Neonatal Intestinal Development and Clinical Correlations

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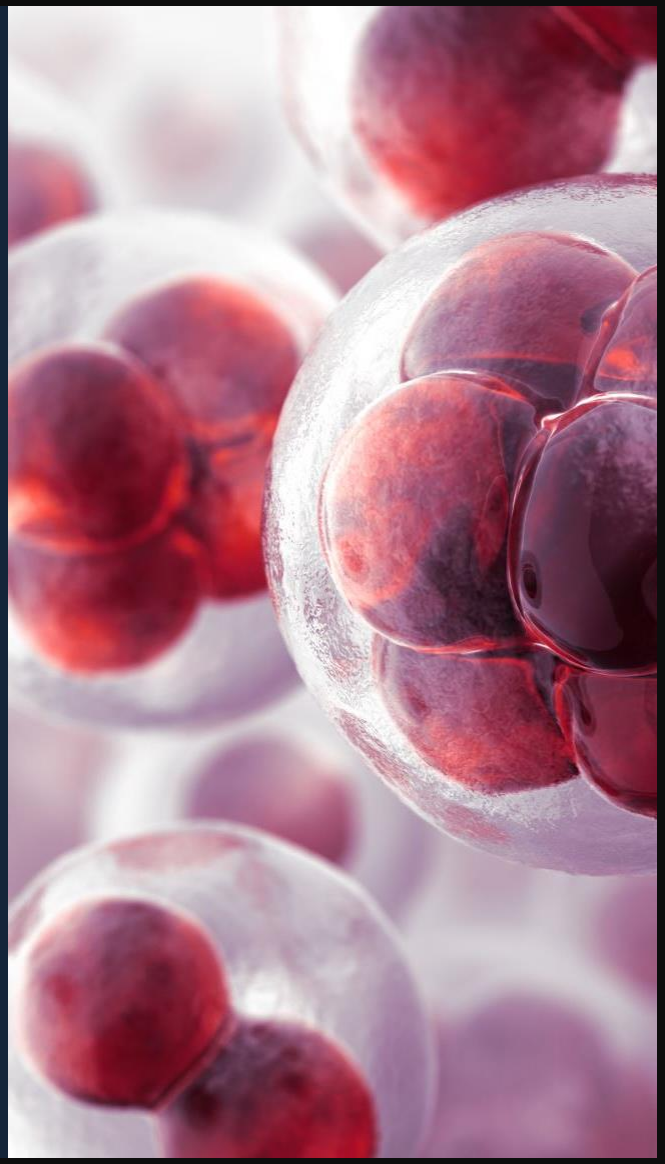
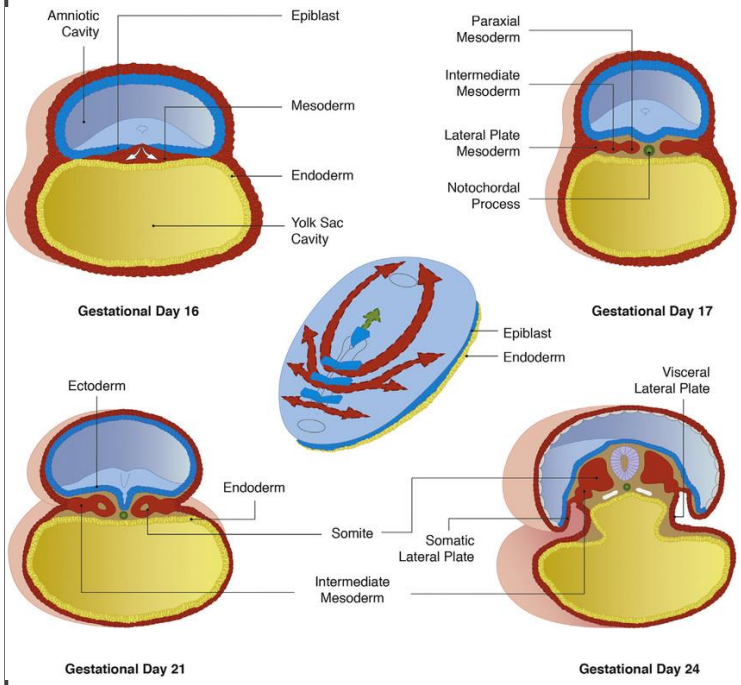
Objectives

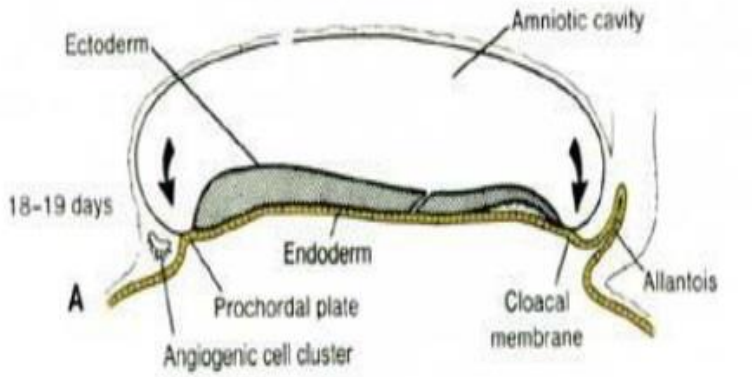
- Associate anatomic developmental features with clinically relevant clinical outcomes seen in the NICU.
- Discuss clinically relevant cases that are commonly seen in the NICU. Differentiate between different forms of intestinal obstruction.

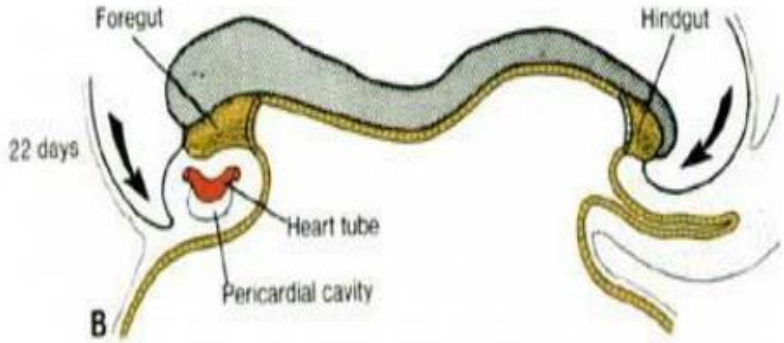


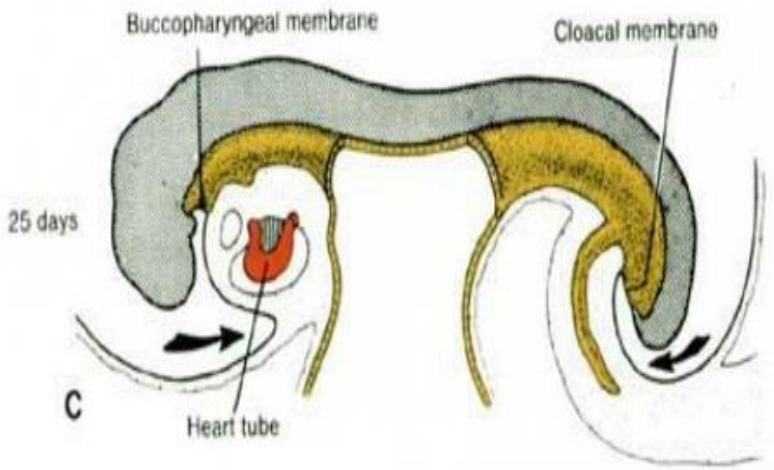
Normal Embryology (gastrula stage-after implantation)

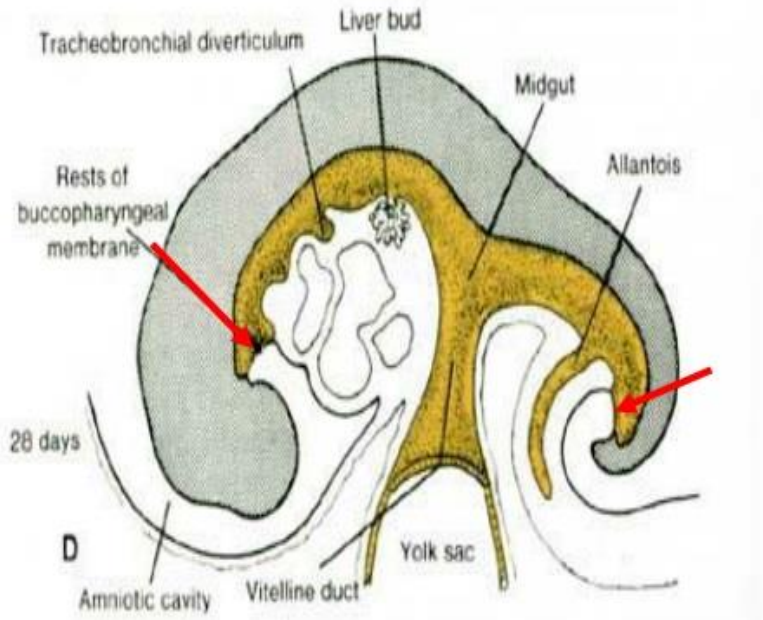


- Endoderm
  - Epithelial lining and glands
- Mesoderm
  - Lamina propria, muscularis mucosa, submucosa, muscularis externa and serosa
- Ectoderm
  - Enteric nervous system and posterior luminal digestive structures









- Embryo folding – incorporation of endoderm to form primitive gut.
- Outside of embryo – yolk sac and allantois.
- Vitelline duct – connects sac + gut

Canalization

- Canalization
  - Week 5 - Endoderm portion of GI tract proliferates
  - Week 6 - Occlusion of the lumen
  - Week 8 - Recanalization due to cell degeneration

  - Abnormalities in this process
    - Stenosis/Atresia
    - Duplications

General Outline of GI organ Development

Foregut—supplied by celiac artery

Midgut—supplied by superior mesenteric artery

Hindgut---suppled by inferior mesenteric artery

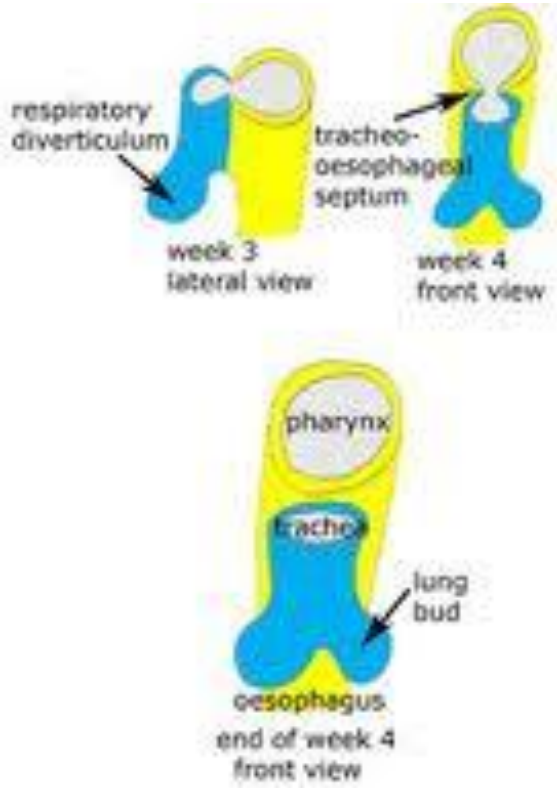
Derivatives of

foregut – pharynx, (+ respiratory diverticle), esophagus
stomach, cranial arm of duodenum
(+ liver, gall bladder pancreas),

midgut – caudal arm of duodenum, small intestine and part of large intestine (cca 1/3 of colon transv.)

hindgut – the rest of large intestine, rectum, upper part of the anal canal

Foregut



- Esophagus – The tracheoesophageal septum divides foregut into the esophagus and trachea

- **Clinical Correlation**
  - Esophageal atresia
  - Tracheoesophageal fistula

Foregut

- Stomach – A fusiform dilation in the foregut occurring during the 4th week. 90-degree clockwise rotation, creates the lesser peritoneal sac.

