Surgical Emergencies in the Newborn

Emergencies

- Types
  - Airway/Respiratory
  - Intestinal Obstruction
  - Intestinal Perforation
- Signs
  - Respiratory distress
  - Abdominal distension
  - Peritonitis
  - Pneumoperitoneum

Airway/Respiratory

- Neck Masses
  - **Cystic Hygromas**
  - Tracheal anomalies
- Thoracic masses/pulmonary lesions
  - **Congenital lobar emphysema**
    - Overdistension of one or more lobes (nl histological lung)
  - **Congenital cystic adenomatous malformation**
    - Multicystic mass of lung tissue, proliferation of bronchial structures at the expense of alveoli
  - Pulmonary agenesis
    - Absence of lung
  - **Congenital diaphragmatic hernia**
  - **Tracheoesophageal fistula**

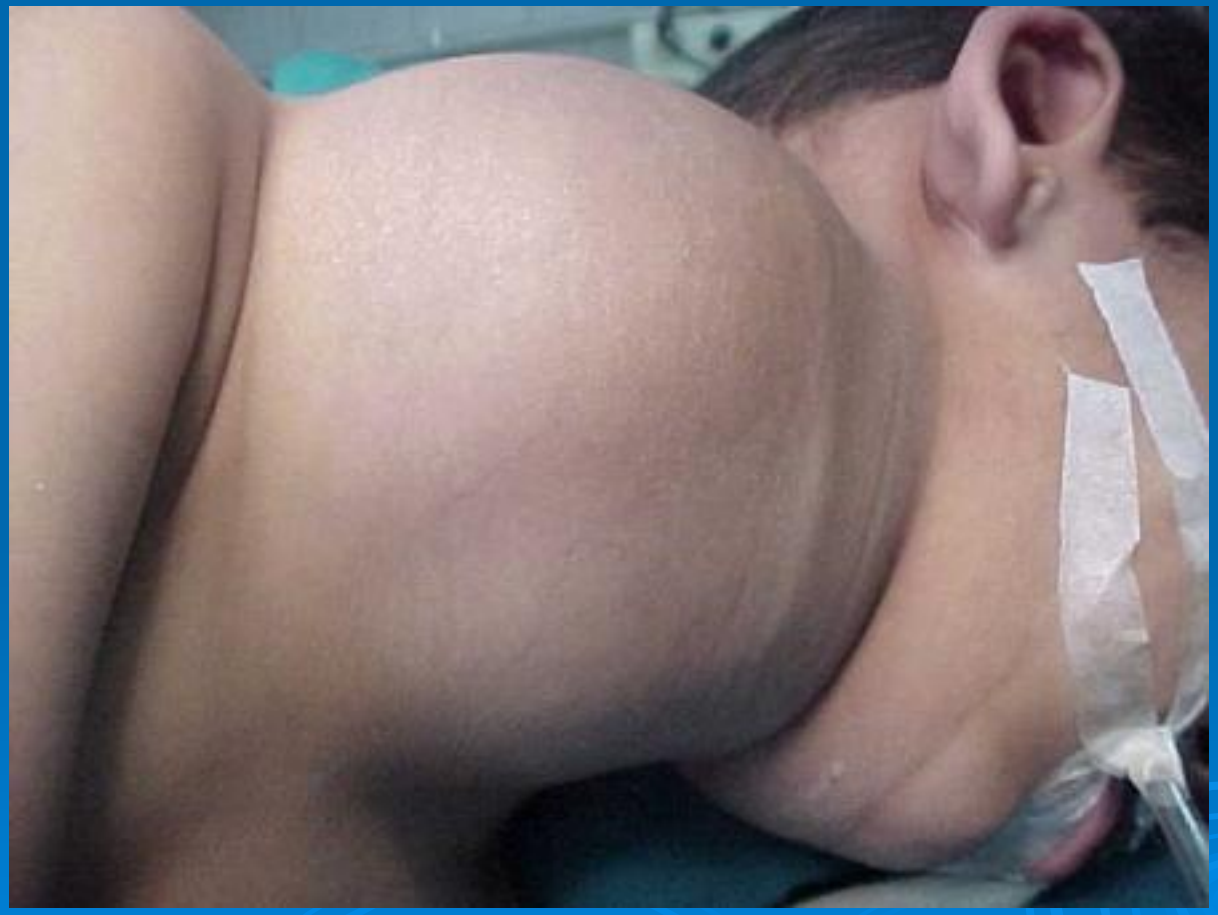
Cystic Hygroma

- Multiloculated cystic spaces lined by endothelial cells
  - Separated by fine walls containing numerous smooth muscle cells
  - Result of maldevelopment of lymphatic spaces
- Incidence about 1 in 12,000 births
  - 50-65% appear at birth, 85-90% appear by age 2
  - Neck-75%, Axilla 20%; can be seen in mediastinum, retroperitoneum, pelvis, groin
