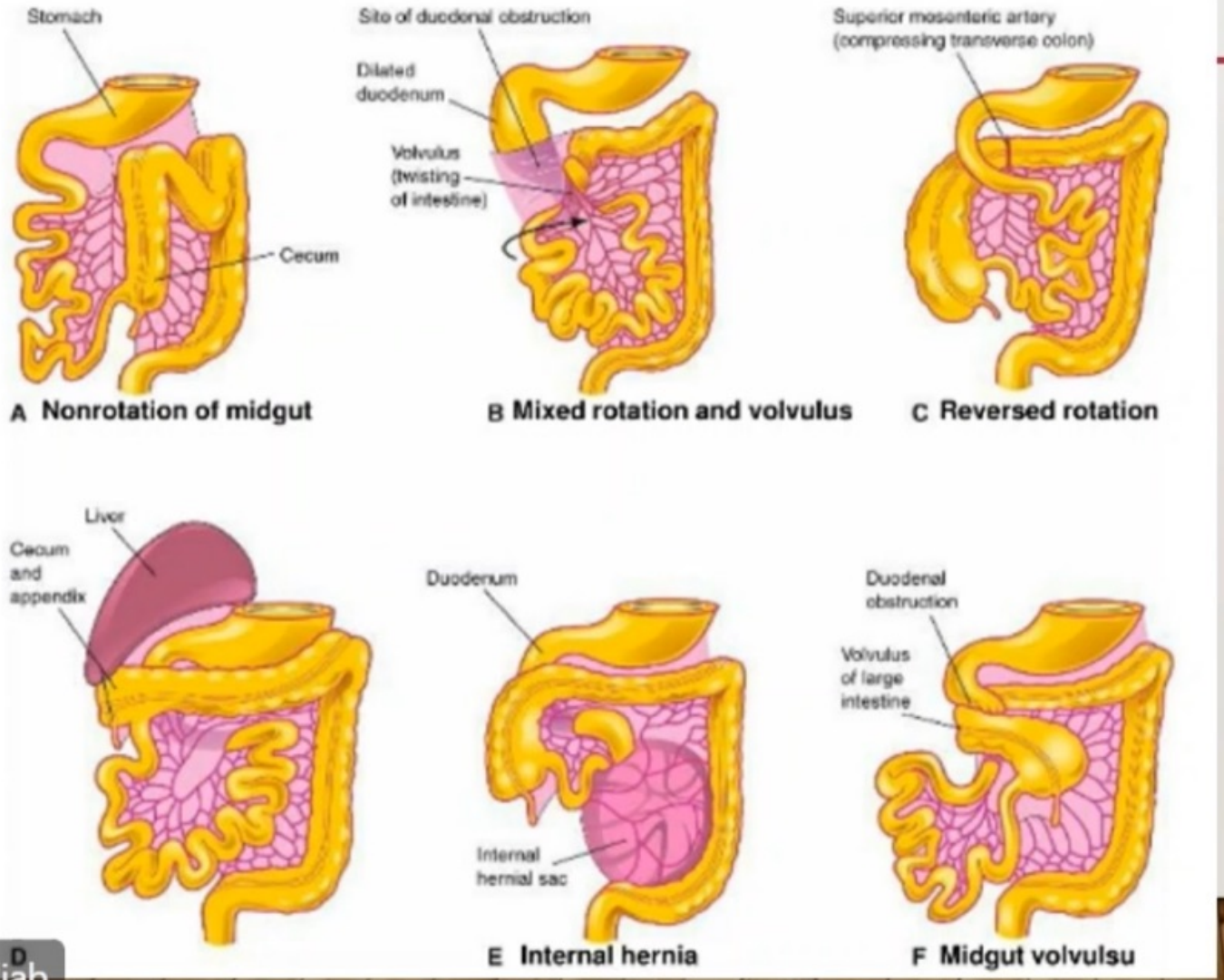
any abnormality in any step will cause malrotation

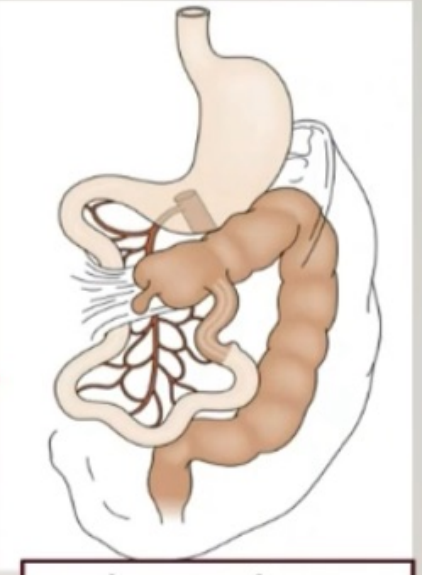
- Embryology

- In total bowel rotates 270 degree counter clock wise around the SMA axis



ITS A THE SPECTRUM

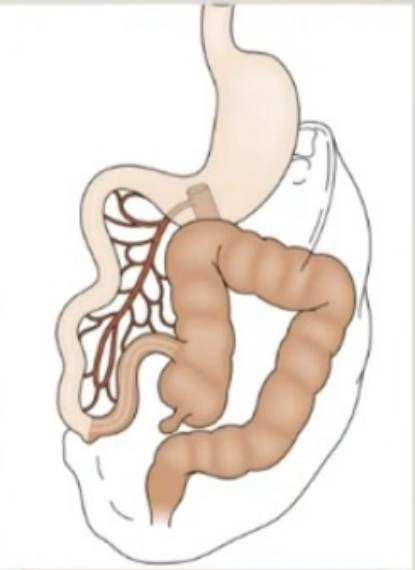




Incomplete rotation

no rotation at all

- no fixation of the colon
- short mesentery
- risk of volvulus



Nonrotation

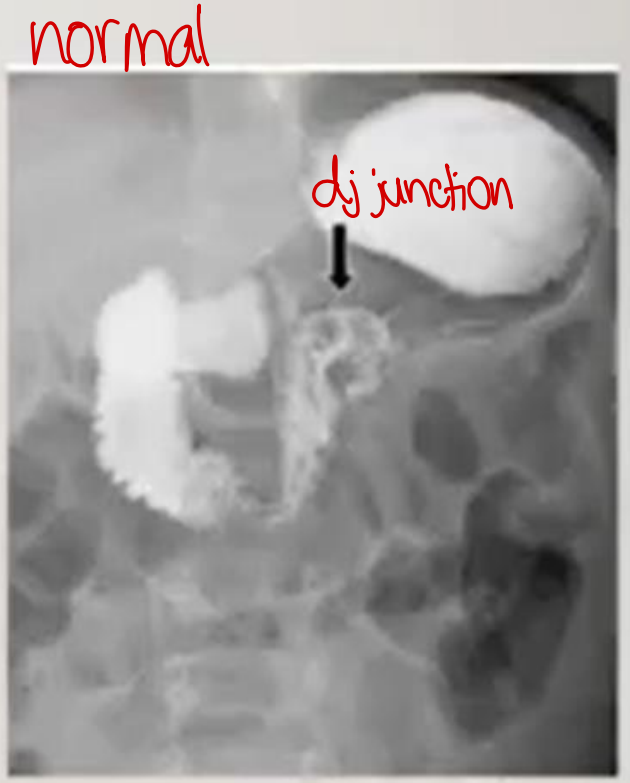
- 1 in 6000 live births
- Classic malrotation with midgut volvulus often develops in a previously healthy term neonate.
- Up to 75% of present during the first month of life and 15% will present within the first year.

billious vomiting: intestinal obstruction + malrotation + volvulus until proven otherwise

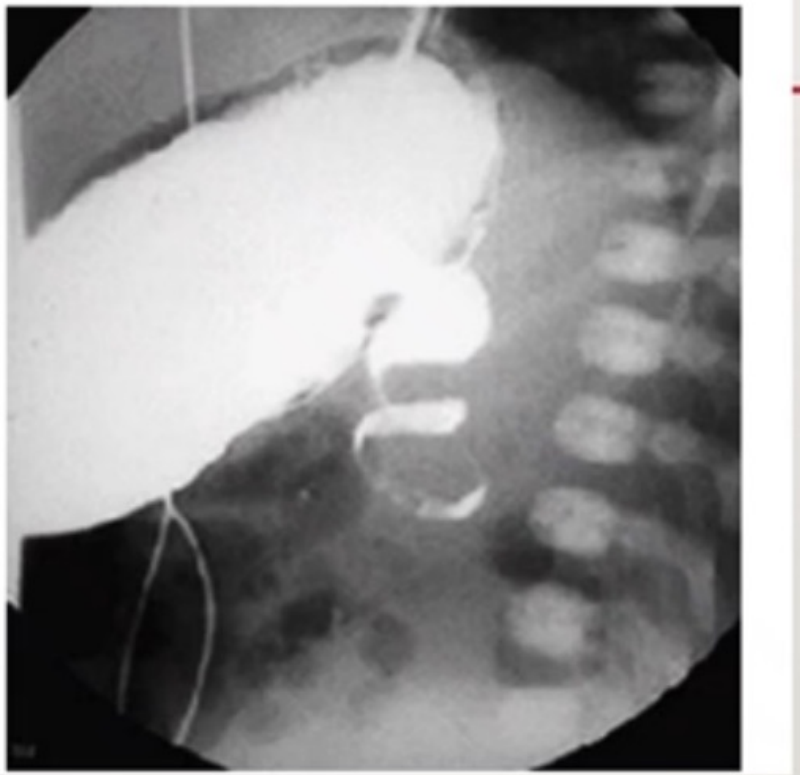
- Sudden onset of bilious vomiting is the cardinal sign of neonatal intestinal obstruction, and malrotation with volvulus must be the presumed diagnosis until proven otherwise