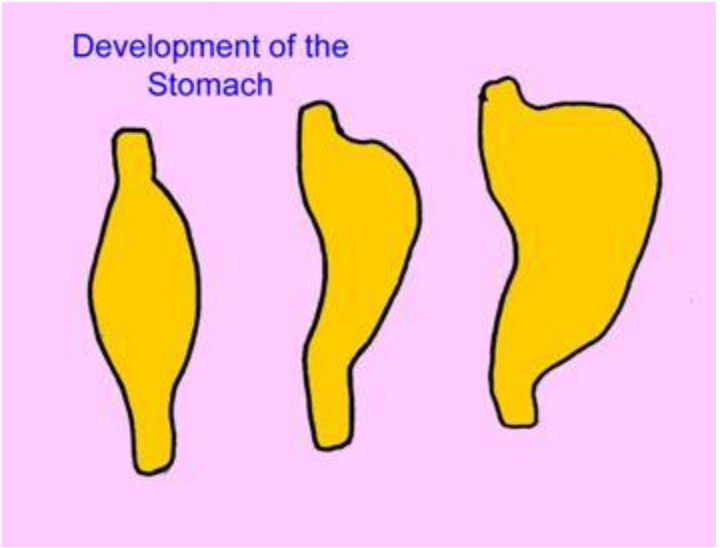


Image from: http://bio.sunyorange.edu

Foregut

- Liver – Develops from an endodermal outgrowth at cranioventral portion of the foregut (hepatic diverticulum).
- Mesoderm surrounds the diverticulum (septum tansversum).

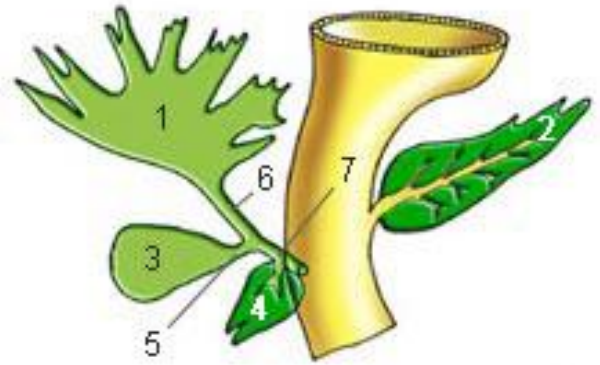


Image from: http://www.embryo.chronolab.com/liver.htm

Foregut

- Gallbladder and bile ducts –
  - Cystic diverticulum develops into gallbladder and cystic duct.

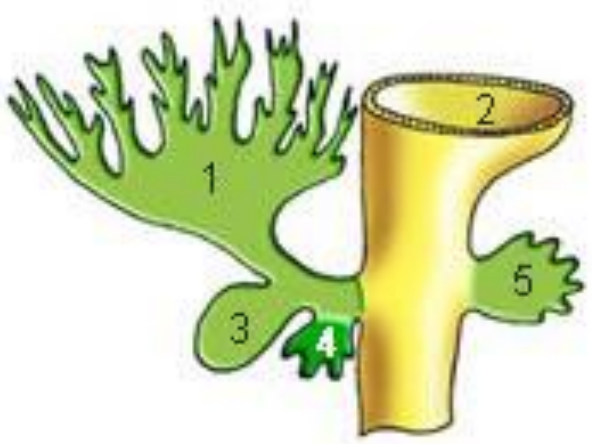


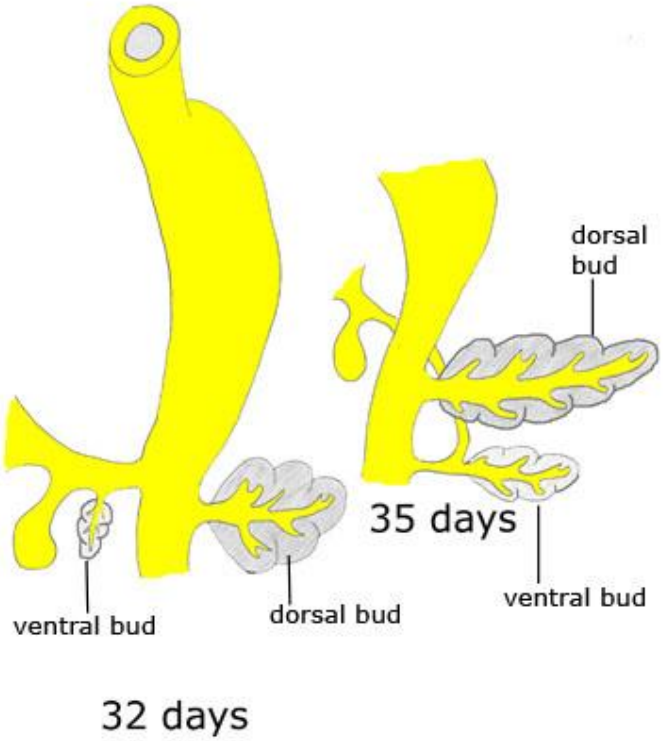
Image from: http://www.embryo.chronolab.com/liver.htm

Foregut

- Pancreas – Develops from 2 pancreatic buds (weeks 4-5).
  - Ventral pancreatic bud forms the uncinate process and the head of the pancreas.
  - The larger dorsal pancreatic bud forms the remaining head, body and tail.

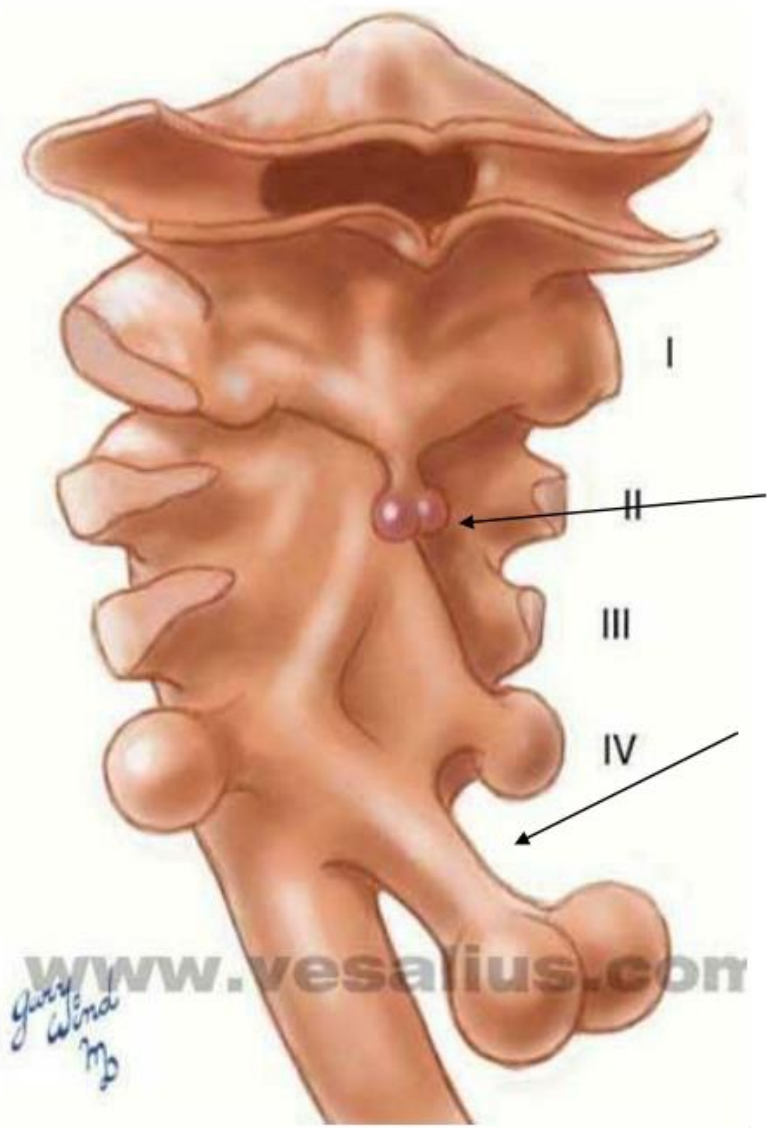
Foregut

- Pancreas continued
  - The ventral pancreatic bud rotates clockwise with the rotation of the duodenum. During 7th week the 2 buds fuse.
- **Clinical Correlation**:
  - Failure of this rotation and fusion results in pancreas divisum.



