Enteric Duplications

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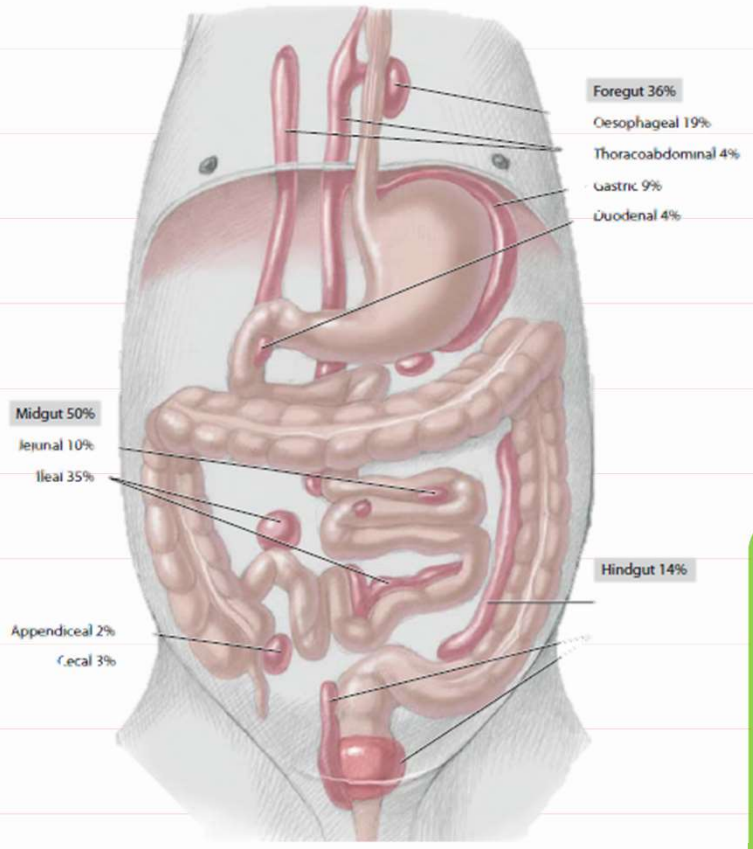
TIKUR ANBESSA

- General Considerations
- Esophageal
- Gastric
- Duodenal
- Small bowel
- Colonic
- Rectal

Introduction

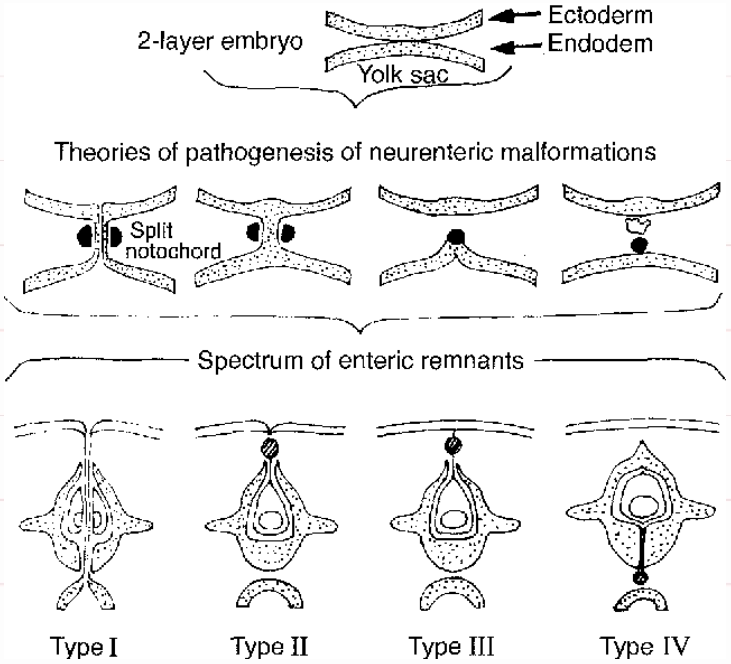
- 1 in 4,500 births
- Can occur anywhere from mouth to anus
  - Small bowel – most common (50%)
  - Esophagus – 2nd most common (15%)
  - Other – colonic>>gastric>>duodenal
- Usually single, cystic
- Variable presentation
- Resection is recommended even if asymptomatic (risk comp. including malignancy)



