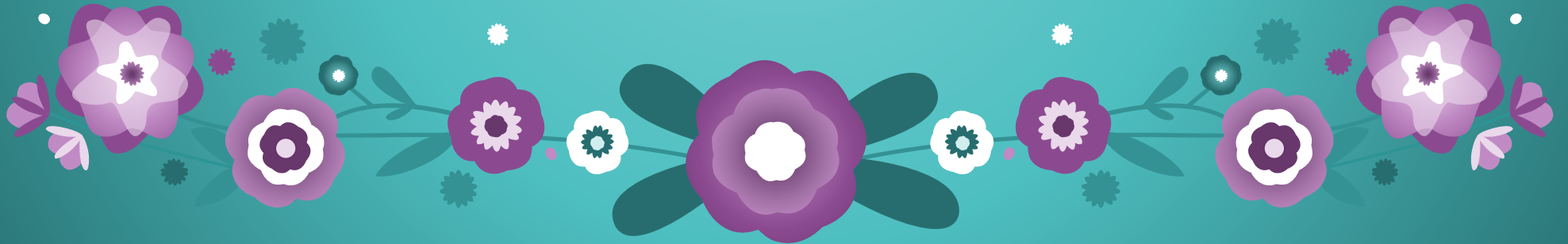


# Intestinal Obstruction in Children

By Raed Al-Taher, M.D.



# How to classify?

Degree
<ul style="list-style-type: none"><li>• Partial</li><li>• Complete</li></ul>

Age
<ul style="list-style-type: none"><li>• Neonatal</li><li>• Infantile</li><li>• Older children</li></ul>

Organ
<ul style="list-style-type: none"><li>• Duodenum</li><li>• Jejunum</li><li>• Ileum</li><li>• Colon</li><li>• Anus</li></ul>

Site
<ul style="list-style-type: none"><li>• External</li><li>• Mural</li><li>• Intraluminal</li></ul>

Cause
<ul style="list-style-type: none"><li>• Mechanical</li><li>• Inflammatory</li><li>• Ischemic</li><li>• Metabolic</li></ul>

*Don't memorize this slide*

# Age

- **Malrotation** – Intestinal obstruction by Ladd band
- Malrotation – Mid-gut volvulus
- **Intestinal atresia**
- **Necrotizing enterocolitis**
- **Meconium ileus**
- Meconium plug
- **Hirschsprung disease**
- **Anorectal malformations**
- Inguinal hernia

Neonatal



- Malrotation – Intestinal obstruction by Ladd band
- Hirschsprung disease
- **Hypertrophic pyloric stenosis**
- **Intussusception**
- Inguinal hernia
- Constipation

Infantile



- Inguinal hernia
- Habitual constipation
- Postoperative adhesions
- Peritonitis
- Hirschsprung disease
- Peritonitis
- Meckel diverticulum (band/ -itis/ intussusception)
- FB ingestion
- Intussusception

Older children



\* Disorders in RED will be discussed in this lecture.

*Don't memorize this slide*

# Site

## External

- Postoperative adhesions
- Meckel diverticulum (vitelline duct/band)
- Abdominal masses/tumors
- Herniation
- **Malrotation** – Intestinal obstruction by Ladd band
- Malrotation – Mid-gut volvulus

## Mural

- **Intussusception**
- **Hypertrophic pyloric stenosis**
- **Hirschsprung disease**
- **Intestinal atresia**
- Malrotation – Mid-gut volvulus
- **Necrotizing enterocolitis**
- **Anorectal malformations**
- Peritonitis
- Meckel diverticulum (diverticulitis/intussusception)
- Paralytic ileus (postoperative, narcotics, electrolyte disturbances)









## Intraluminal

- Constipation
- **Meconium ileus**
- Meconium plug
- FB ingestion (bezoars)

\* Disorders in RED will be discussed in this lecture.

*Don't memorize this slide*

# Summary

	Short definition	M.C. Presentation	Best Dx & specific signs	Types	Best Tx
 <p><b>Intestinal atresia</b></p>	Congenital atretic segment of (small or large) bowel	Newborn with bilious vomiting + abdominal distension	<ul style="list-style-type: none"> <li>Clinical</li> <li>AXR (double bubble sign for duodenal atresia)</li> </ul>	I, II, IIIa, IIIb, & IV	<p><b>Surgery:</b></p> <ul style="list-style-type: none"> <li>Resection + anastomosis</li> <li>Duodeno-duodenostomy without resection for duodenal atresia</li> </ul>
 <p><b>Malrotation</b></p>	Congenital abnormal rotation of (small & large) bowel, results in short mesentery & midgut volvulus	Neonate with bilious vomiting	<ul style="list-style-type: none"> <li>Clinical</li> <li>Upper GI contrast study</li> </ul>	-	<p><b>Emergent surgery:</b></p> <ul style="list-style-type: none"> <li>Ladd's procedure</li> </ul>
 <p><b>MI</b></p>	Congenital thick meconium obstructing distal small bowel	Newborn with bilious vomiting + abdominal distension + delayed passage of meconium	<ul style="list-style-type: none"> <li>Clinical</li> <li>Contrast enema (Neuhauser sign)</li> <li>Testing for CF</li> </ul>	<ul style="list-style-type: none"> <li>Simple</li> <li>Complicated</li> </ul>	<p><b>Conservative:</b></p> <ul style="list-style-type: none"> <li>Rectal washouts</li> <li>Pancreatic enzymes</li> </ul> <p>+/- Ileostomy for irrigation +/- Surgery for complications</p>
 <p><b>NEC</b></p>	Acquired (small &/or large) bowel hypoperfusion and necrosis	Premature neonate in sepsis + bilious vomiting + abdominal distension +/- bloody stool	<ul style="list-style-type: none"> <li>Clinical</li> <li>AXR (pneumatosis)</li> </ul>	Bell's staging: I. Suspected II. Definite III. Advanced	<p><b>Conservative:</b></p> <ul style="list-style-type: none"> <li>NPO + NGT + IVF + ABx</li> </ul> <p>+/- Surgery for complications + Ileostomy</p>
 <p><b>HD</b></p>	Congenital aganglionosis of distal large bowel	Newborn with bilious vomiting + tense abdominal distension + delayed passage of meconium + gush of liquid stool on DRE	<ul style="list-style-type: none"> <li>Clinical</li> <li>Contrast enema (transitional zone)</li> <li>Rectal biopsy</li> </ul>	<ul style="list-style-type: none"> <li>Ultrashort segment</li> <li>Short segment (recto-sigmoid)</li> <li>Long segment (includes total colonic &amp; total GI)</li> </ul>	<p><b>Surgery:</b></p> <ul style="list-style-type: none"> <li>+/- Colostomy</li> <li>Pull-through</li> </ul>
 <p><b>ARM</b></p>	Congenital absence/abnormality of the anus	Newborn with abnormal anus +/- meconium via urethra or vestibule	<ul style="list-style-type: none"> <li>PEx (perineal inspection)</li> <li>+/- Cross-table x-ray</li> <li>+/- Colostography</li> </ul>	<ul style="list-style-type: none"> <li>High/low</li> <li>With/without fistula</li> </ul>	<p><b>Surgery:</b></p> <ul style="list-style-type: none"> <li>+/- Colostomy</li> <li>Anorectoplasty</li> </ul>
 <p><b>HPS</b></p>	Acquired hypertrophy of pyloric muscle	2-8 weeks term neonate with progressive non-bilious projectile vomiting + hypochloremic hypokalemic metabolic alkalosis	<ul style="list-style-type: none"> <li>Clinical (olive sign)</li> <li>Abdominal US</li> </ul>	-	<p><b>Surgery:</b></p> <ul style="list-style-type: none"> <li>Pyloromyotomy</li> </ul>
 <p><b>Intussusception</b></p>	Acquired invagination of proximal bowel into distal bowel (small &/or large)	3mo-3yrs infant/toddler with abdominal colics +/- vomiting +/- currant jelly stool + Hx of recent URTI or GE	<ul style="list-style-type: none"> <li>Clinical (Dance sign)</li> <li>Abdominal US (target or donut sign, &amp; pseudokidney sign)</li> </ul>	<ul style="list-style-type: none"> <li>Primary (idiopathic)</li> <li>Secondary</li> </ul>	<p><b>Conservative:</b></p> <ul style="list-style-type: none"> <li>Reduction enema</li> </ul> <p>+/- Surgical reduction (if enema failed or C/I)</p>

# Intestinal Atresia (Congenital intestinal obstruction)

- 1 in 3,000 | black > white | jejunoileal
- 1 in 5,000 | F>M (slight) | duodenal
- 1 in 50,000 | colonic

**\* Jejunioileal > duodenal > colonic**

*Don't memorize this slide*

# Intestinal Atresia

## Pathology *(mostly unknown)*

- Theory:

Intrauterine vascular insult to intestinal segment after being completely developed

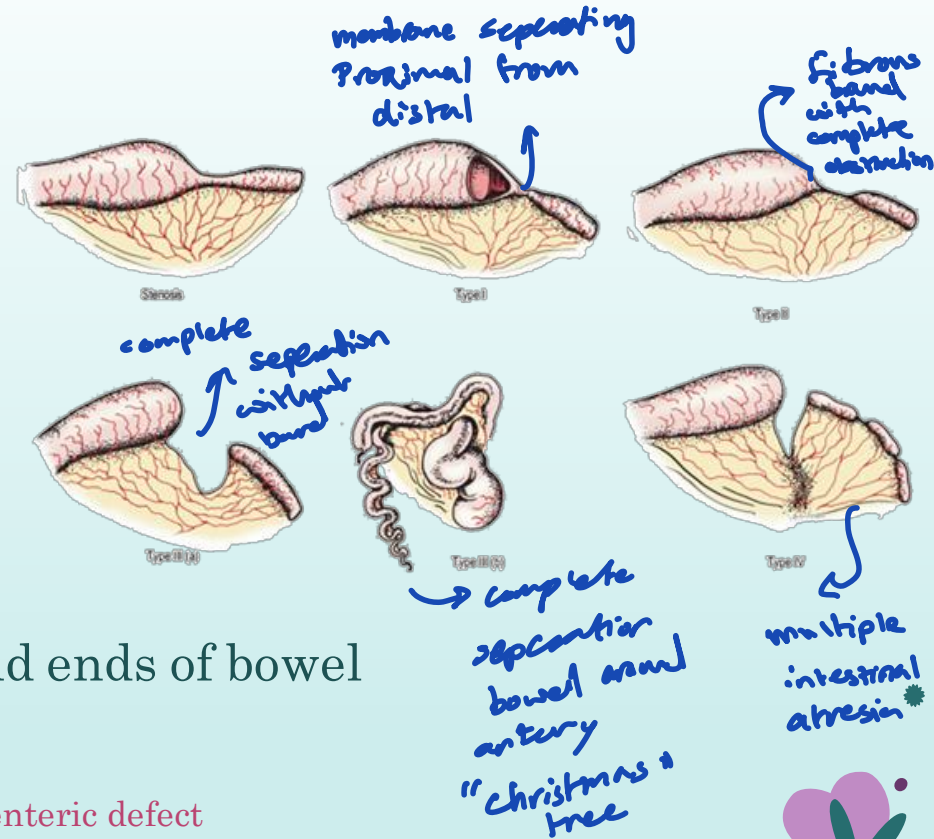


Meconium can be found in distal bowel beyond the site of obstruction

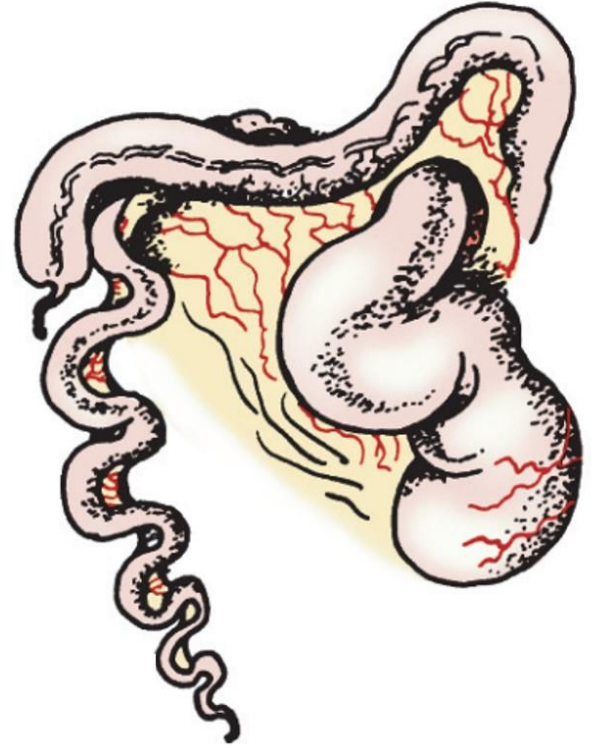
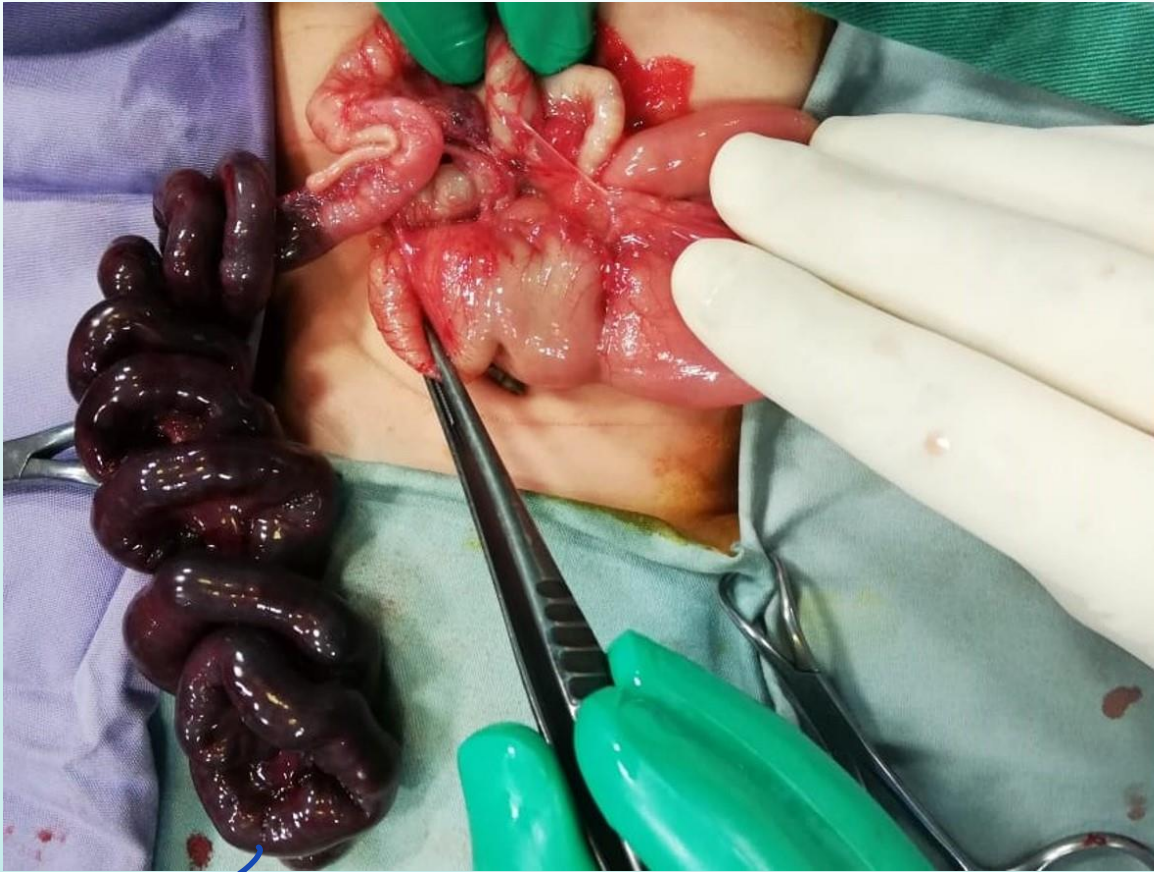
# Intestinal Atresia