DIAGNOSIS

- **The upper contrast study is the gold standard study**
- Color Doppler US



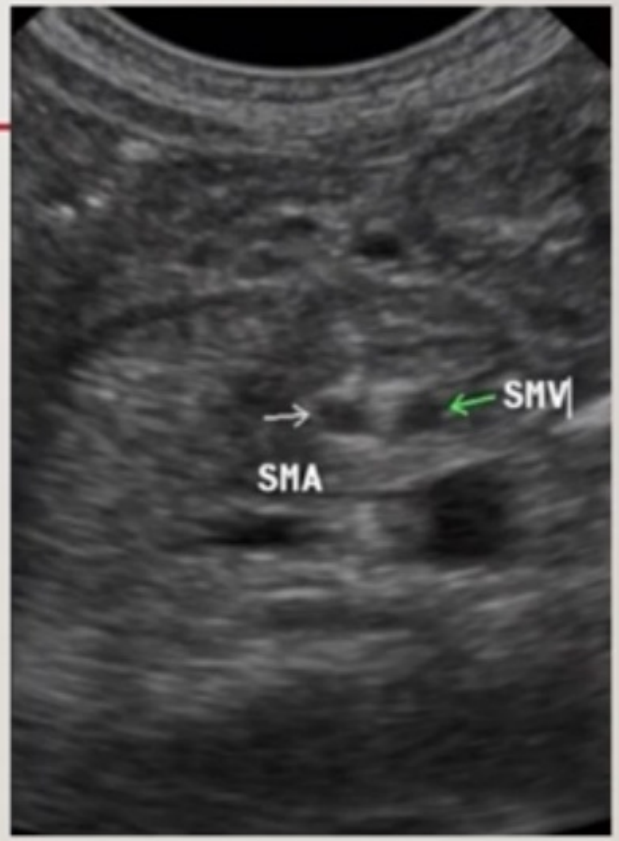
normally:
Rt: SMV
Lt: SMA



Coil spring: incomplete obstruction bc of malrotation & volvulus

beak appearance: complete obstruction

"Coil spring" or "corkscrew" configuration with incomplete obstruction and the "beak" appearance in the duodenum with complete obstruction



inversion of SMA & SMV → acute volvulus

Dilated duodenum with inversion of the SMA and vein (the whirlpool sign) in cases of acute volvulus

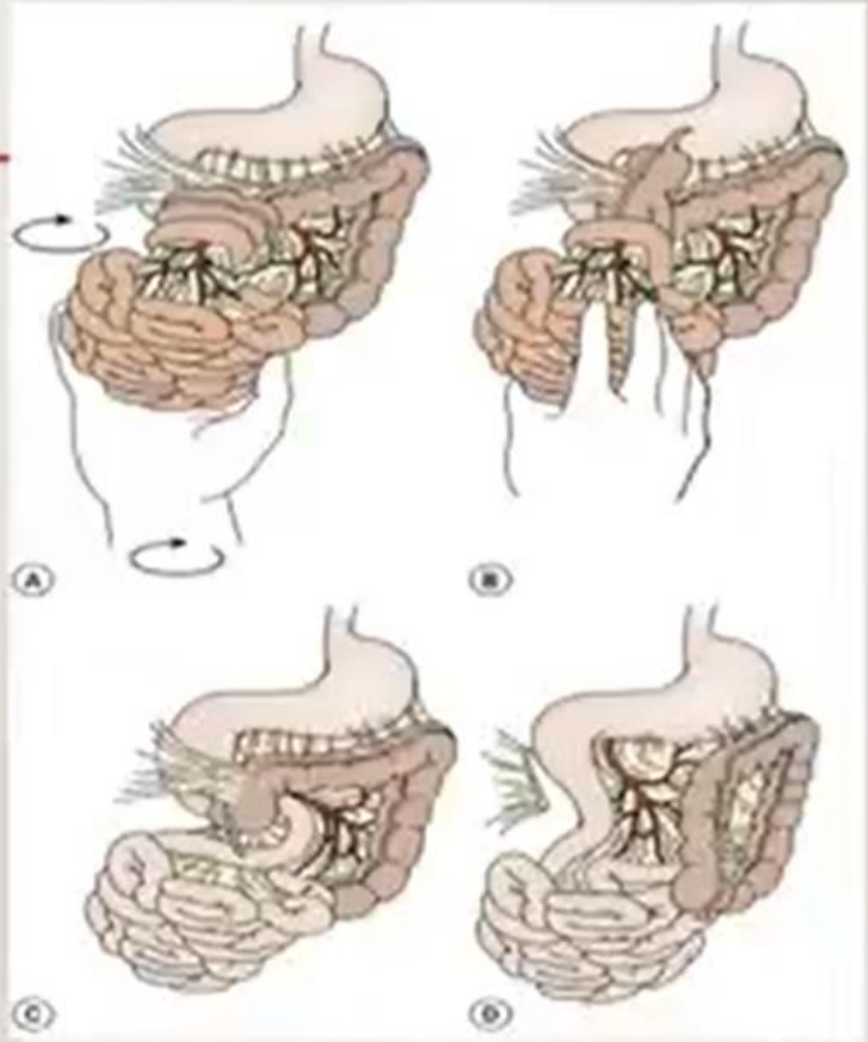
MANAGEMENT

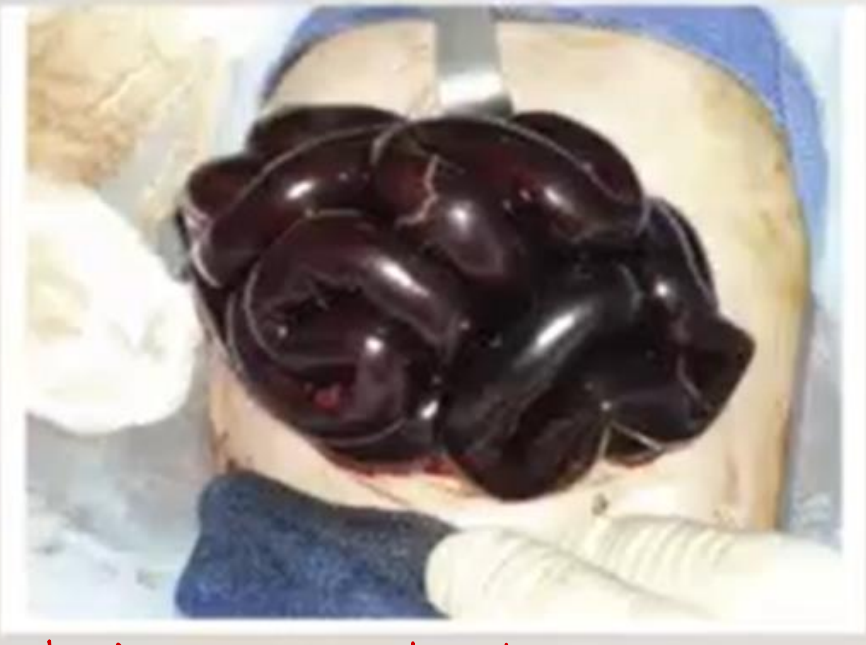
- Resuscitation , gastric decompression , broad spectrum Abx
- Surgery : open/laparoscopic

urgent surgery: can have cut of blood supply & gangrene

Box 31.1 Six Key Elements in Operative Correction of Malrotation

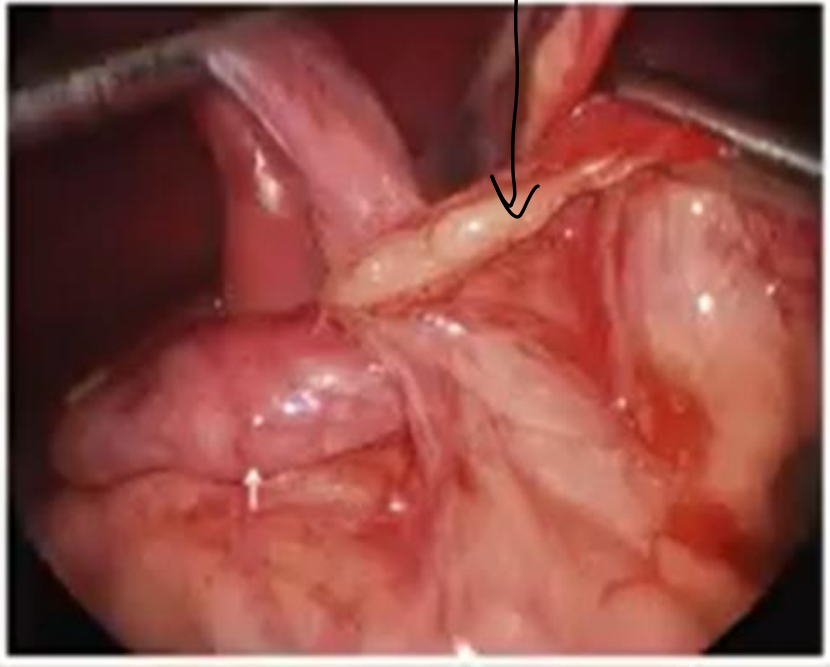
1. Entry into abdominal cavity and evisceration (open)
2. Counterclockwise detorsion of the bowel (acute cases)
3. Division of Ladd cecal bands
4. Broadening of the small intestine mesentery
5. Incidental appendectomy
6. Placement of small bowel along the right lateral gutter and colon along the left lateral gutter