Embryology

One theory doesn't explain all types & ass. anomalies

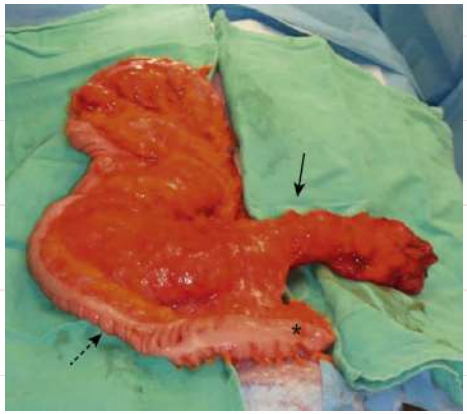
- Partial twinning: explain GU duplication in hindgut
- Split notochord: explain ass. with spinal defect
- Canalization defect: persistent fetal diverticula
- Vascular accident: explain ass, with atresia



Split notochord theory: Abnormal adherence of the endoderm of the roof of the primitive gut to the notochord

Pathology

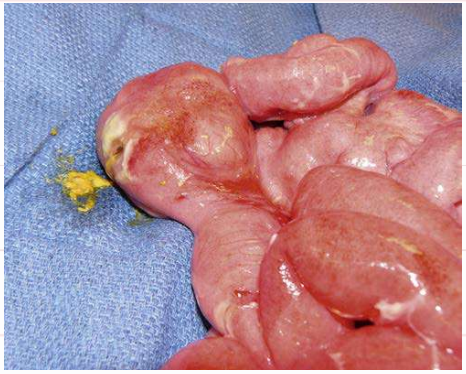
- **2 Types**
  - Cystic (majority)
  - Tubular (ectopic mucosa more common)
- Always found on **mesenteric side** (unlike Meckel's)
  - May have communication with lumen
  - can be separate from GI tract (no common wall)
- Three common findings
  - Well developed smooth muscle coat
  - Epithelial lining
  - Attachment to alimentary tract
- **Ectopic mucosa** in 1/3 (Gastric, pancreatic, Respiratory)



Native colon (dotted arrow) and Tubular duplication on mesenteric side (solid arrow)

Presentation

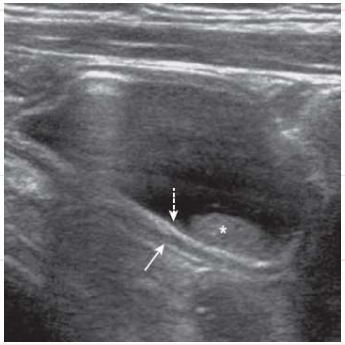
- Mass - Asymptomatic (prenatal US, <2yrs)
  - Compressive symptoms
    - Thoracic - Dysphagia, breathing difficulty
    - Abdomen - GOO, intestinal obstruction
  - Intussusception, segmental volvulus
- Heterotopic mucosa (1/3- gastric, pancreatic, resp.)
  - Bleeding, Pain, perforation
  - Late malignant degeneration (gastric, rectal)
- Ass. Anomalies (half),
  - Foregut – vertebral (bifid/hemivertebre)
  - Midgut – malrotation, atresia
  - Hindgut – GU anomalies, exstrophy



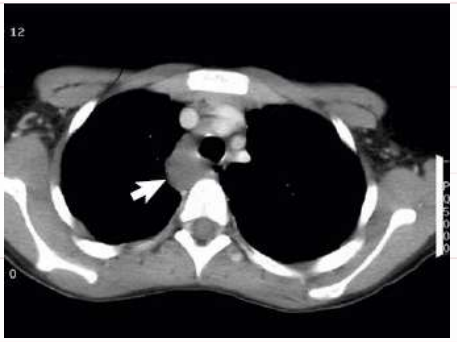
Ectopic gastric mucosa can lead to ulceration with bleeding or perforation. Pressure necrosis from an adjacent duplication also can lead to hemorrhage or perforation.