## 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)
- Type IV – Multiple atresia (“string of sausages” appearance)







no mesentery (mesentery defect)

Jejunal atresia – type IIIb  
“Apple-peel” atresia complicated by necrosis.

# Intestinal Atresia

## Clinical Features

### • Antenatal:

#### • US may show:

- polyhydramnios (↑ with the more proximal atresias).
- “double bubble”
- dilated proximal loops
- echogenic bowel (distal)

### • Postnatally:

- Bile-vomiting (ampulla of Vater is included)
- Varying degrees of distension (depending on level of obstruction)

# Intestinal Atresia

## Investigations

- AXR

- Features of obstruction *(nothing specific)*



- **“Double bubble” and no distal gas** (classical feature in duodenal obstruction)

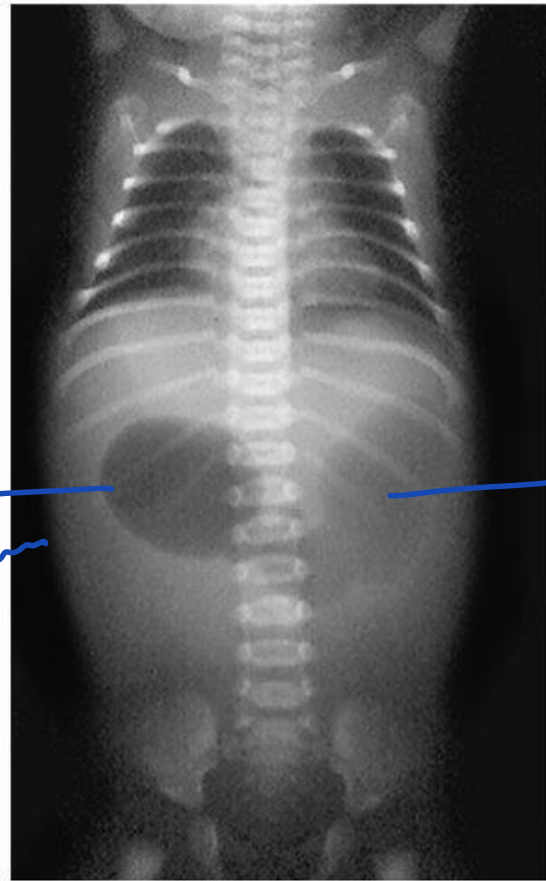
- **Peritoneal calcification:** suggests perforation and meconium cyst formation

- Contrast enema

- may show **microcolon**

- helps ruling out Hirschsprung disease or meconium plug

part of  
the duodenum



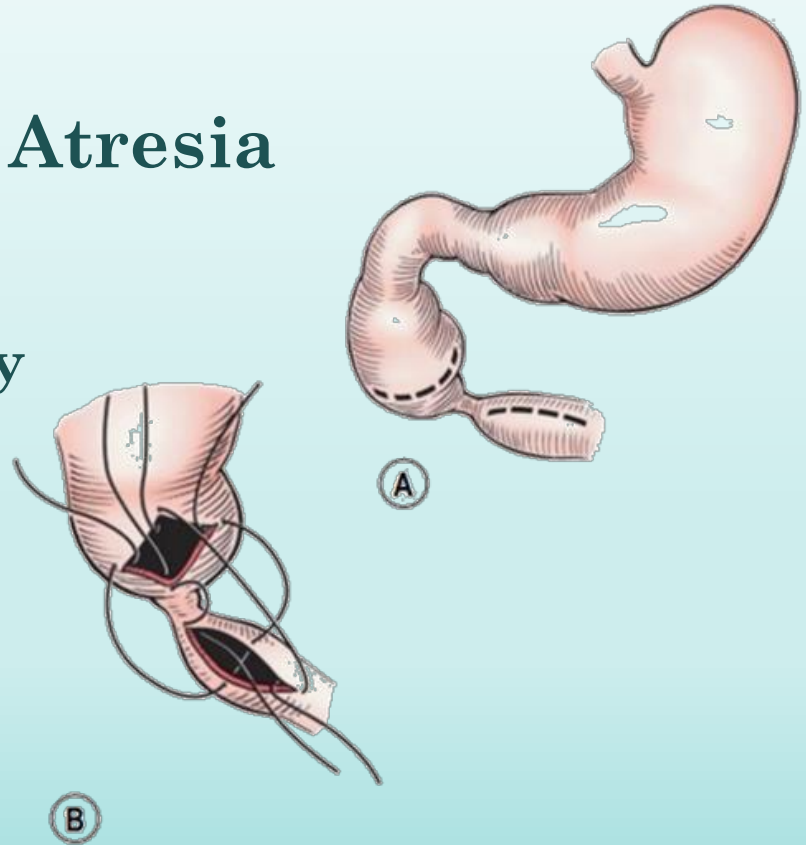
→ stomach

“**Double bubble**” and no distal gas  
(classical feature in duodenal atresia)

# Intestinal Atresia

## Surgery for Duodenal Atresia

- Duodeno-duodenostomy
- without any resection



# Intestinal Atresia

## Surgery for Jejunio-ileal Atresia

### ➔ Resection & re-anastomosis

1. Assess viability and length of residual bowel
2. Assess patency of distal bowel lumen
3. Resection of atretic segment(s) and re-anastomosis

# Intestinal Atresia

## Surgery for Colon Atresia

- Resection of atretic segment + either one of these:
  - Primary anastomosis
  - Colostomy creation and staged anastomosis

# Malrotation

A spectrum of anomalies of  
rotation and fixation of the intestines

(principally the midgut)



# Malrotation

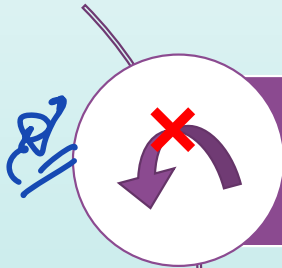
Associated with:

- Gastroschisis and omphalocele
- Diaphragmatic hernia
- Duodenal atresia and biliary atresia
- Intussusception (Waugh's syndrome)
- Dysmotility and pseudo-obstruction syndromes

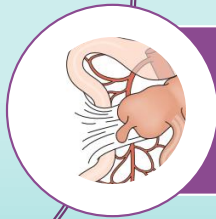
*Don't memorize this slide*

# Malrotation

- The commonest variant:



Failure of final 90° anticlockwise rotation taking the cecum from RUQ to RLQ



Cecum is fixed to retroperitoneum by peritoneal bands running anteriorly to the 2<sup>nd</sup> part of duodenum (Ladd's bands).

*cecum stays  
in the RUQ*



# Malrotation

- The key pathology is:

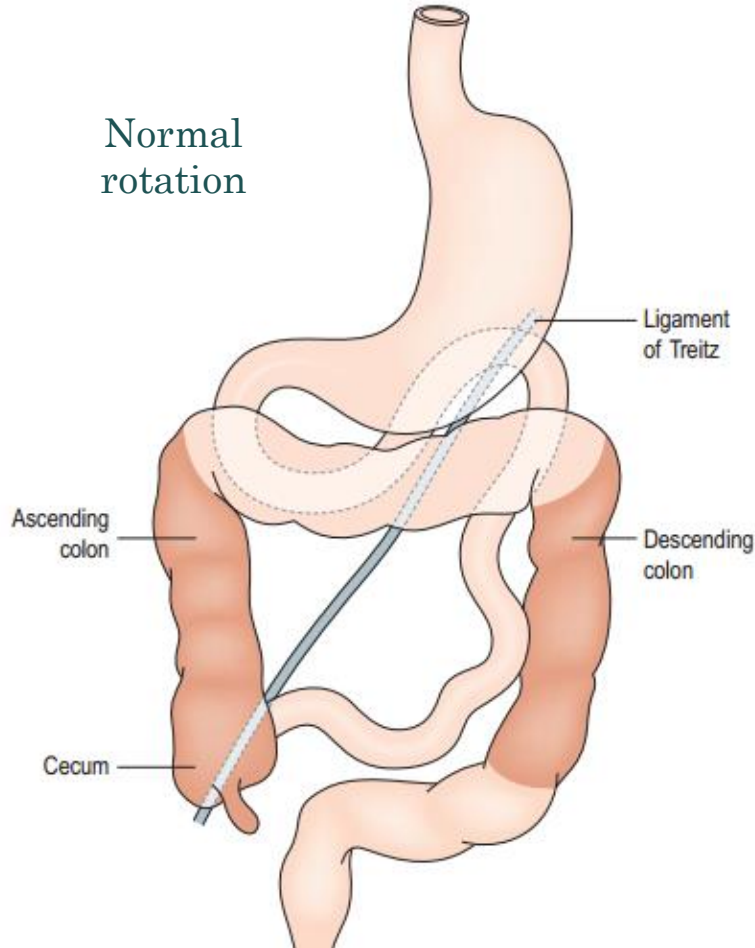
- The distance between the two ends of the small bowel mesentery

(between DJ junction and IC valve)

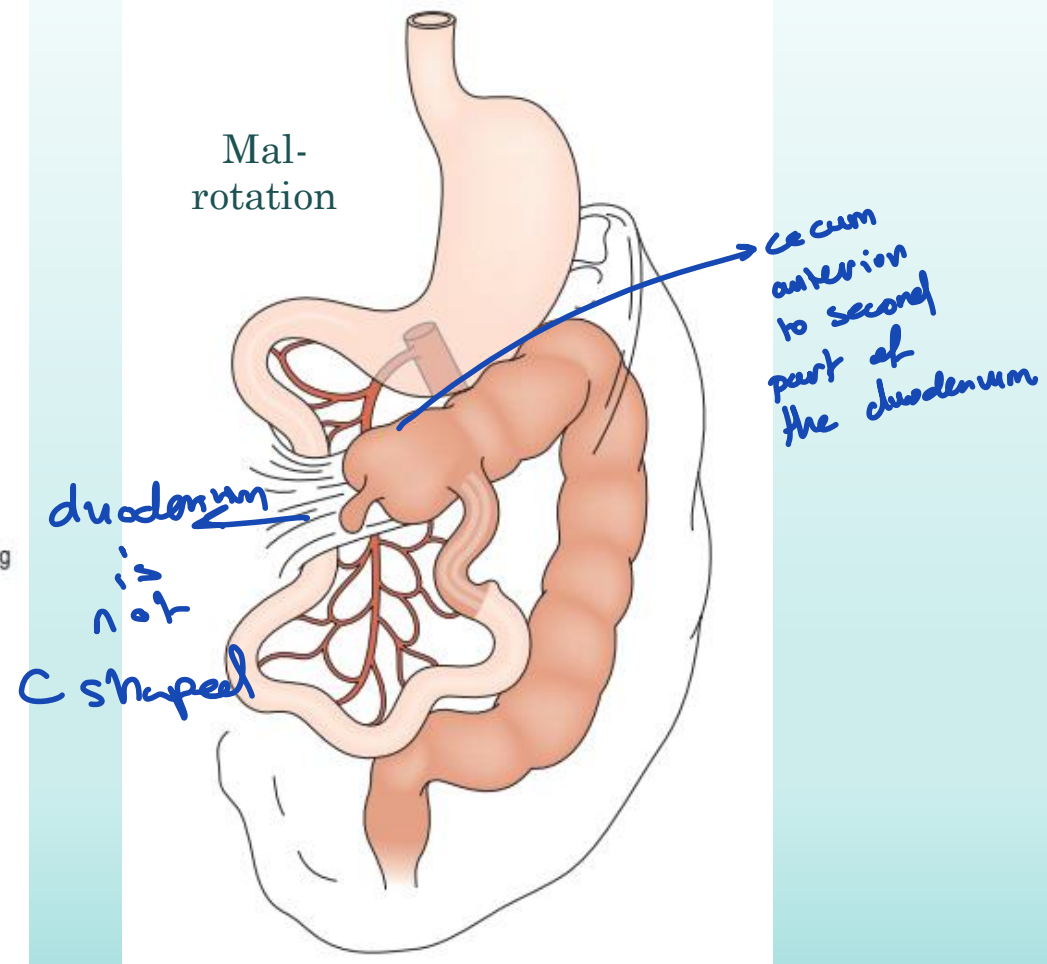
→ when diminished

→ **↑ risk of volvulus**

## Normal rotation



## Mal-rotation



# Malrotation

## Clinical Features

- Can present at any age.
- Classic presentation:

Early:

- bile vomiting *caused by obstruction either from Ladd's bands or the twist of the volvulus*

Late:

- ✓ intermittent or acute abdominal pain
- ✓ diarrhea then constipation
- ✓ failure to thrive
- ✓ passage of altered blood *(bloody stool or bloody meconium)*

*not specific* ←

# Malrotation

## Clinical Features

On physical examination:

- Malrotation without volvulus:
  - Abdomen is soft and non-tender
- Malrotation with volvulus:  $\pm$  bowel strangulation
  - Abdomen is rigid and tender
  - Upper abdominal distension
  - Blood-stained stools

# Malrotation

## Clinical Features

- Chronic midgut volvulus (<10% of cases)
  - mesenteric thickening
  - lymphatic obstruction leading to chylous ascites

# Malrotation

## Investigations

### 1. AXR

- “Normal” in most cases.