  - Nuchal/post cervical CH's have been associated with chromosomal abnormalities—high mortality rate

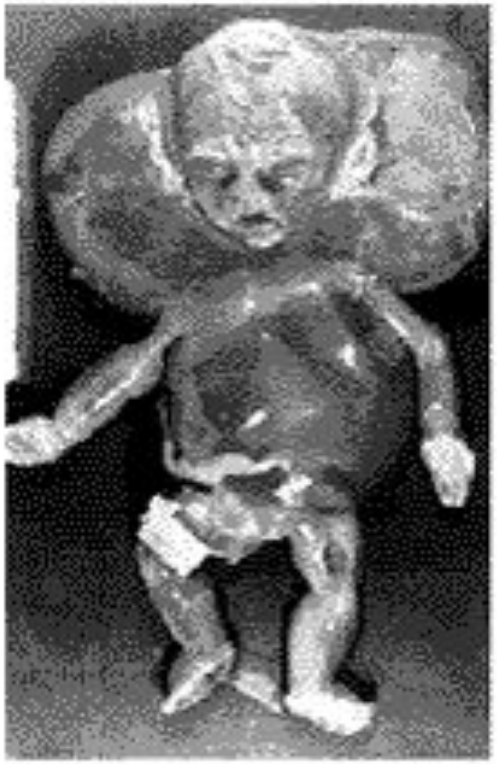
Cystic Hygroma

- Complications
  - Respiratory—large hygromas can extend into oropharynx and trachea
  - Inflammation/Infection
  - Hemorrhage
- Treatment
  - Dependent on size, location, symptoms/complications
  - Some pts require emergent surgery due to airway compromise
  - Best treatment is complete excision
  - Aspiration typically not effective due to rapid refilling of fluid
  - Sclerotherapy—Bleomycin, OK-432 (no longer available in US), doxycycline, fibrin glue

Cystic Hygroma



Cystic Hygroma



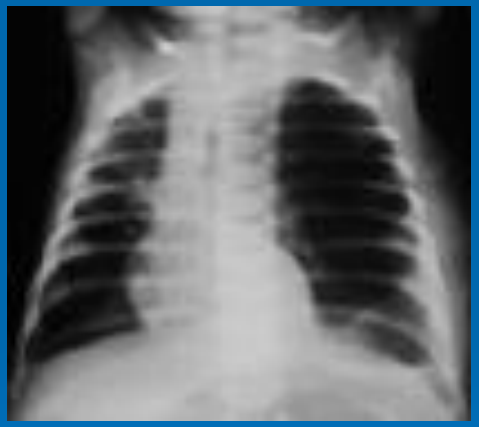
Congenital Lobar Emphysema

- Postnatal overdistension of one or more lobes of histologically normal lung
  - Probably due to cartilaginous deficiency in the tracheobronchial tree
  - Obstruction causing the overdistension may be due to
    - 1—chondromalacia of bronchi
    - 2—extrinsic pressure on bronchus by anomalous pulmonary vein or abnormally large PDA
    - 3—idiopathic
- Location
  - LUL 47%, RML 28%, RUL 20%; lower lobes <5%; Bilat rare