Imaging

- **Abdominal Ultrasound**
- **Chest CT**
- **Spine MRI-** if suspect communication
- **Barium enema** - may show communication to GI or GU tract in hindgut lesion



Typical US shows double wall sign (hypoechoic muscle layer b/n two hyperechoic layers - serosa and mucosa).



Foregut lesion appear as mediastinal mass on x-ray and require further workup with CT for preop planning.
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Surgery

Procedure should not be more radical than necessary to eliminate complaints and prevent recurrence

- **Cyst resection/Enuclation** (difficult without violating native bowel b/c common blood supply)
- **Resection and anastomosis** (more plausible)
- **Partial excision with mucosal stripping** (long tubular duplication)

Prophylactic surgery done after a few months in asymptomatic neonates

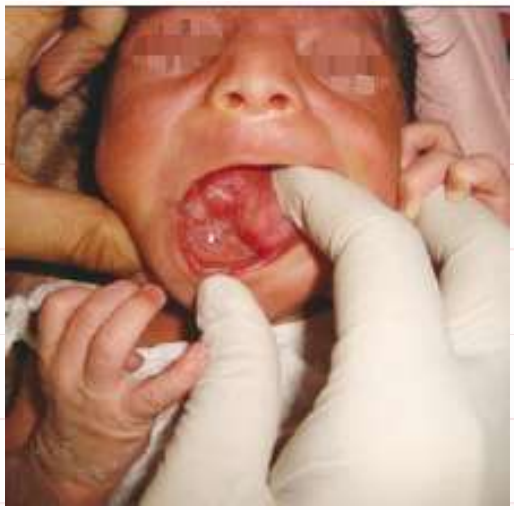
Complications

- **Incomplete excision**
  - cyst recurrence
  - infection, meningitis (neurenteric cysts)
  - GI bleeding and perforation

Lingual

- Mostly in anterior 2/3
- May cause airway comprise
- May contain gastric mucosa
- Best treated by complete excision

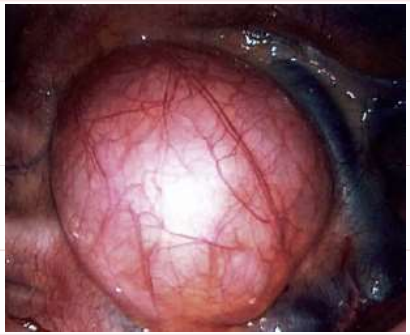
  (intraoral, sublingual approach)



Esophageal

- Mostly asymptomatic
- 40% contain ectopic mucosa
- Majority are …
  - Thoracic (rt side)
  - cystic
  - Share a wall but no communication
- Abdominal US
  - 25% synchronous duplication
- MRI
  - neuroenteric cyst (extension to spine) in 50%