- Abnormal features:

- ✓ Malposition of the bowel (“small bowel” to the right and “colon” to the left)
- ✓ Lack of distal bowel gas (“gasless” abdomen or a “double bubble” appearance)
- ✓ “Whirled” appearance of mid-abdominal bowel
- ✓ Thick-walled, tubular bowel loops (suggesting chronic volvulus)

→ not accurate sign

could be: duodenal  
malrotation  
stenosis

- ### 2. Upper GI contrast study
- will show either the obstruction or the state of the bowel

- investigation of choice (if time permits!)

gi contrast study



# Contrast studies in malrotation

	Normal features	Abnormal features
Overall	“C” shaped duodenum	Redundant duodenum, right-sided jejunal loops
Lateral appearance	Overlapping, posterior position of second and fourth parts	Anterior position of fourth part
Duodeno-jejunal flexure	Left of spinal pedicles; rising to at least level of L1/L2 disk space	Right-sided or centrally placed. Failure of ascent to level of pylorus

*Don't memorize this slide*

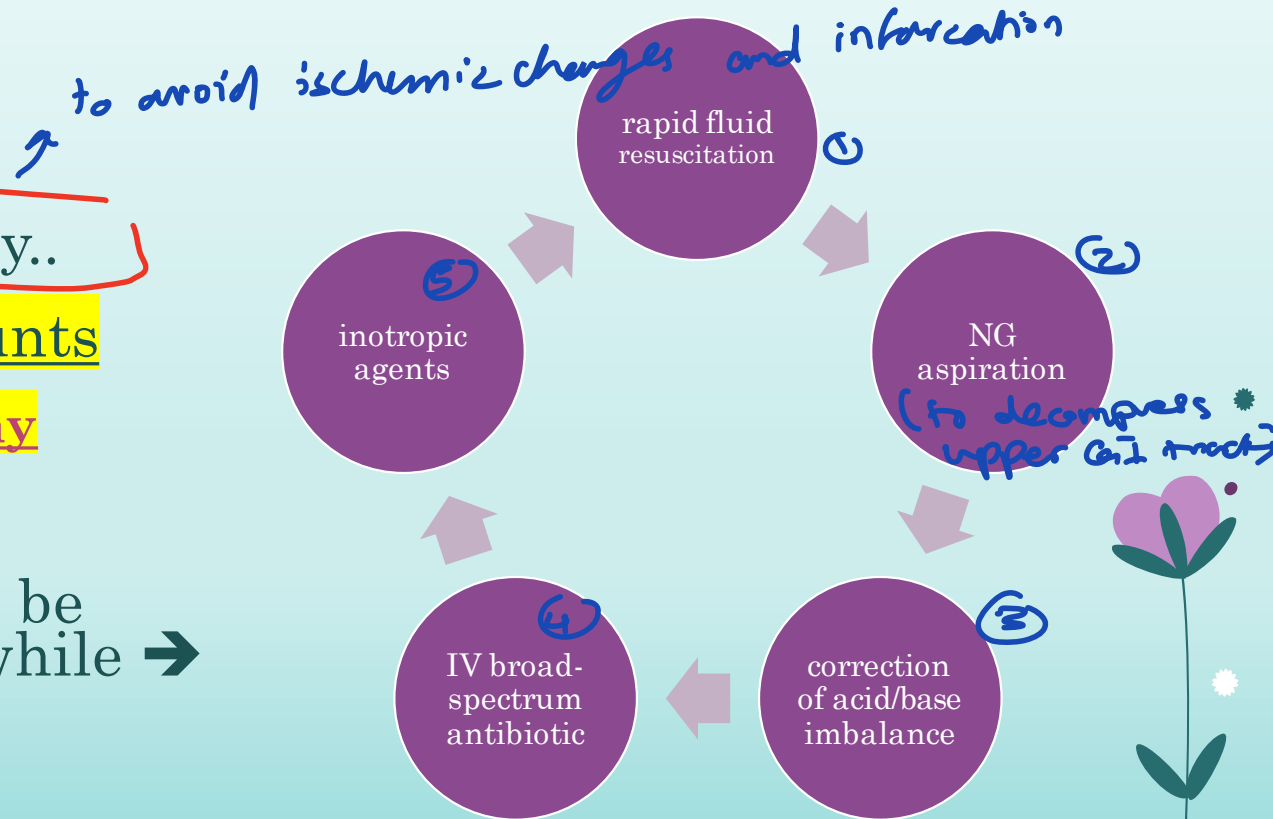
# Malrotation Management

- If volvulus is likely..

- every minute counts

- Urgent laparotomy

- Other elements to be considered meanwhile →



# Malrotation

## Management: Surgery (Ladd's Procedure)

1. Division of Ladd's bands
2. Widening of mesenteric base
3. Position bowel (small bowel right and large bowel left)
4. ± Appendectomy *if left will deftate in horizontal position so it's better to be removed*

# Meconium Ileus (MI)

Distal ileal intraluminal obstruction due to presence of abnormally thick meconium.

~90% of infants with MI will have cystic fibrosis

# Meconium Ileus

## Investigation

### 1. AXR:

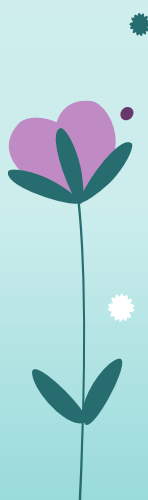
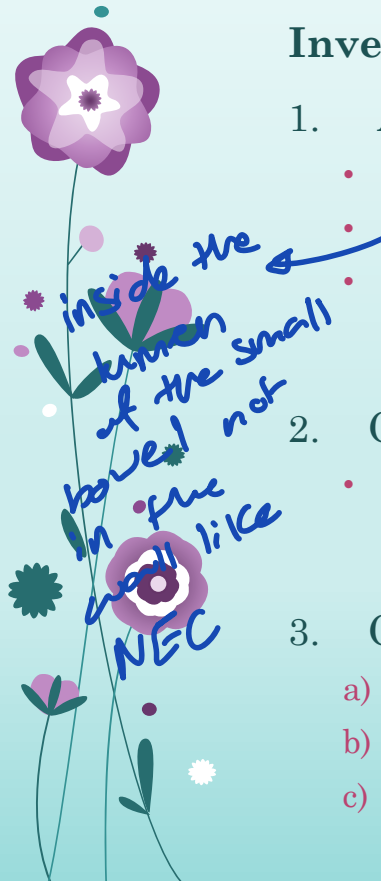
- Dilated proximal bowel loops
- “Soap-bubble” appearance or “Neuhauser’s sign” (in the loops filled with meconium)
- Calcification (in meconium peritonitis)

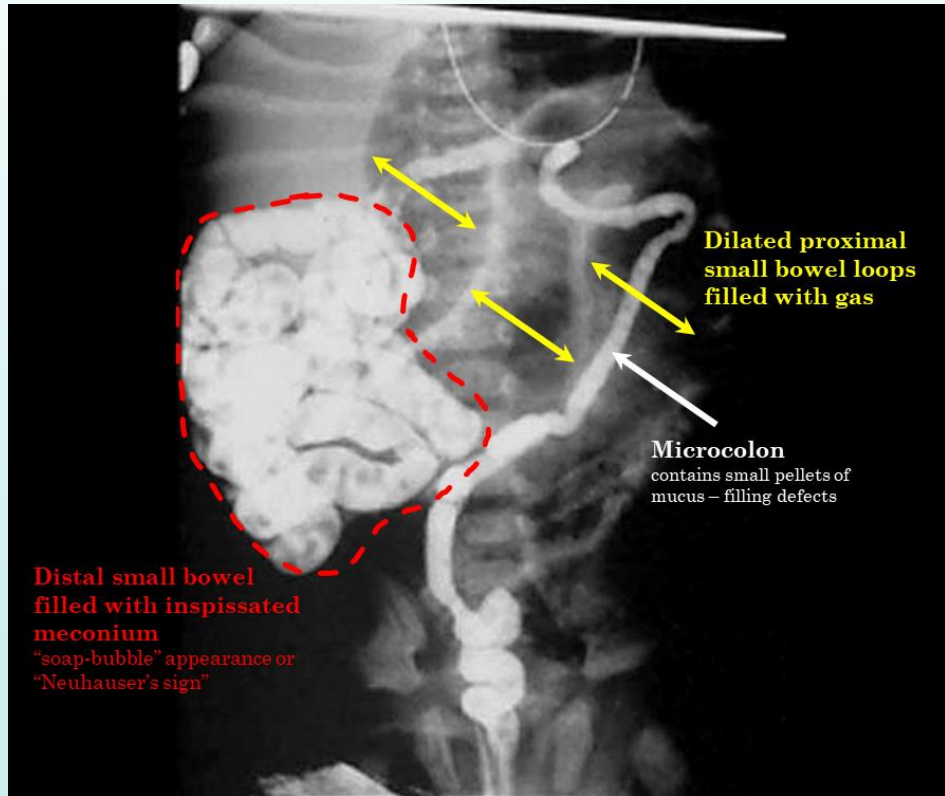
### 2. Contrast enema

- **Microcolon** (contain small pellets of mucus)

### 3. Confirmation of CF:

- a) **Sweat-Chloride test** (normal  $<40$ , diagnostic  $\geq 60$  mmol/L)
- b) **Gene mutation analysis**
- c) **Immunoreactive trypsinogen** (basis for screening -  $\uparrow\uparrow$  levels in CF)





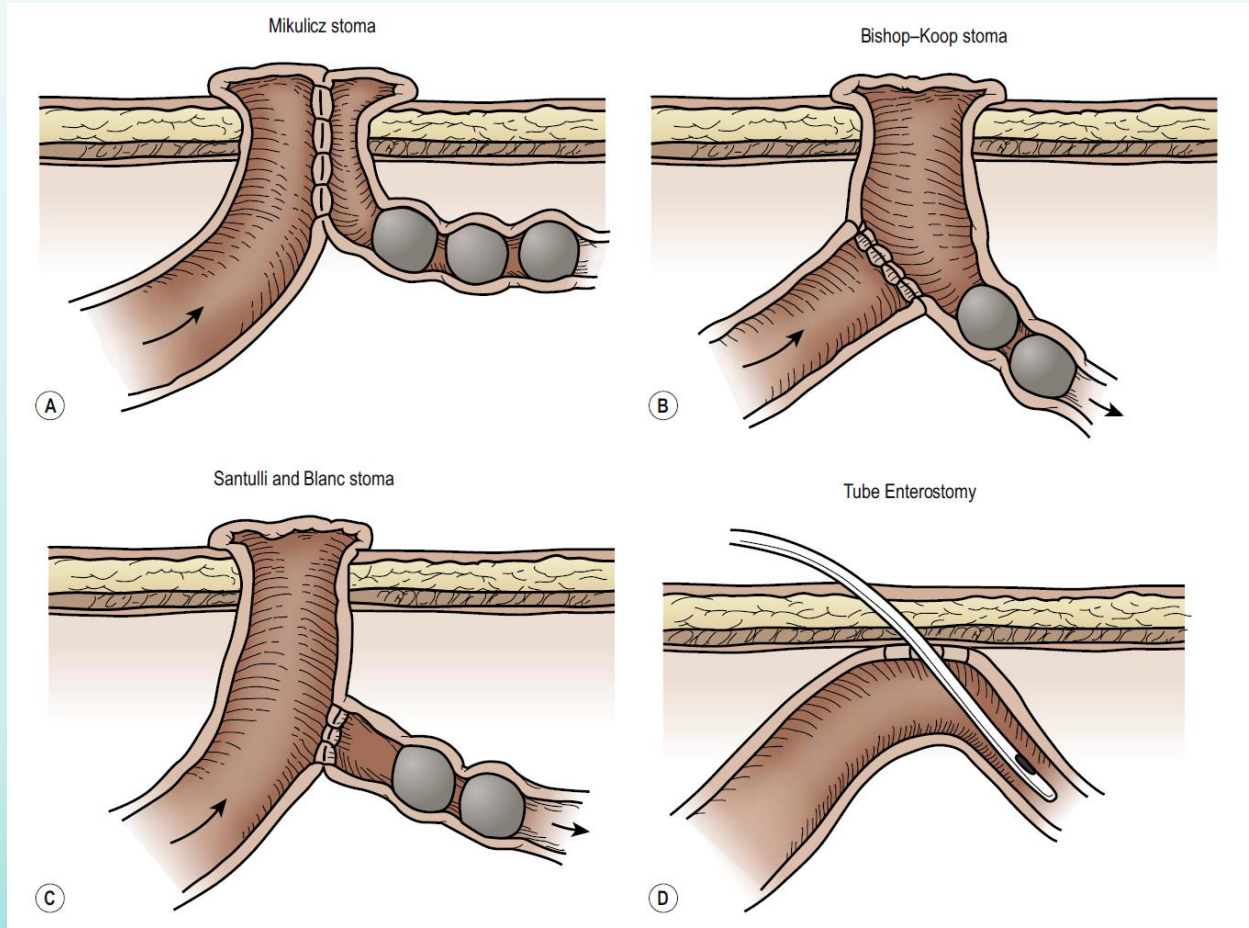
**Contrast Enema of two MI cases**

# Meconium Ileus

## Management

- Conservative:
  - Water-soluble contrast enema (success 60–70% in simple MI)
- Surgery:
  - **Simple MI:** ileotomy and irrigation using N-acetylcysteine or normal saline, followed by either:
    - Simple closure and return
    - Enterostomy tube
    - Ileostomy creation (Double-barrelled Mikulicz, Bishop-Koop, or Santulli)
  - **Complicated MI:** Resection of ischemic bowel + diverting stoma or primary anastomosis