Congenital Lobar Emphysema

- Diagnosis
  - Usually can be made by plain CXR; Chest CT and V/P scans may be helpful
- Treatment
  - May require urgent surgical decompression with lobectomy
  - Selective bronchial intubation
  - Sometimes see spontaneous resolution—need close observation

Congenital Lobar Emphysema



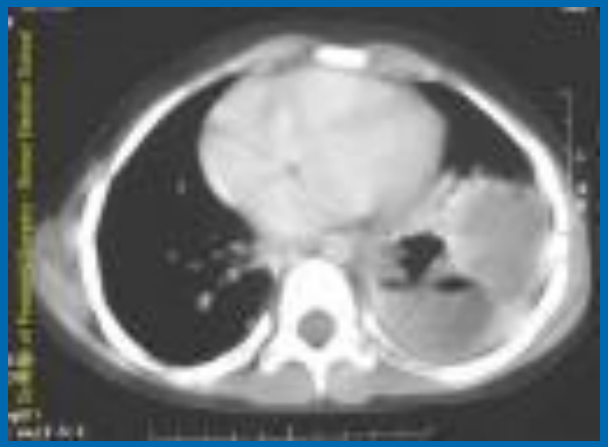
Congenital Cystic Adenomatous Malformation (CCAM)

- Mass of cysts lined by ciliated cuboidal or columnar pseudostratified epithelium
- Three types
  - I—few large cysts >2cm; thick walls, normal alveoli between the cysts; ciliated pseudostratified columnar epithelium
  - II—numerous small cysts <1cm, thin muscular coat, large alveolar-like structures between the cysts; ciliated cuboidal to columnar epithelium; assoc w/other congenital anomalies
  - III—bulky firm masses of folded ciliated and non-ciliated cuboidal epithelium and thick layer of smooth muscle; often occupy the entire lobe or lobes of lung
- More common on the left side, 2% bilateral

CCAM

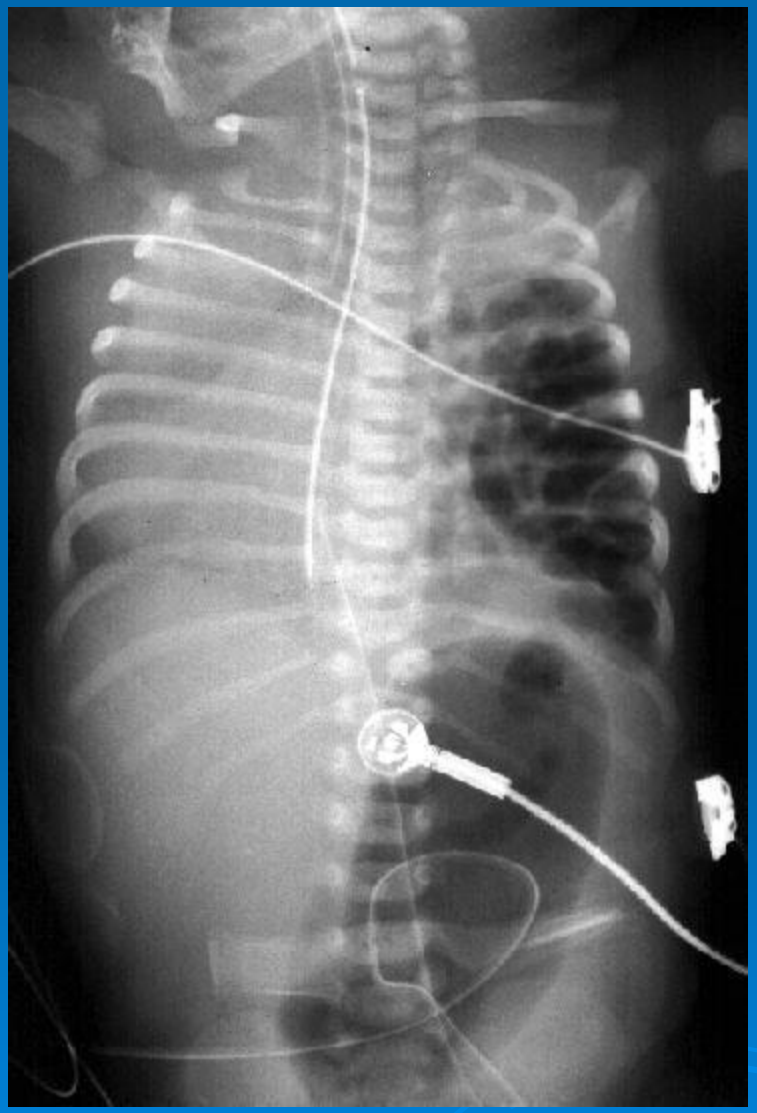
- Diagnosis
  - CT scan allows differentiation of types
  - Some can be diagnosed on prenatal US
- Treatment
  - Surgical excision, typically anatomical lobe resection, due to risk of infection, malignant transformation
  - Some are performing fetal aspiration