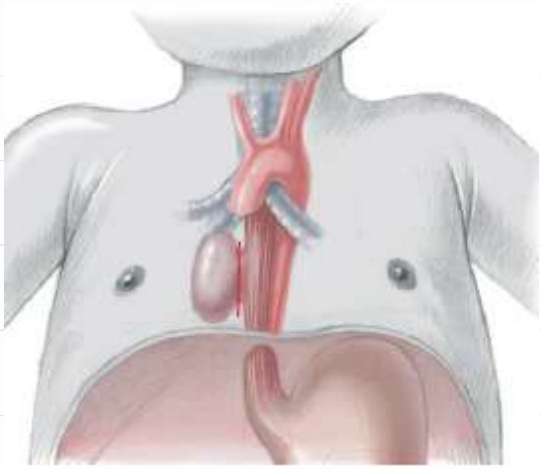
    thoracic lesions have communication

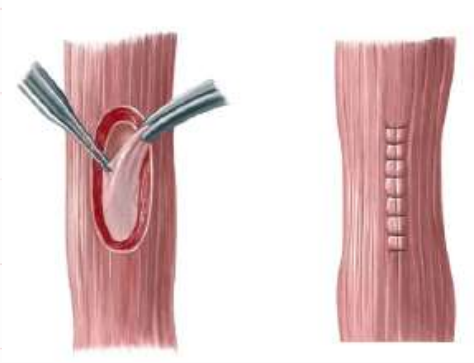


Cystic esophageal duplication

Esophageal: Resection

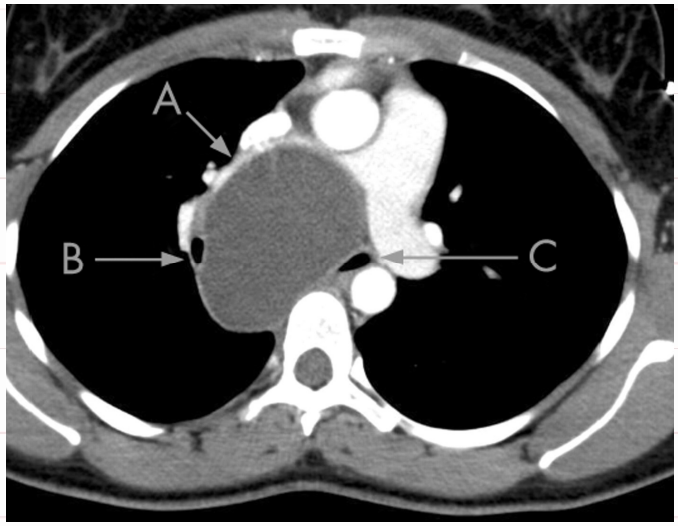
- Thoracic - rt side thoracoscopy/ open
- Cervical – supraclavicular, neck extended
- **Principles**
  - Mobilize close to its wall
  - Avoid vagus, phrenic, lymphatic duct
  - Transect close to esophagus
  - Remove residual mucosa
  - Repair residual muscular defect
  - resect adjacent esophageal stricture or ulcer





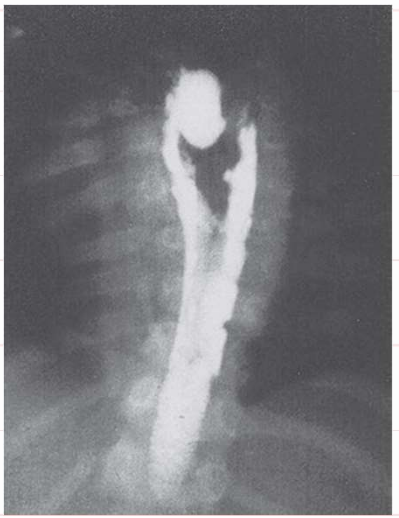
**Bronchogenic cyst

- variant with cartilaginous wall and ciliated respiratory epithelium.
- Thought to arise from forgot destined for resp.
- noncommunicating
- often located near the tracheal bifurcation but can occur anywhere along the alimentary canal



Thoracoabdominal

- esophageal duplication extending to abdomen (2%)
  - communicate with stomach/intestine
- Higher risk of ectopic mucosa
- Majority are
  - Tubular
  - Communicate at jejunum
- MRI
  - High incidence of vertebral anomalies (88%)