ischemic volvulus

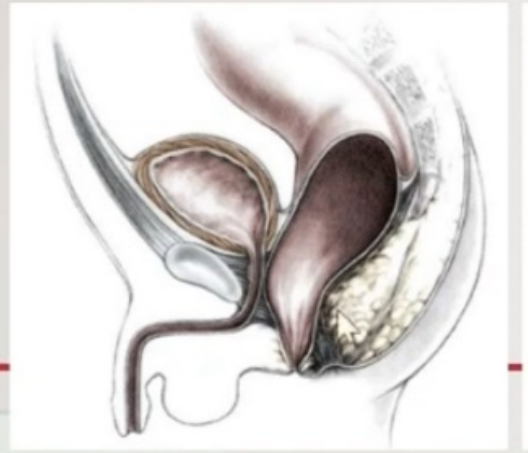
Ladd band

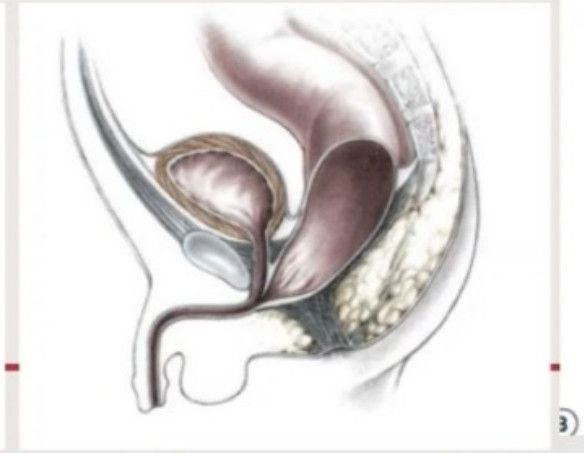


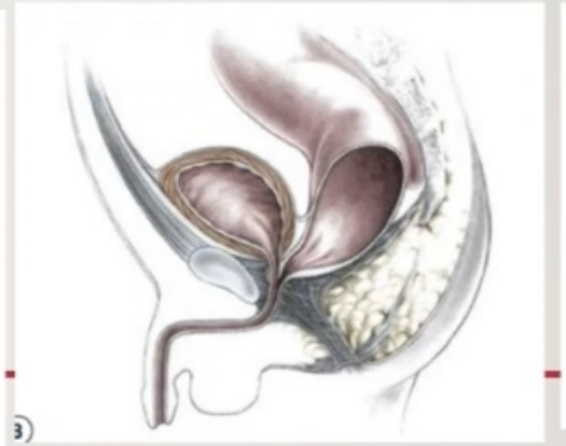
3- Anorectal malformation

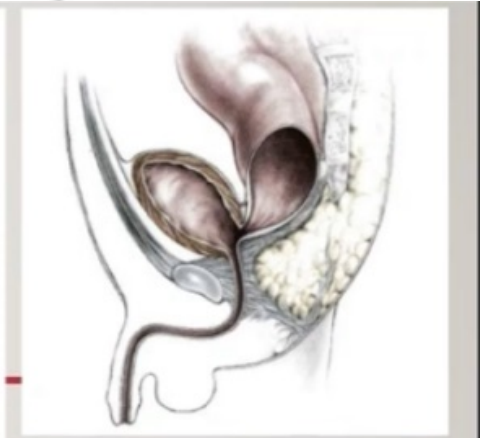
imperforate anus ± rectum

- 1 in 4000–5000
- slightly more common in males M > F
- The estimated risk for a couple having a second child with an anorectal malformation is approximately 1%
- Present as imperforate anus either with fistula (to GU tract) or without fistula (5%, Down syndrome)







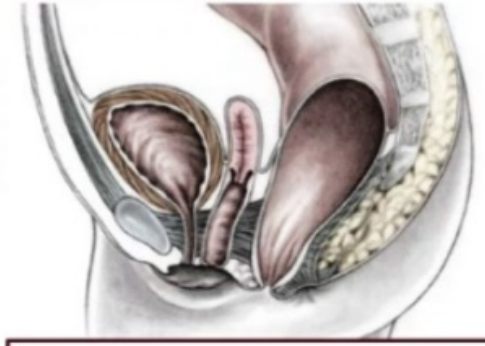


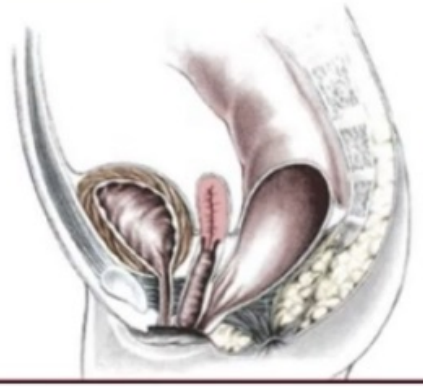
Males
- Rectoperineal fistula
- Rectourethral bulbar fistula
- Rectourethral prostatic fistula
- Rectobladderneck fistula
- Imperforate anus without fistula
- Rectal atresia/rectal stenosis

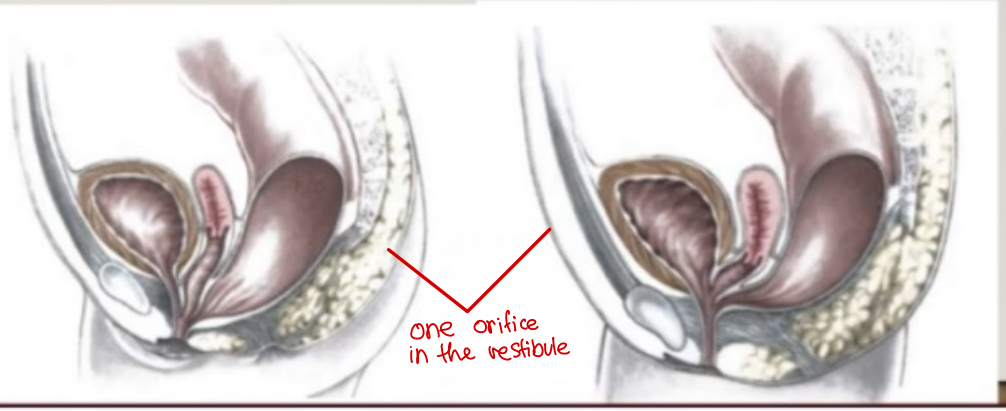
Females
- Rectoperineal fistula
- Rectovestibular fistula
- Cloaca
- Complex malformations
- Imperforate anus without fistula
- Rectal atresia/rectal stenosis

Recto perineal fistula — ant. to anal sphincter → incontinence | **Rectourethral bulbar fistula** | Rectourethral prostatic fistula | Rectobladder neck fistula

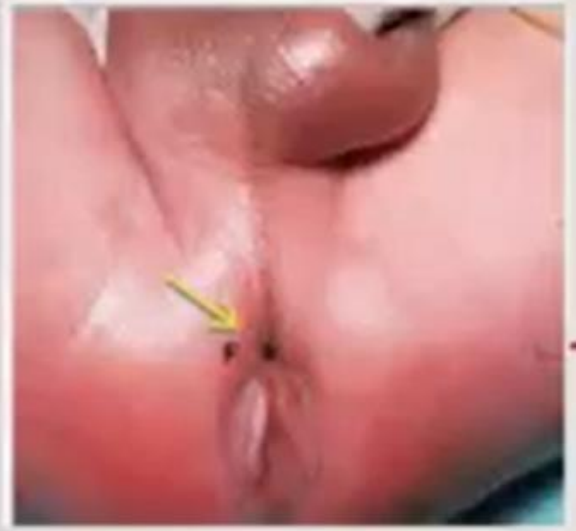
Should know the type to know if the patient will have continence & see if stool passes with urine





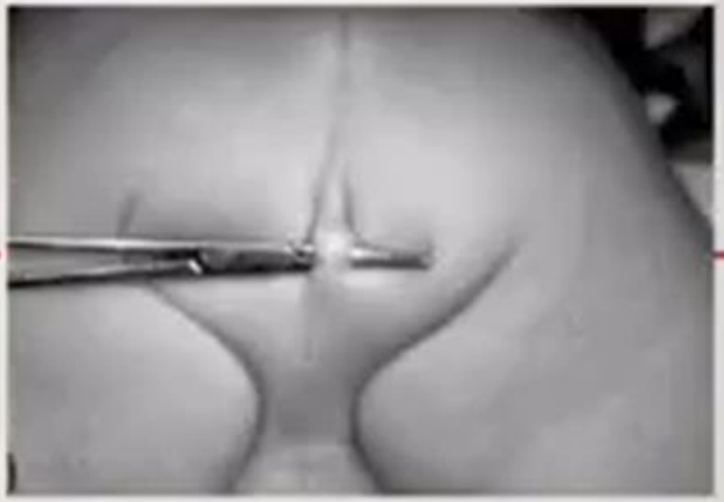


Recto perineal fistula | **Rectovestibular fistula** | cloaca (short channel) | cloaca (long channel)

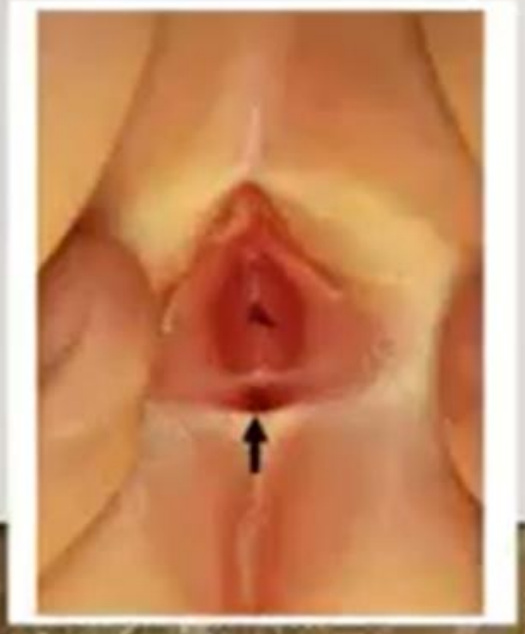


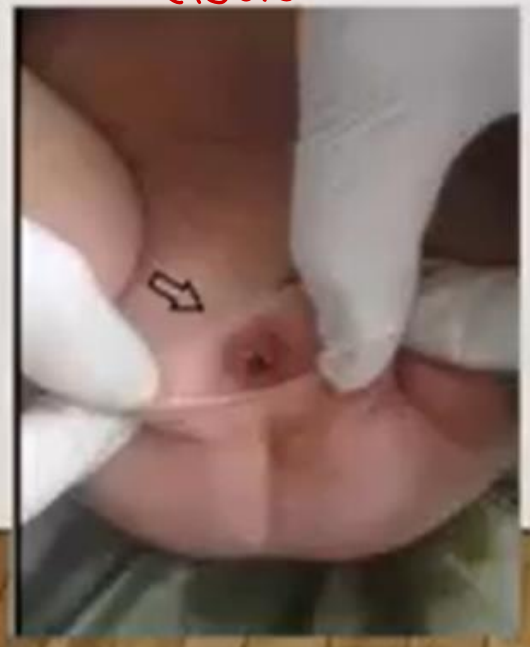
recto perineal

cloaca



recto-vestibular





- Clinical examination is the most important step and you can diagnose most cases (except with rectal atresia type/ normal looking anus)

- Initial management :
  - Nasogastric tube (for decompression, & to rule out esophageal atresia)
  - NPO + IV fluids
  - Antibiotic prophylaxis
  - Watchful waiting (for 12-24 hrs)
  - Rule out VACTERL associations

V A C T E R L

Vertebral
anorectal
cardiac
tracheo-
esophageal
renal
limb

MALE Anorectal Malformation

Newborn male infant

No anus seen at birth

Inspect perineum @ 24 hours

Meconium in urine or cross-table lateral film showing rectum further than 1 cm

Meconium on perineum or cross table lateral film showing rectum within 1 cm

Primary PSARP

Stoma
Divided proximal sigmoid colostomy with skin-bridge and mucous fistula

next page....