Esophagus development

below respiratory diverticle,
behind larynx and trachea

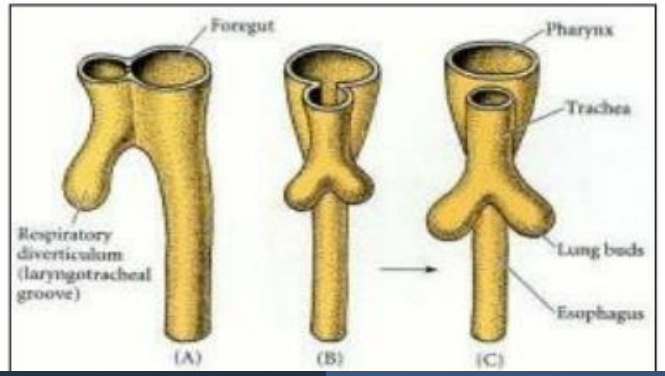


primitive pharynx

thyroid gl.

laryngotracheal diverticle
(respiratory diverticle)

esophagus



Teratology

Esophageal atresia – failure of recanalization or septum deviation

Susp.: polyhydramnios, fetus cannot swallow

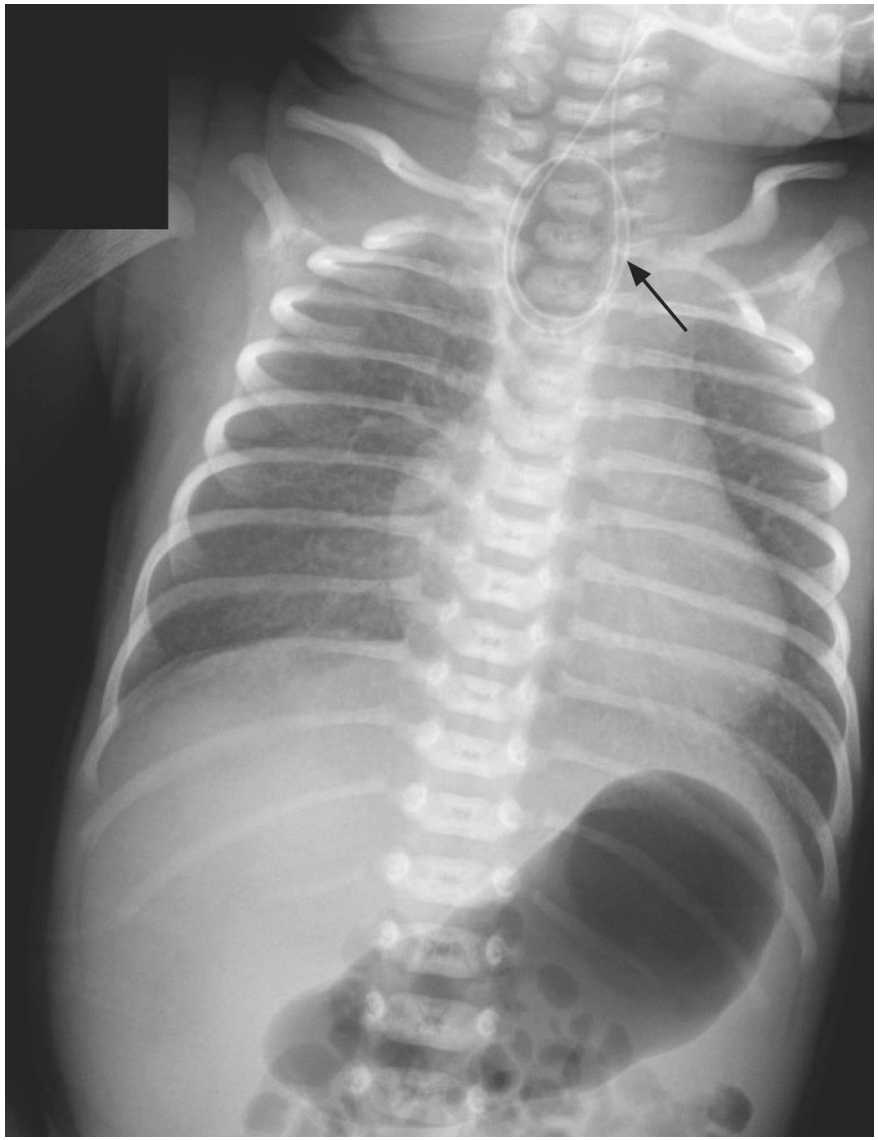
Esophageal stenosis – narrow lumen, incomplete recanalization

Tracheoesophageal fistula – defect in septum

Case: Term infant, normal delivery, no known antenatal problems other than polyhydramnios

- Frothing or drooling from the mouth noticed shortly after birth.
- Coughing and choking while nursing or taking a bottle.
- Difficulty breathing while feeding.
- Blue-tinged skin while feeding.

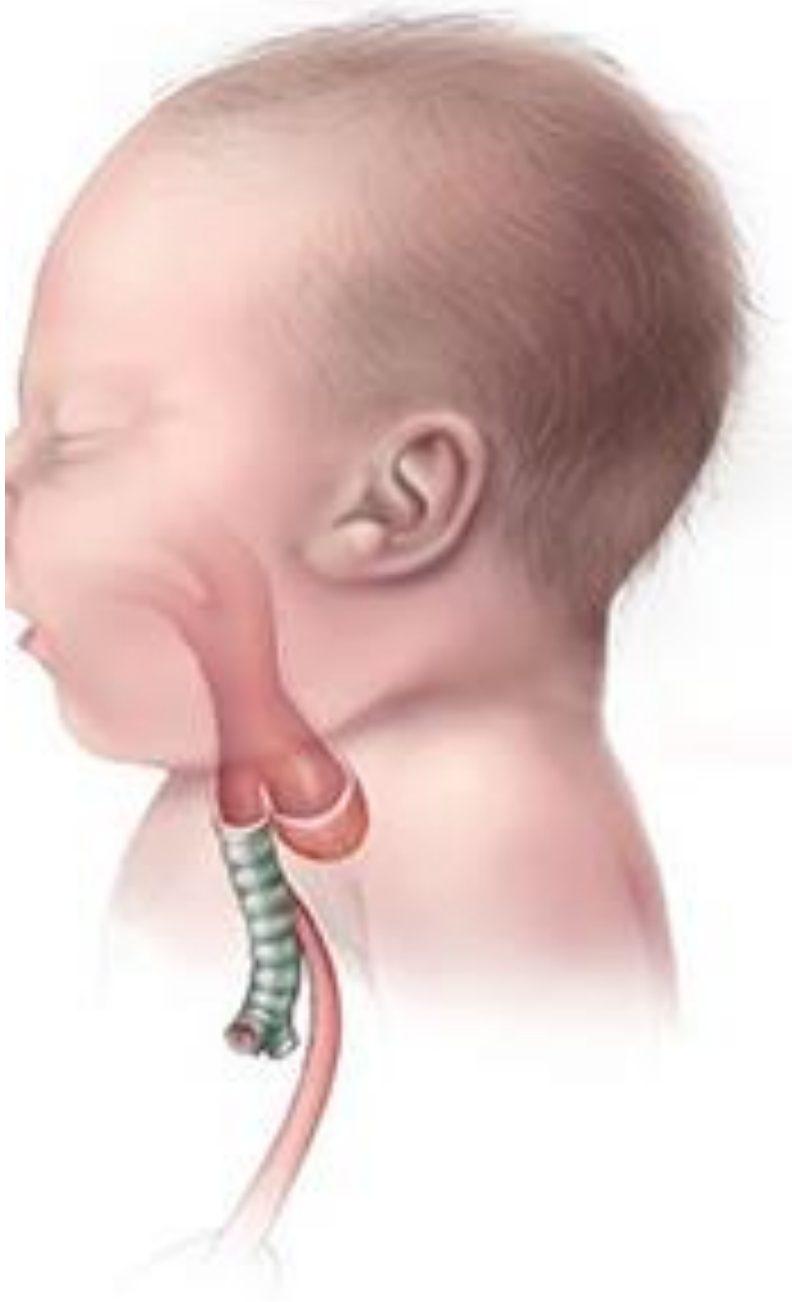
What would you like to do as a next step?

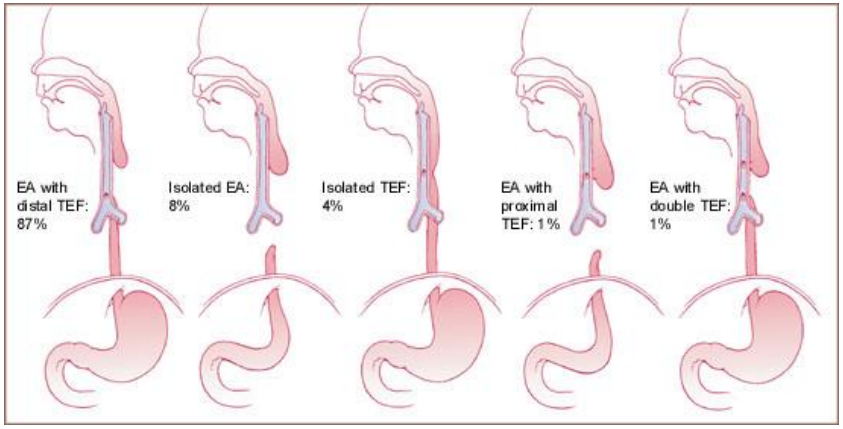


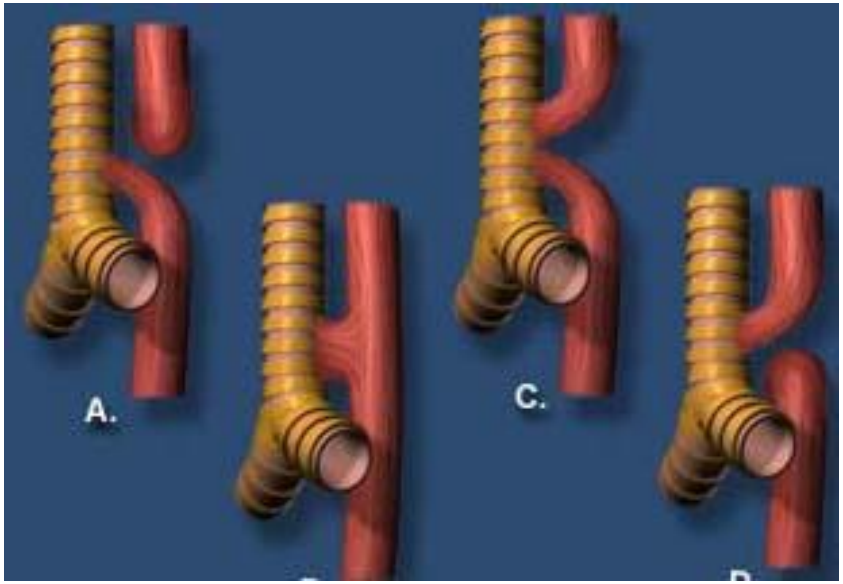
Chest X ray

Tracheo-Esophageal Fistula (TEF)

- Esophageal atresia with TEF is most common (85%)---"proximal pouch with distal fistula".
  - Diagnosis may be suspected antenatal with absence of stomach bubble and polyhydramnios.
  - Often associated with other anomalies: VATER and chromosomal




EA with distal TEF: 87%
Isolated EA: 8%
Isolated TEF: 4%
EA with proximal TEF: 1%
EA with double TEF: 1%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Operative management

- Ligation of fistula at trachea.
- Mobilization of distal esophageal segment with primary anastamosis to proximal pouch.
- NG tube left in place to stent open anastamosis while healing.
- Chest tube left in for serous drainage usually.