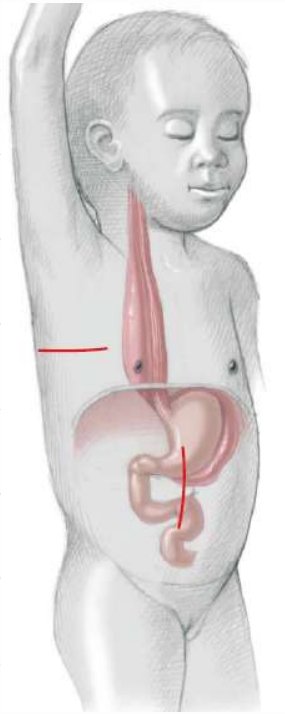


myelogram shows the filling defect caused by a neuroenteric cyst

Thoracoabdominal: Resection

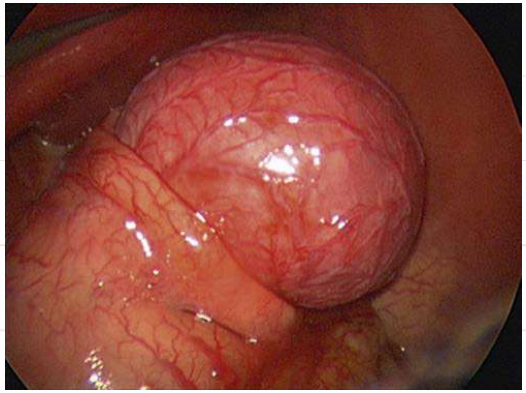
One stage thoracoabdominal approach (incomplete excision may cause meningitis, GI bleeding, perforation, and respiratory comp.)

- **Posteriolateral thoracotomy**
  - Dissect free from bony attachment
  - Fistula ligation/lobectomy (Some PUD may erode to lung- hemoptysis)
  - Trace distal to diaphragm, pull up and ligate
- **Laparotomy**
  - Alternatively thoracic component passed through diaphragm and ligated in abd
- **Laminectomy** – if intraspinal component



Gastric

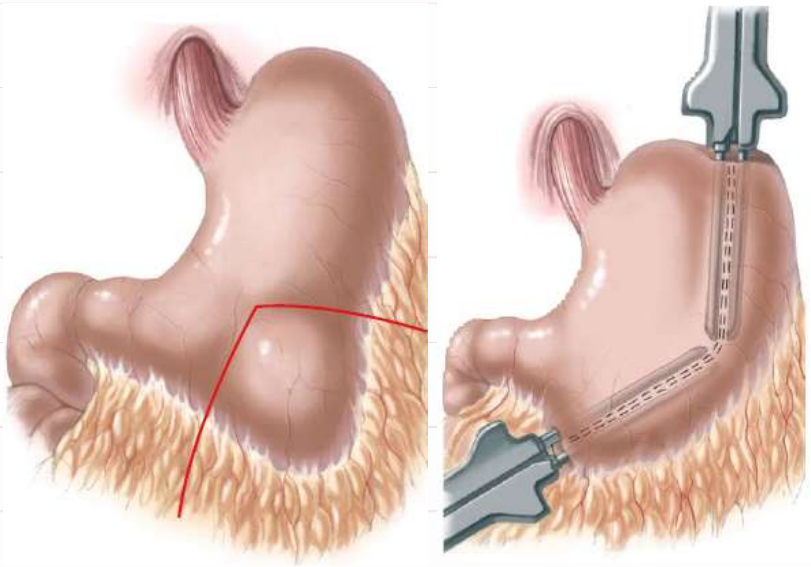
- Become symptomatic earlier in life
- Majority are
  - cystic
  - From greater curvature
  - Don’t communicate with lumen
- CT
  - May confuse with choledochal cyst or pancreatic pseudocyst



large gastric duplication from greater curvature

Gastric: Resection

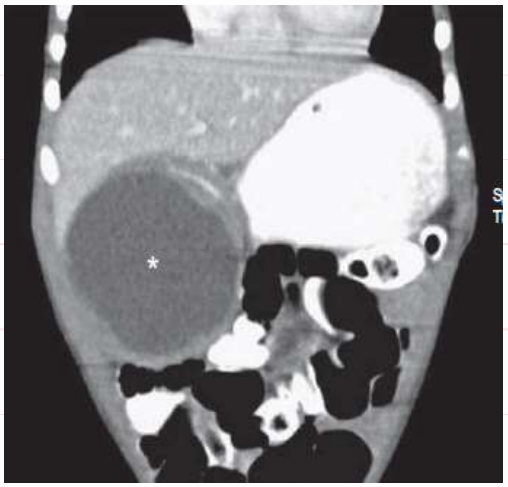
- may need **wedge resection** of stomach
- ***Partial excision and mucosal striping** for difficult location (lesser curve, GEJ, pylorus) or if large
- **Laparoscopic stapling** of septum via gastrostomies for tubular duplication (?may leave mucosa behind)