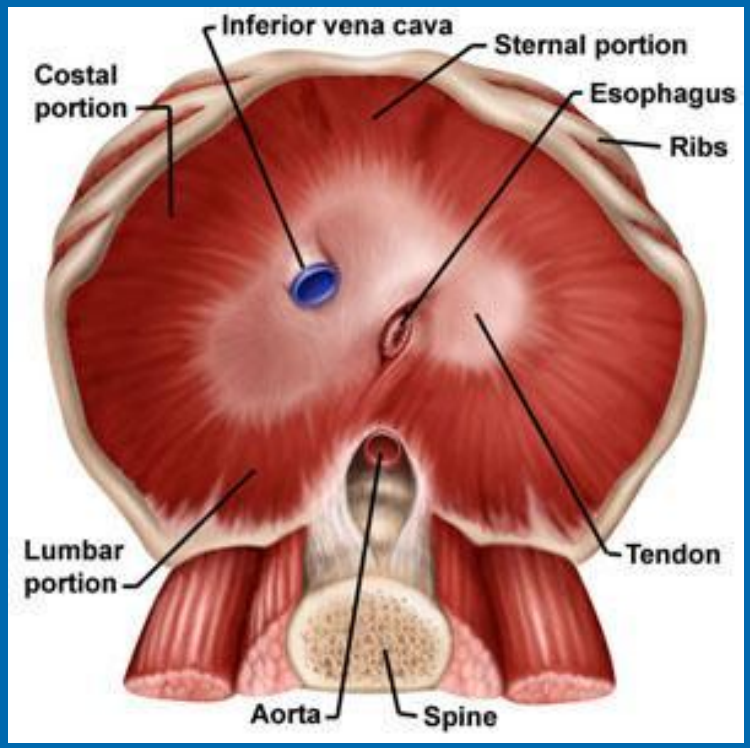
CCAM

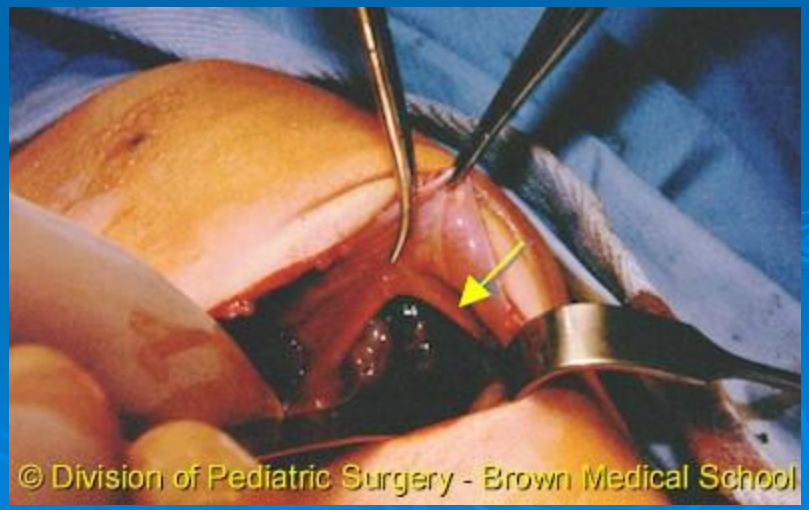


Congenital Diaphragmatic Hernia

- Intro
  - 1 in 200-5000 live births, females >males
  - Etiology unknown
  - Large percentage of fetuses are stillborn
  - Still high mortality of those that make it to birth
- DX
  - Frequently made prenatally
  - CXR
- Treatment