# Variations in ileostomy for meconium ileus



*Don't memorize this slide*



# Meconium Ileus

## Management

Postoperative care:

- Parenteral nutrition
- N-acetylcysteine (10%) enterally (5–10 mL)
- Enteral pancreatic enzymes (e.g., Creon®, Pancrease®)
- Antibiotics
- Involvement of CF team

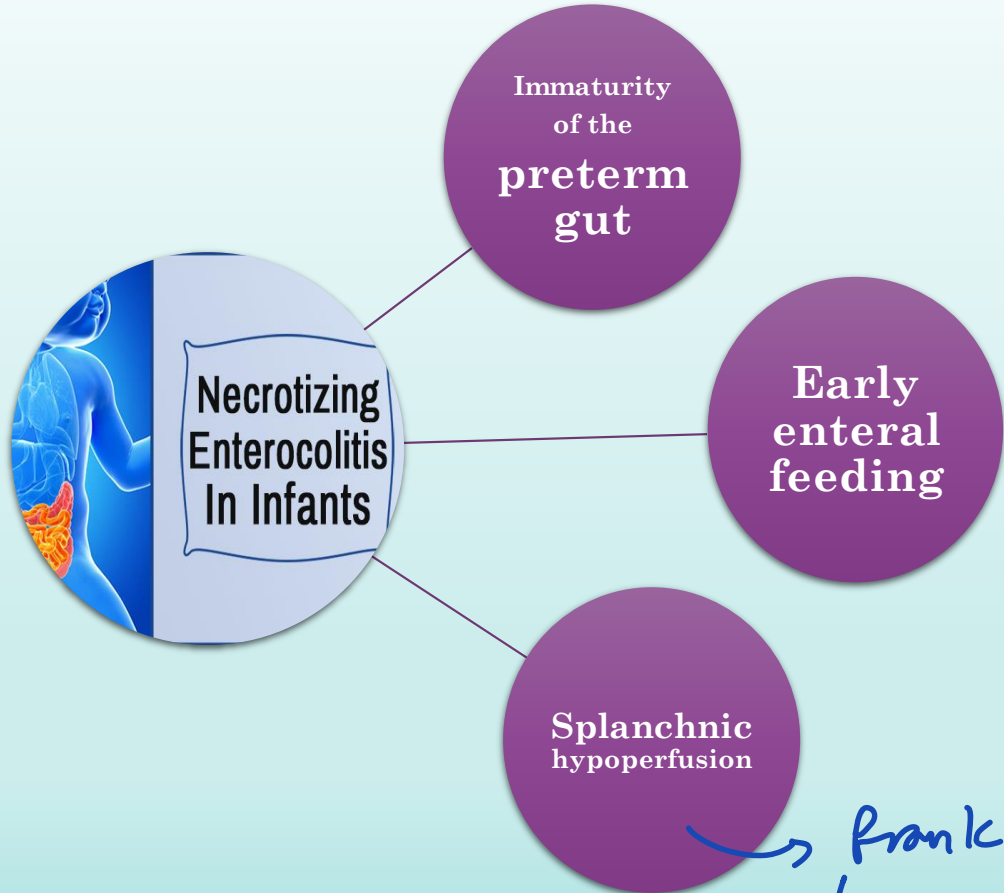
# Necrotizing Enterocolitis (NEC)

- 1–3/1,000 live-births | M:F = 2:1 | Black > white
  - **>90% occur in preterm infants**
  - **Term infants (10%)** – associated with congenital heart disease or birth asphyxia
- ✓ **↓ incidence in breast-fed infants**

# Necrotizing Enterocolitis

## Pathophysiology

- Multifactorial; a unique response in the immature, neonatal gut to “stress”
- NEC may be:
  - Focal → small segment
  - Diffuse → all small + large bowel
- Most commonly affected sites:
  - Terminal ileum
  - Colon



Possible causal factors in NEC

frank sepsis  
(away shunting from  
mesenteric vessels)

*Important slide*

# Necrotizing Enterocolitis

## Pathophysiology

- Submucosal and subserosal gas-filled cysts  
(N<sub>2</sub> or H<sub>2</sub> from gas-forming bacteria)
- Histologically:
  - ✓ Mucosal ulceration
  - ✓ Transmural coagulative necrosis
  - Microthrombi in mesenteric vessels

# Necrotizing Enterocolitis

## Clinical Features

- **Nonspecific signs**

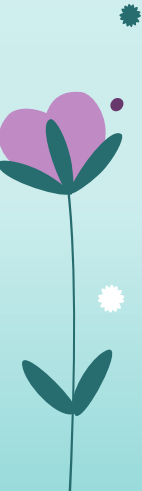
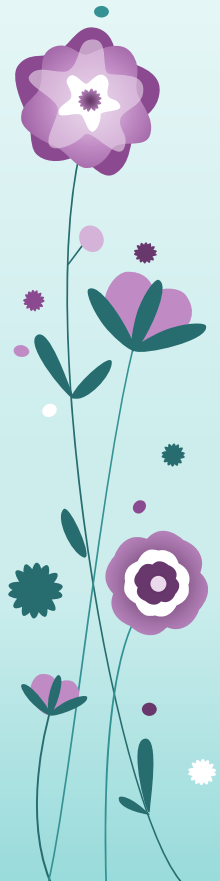
related to sepsis and ischemia

- Tachycardia
- Hypotension
- Metabolic acidosis
- Unstable body temperature
- Increasing O<sub>2</sub> requirement
- Thrombocytopenia
- Coagulopathy

- **Specific local signs**

related to the affected bowel loops

- Peritonism
- Abdominal wall erythema
- Bile-vomiting
- GI bleeding
- Abdominal mass formation





Abdominal wall erythema

↳ severe inflammation inside.  
\* Preterm baby think of Necrotizing enterocolitis

# Bell's Staging of NEC

	Clinical	Radiology
I – suspected	Irritable, apnea, bile-aspirates, abdo. Distension, +ve fecal occult blood	Intestinal distension
II – definite	GI bleeding ± Abdo. mass	Pneumatosis, “fixed” loops, portal venous gas
III – advanced	Shock, marked GI bleeding	Pneumoperitoneum, severe ascites

↓  
surgical  
intervention  
needed



# Necrotizing Enterocolitis

## Investigations

1. Blood work-up for signs of sepsis
  - a) CBC
  - b) Coagulation screen (INR, fibrinogen, D-dimers)
  - c) Lactate and ABGs

# Necrotizing Enterocolitis

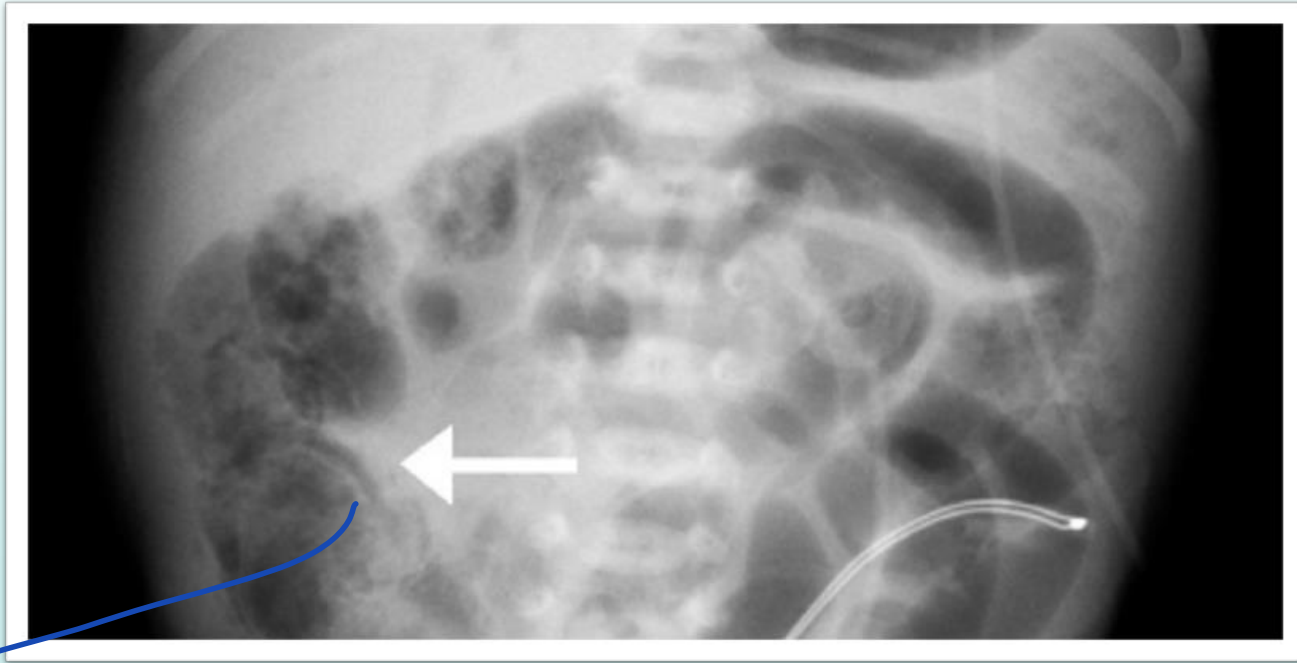
## Investigations

### 2. AXR (AP $\pm$ lateral)

- a) Early signs – nonspecific distension
- b) Late signs:



- **Pneumatosis** (linear radiolucent bands/bubbles parallel to the bowel wall)
- **Portal venous gas**
- **Extravisceral free air** (air under diaphragm or “football sign”)



band of  
gas

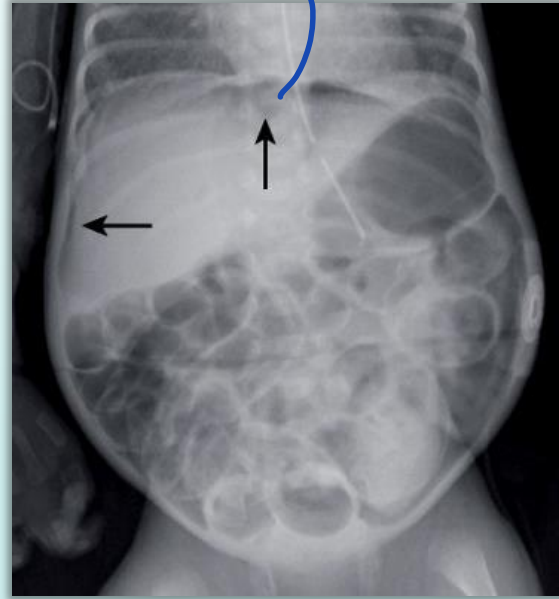
### **Pneumatosis Intestinalis**

seen as linear radiolucent bands parallel to the wall of the bowel or “soap-bubble” appearance

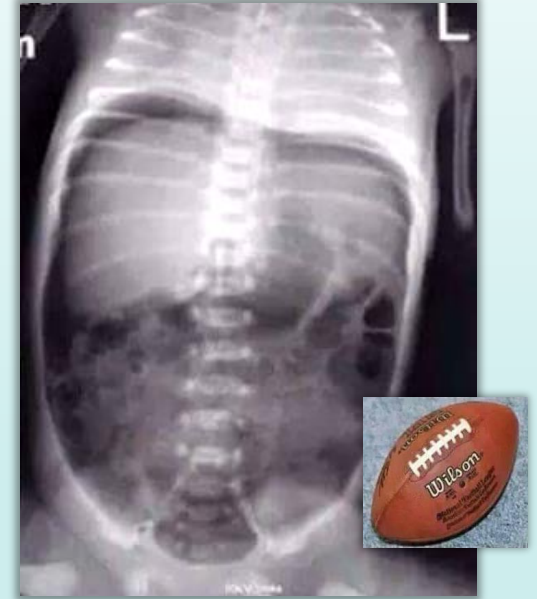
black shadows especially around  
the hepatic margins



Portal venous gas



Pneumoperitoneum  
(Extravisceral free air)



Pneumoperitoneum  
(Football sign)\*

\*From <https://meducation.net>

# Necrotizing Enterocolitis

## Management

- Supportive:
  - Resting the gut
  - Optimizing hemodynamic and metabolic conditions

Nil by mouth  
(NPO)  
And  
NG tube

Restoration of..