Duodenal

- May cause obst jaundice or pancreatitis
- Majority are
  - cystic
  - Don’t communicate with lumen
- CT /MRCP, ?ERCP?
  - Delineate anatomy prior to surgery
  - 30% communicate with pancreatobiliary

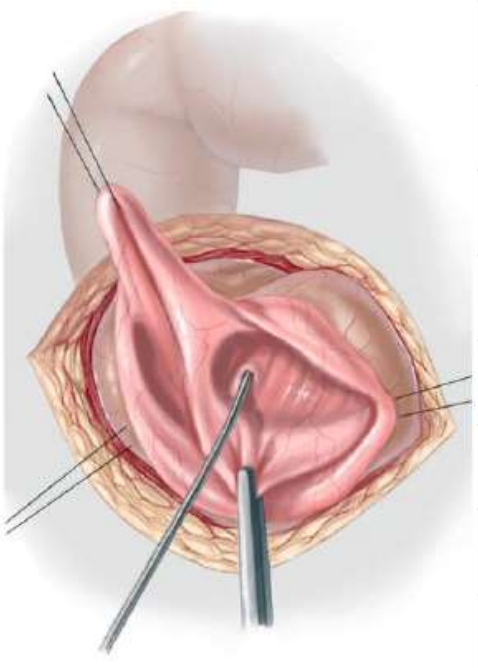
*if opening of duct is uncertain- intraop cholangiography or cholecystectomy & pass probe



CT shows large hypodense cystic structure in the right upper quadrant which represents a large duodenal duplication

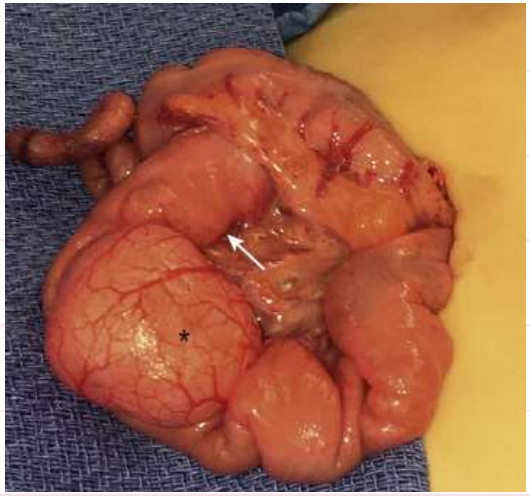
Duodenal: Resection

- **complete excision**, division of duct communication
- **partial excision and mucosectomy** of remaining
- **fenestration into the duodenal lumen** (if adjacent to the ampulla of Vater, and r/o presence of gastric mucosa with intraop biopsy) * window shold be sufficient size for free dependent drainage
- **Internal endoscopic drainage** (intraluminal duplication with no gastric mucosa)
- **roux-en-y cystjejunostomy** (intimate r/n to biliary)
- **Whipples/distal pancreatectomy** (located in pancreas and cause recurrent pancreatitis)



Small bowel

- Share common blood supply
- May share common wall or be separate
- Majority are…
  - Cystic
  - In ileum
- Ectopic mucosa (80% in tubular, 20% in cystic)
- ass. With intestinal atresia and malrotation
- 10% may have additional Abdominal or thoracic duplication



cystic small bowel duplication (asterisk) in the terminal ileum, and a second smaller duplication at the ileocecal valve (arrow)

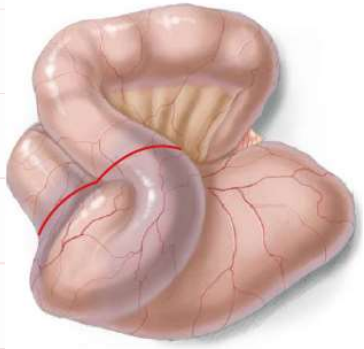
Small bowel: Resection

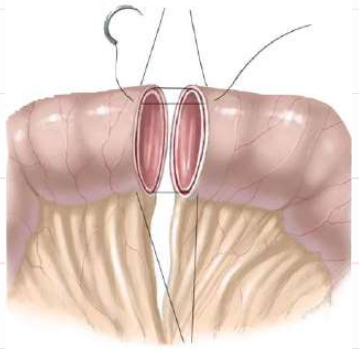
- depend on intricacy of blood supply and length
- exact point of termination of the duplicated bowel can sometimes be difficult to determine.