Distal colostogram @ 8–10 weeks

low malformation ↳to know the type high malformation

Rectum reachable through posterior sagittal incision

Rectum NOT reachable through posterior sagittal incision

PSARP

Laparoscopic assisted PSARP

Exam of anus to determine size of anoplasty

Dilations started at 2 weeks post op

VACTERL SCREENING:
- **Spinal radiograph**
- **Sacral radiographs**
  - o AP & Lateral
- **Spinal & Pelvic US** (<3 months old)
- **MRI spine** (>3 months old) or if spinal USS abnormal
- **ECHO**
- **NG tube** and AXR to confirm in stomach
- **Renal USS**
- **Limbs radiographs** (as clinically indicated)

R/O VACTREL

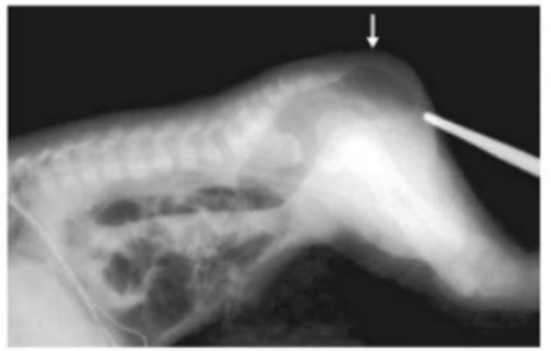
Follow up:
- 6 months
- 1 year
- Annually

Continence assessment @ 4 years

Sacral deformities are the most frequently associated defect.

Spinal abnormalities genitourinary defects varies from 20–54%

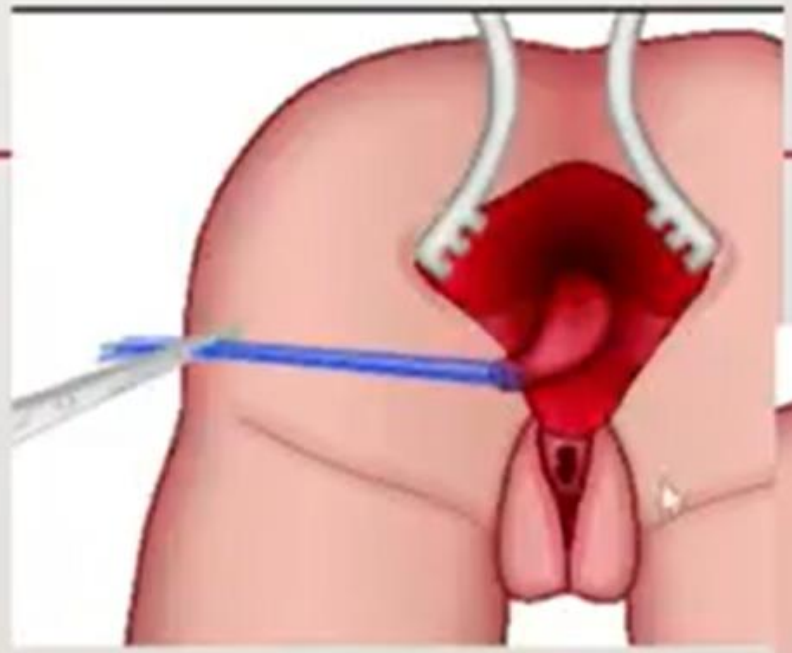
Click to see details

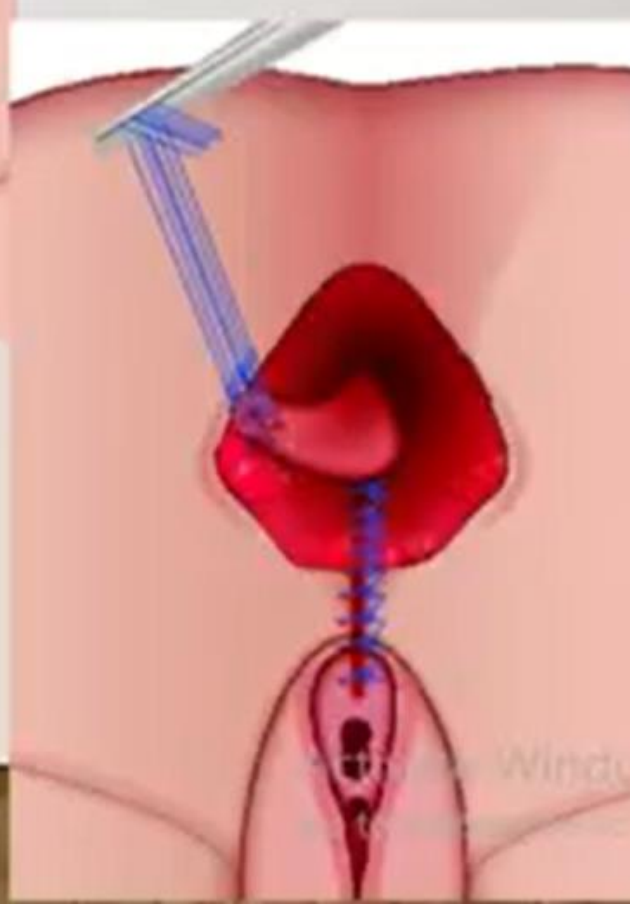


PSARP: Post. sagittal ano-rectoplasty

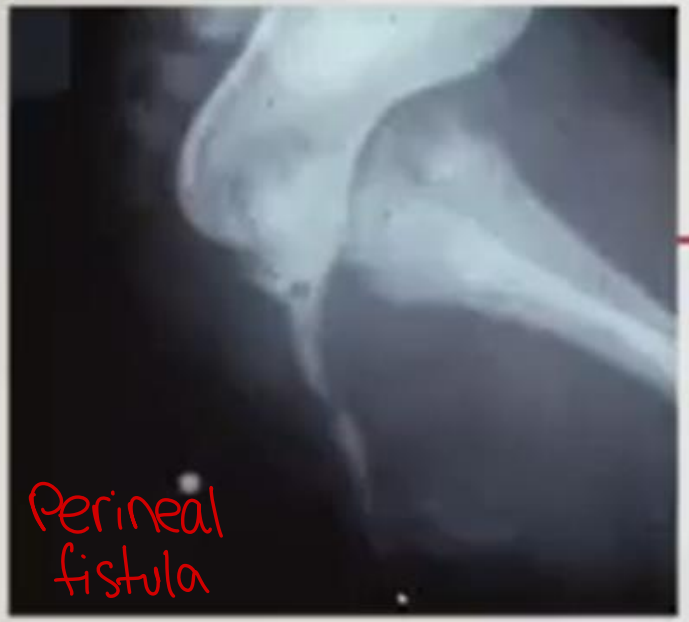
a: empty the bowel
b: distal colostogram

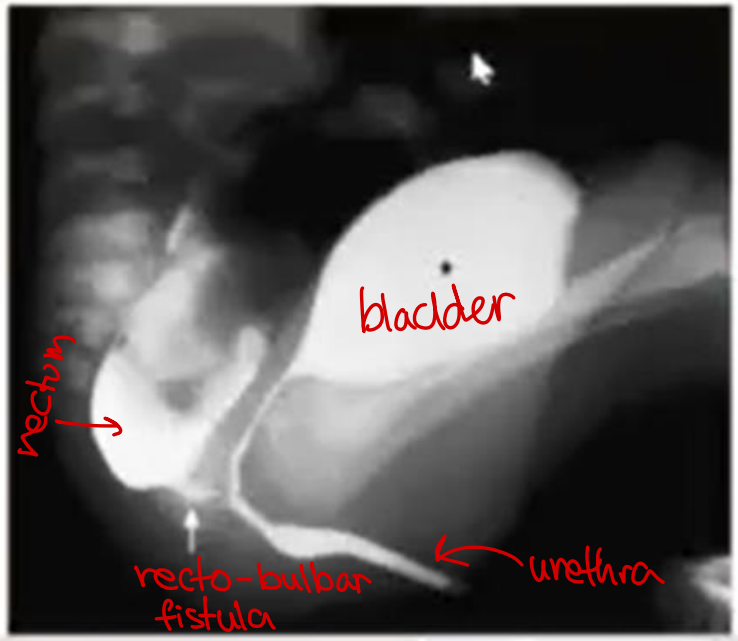
Put contrast in the arrow to know the type & if there is a fistula or no