  - Respiratory support
  - ECMO
  - Primary closure or patch closure when pt stable




Inferior vena cava
Sternal portion
Costal portion
Esophagus
Ribs
Lumbar portion
Tendon
Aorta
Spine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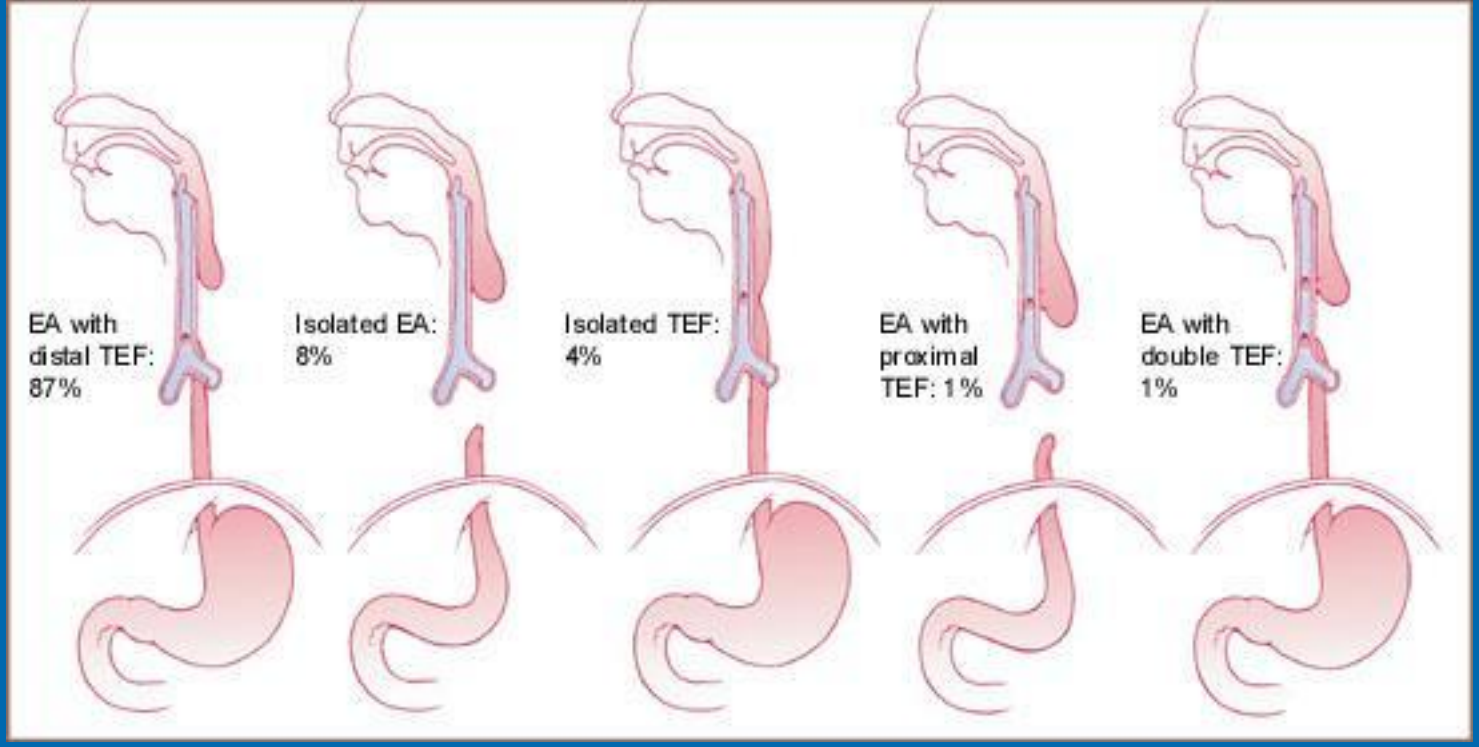
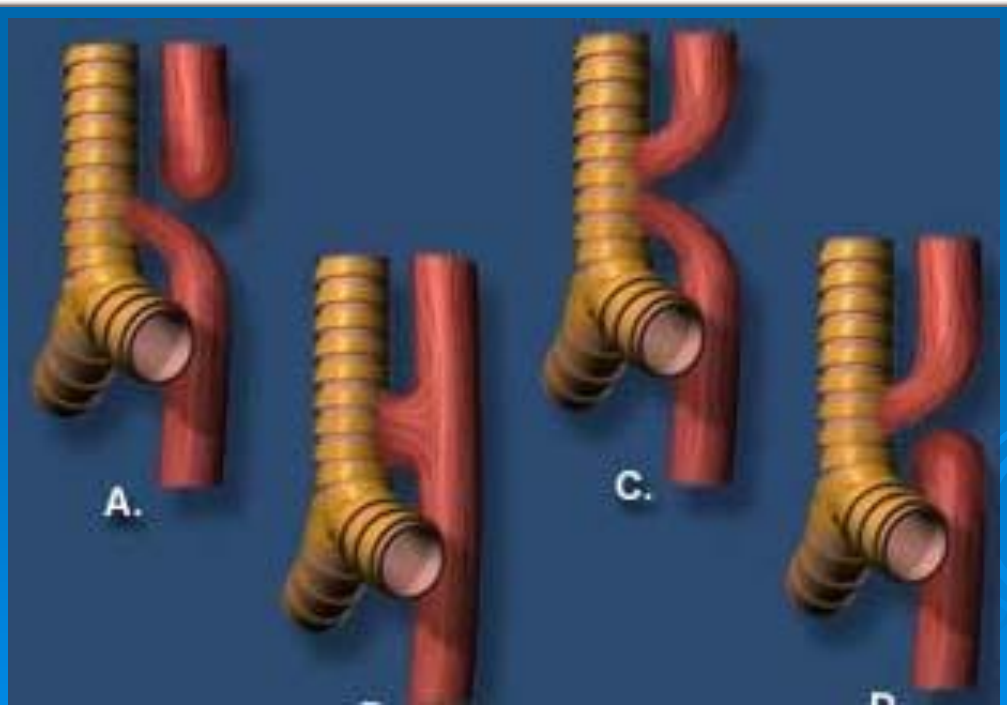
Tracheoesophageal Fistula and Esophageal Atresia