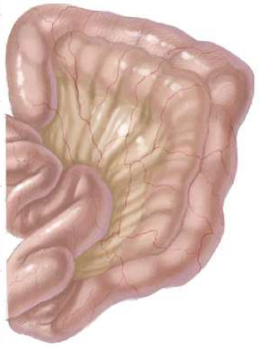
- Localized – **resected** with adjacent intestine

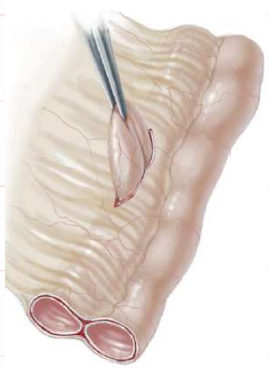
- Long tubular – **mucosal stripping** through multiple enterotomies (cautery or blunt dissection)

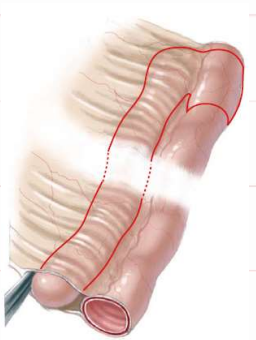
- Tubular in mesentery separate from intestine – **enuclation** without jeopardizing bowel (careful separation of the two leaves of the mesentery and division of vessels on one side only)





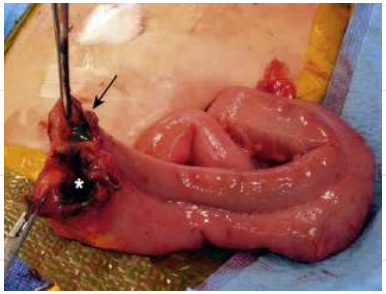


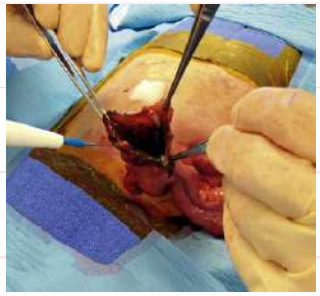




Colonic

- Rarely contain ectopic gastric mucosa
- Majority are…
  - cystic
  - In cecum
- Long tubular duplication are ass with GU anomalies
- Duplication may lie lateral or medial to the normal colon and often has a proximal connection.
- CT/MRI/contrast
  - May communicate with native colon
  - May have fistula to perineum/ GU

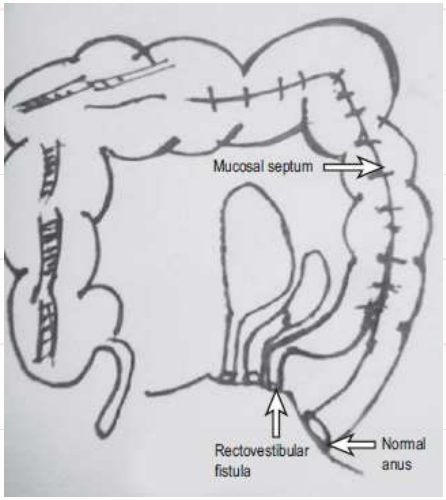




A stapler is inserted via distal enterotomy to create a common channel between the two lumens of duplicated colon

Colonic: Resection

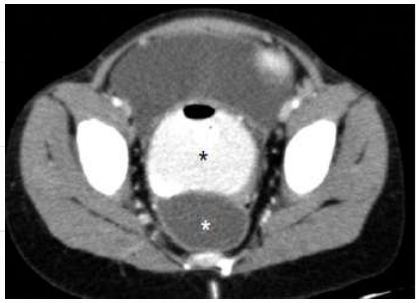
- difficult if long tubular..
- mucosal stripping not routinely needed.
- Distal communication may be created if obst
- Fistulous tracts need to be closed