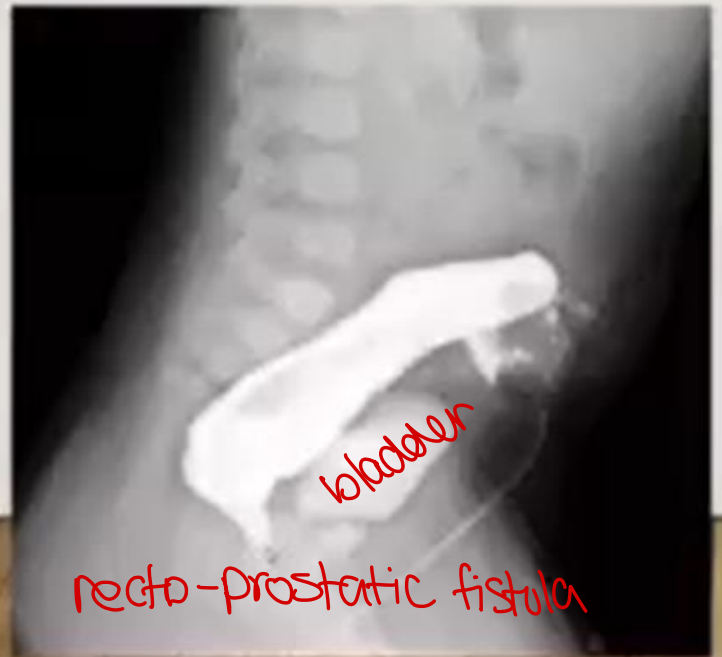


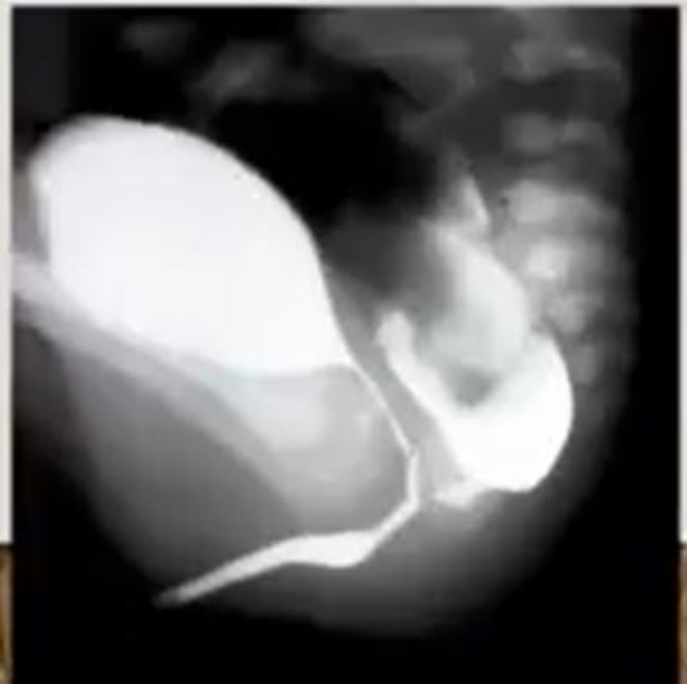


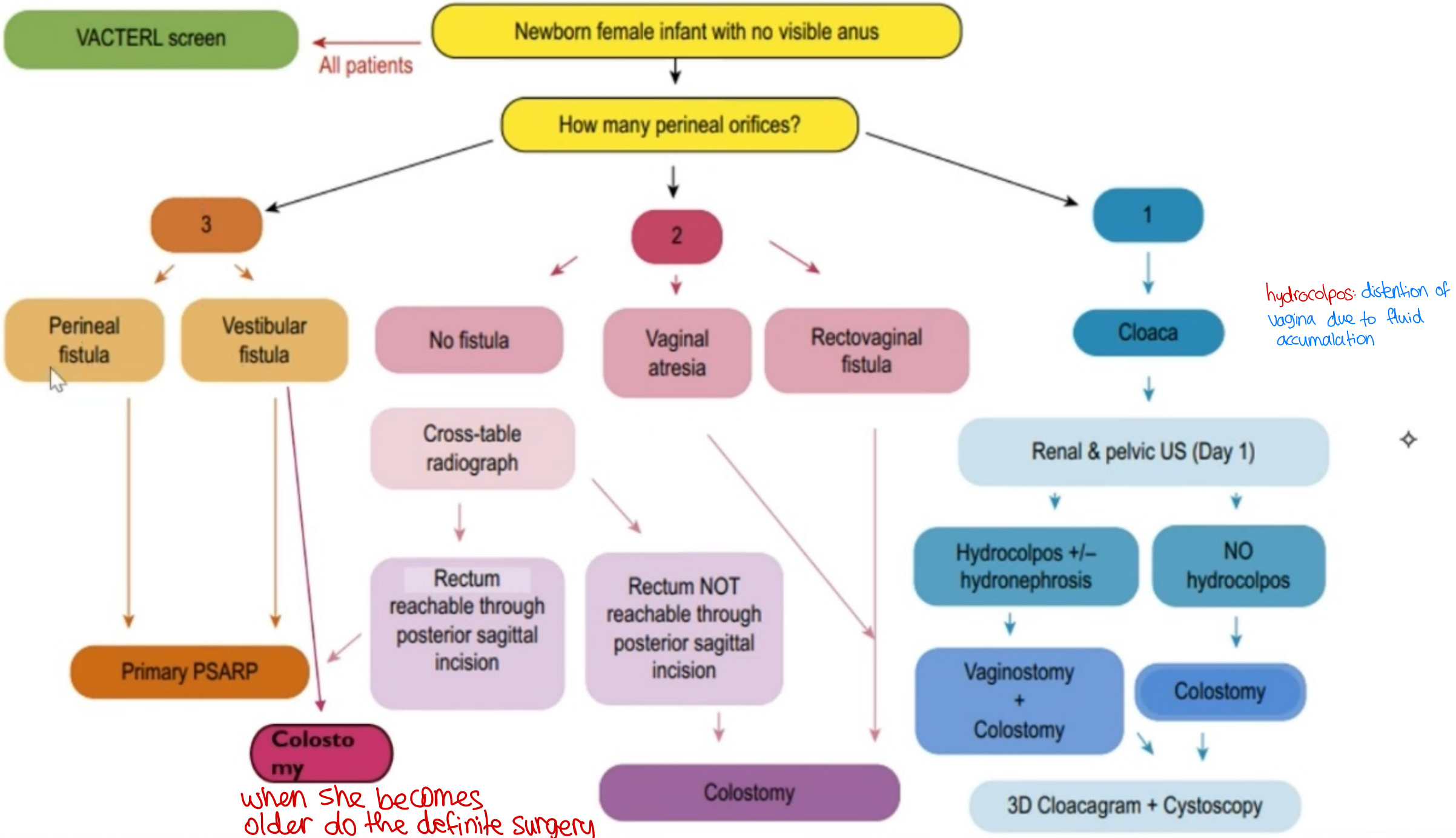
low malformation: more consitipation (below level of levator ani)
high malformation: more incontinence (above level of levator ani)











hydrocolpos: distention of vagina due to fluid accumulation

when she becomes older do the definite surgery

OUTCOME

- Fecal and urinary Incontinence

(high in high anorectal malformation , associated sacaral and spinal abnormality)

- Constipation :in low type malformation

- Recurrent fistula

4- Hirshprung disease

ماغي أعصاب

- Absence of ganglion cells in the myenteric and submucosal plexuses of the intestine

no descendant of ganglionic cells, or died after descending

- 1 in 5000 live births
- ¬ 80% of children have a "transition zone" in the rectum or rectosigmoid colon

¬ 10% have more proximal colonic involvement

5–10% have total colonic aganglionosis with variable involvement of the distal small intestine.

Near total intestinal aganglionosis is rarely encountered

- Associated syndromes : trisomy 21, congenital central hypoventilation syndrome, Goldberg–Shprintzen syndrome, Smith–Lemli– Opitz syndrome, neurofibromatosis, and neuroblastoma , MEN2 , Neurocristopathy (e.g Waardenberg-Shah syndrome).

- Associated with heart disease , malrotation , UT anomalies , CNS anomalies.

Urinary tract

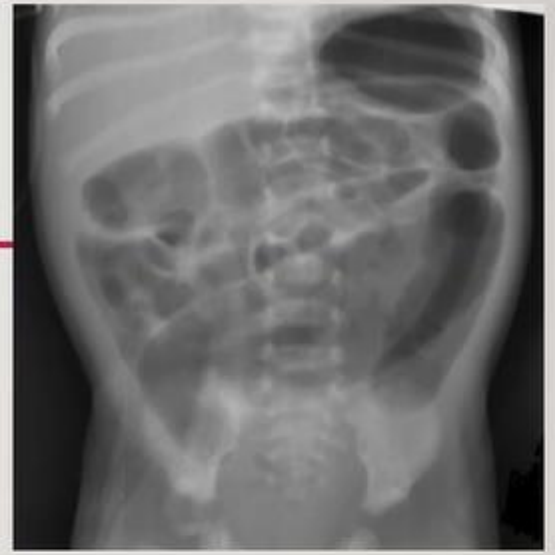
- Genetic :most common is the RET proto-oncogene (esp familial and long-segment involvement) .other like SOX10, EDNRB, GDNF , EDN3, ECE1, NTN, SIP1 don't memorize

DIAGNOSIS

- Prenatal diagnosis of HD is rare

- Abdominal distension, bilious vomiting, and feeding intolerance. Delayed passage of meconium beyond the first 24 hours is present in approximately 90%.
- ¬ 10% of neonates with HD present with Hirschsprung-associated enterocolitis (HAEC)
  ↳ inflammatory/infectious process, stasis of normal flora & growth → diarrhea
- Patients presenting later in childhood have severe chronic constipation

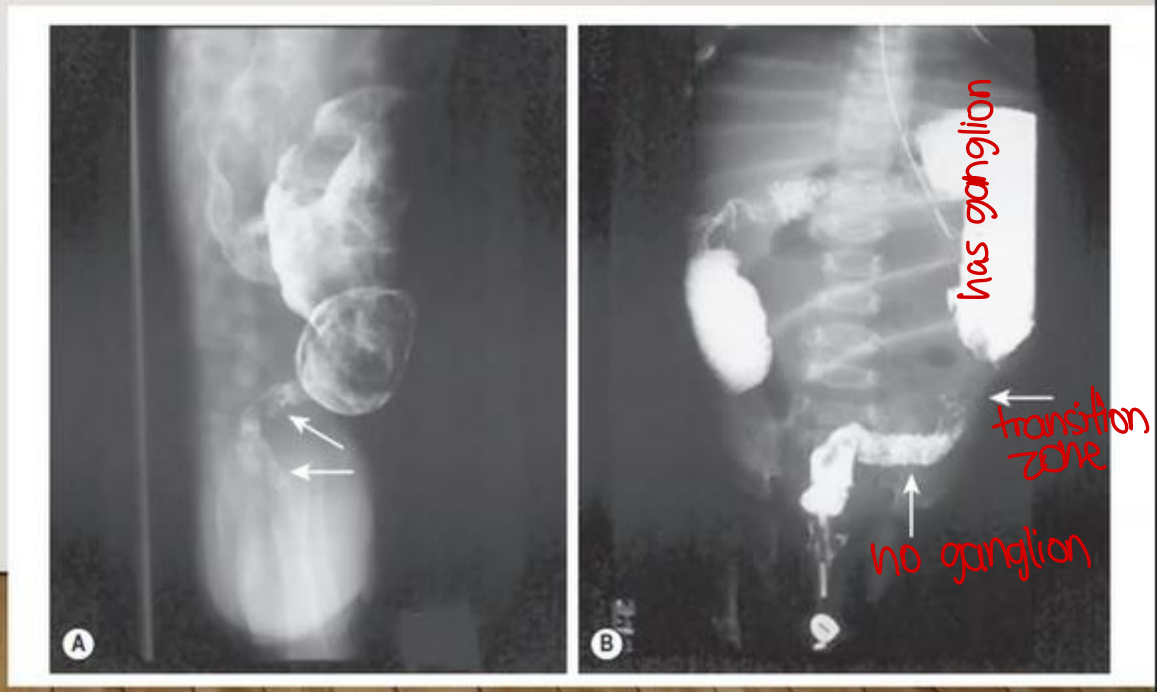
- failure to thrive
- distended abdomen
- recurrence of enterocolitis