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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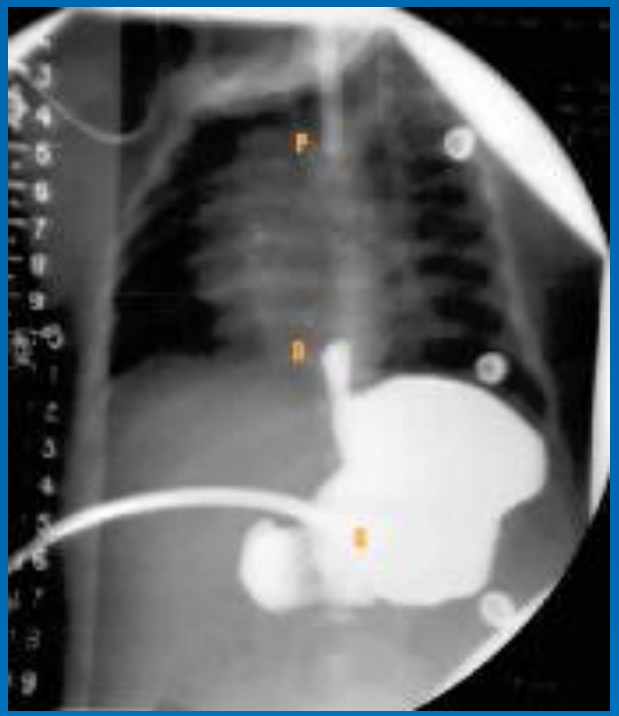