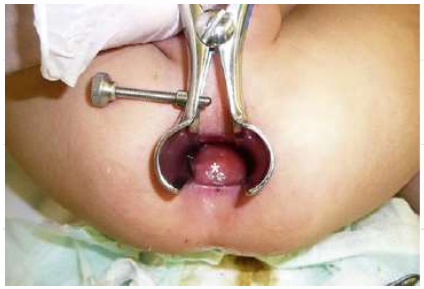


Colostomy is recommended if there are 2 perineal openings. Later the duplication can be resected at level of rectum and anastomosed to normal rectum. The mucosa in distal segment (RVF) can be excised using an endorectal pull-through technique, and the remaining muscular cuff plicated and closed.

Rectal

- Mostly found in presacral space
- Present with constipation
- Also as mucosal swelling/prolapse
- Can be ass. With ARM or fistula to other organs
- Risk of malignancy

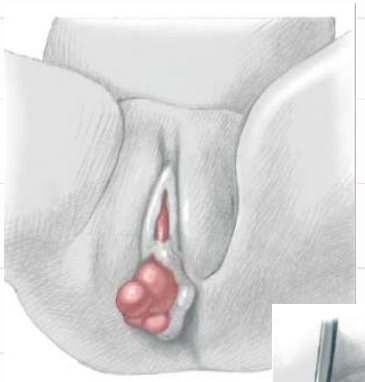


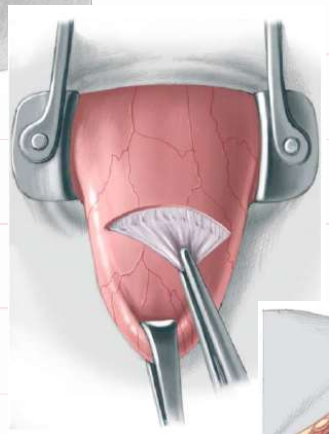


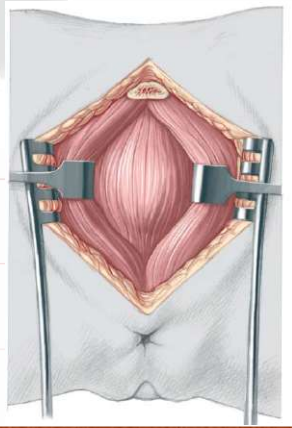
Cyst posterior to native rectum. bulge created by the duplication can be seen through the posterior wall of the native rectum

Rectal: Excision

- Complete resection b/c risk of late malignancy
- –require colostomy if large/complex
- Limited perineal excision – localized small, mucosal
- Transanal – submucosal rectal lesion
  - incise rectal mucosa over cyst and working within the submucosal plane of the rectum, the cyst is gradually dissected
- Posterior sagittal – if more extensive (extend up)

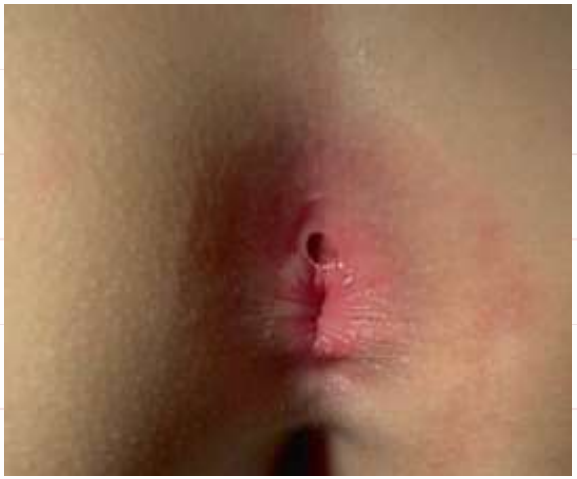






Anal duplication

- 40 cases reported in literature
- Lie posterior to normal anus
- Variable length
- More common in females
- Excision via posterior sagittal approach

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