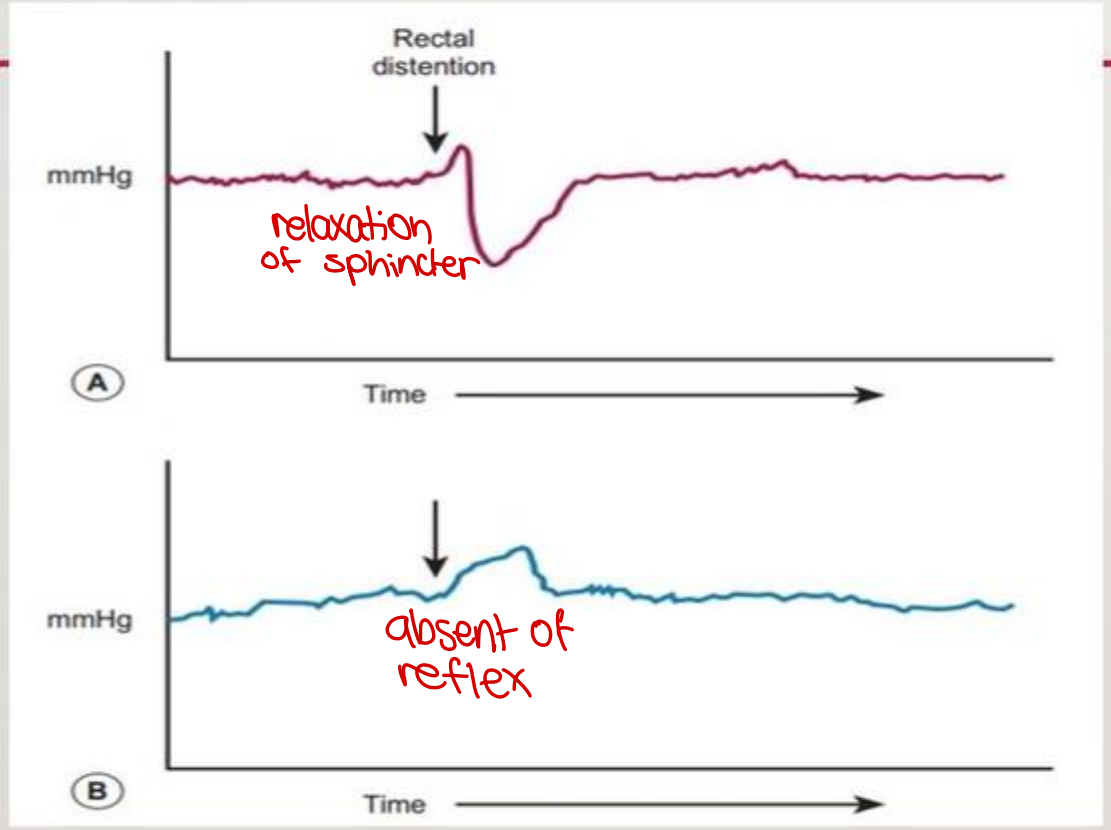
- Abd Xray
- Lower contrast study/ enema. diagnostic
- Anorectal manometry

confirmation

- **Rectal biopsy (suction or strip) is the gold standard**
  - ↳ no ganglion cells in mucosa/submucosa + abnormal acetylcholine staining + absent calretenin



anorectal manometry:



MANAGEMENT

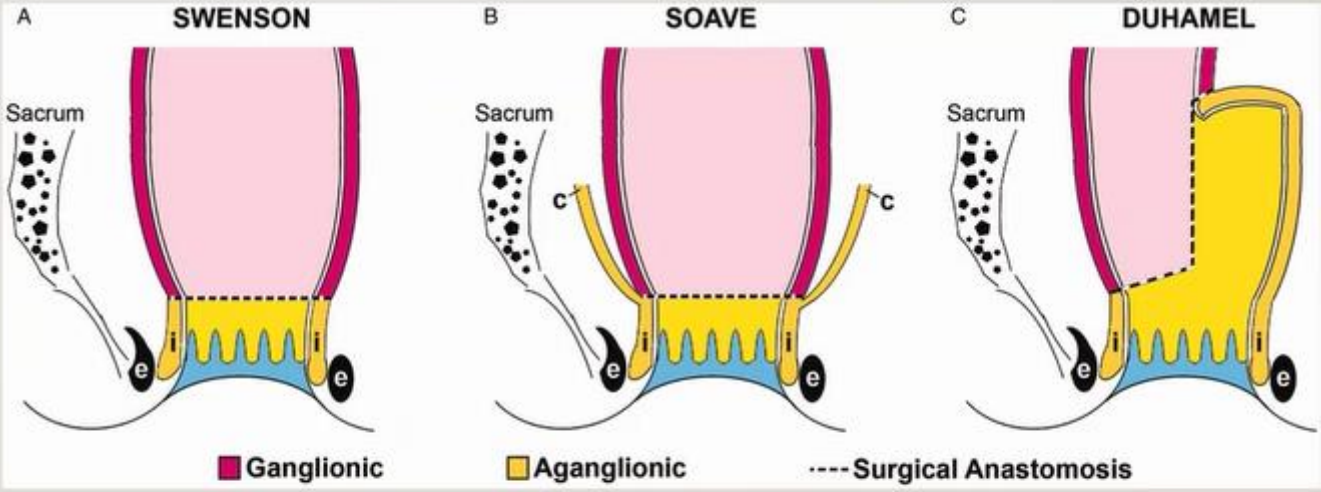
- Rectal washouts
- Colostomy (if unstable baby ,failure of washout , enterocolitis)
- Antibiotics (enterocolitis)
- **Definitive management :**

resect aganglionic segment & pull through a segment that has ganglionic cells

Pull-through procedure

- Swenson's pull-through
- Duhamel's retrorectal pull-through
- Soave's endorectal pull-through
- Trans-anal pull-through

→ Lap or open



don't memorize names only the principle

LONG TERM OUTCOME

- Obstructive symptoms (mechanical obstruction, recurrent or acquired aganglionosis, disordered motility in the residual colon or small bowel, internal sphincter achalasia, or functional megacolon caused by stool-holding behaviour)
- Fecal soiling (abnormal sphincter function, abnormal sensation, or pseudo-incontinence)
- Enterocolitis (Clostridium difficile or rotavirus and other . more common in younger children, longer segment disease, and trisomy 21)
- Hirschsprung associated inflammatory bowel disease (unknown etiology , Risk factors for this condition include long-segment disease and trisomy 21)

5- MECONIUM ILEUS

intraluminal obstruction , most commonly in ileocecal valve

- One of the most common causes of intestinal obstruction in the newborn (accounting for 9–33% of IO)
- Extremely viscid, protein-rich, inspissated meconium causing an intraluminal obstruction in the distal ileum(usually at the ileocecal valve)

- It is often the earliest clinical manifestation of cystic fibrosis (CF), occurring in approximately 16% of patients with CF
- CF ,AR disease due to mutations in the CF transmembrane regulator (CFTR) gene , located at chromosome 7q31

- most common mutation is F508del

DIAGNOSIS

- Antenatally :

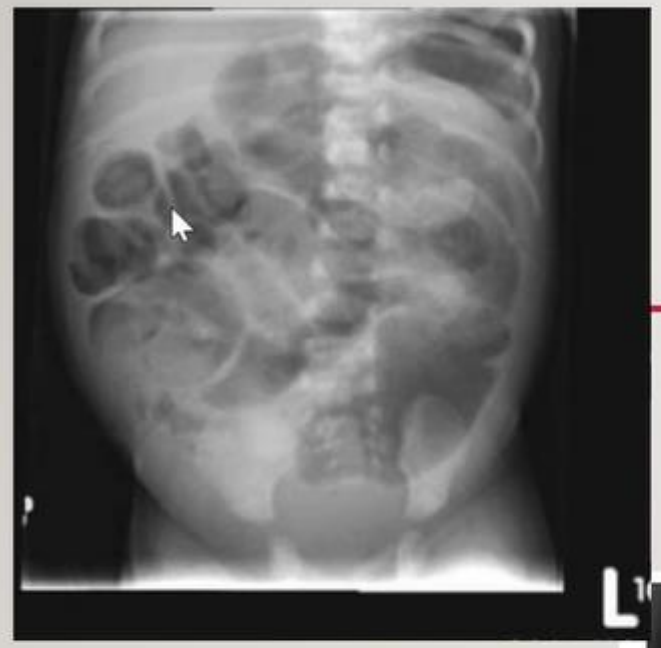
Fetal US : hyperechoic, intra-abdominal mass (inspissated meconium) , dilated bowel, and nonvisualization of the gallbladder

Presence of family history

CLINICAL PRESENTATION :

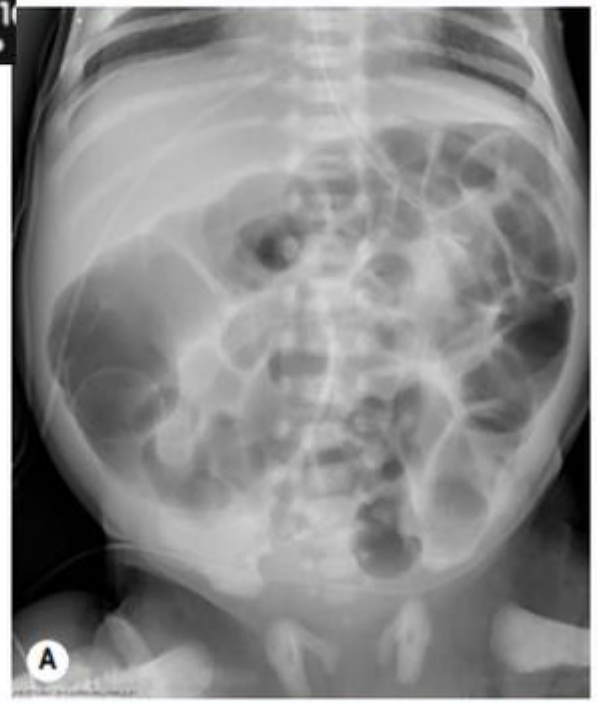
- Usually healthy immediately after birth. However, within 1–2 days, they develop abdominal distension and bilious emesis.
- Normal meconium will not be passed.
- Eventually, dilated loops of bowel become visible on exam and have a "doughy" character that indent on palpation.
- The rectum and anus are often narrow, a finding that may be misinterpreted as anal stenosis..