Intestinal Obstruction

- Incidence approx 1 per 500-1000 live births
- Approx 50% due to atresia or stenosis
- Majority of neonates present shortly after birth

Anatomic Differentiation

- Upper GI
  - Duodenal atresias/webs
  - small bowel atresias
  - malrotation/midgut volvulus
  - GERD
  - Meconium ileus
  - pyloric stenosis
  - Inguinal hernia
  - NEC

Anatomic Differentiation

- Lower GI
  - Colonic atresia
  - Meconium plug
  - Hirschsprung's
  - Small Left Colon Syndrome
  - Magalocystis-Microcolon-Intestinal Hypoperistalsis Syndrome
  - Imperforate anus

Urgency to Treat

- Emergencies
  - Free air on KUB
  - Peritonitis
  - Acute increase in abd distension
  - Clinical deterioration (incr pressors, dec platelets, worsening acidosis)
  - Abd wall cellulitis/discoloration

Urgency to Treat

- Further workup
  - Contrast enemas for distal obstructions
  - KUB/Cross-table lateral
  - Milk Scans for GERD
  - UGI for malrotation/proximal atresias

Common Disorders

- NEC
- Duodenal Atresia
- Small Bowel Atresia
- Malrotation/Volvulus
- Hirschsprung's

NEC Con't

- Presentation
  - distension, tachycardia, lethargy, bilious output, heme pos stools, oliguria
- DX
  - clinical
  - KUB may show pneumatosis, fixed loop, free air, portal venous gas, ascites