- fluid losses (3<sup>rd</sup> space)
- hematocrit
- temperature instability
- glucose instability
- acid-base imbalance

Appropriate  
oxygenation

Broad-  
spectrum  
antibiotics

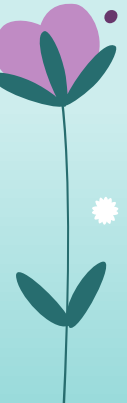
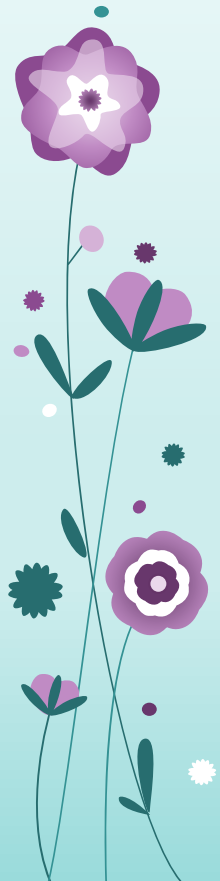
*perforation of  
NEC*

# Necrotizing Enterocolitis

## Management

- **Serial radiography** to assess onset of complications (as perforation)
- If improving.. continue for 7–10 days before restarting enteral nutrition

*serial abdominal x ray either every day or other day*



# Necrotizing Enterocolitis

## Management

### Indications of Surgery:

- Pneumoperitoneum (perforation) (Stage III Bell's)
- Failure to progress (after 24 hr of full supportive management)
- Obstructive features (↑ distension, ↑ bile-aspirates)
- “Fixed-loop” on serial imaging
- ↑ Abdominal wall erythema
- Palpable abdominal mass

# Necrotizing Enterocolitis

## Management

### Surgery options:

- Resection + ileostomy creation
- “Clip and drop” technique (for pan-intestinal disease)  
+ 24-hour second-look laparotomy
- Primary peritoneal drainage (PPD) (if unstable)






Part of surgical gloves  
used as a drain for open  
peritoneal drainage

**Primary peritoneal drainage (PPD)**  
in an ELBW newborn



# Necrotizing Enterocolitis

## Outcome and Complications

- **Mortality 20-50%** (higher in VLBW and ELBW)
  - Short-gut syndrome (~25%)
  - Strictures (20%)
- 

# Necrotizing Enterocolitis

Possible Prevention Strategies (little evidence yet)

1. Breast milk (fourfold reduction in NEC incidence vs. formula)
2. Avoidance of indomethacin/ibuprofen/ranitidine and maintenance of gastric acidity
3. Probiotics (e.g., lactobacillus/bifidobacterium)
4. Oral antibiotics
5. Oral/IV immunoglobulin (IgA/IgG)
6. Amino-acid supplementation (glutamine)

*Don't memorize this slide*

# Hirschsprung's Disease

## Embryology

- Migration of neuroenteric cells from the neural crest to GI tract, reach:

1. Esophagus - 5<sup>th</sup> week
2. Mid-gut - 7<sup>th</sup> week
3. Distal colon - 12<sup>th</sup> week

→ most affected (recto sigmoid)

## Anatomy

- The normal intestine contains two distinct nerve plexi:

1. Submucosal plexus (of Meissner)
2. Myenteric or intermuscular plexus (of Auerbach)

→ between two muscular layers

# Hirschsprung's Disease

## Genetics

- Risk in siblings is 3–4% (↑ with long-segment disease)
- Gene mutation (50% familial and 15–35% isolated)
  - **RET gene** (Ch 10q11 | associated with Down's syndrome)
  - Other genes (SOX10, EDNRB, GDNF , EDN3, ECE1, NTN, SIP1).

## Pathology

- Lack of progression of peristaltic wave into the aganglionic segment of intestine
- Absent or abnormal internal anal sphincter relaxation

# Hirschsprung's Disease

## *Associated anomalies* (variable incidence ~10%)

- **Down's syndrome (~5%)**
- Neurocristopathies as:
  - *Waardenburg-Shah* syndrome; white forelock, bicolored iris, deafness
  - *Hypoventilation syndrome* (Ondine's curse) – association with HD termed Haddad syndrome
- Mental retardation syndromes
  - *Smith-Lemli-Optiz* syndrome; mental retardation, polydactyl, defect in cholesterol metabolism
  - *Mowat-Wilson syndrome*; mental retardation, characteristic facies
- Development colon anomalies
  - Colon atresia, anorectal atresia
- Miscellaneous
  - *Kaufman-McKusick syndrome*; hydrometrocolpos, hypospadias, polydactyl

[N.B. MEN type 2B (Marfanoid habitus, medullary thyroid cancer, café au lait spots, mucosal neuroma) is associated with *hyperganglionosis* (functionally similar to HD)]

Don't memorize this slide

# Hirschsprung's Disease

## Variable Affected Segment

- Short segment (recto-sigmoid) (75%)
- Long segment:
  - Total colonic +/- ileal involvement
  - Total intestinal aganglionosis (incompatible with life)
- Ultra-short segment disease (rare)
- Segmental disease or “skip” lesions (extremely rare)

# Hirschsprung's Disease

## Clinical Features

Two overlapping scenarios

1. Neonatal bowel obstruction:

- ✓ • Delayed passage of meconium
- ✓ • Abdominal distension
- ✓ • Bile vomiting
- ✓ • ± Enterocolitis

2. Chronic constipation (no encopresis/soiling)

- ± Enterocolitis
- Failure to thrive
- Explosive discharge of liquid fecal matter after DRE



# Hirschsprung's Disease

## Investigations

1. AXR (non-specific signs of intestinal obstruction)

2. Contrast enema

- dilated proximal colon
- small-caliber distal colon
- transitional zone
- retained contrast on a 24-hour film

3. Rectal biopsy (suction or open under GA)

- 1, 2, and 3 cm above dentate line

Characteristic HP features:

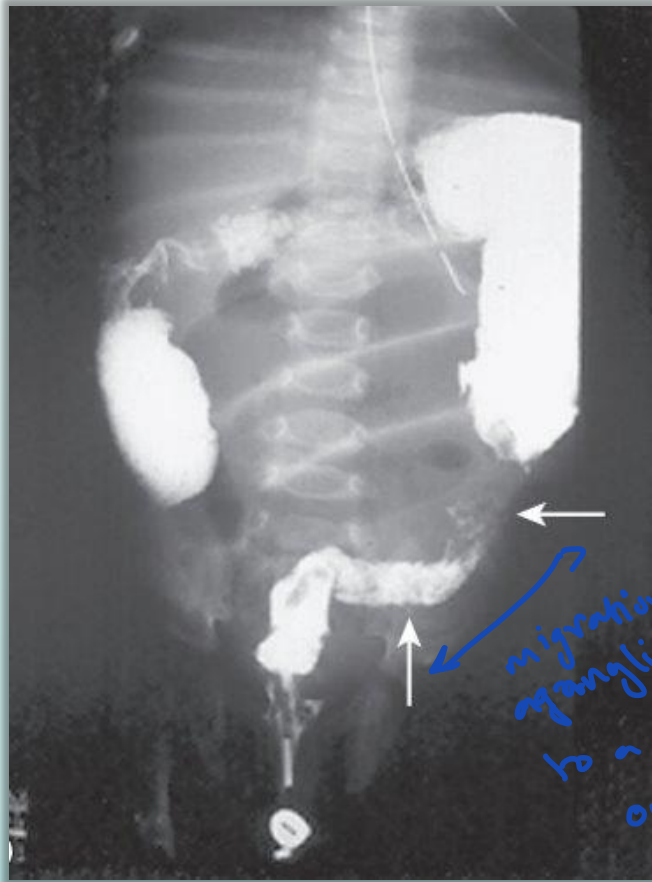
- ✓ Absence of ganglion cells
- ✓ Hypertrophied nerve bundles
- ✓ Acetyl cholinesterase staining
- ✓ Absence of Calretinin staining → most common method now
- ✓ Immunohistochemistry (e.g., LDH, S100, SDH, etc.)

4. Anorectal manometry (absence of Recto-Anal Inhibitory Reflex)

↳ not needed in most cases, only in query cases

↳ bigger children

neurology



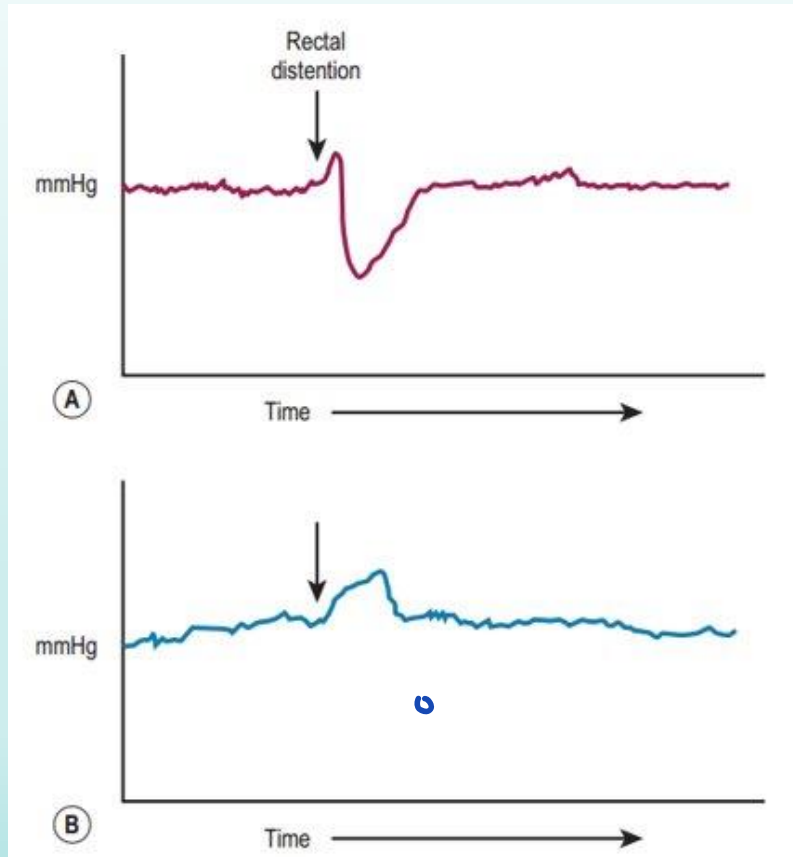
### Contrast Enema

showing dilated proximal segment and distal narrow segment with transitional zone in between



### Contrast Enema

showing narrow colon, suggesting "Total Colonic HD"



## Anorectal Manometry

A. Presence of Recto-Anal Inhibitory Reflex → Normal

B. Absence of Recto-Anal Inhibitory Reflex → HD

# Hirschsprung's Disease

## Management

- Daily rectal washouts by parents
- Colostomy (if unstable)
- Pull-through procedure (resection of aganglionic segment, bringing the ganglionic bowel through the pelvis and anastomosing it to the anus)
  - Swenson's pull-through
  - Duhamel's retrorectal pull-through
  - Soave's endorectal pull-through
  - Laparoscopic-assisted pull-through
  - Trans-anal pull-through

# Hirschsprung's Disease

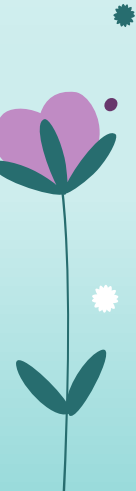
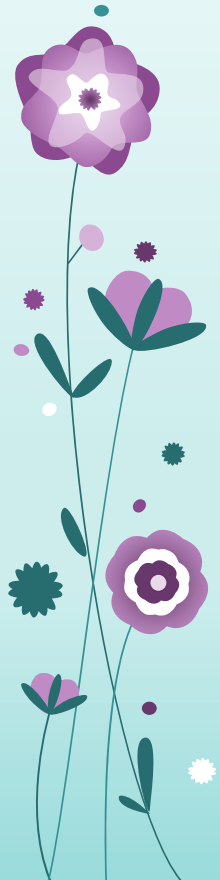
## HD Associated Enterocolitis (HAEC)

= HD + sepsis, fever, diarrhea, +/- bloody stool

- Active intervention is required:
  - **Rectal washouts** (10–20 mL/kg of normal saline x3 daily)
  - **Antibiotics** (e.g., vancomycin, metronidazole)
  - **± Colostomy**

# Anorectal Malformations

The terminal part of the hindgut  
is abnormally placed



# Anorectal Malformations

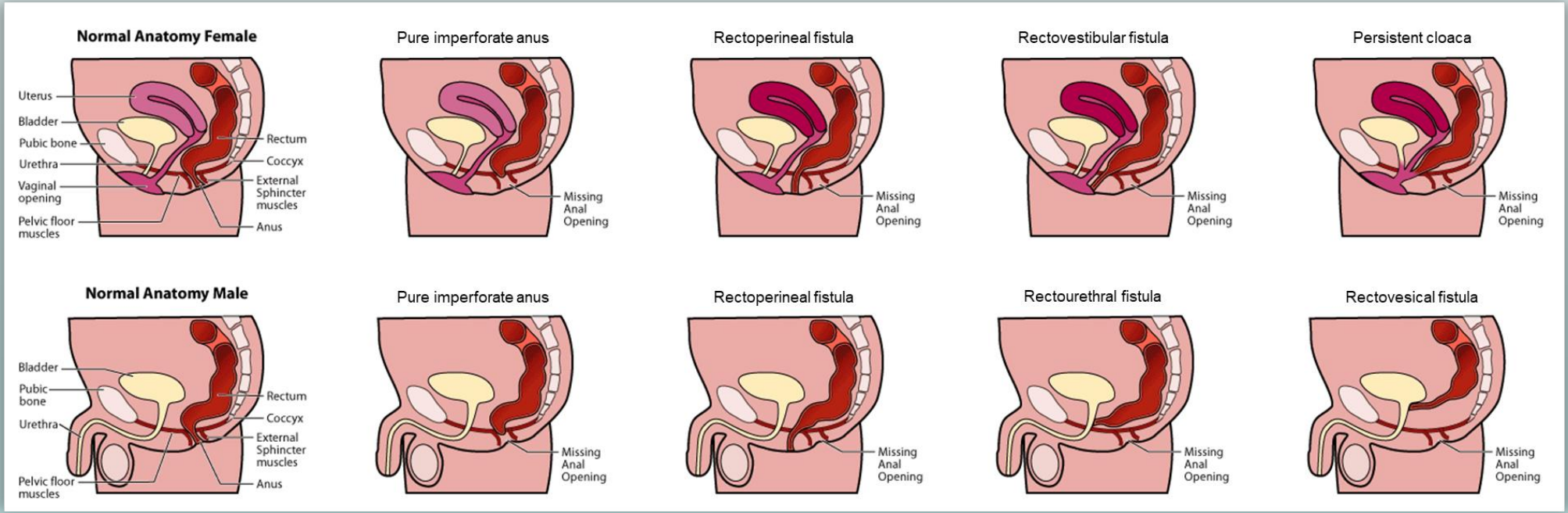
- Incidence ~1 in 5,000
- Male>female (60:40)
- Present as imperforate anus:
  - with fistula (majority) (connection between distal rectum and genitourinary tract)
  - without fistula (5%) (associated with Down's syndrome)