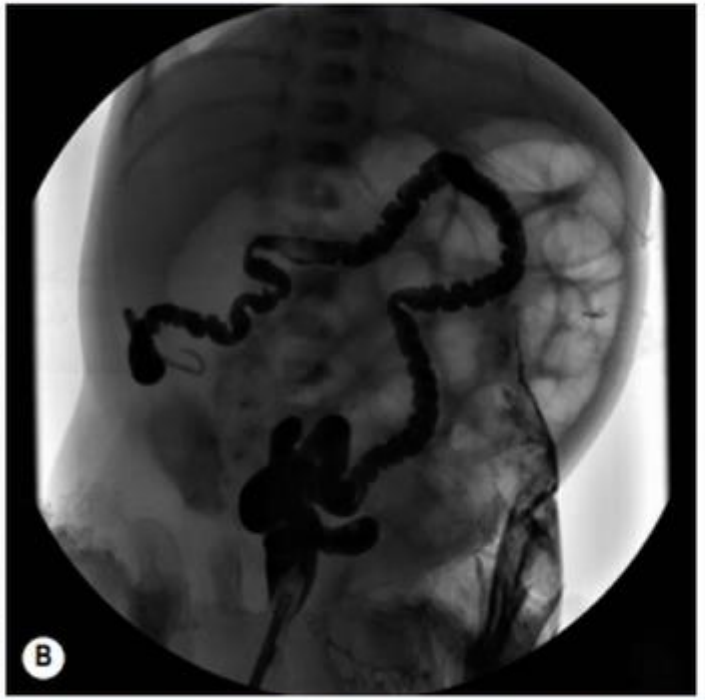
- Infants with complicated MI present with symptoms within 24 hours of birth or even immediately after birth as a result of in utero perforation or bowel compromise.
- Signs of peritonitis, including distension, tenderness, abdominal wall edema and erythema +/- of sepsis,
- A palpable mass suggests pseudocyst formation, which results from in utero bowel perforation

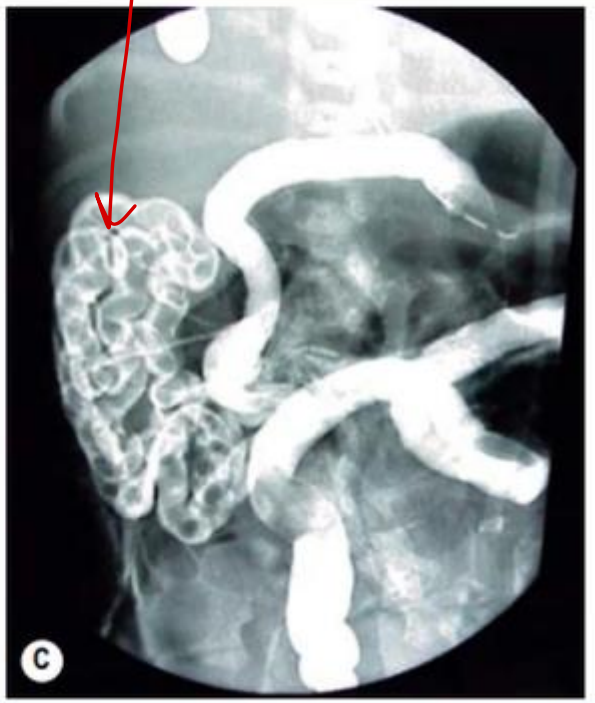


contrast enema:

filling defect suggestive of MI







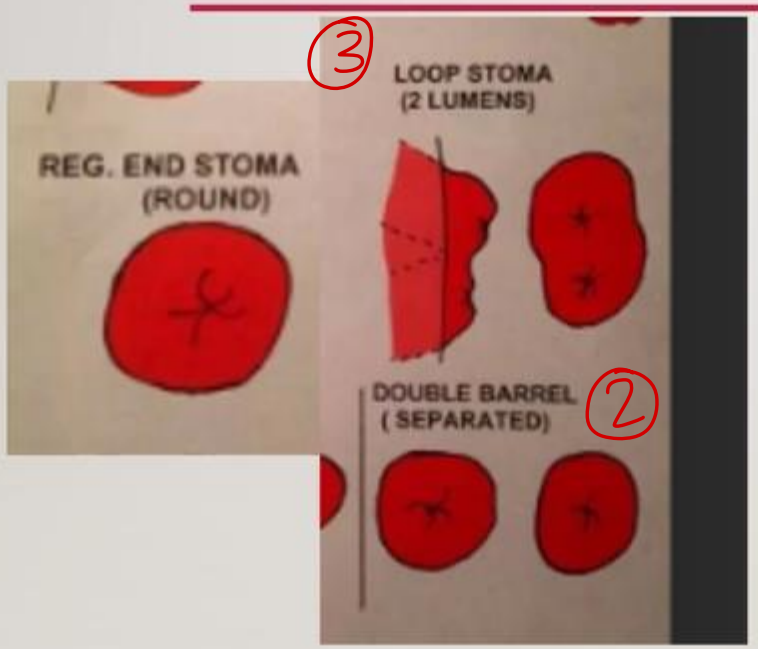
MANAGEMENT

- Conservative : simple MI
  - Water-soluble contrast enema , success rate ¬2/3 cases
    → breaks down the meconium
- Surgery:
  - **Simple MI: failed conservative**

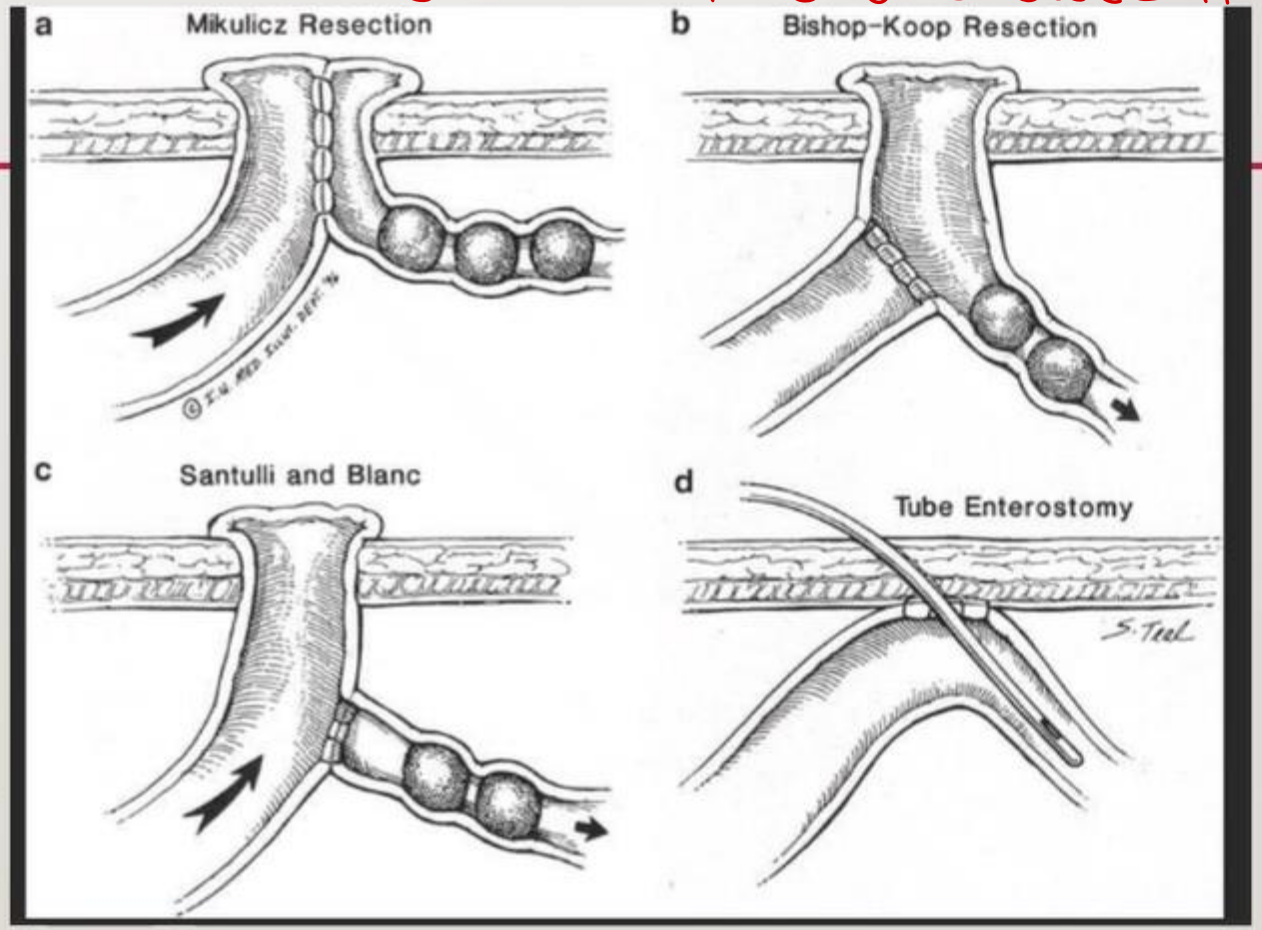
    Enterotomy and irrigation (N-acetylcysteine or normal saline) followed by enterotomy closure OR enterostomy tube Or ileostomy formation

  - **Complicated MI:** Resection of ischemic bowel + diverting stoma or primary anastomosis

These are what we do now



These are historical no one does it now



*confirm CF patient

Postoperative care:

- Nutrition
- N-acetylcysteine (10%) enterally (5–10 mL)
- Enteral pancreatic enzymes (e.g., Creon®, Pancrease®)
- Antibiotics
- Involvement of CF team (Resp/GI/Genecisit/social worker)

COMPLICATION OF MI AND CF

- Respiratory problems
- Gastroesophageal reflux
- Biliary tract disease
- Distal Intestinal Obstruction Syndrome (DIOS)
- Appendicitis
- Fibrosing colonopathy
- Intussusception : 1% of children with CF with the average age of onset of 9.5 years

6- Necrotizing Enterocolitis (NEC)

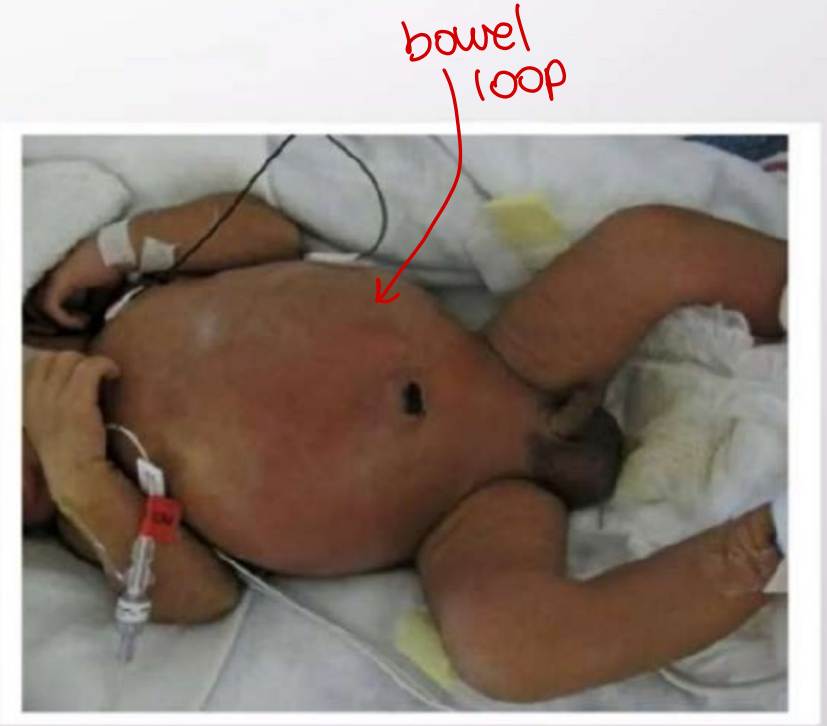
exclussive for neonate, mainly premature

- Disease of premature neonates.
- NEC affects about 10% of VLBW very low birth weight
- Incidence is inversely proportional to birth weight.
- The overall mortality of NEC probably approaches 30%
- Lower birth weight and younger gestational age correlate with higher risk of death.