Post-operative Management

Careful airway management to prevent trauma to the fistula ligation site in the trachea.

Prior to feedings, must make sure that the esophageal anastamosis does not leak. (swallow study)

Often have on going feeding problems. May need dilation procedures periodically

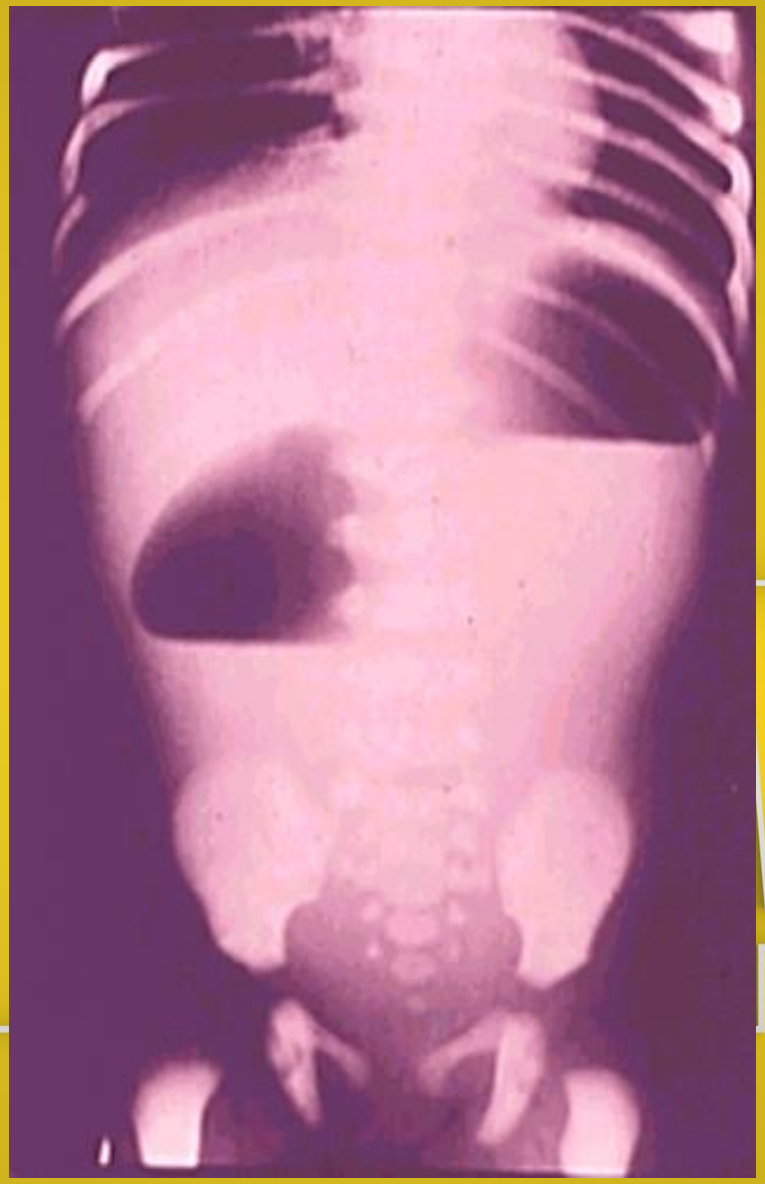
Midgut

The derivatives

- the distal duodenum, jejunum, and proximal ileum +
- the distal ileum, cecum, appendix, ascending colon, and proximal 2/3 of transverse colon.

the midgut grows faster than the embryo, creating:

- **duodenal loop**
- **umbilical loop**



Case

- Spontaneous vaginal delivery term infant
- History of polyhydramnios
- Bilious emesis day 1
- Abdomen not distended
- What do you do?

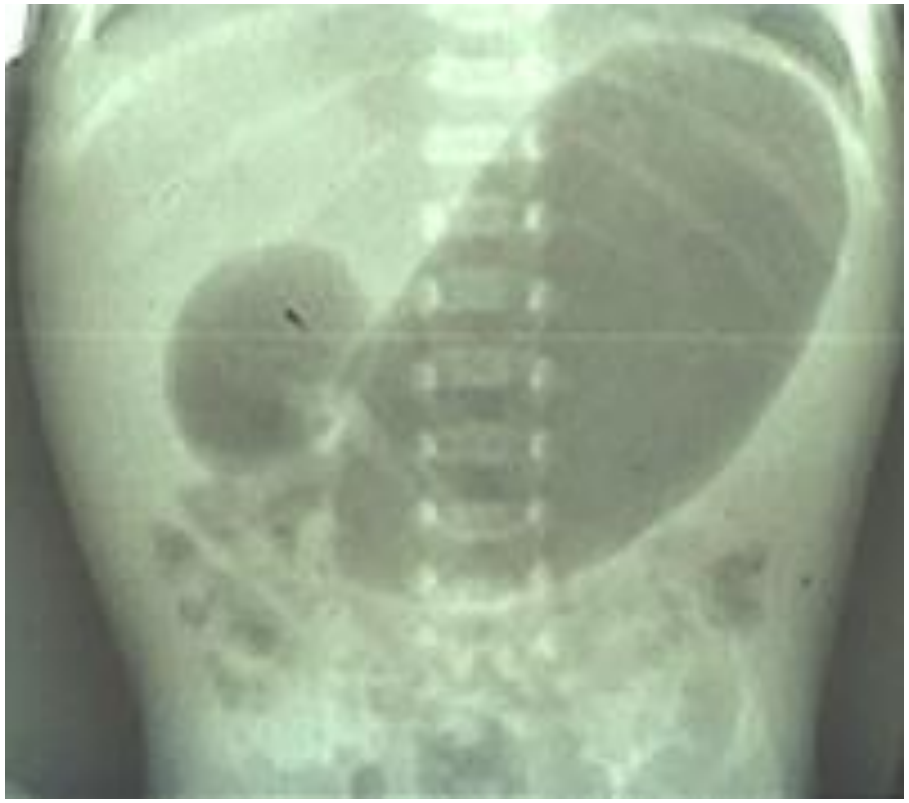
Duodenal Atresia

- Incidence--1 in 5,000 to 10,000 live births
- 75% of stenoses and 40% of atresias are found in Duodenum
- Multiple atresias in 15% of cases
- 50% pts are LBW and premature
- Polyhydramnios in 75%
- Bilious emesis usually present

Duodenal Atresia Con't

- Associated Anomalies
  - **Down's (30%)**
  - Malrotation
  - Congenital Heart Disease
  - Esophageal Atresia
  - Urinary Tract Malformations
  - Anorectal malformations
  - VACTERL

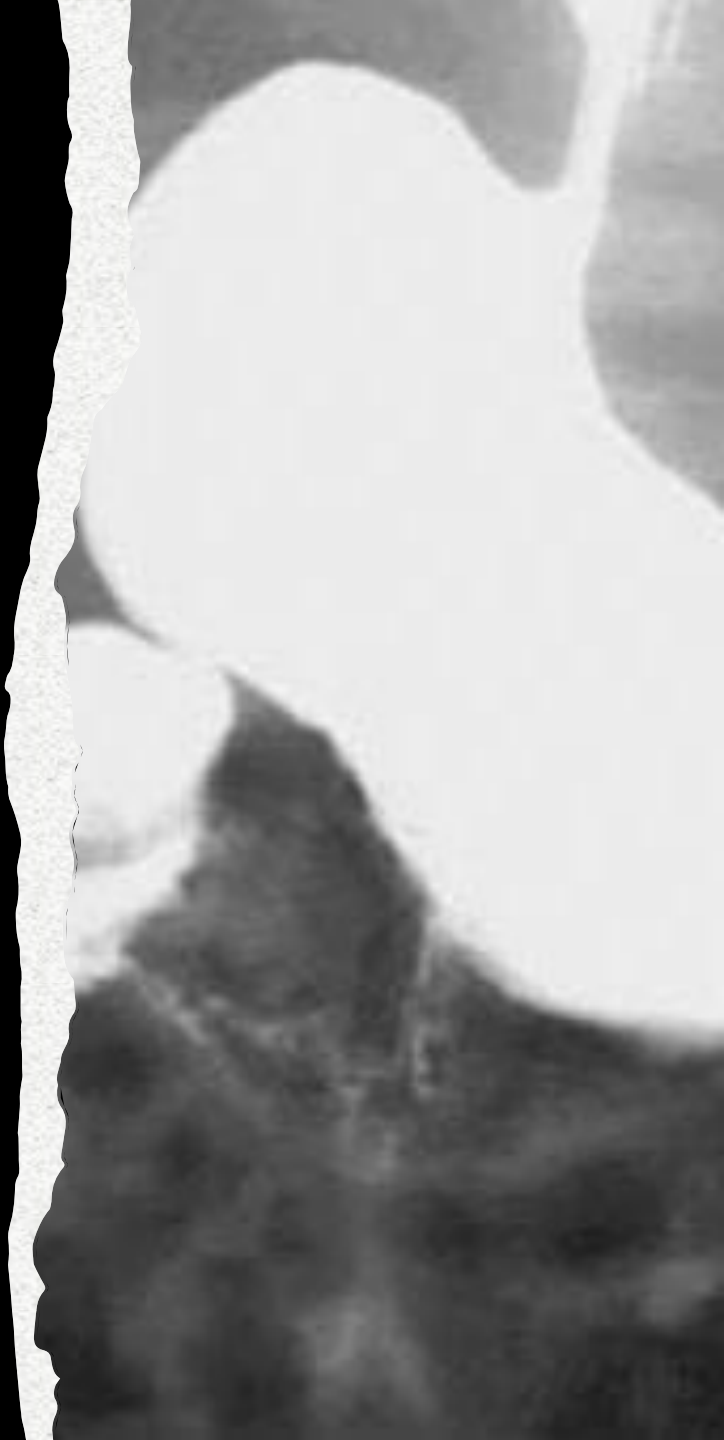
What’s this?