# Anorectal Malformations

## Anatomical classification

Males	Females
Anteriorly displaced anus	Anteriorly displaced anus
Recto-perineal fistula	Recto-perineal fistula
Recto-bulbar urethral fistula (most common)	Recto-vestibular fistula (most common)
Recto-membranous urethral fistula	
Recto-prostatic urethral fistula	Recto-vaginal fistula
Recto-bladder neck fistula	
Imperforate anus without fistula	Imperforate anus without fistula
Rectal atresia (normal anal opening)	Rectal atresia (normal anal opening)
-	Persistent cloaca





\* Source: <http://surgery.ucsf.edu/conditions--procedures/anorectal-malformation.aspx>

## Clinical Features

- <sup>physical</sup> Clinical examination is the most important part  
(makes the Dx in 90% of cases)



“Bucket-handle” deformity, suggesting a low type ARM



Imperforate anus with flat perineum, suggesting a high type ARM



Single UG orifice with no anus suggesting a Persistent cloaca\*

# Anorectal Malformations

## 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

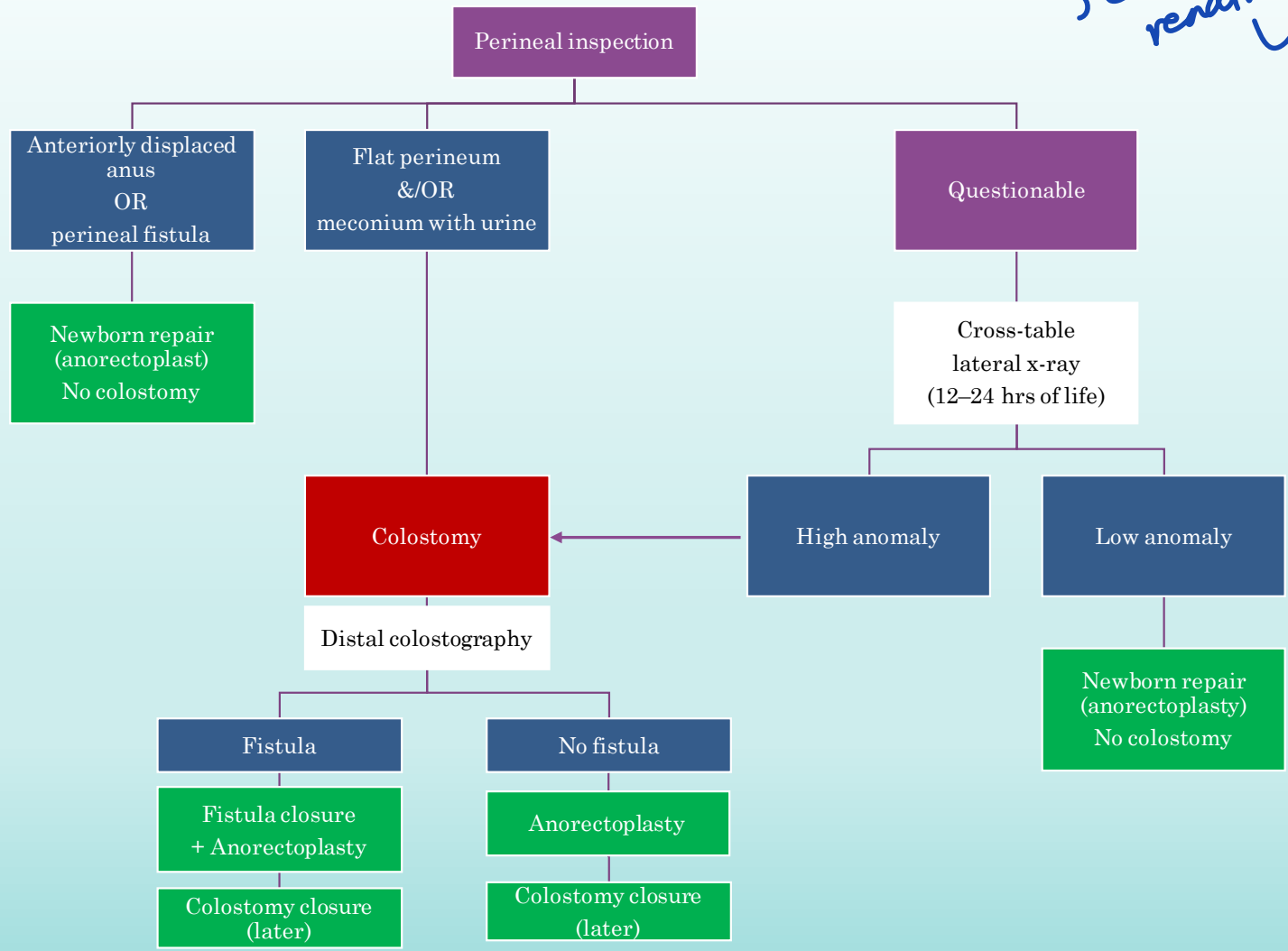
- Vertebral PEx & imaging
- Echocardiography
- NGT + CXR
- Renal US
- Limbs PEx

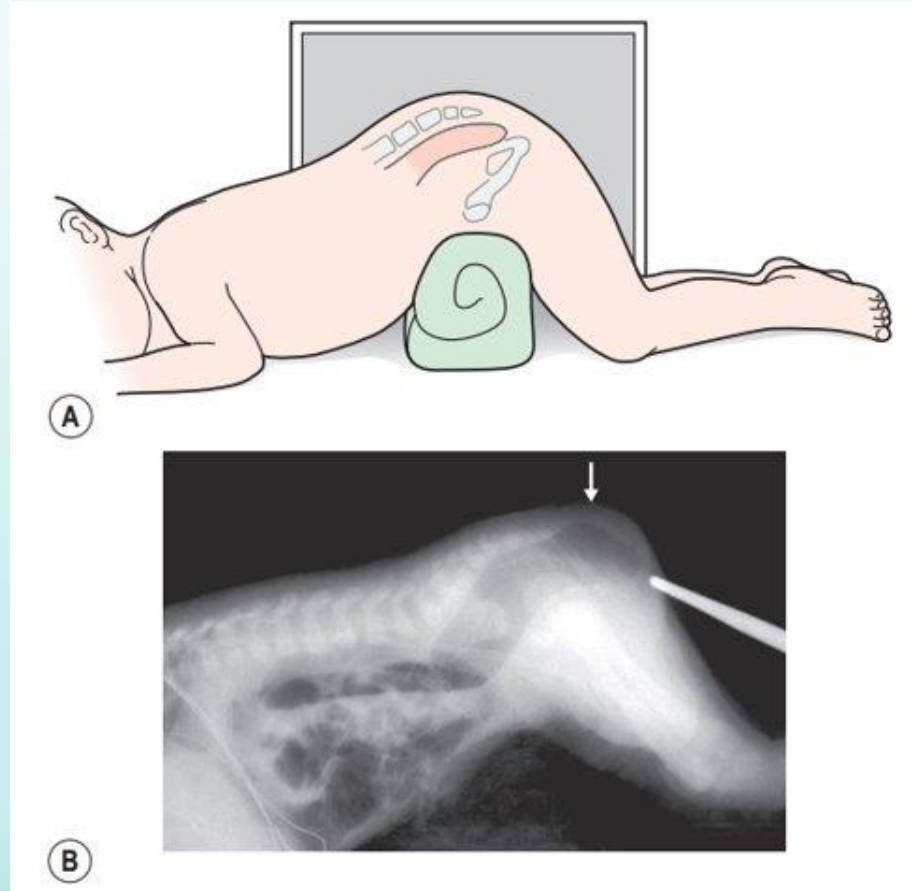
Then..

- Follow the management **algorithms**

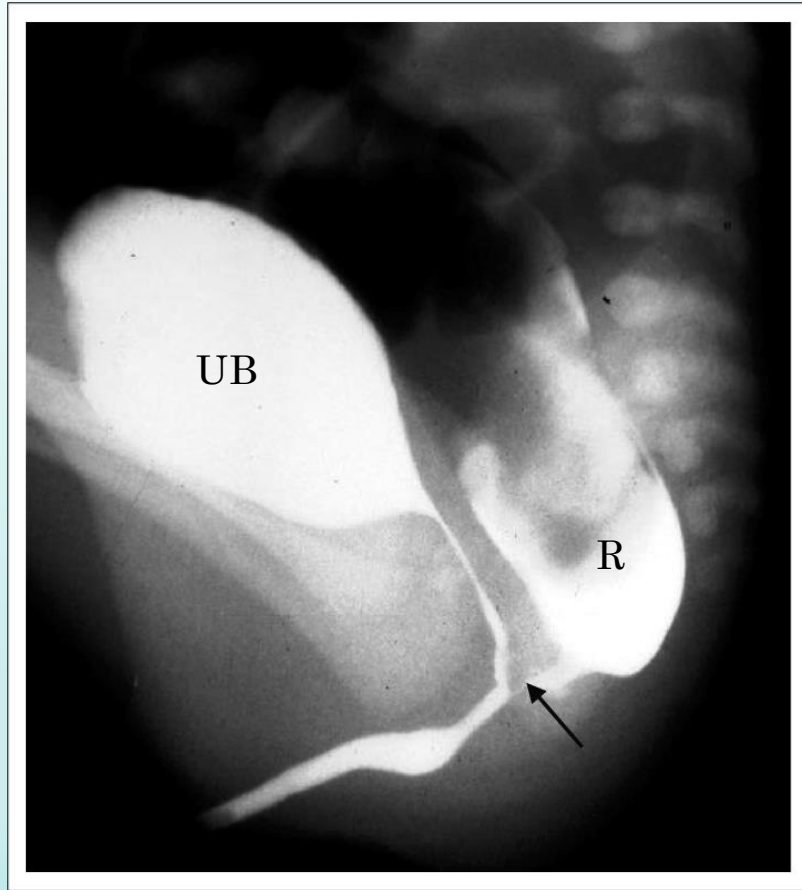
*self  
reading*

# Algorithm for male infant





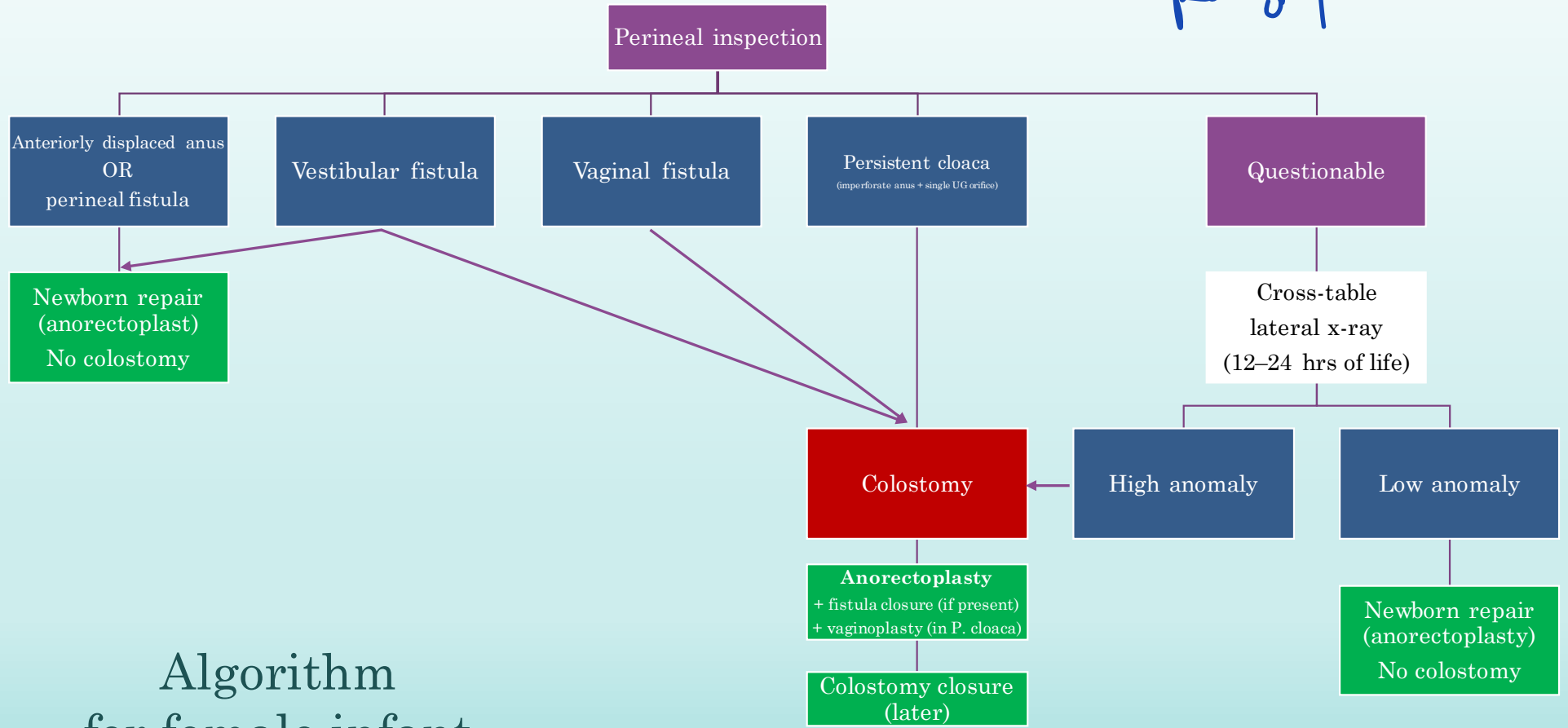
**Cross-table lateral film (after 12–24 h of life) in Jackknife position**  
to localize the distance of rectal gas from perineum