Presentation

- NEC presents with feeding intolerance(vomiting or high gastric residuals), abdominal distention. Bleeding per rectum is a late sign .

- On exam : Abdominal distention
  - visible +/- palpable dilated bowel loops
  - skin discoloration (dusky)
  - signs of peritonitis



Diagnosis

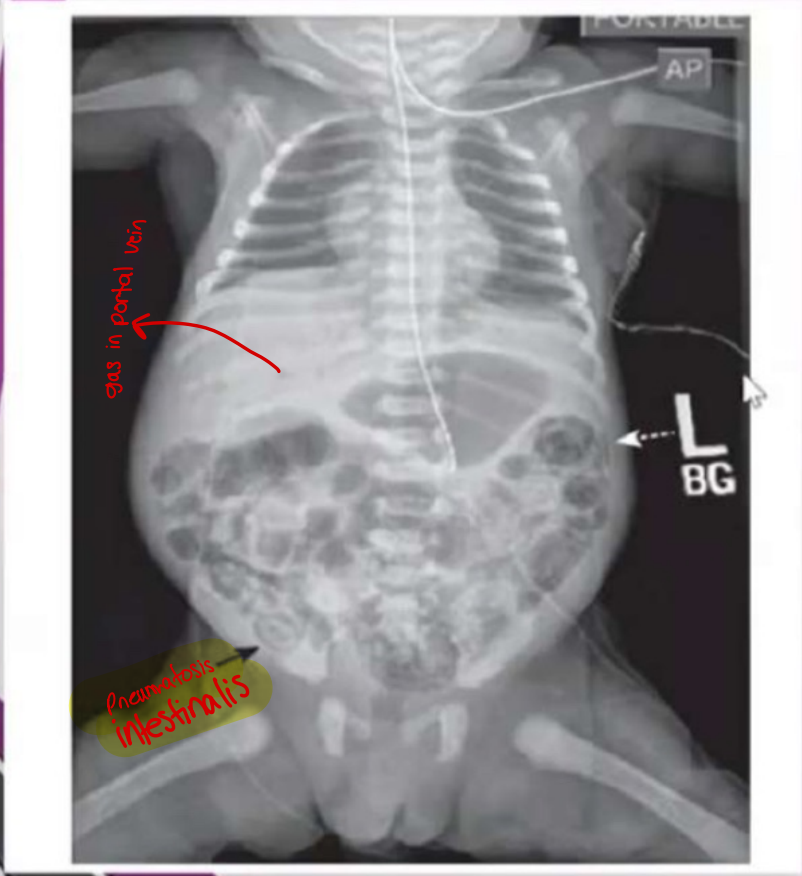
→high Suspicious

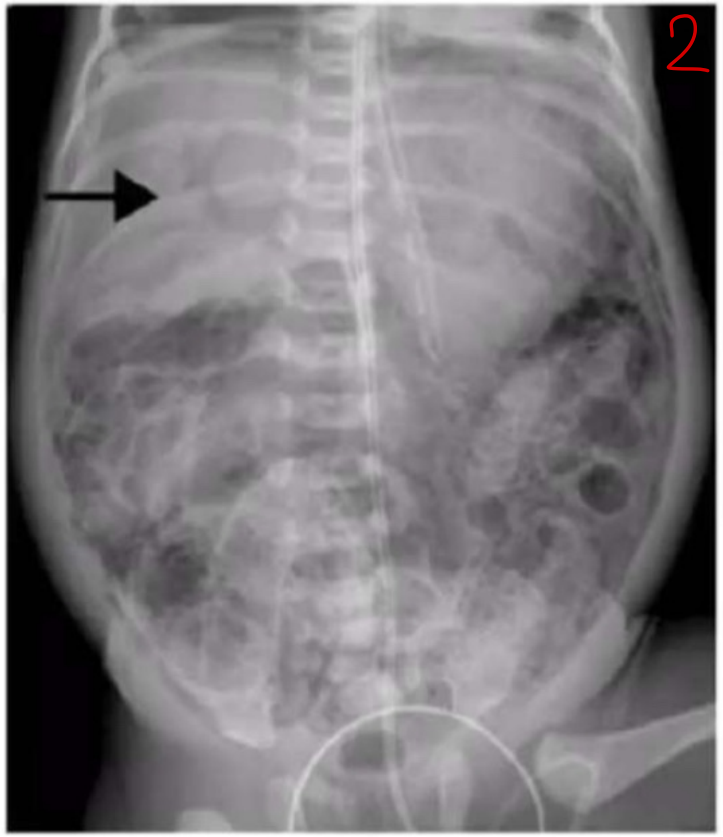
- Clinical and radiological
- History and physical examination
- Lab : High WBC, CRP , hyponatremia , thrombocytopenia , high lactate , metabolic acidosis .

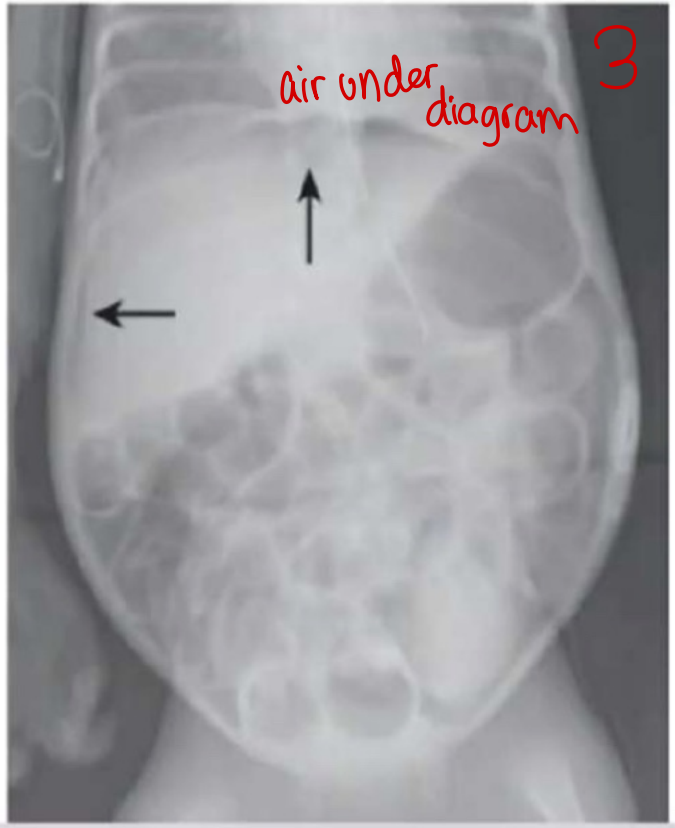
Table 33.1 Modified Bell Classification for NEC

	Clinical Findings	Radiographic Findings	Gastrointestinal Findings
Stage I	Apnea, bradycardia, and temperature instability	Normal gas pattern or mild ileus	Mild abdominal distention, stool occult blood, gastric residuals
Stage IIA	Apnea, bradycardia, and temperature instability	Ileus with dilated bowel loops and focal pneumatosis	Moderate abdominal distention, hematochezia, absent bowel sounds
Stage IIB	Metabolic acidosis and thrombocytopenia	Widespread pneumatosis, portal venous gas, ascites	Abdominal tenderness and edema
Stage IIIA	Mixed acidosis, coagulopathy, hypotension, oliguria	Moderate to severely dilated bowel loops, ascites, no free air	Abdominal wall edema, erythema, and induration
Stage IIIB	Shock, worsening vital signs and laboratory values	Pneumoperitoneum	Bowel perforation

- **Abd Xray** :Pneumatosis intestinalis is the classic radiographic finding in NEC. + air under diaphragm
- US

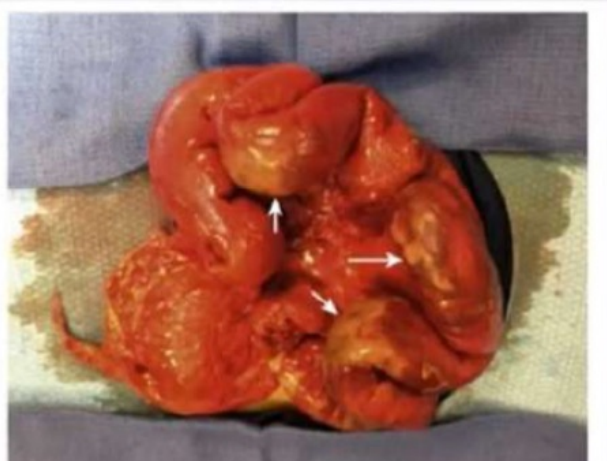






Management

- primary management is supportive (bowel rest, gastric decompression, IVF,IV antibiotic and parenteral nutrition, cardiopulmonary support if needed)
- Surgery is absolute indication in cases with pneumoperitoneum
- Surgical options : Laparotomy with resection /anastomosis laparotomy with resection/ stoma formation or peritoneal drainage (VLBW)



Outcome

- Recurrence 10 %
- Mortality ¬30%(inversely proportional to birth weight and gestational age) Medical NEC carries a mortality of 20%, surgical NEC mortality is 35- 50%)
- Intestinal failure :NEC is the leading cause of pediatric intestinal failure (IF) resulting in more than 1/3 of IF patients
- Stoma complications
- Intestinal stricture
- Neurodevelopmental delay :intellectual delays,moderate-to-severe developmental delay with speech and motor impairment


THANK YOU