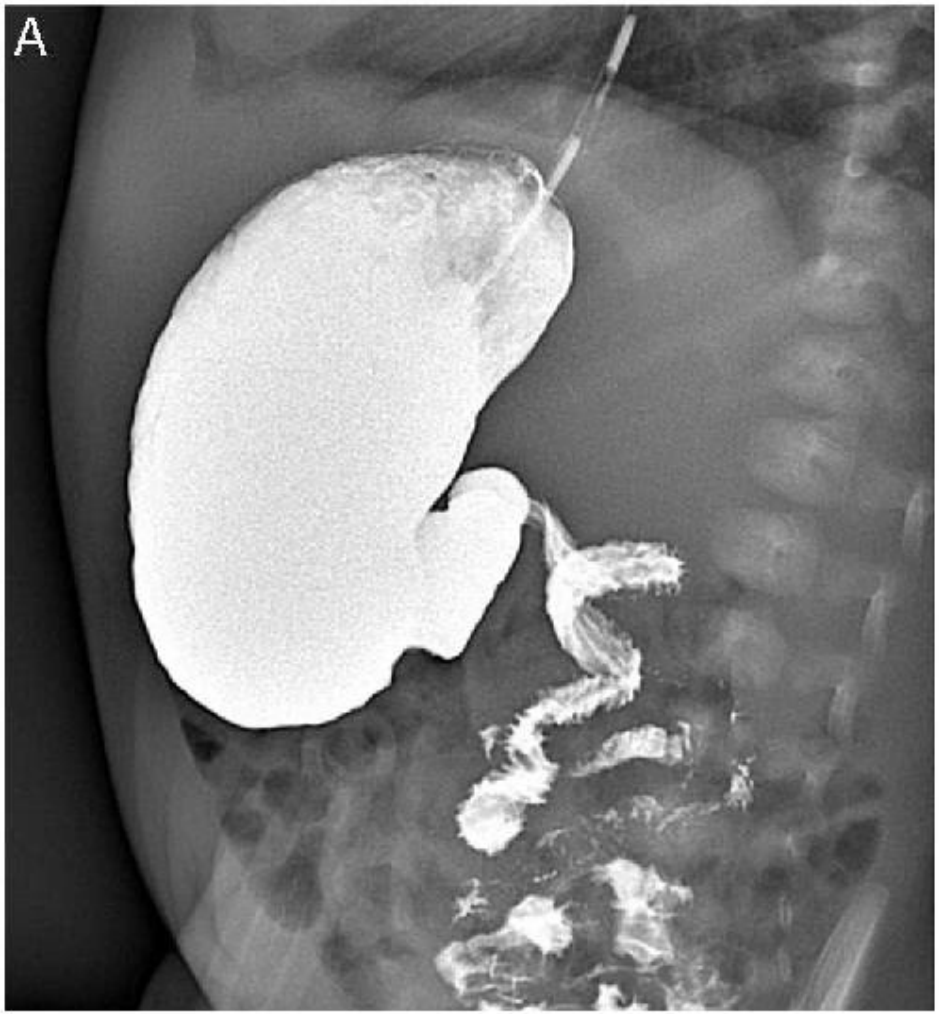


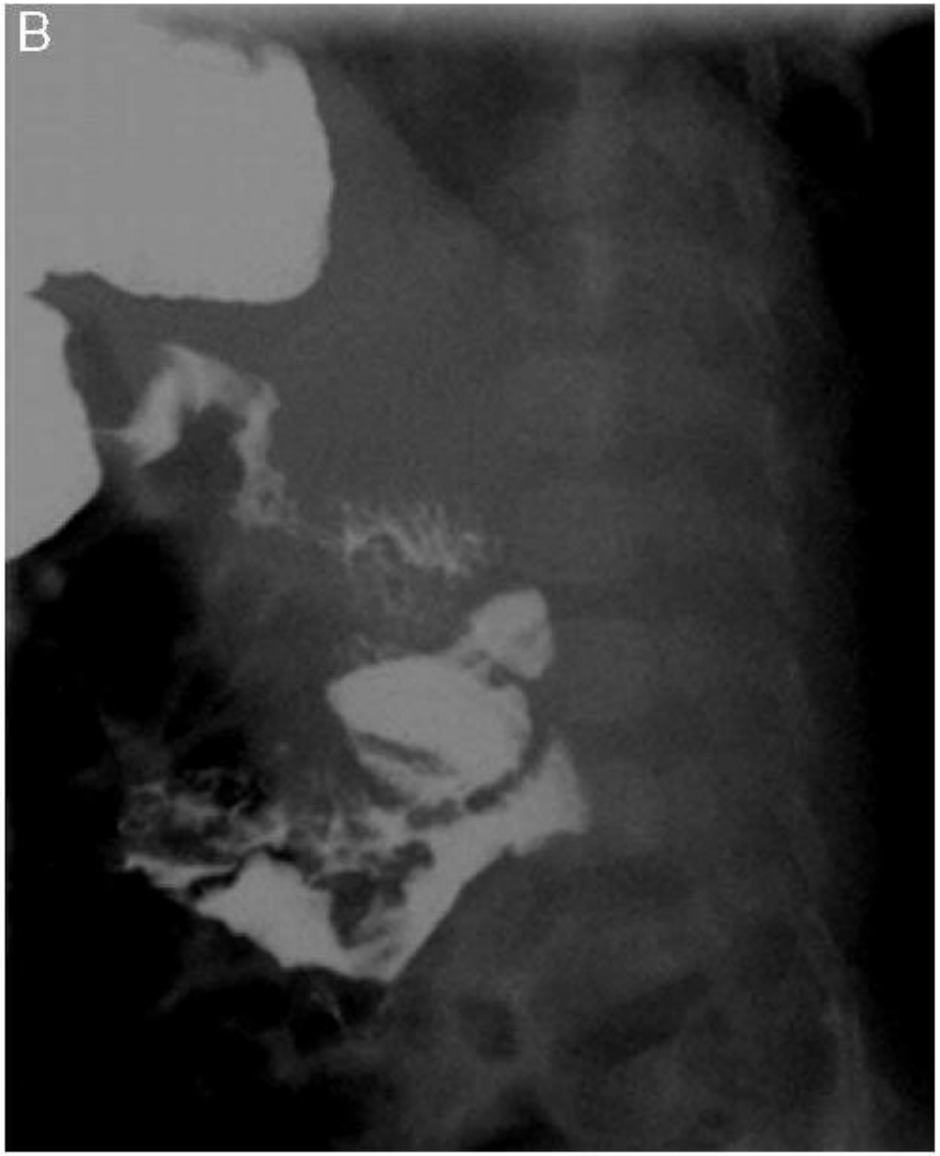
Case 6

- Five-day old infant at home after uneventful neonatal course begins to have bile-stained vomiting. On PE abdomen is slightly protuberant but not bulging or hard.
- What do you do?

Upper GI contrast study

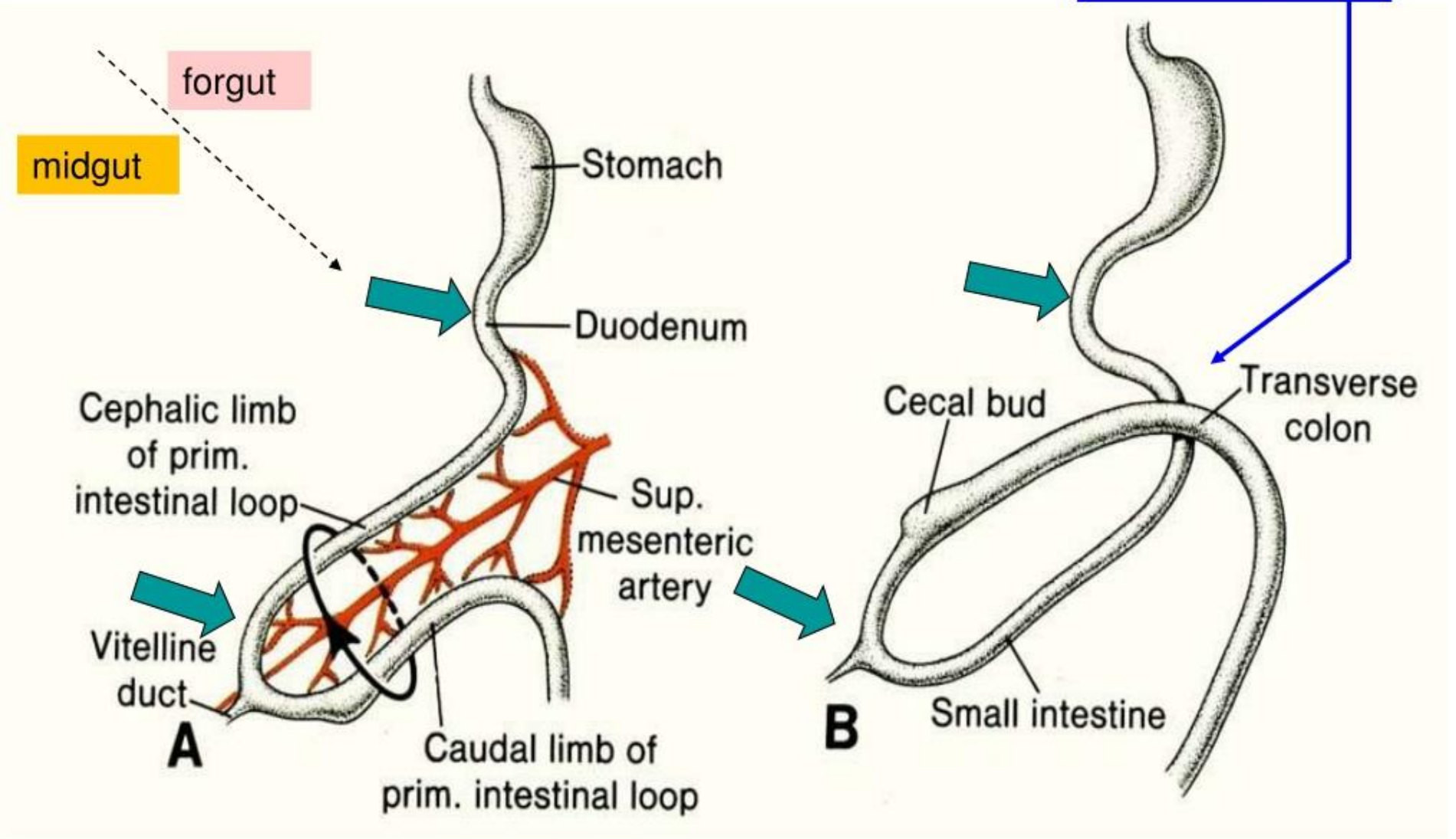




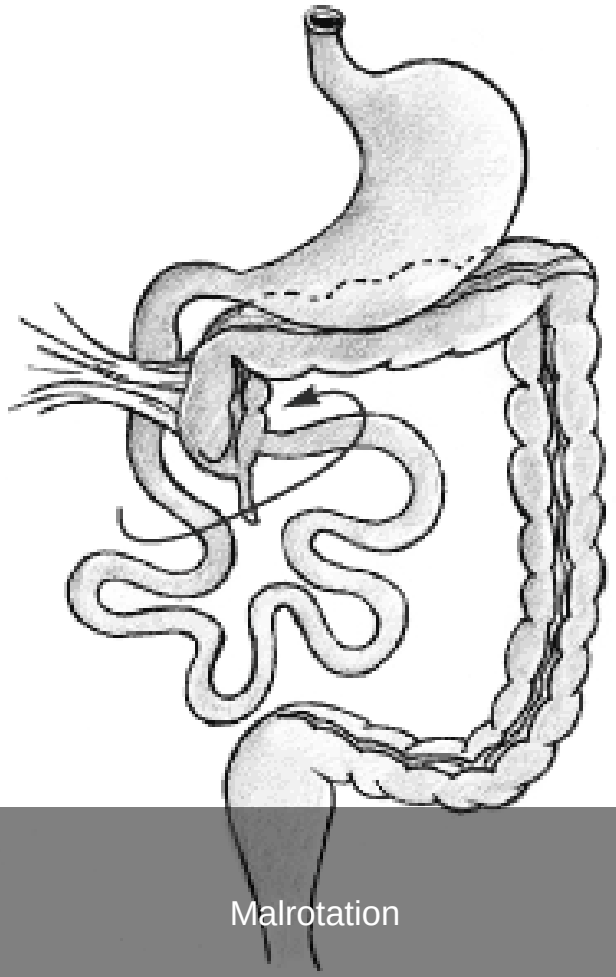


tinal transit. (A) Gerkegray sign in a patient with intestinal malrotation and volvulus of Ml. (B) M