## Distal Colostogram

Shows that contrast is passing through a recto-membranous urethral fistula (black arrow) filling the urinary bladder and urethra

*Read only*



Algorithm for female infant

# Anorectal Malformations

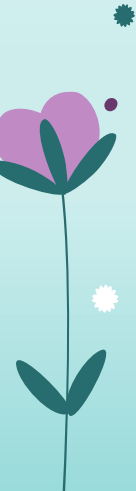
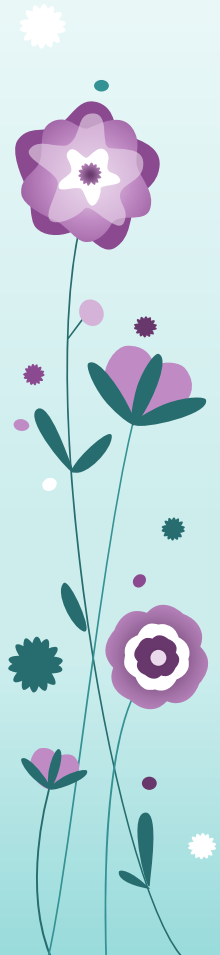
## Outcome

- Lower long-term continence:
  - In higher anomalies
  - If associated sacral anomaly
- Long-term constipation in lower anomalies



# Hypertrophic Pyloric Stenosis (HPS)

- M:F = 4 : 1
- Risk factors:
  - Family history
  - Male gender
  - Younger maternal age
  - Being a first-born infant
  - Maternal feeding patterns



# Etiology

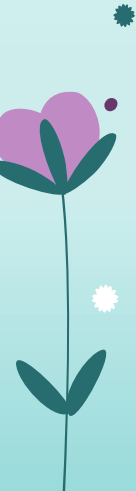
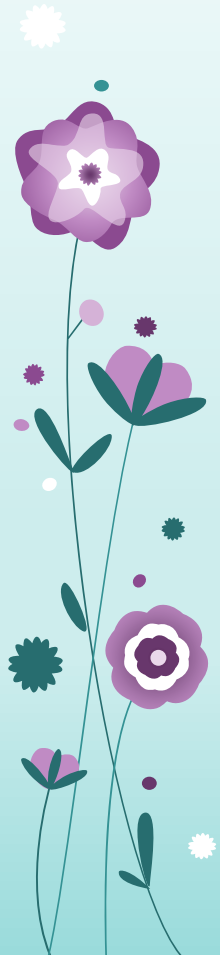
## Unknown (multifactorial with environmental influences)

- Genetic factors
  - race discrepancies
  - increased frequency in **males**
  - birth order (**first-born infants** with a positive family history)
- Environmental factors
  - method of feeding (breast vs **formula**)
  - seasonal variability
  - exposure to erythromycin
  - **transpyloric feeding** in premature infants
- Other factors
  - excessive substance P
  - decreased neurotrophins
  - deficient nitric oxide synthase
  - gastrin hypersecretion

# Diagnosis

## Classic presentation:

- nonbilious, progressive projectile vomiting (of recent feedings)
- full-term neonate
- 2-8 weeks old



# Diagnosis

## PEx:

- **General:**
  - Usually appears well (early)
  - Dehydration, somnolence (late)
- **Abdominal Ex:**
  - Visible gastric peristaltic waves
  - Palpable pylorus “olive sign” (70–90% of patients)

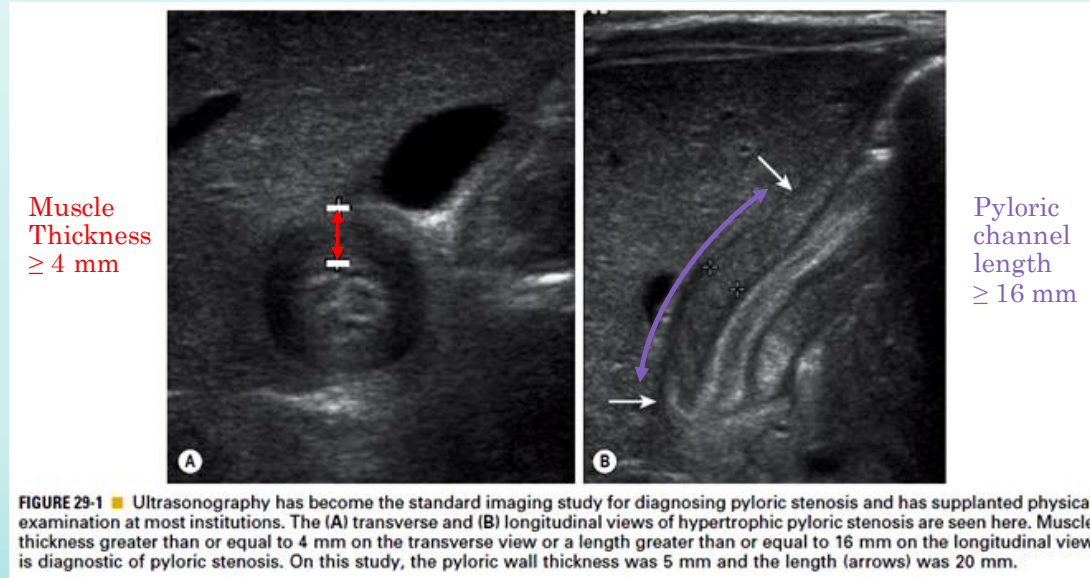
## Labs:

- Hypochloremic
- Hypokalemic
- Metabolic alkalosis

# Diagnosis

## Ultrasound:

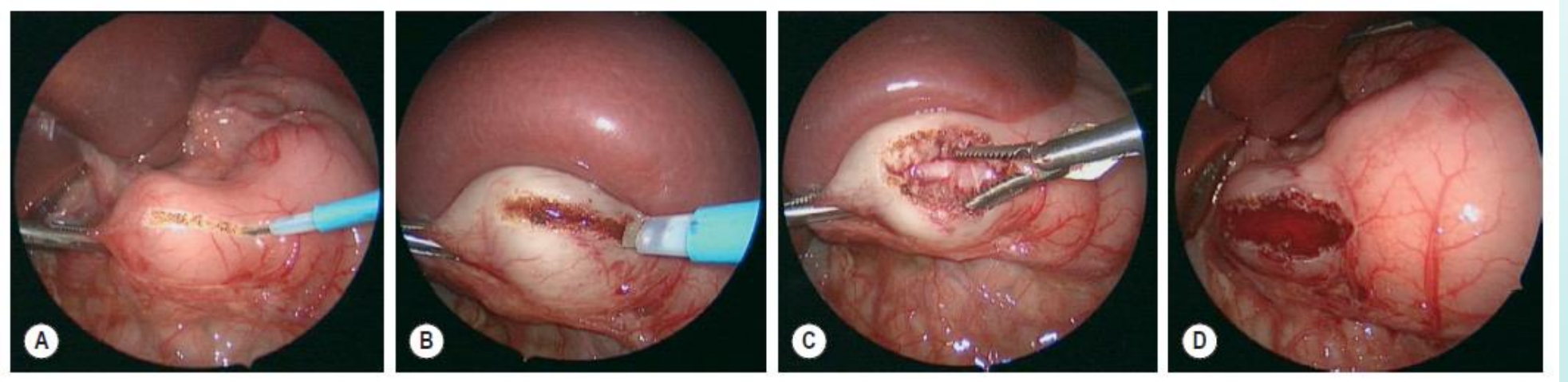
- Standard technique for diagnosis



# Treatment

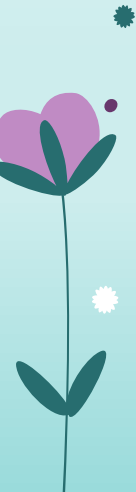
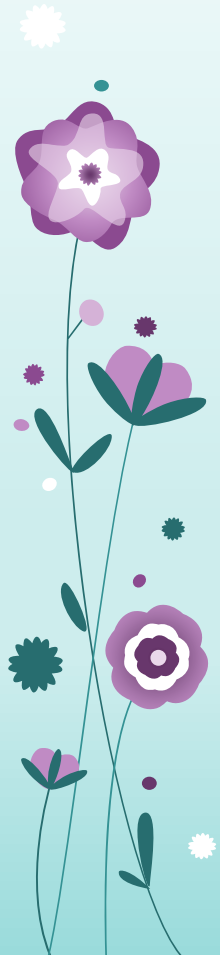
- Preop. supportive measures:
  - NPO
  - IV fluid resuscitation
  - Correction of electrolytes
- Surgery:
  - Non-emergent
  - Laparotomic or laparoscopic “Pyloromyotomy”
- Other reported modalities (require long periods, and often not effective):
  - medical treatment with atropine
  - pyloric dilation

to incise (otomy) the muscle (myo) of the pylorus (pyloro)



# Complications of Pyloromyotomy

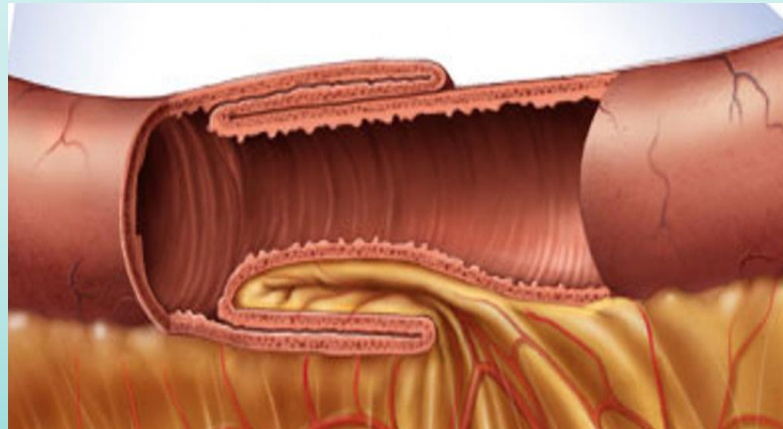
- Mucosal perforation (1-2%)
- Postoperative emesis (occur in most infants)
- Prolonged postoperative emesis  
(less common | due to GER or incomplete myotomy)





# Intussusception

An acquired invagination of the proximal bowel (intussusceptum) into the distal bowel (intussuscipiens)

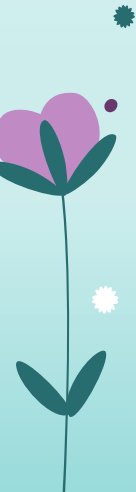
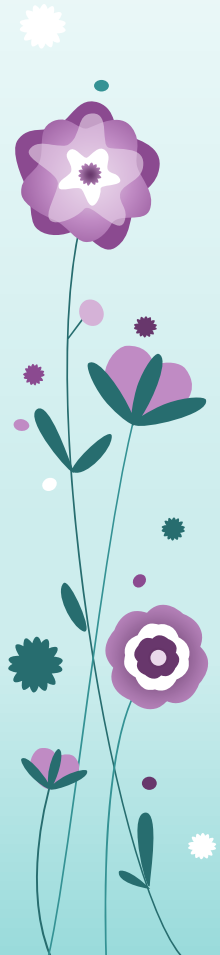


# Consequence

The **mesentery** of the proximal bowel is compressed..

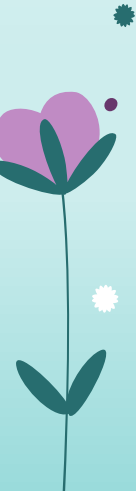
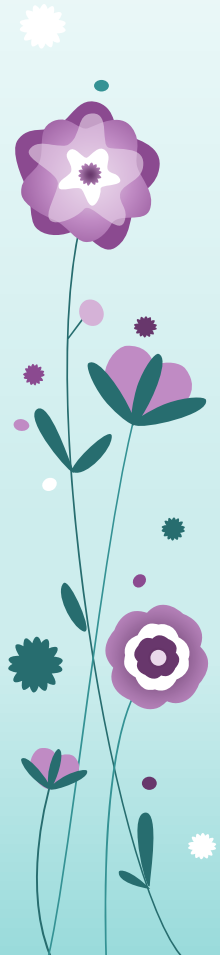
→ venous obstruction + bowel wall edema

→ may progress into **arterial insufficiency** + ischemia and bowel wall necrosis



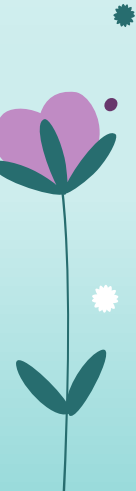
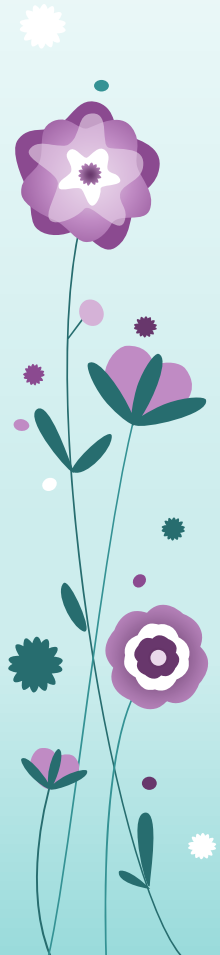
# Types

- Primary (idiopathic) **most common**
- Secondary (lead point)



# Primary Intussusception

- Generally attributed to **hypertrophied Peyer patches**
- Frequently after a recent **URTI** or **gastroenteritis** (adenoviruses and rotaviruses in 50% of cases)
- Incidence
  - can occur at any age
  - most are well-nourished, healthy infants
  - two-thirds are boys
  - highest incidence in infants 4-9 months
  - uncommon < 3 months and > 3 years of age



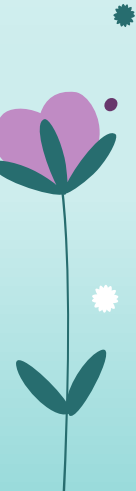
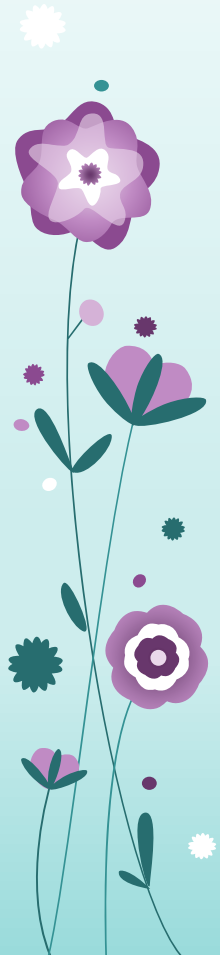
# Secondary Intussusception

- There is an identifiable lesion → serves as a lead point:
  - Meckel diverticulum (most common)
  - Polyps
  - Duplications
  - Appendix
  - Hemangiomas
  - Carcinoid tumors
  - Foreign bodies
  - Ectopic pancreas or gastric mucosa
  - Hamartomas from Peutz–Jeghers syndrome
  - Lipomas
  - Malignant causes (rare | ↑ with age | as lymphomas and small bowel tumors)
  - Systemic diseases (as Henoch–Schönlein purpura and cystic fibrosis)
  - Celiac disease
  - *Clostridium difficile* colitis

# Clinical presentation

- Classic presentation (infant or young child):
  1. intermittent, crampy abdominal pain
  2. 'currant jelly' stools (bowel ischemia and mucosal sloughing)
  3. palpable abdominal mass

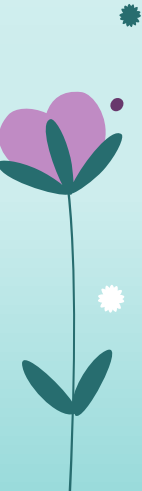
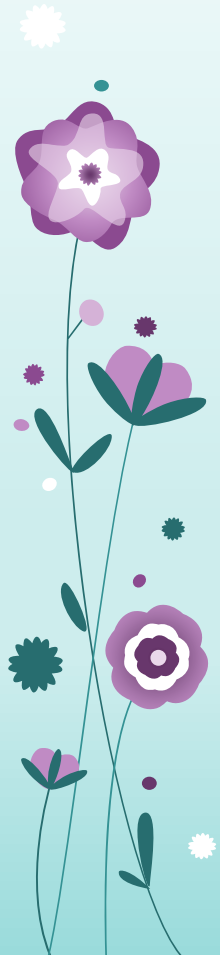
→ this triad is seen in <25% of children





# Clinical presentation

- Abdominal pain:
  - **Sudden** (often ceases as suddenly as it started every 15-30 minutes)
  - Child may stiffen + pulls the legs up to the abdomen
  - Between attacks → may appear comfortable
- Associated with:
  - Hyperextension
  - Breath holding
  - Vomiting (gastric early | bile later)
  - Abdominal distension
  - Lethargy (later)
  - Red currant jelly stools (later)



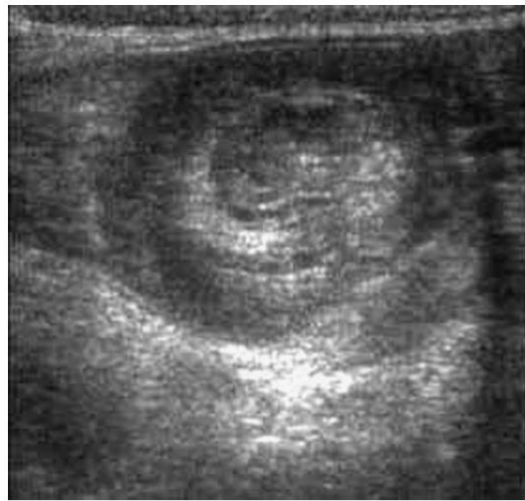


# Physical examination

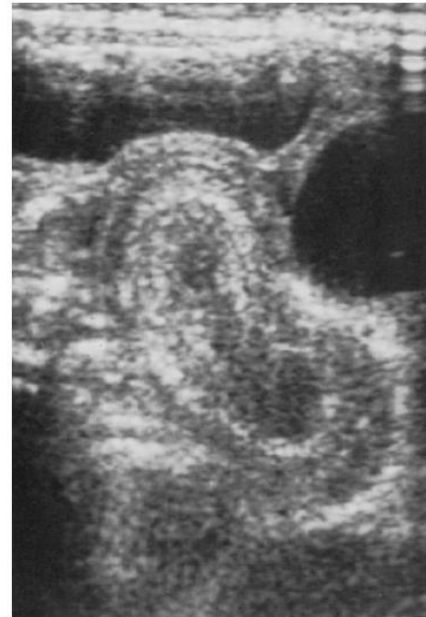
- Vital signs
  - Normal (early)
  - Hypotension, fever, &/or tachycardia (late)
- Signs of dehydration (early &/or late)
- Abdominal PEx:
  - RLQ can appear flat or empty (Dance sign)
  - RUQ mass (Sausage-shaped or curved mass)
  - Audible peristaltic rushes (early)
  - On rectal Ex: bloodstained mucus or blood (late)
- +/- Prolapse of the intussusceptum through the anus (late)

# Ultrasonography (US)

- ‘Target’ or ‘donut’ lesion (in transverse plane)
- ‘Pseudokidney’ sign (on longitudinal plane)



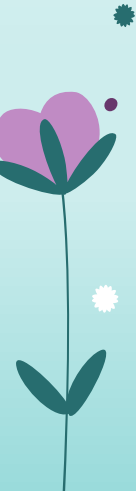
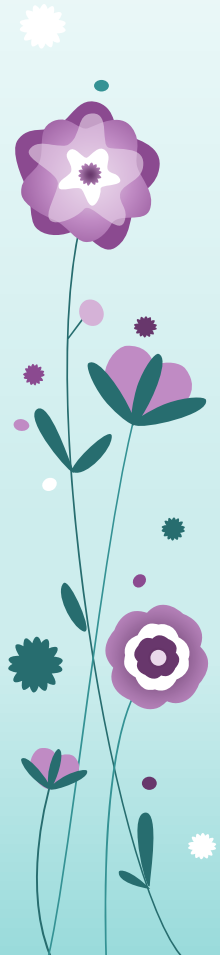
**FIGURE 38-4** ■ This transverse sonographic image shows the alternating rings of low and high echogenicity due to an intussusception. This finding has been called a ‘target’ sign.



**FIGURE 38-5** ■ Sonogram showing the ‘pseudokidney’ sign seen with intussusception on longitudinal section.

# Nonoperative management

- NGT (to decompress the stomach)
- Bowel rest (NPO)
- IV fluid resuscitation
- CBC and serum electrolytes



# Nonoperative management

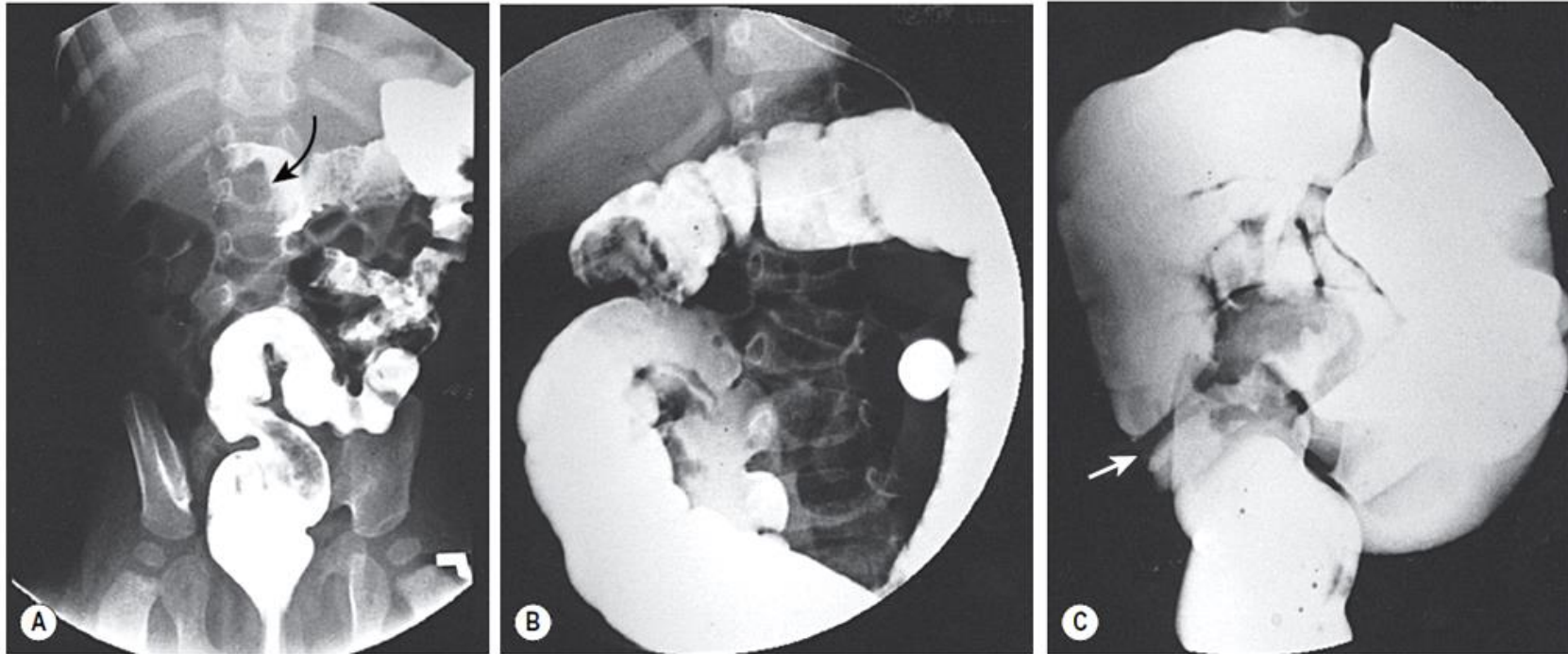
## Hydrostatic/ Pneumatic Reduction

- Under fluoroscopy or ultrasound guidance
- When there are no contraindications

### Contraindications:

- Intestinal perforation
- Peritonitis
- Persistent hypotension

- Successful reduction ( $\approx 85\%$ ), followed by:
  - Admission for observation
  - Short period of bowel rest (NPO)
  - IV fluids

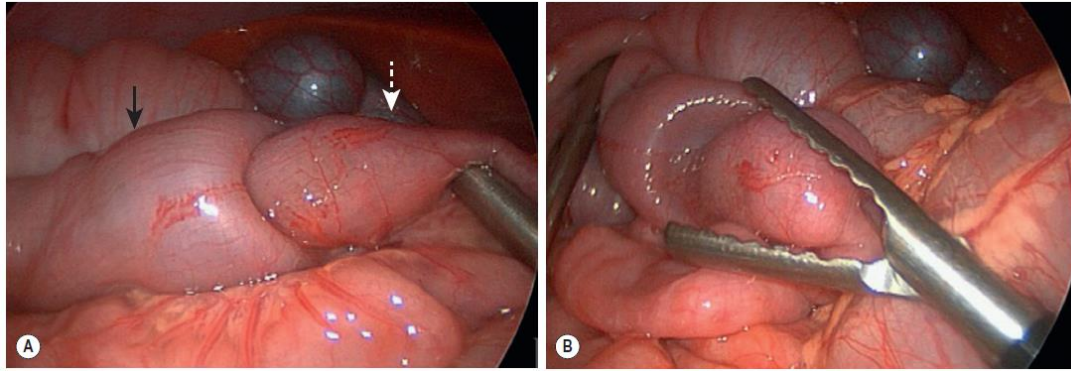


**FIGURE 38-7** ■ Fluoroscopic examination using isotonic contrast for hydrostatic reduction of intussusception. (A) Intussusception (arrow) seen in midtransverse colon. (B) Reduction has occurred to the hepatic flexure. (C) Complete reduction with reflux of contrast medium into the terminal ileum. Note the edematous ileocecal valve (arrow).

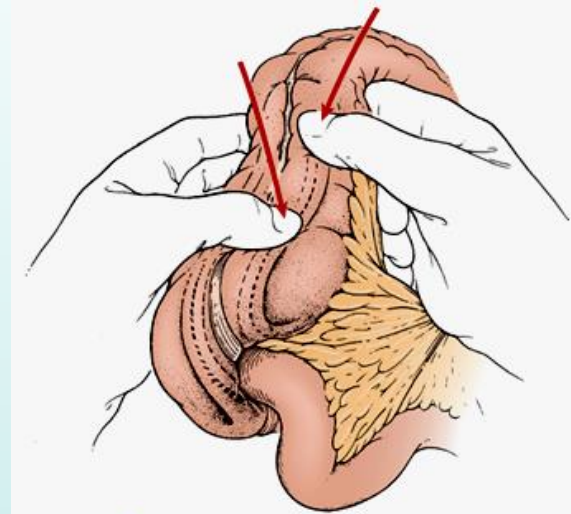
# Operative management

## • Indications:

- Nonoperative reduction is unsuccessful or incomplete
- Signs of peritonitis/ pneumoperitoneum
- Presence of a lead point (secondary intussusception)



Lap. reduction



Open reduction

**FIGURE 38-9** ■ A right lower quadrant muscle-splitting incision allows delivery of the intussusception through the incision. Gentle and continuous massage from distal to proximal usually results in reduction of the intussusception.

# THANK YOU!

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