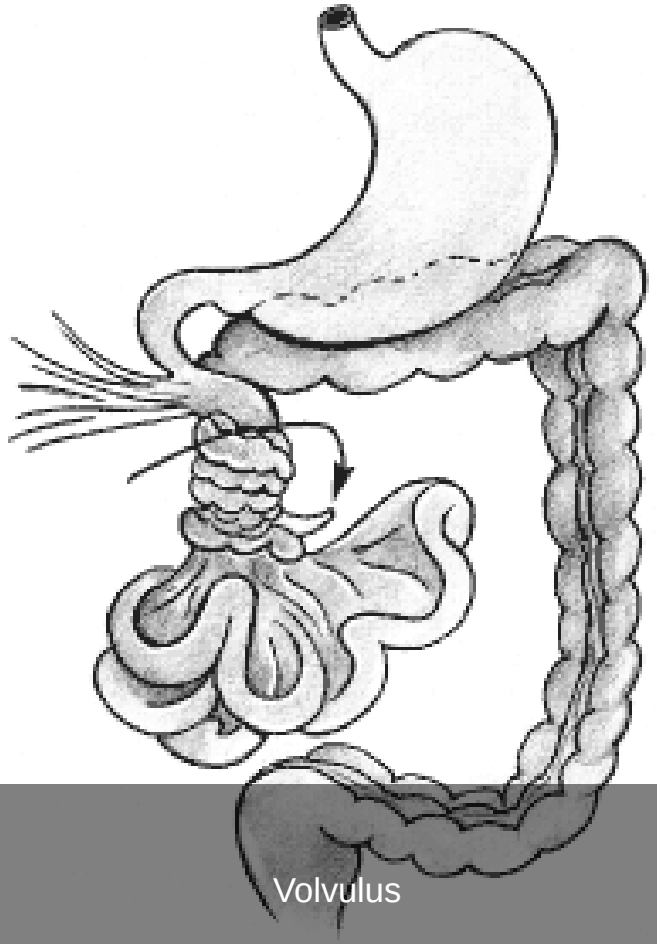
Duodenal loop and umbilical loop



Umbilical loop herniates into the umbilical cord (**physiologic herniation**, in week 6-10)

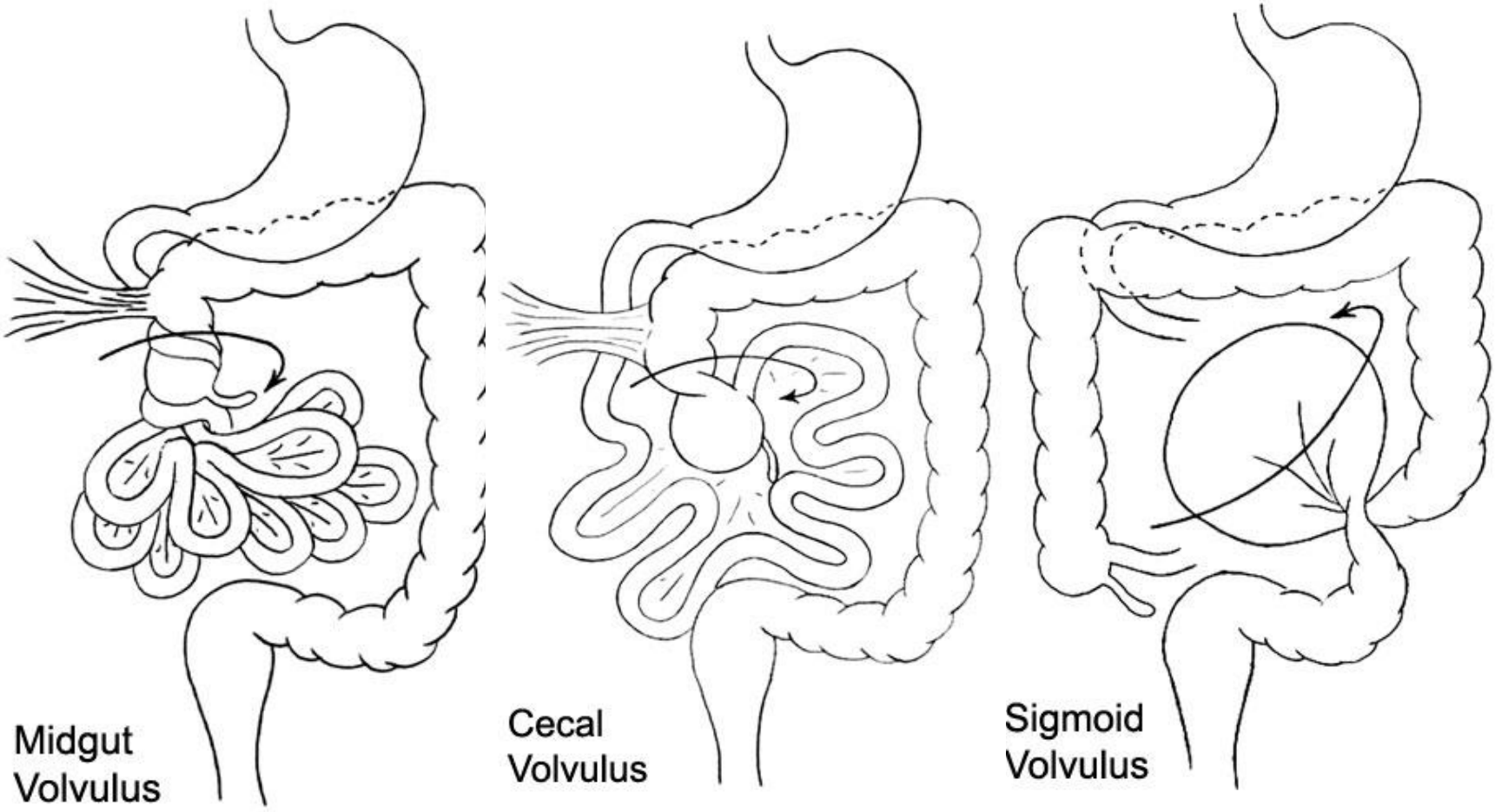


Malrotation



Volvulus

Malrotation



Slide from S Baker, MD

Malrotation with volvulus

May present with sudden bilious vomiting shortly after birth. This must be considered as an emergency until proven otherwise.

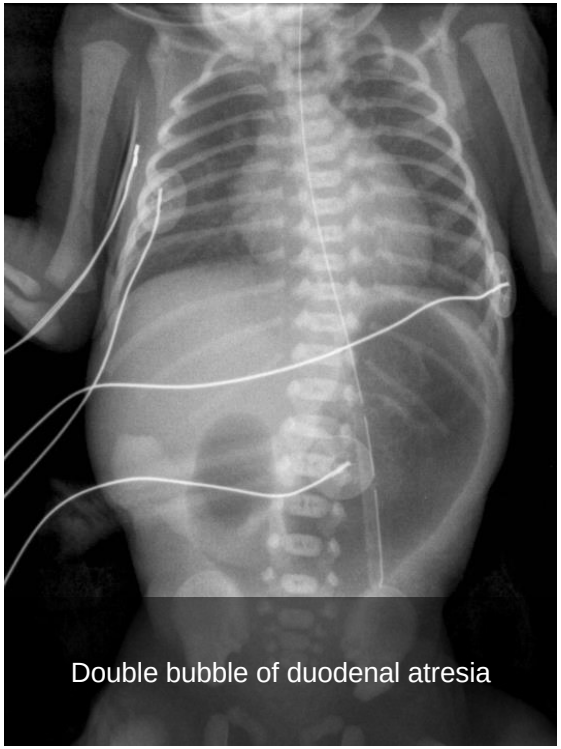
Surgical emergency since intestinal viability is at stake.

Can occur in the fetus – large calcified shadow in midabdomen on x-ray

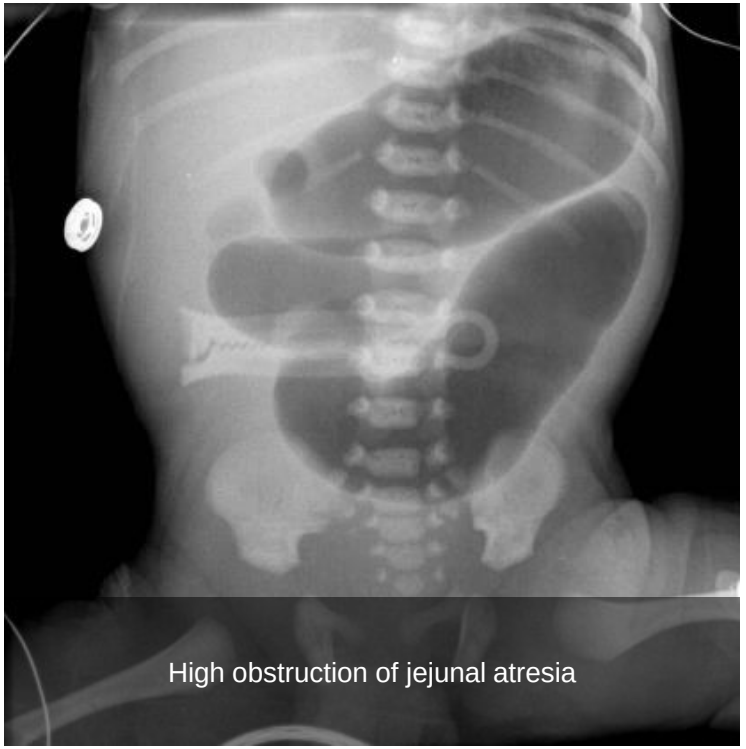
Signs of shock and sepsis can be present

UGI to evaluate for position of ligament of Treitz

Atresias



Double bubble of duodenal atresia



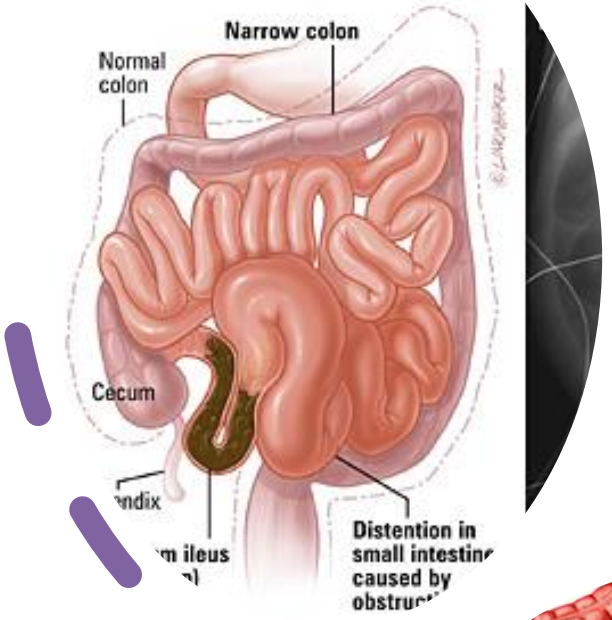
High obstruction of jejunal atresia

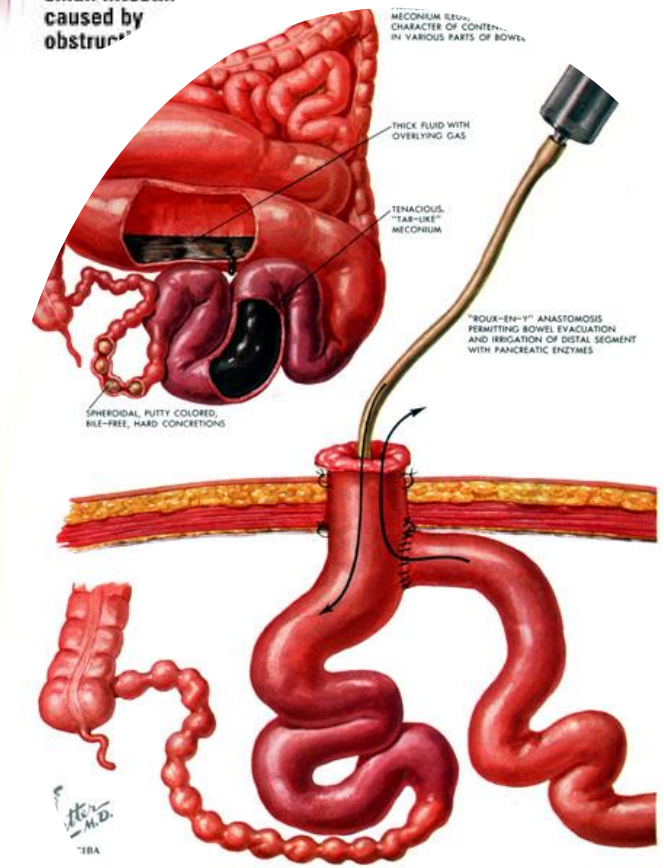
Midgut Anomalies

- **Clinical correlation** –
  - Duodenal atresia is due to failed canalization.
  - Omphalocele results from failure of the midgut loop to return to the abdomen.
  - Meckel's diverticulum occurs when a remnant of the yolk sac (Vitelline duct) persists.
  - Malrotation occurs if the midgut does not complete the rotation prior to returning to the abdomen.

Meconium Ileus

- Obstruction of bowel by thick tenacious meconium
- 30% of intestinal obstruction in neonates
- Frequent cause of meconium peritonitis
- Most are associated with cystic fibrosis (but only 15% of infants with CF will have meconium ileus)
- Abdominal distention is typically present at birth





Meconium Ileus (cont)

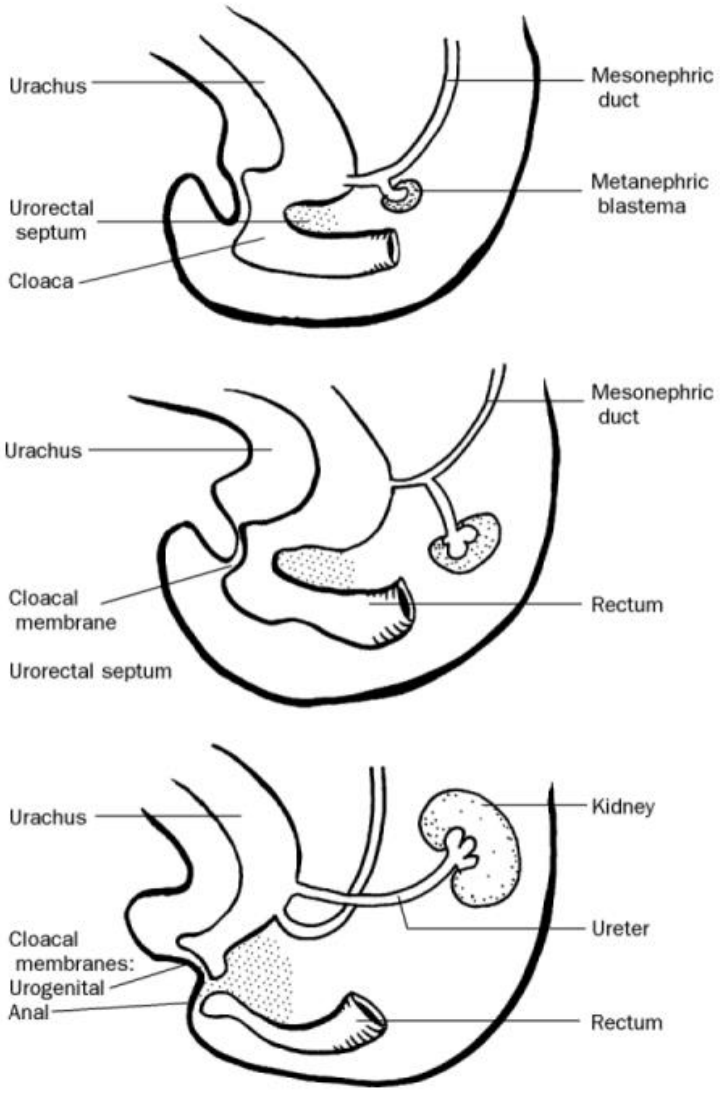
- Diagnosis made with contrast enema
- Gastrografin enema with aggressive hydration can be used to treat some
- Operative evacuation of meconium
- May require ostomy
- Proximal bowel dilated and distal bowel may be very small (microcolon) and require time to dilate with use

Meconium plug

- Difference between meconium ileus and meconium plug is site and severity of obstruction
- Preterm infants, infants of diabetic mothers, IUGR babies, otherwise ill babies
- Treatment with glycerin suppositories and warm saline enemas
- May require contrast enema to make diagnosis
- Normal stooling pattern should follow evacuation of plug

Hindgut

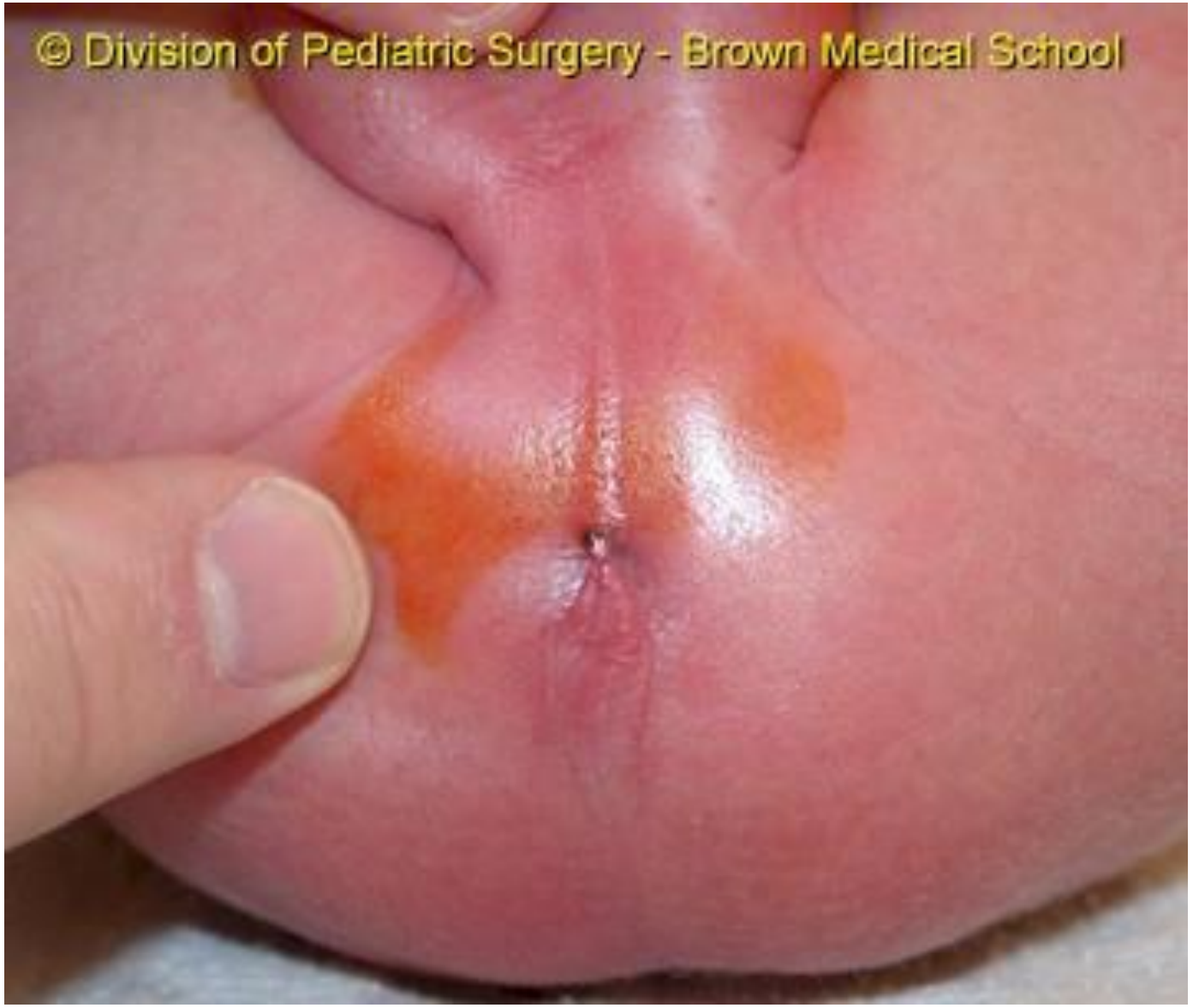
- Distal 1/3 of the transverse colon, descending colon and sigmoid colon develop from the cranial end of the hindgut.
- Upper anal canal develops from the terminal end of the hindgut with the urorectal septum dividing the upper anal canal and the urogenital sinus.



http://www.med.umich.edu

Hindgut Anomalies

- **Clinical Correlation**
  - Anorectal agenesis occurs if the urorectal septum does not develop appropriately.
  - VACTERL Association
    - Vertebral anomalies, anal defects, cardiac defects, TEF, Renal and Limb defects
  - Hirschsprung disease – failure of the neural crest cells to form the myenteric plexus (see Enteric Nervous System).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Imperforate Anus

May pass meconium if a rectovaginal or rectourinary fistula exists.

Low imperforate anus: the rectum has descended through the puborectalis sling and exists as a fistula on the perineum.

- May see mec on the perineum, may be seen in the rugal folds or scrotum of males and vagina of females.
- These fistula may be dilated to temporarily relieve obstruction

Imperforate Anus (cont)

- High imperforate anus: rectum ends above the puborectalis sling.
- No perineal fistula, but may have urinary fistula
- Temporary colostomy is necessary in all babies with high imperforate anus.

Enteric Nervous System

Collection of neurons in the GI tract.

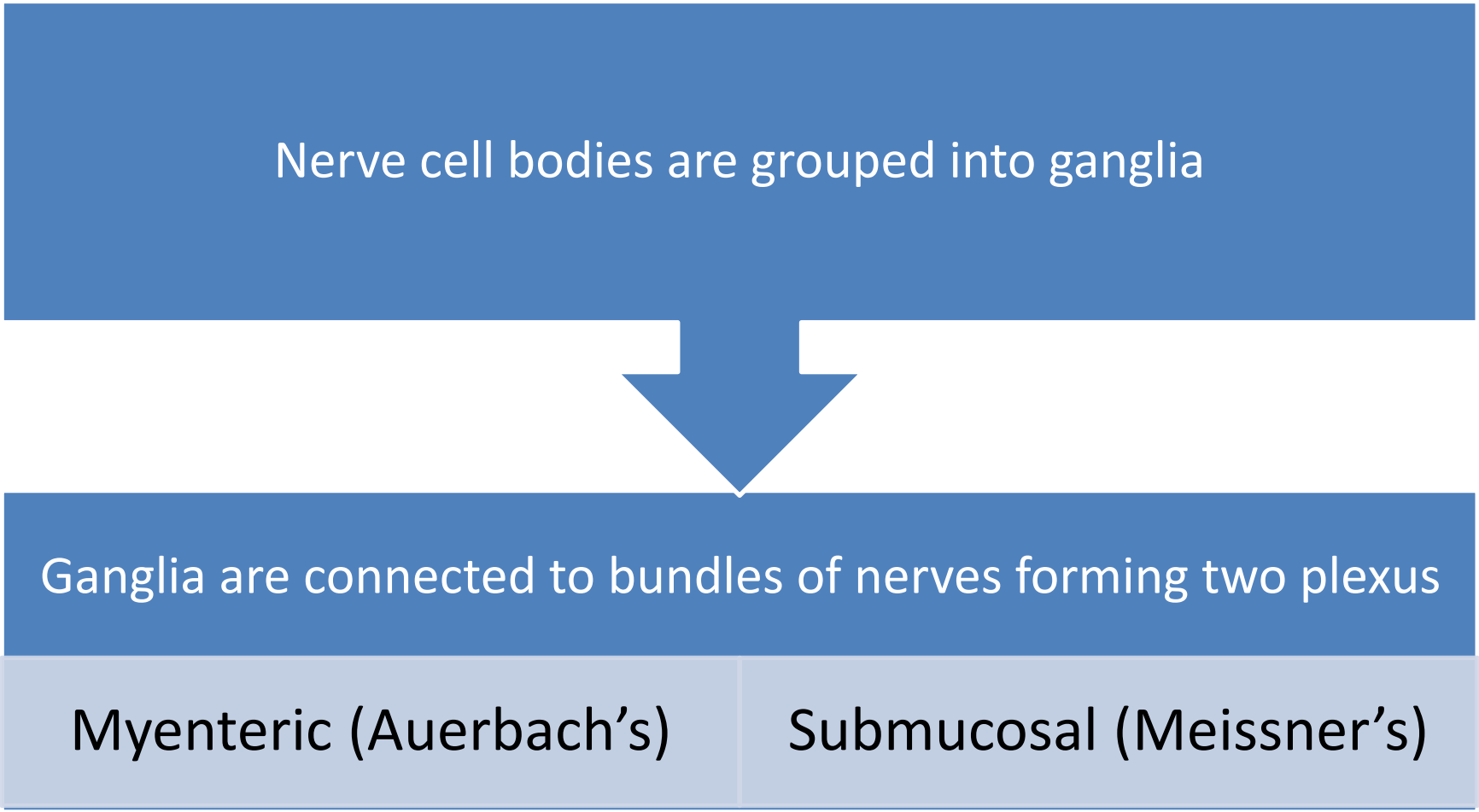
Controls motility, exocrine and endocrine secretion and microcirculation.

Regulates immune and inflammatory process.

Functions independent of CNS.

Image from: Young. Gut 2000

Development of the Enteric Nervous System



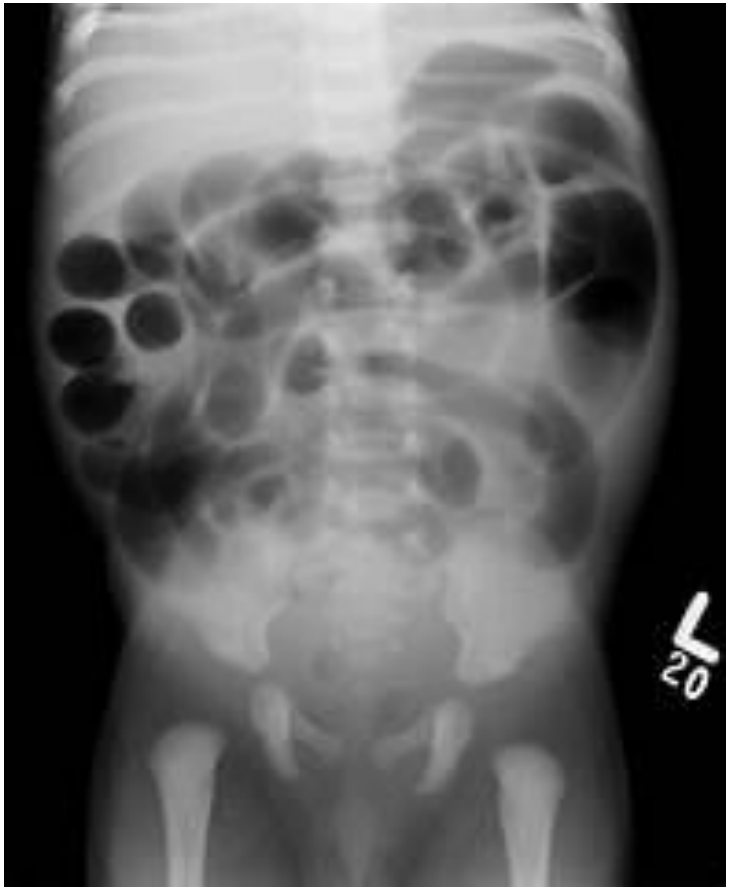
http://en.wikipedia.org/wiki/Enteric_nervous_system

Case

- Call from a peripheral hospital----48 hour old term infant. No stool. Abdomen slightly distended, no longer tolerating feedings.
- Otherwise looks fine.

Hirschsprung's Disease

- Congenital disorder
- 1:5000 live births
- Failure of neural crest cells to colonize the entire gut resulting in an aganglionic zone
  - Tonic constriction of aganglionic section
- Long (20%) and Short Segment (80%)
  - Short segment 4:1 male:female
- Isolated anomaly in 70% of cases
- Multiple genes and modifier genes identified
  - Not mendelian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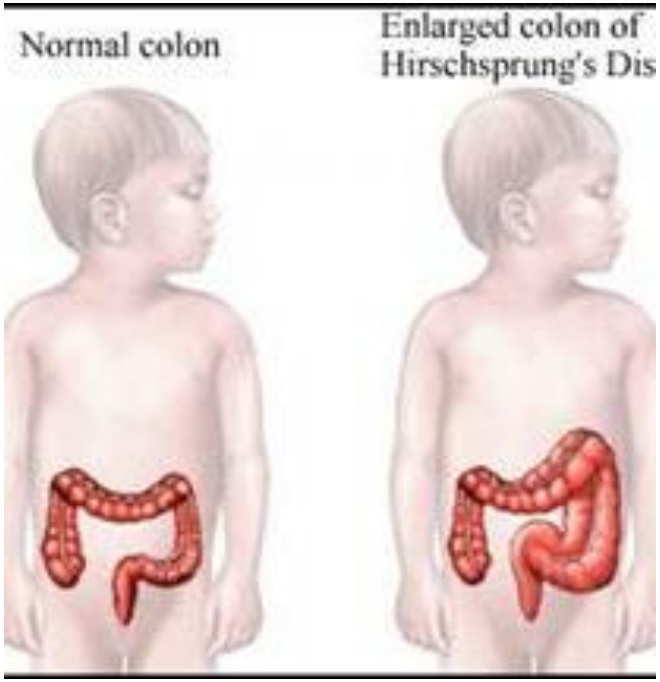
Hirschsprung's Disease

- Colonic agangliosis
- Extent can vary from very short segment of rectal tissue to entire colon
- Should be considered in any baby who does not pass stool spontaneously by 24 hours of age.
- Diagnosis by rectal biopsy to look for ganglion cells



- Barium enema can show transition zone
- Short segment disease can be treated with rectal irrigations followed by primary pull through procedure
- Longer segment disease requires ostomy followed by pull through when older (months usually).

Hirschsprung's Disease (cont)

Intestinal obstructions

- Atresias – duodenal, jejunal, colonic
- Meconium ileus
- Meconium plug
- Hirschsprung's disease
- Imperforate anus
- Malrotation with volvulus
- Adhesions and strictures