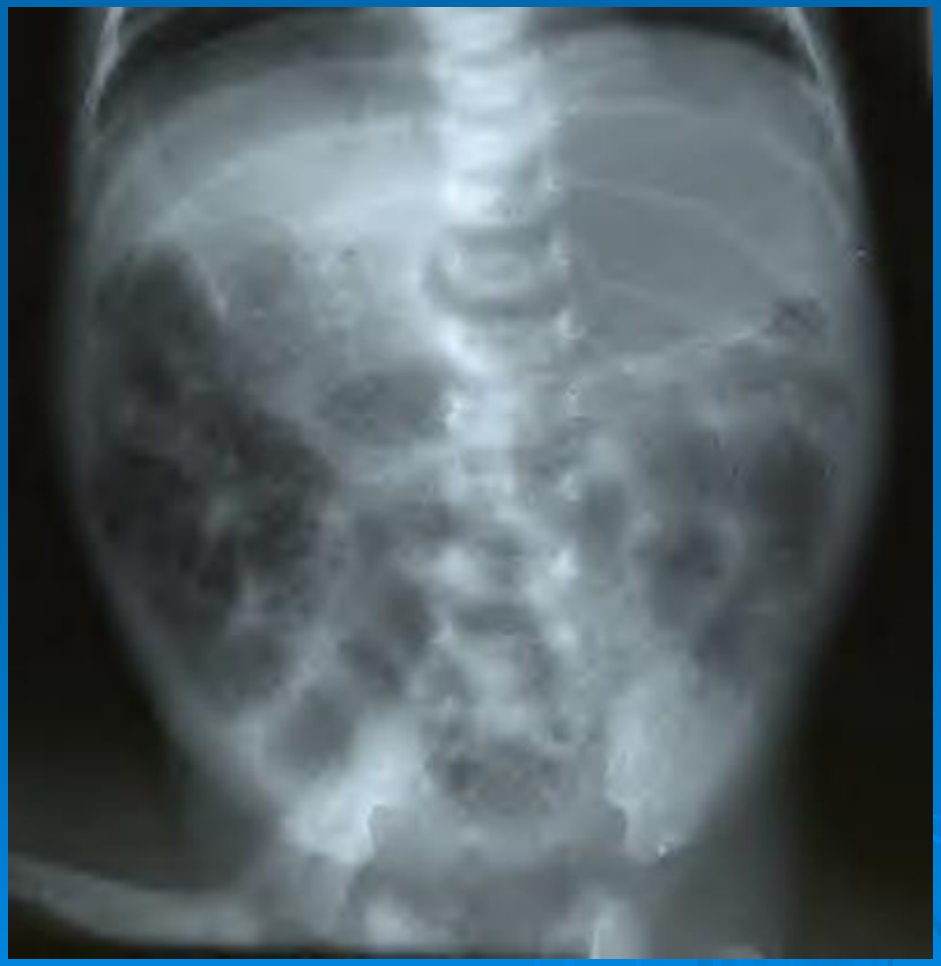
NEC Treatment

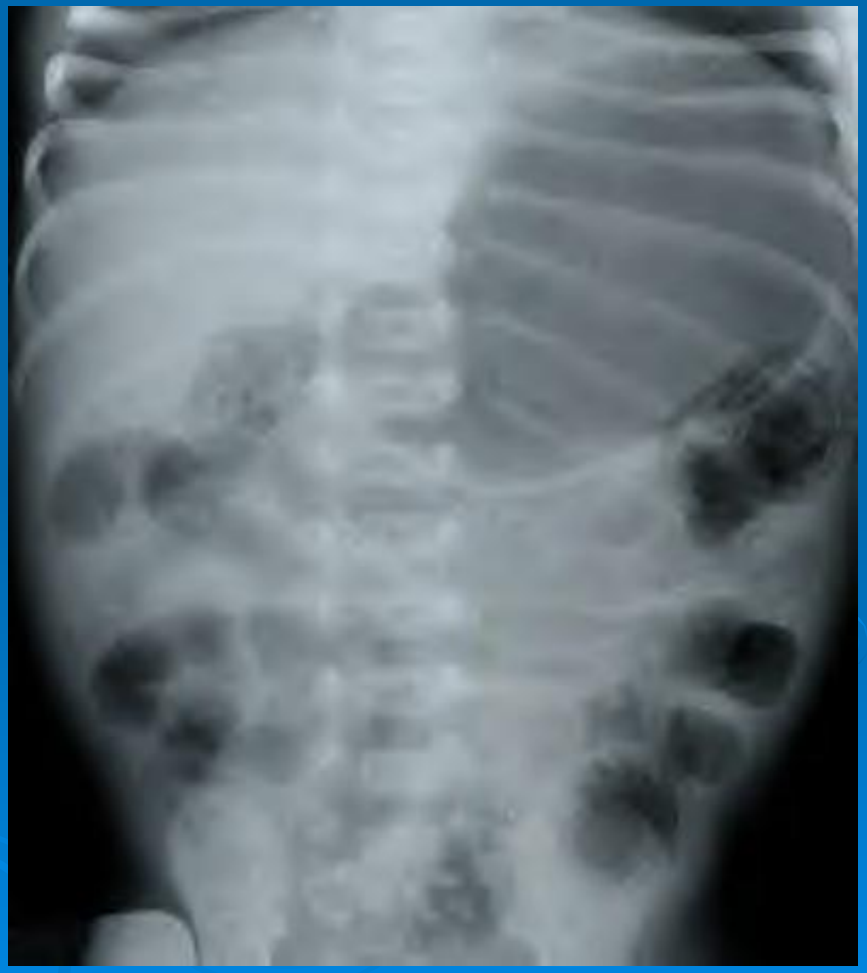
- Medical
  - NPO, sump tube, Broad Abx after cx's drawn, serial KUB/lateral x-rays, frequent abd exams
- Surgical indications
  - Free air
  - Abd wall Cellulitis
  - Fixed loop on KUB
  - Clinical deterioration

NEC Outcomes

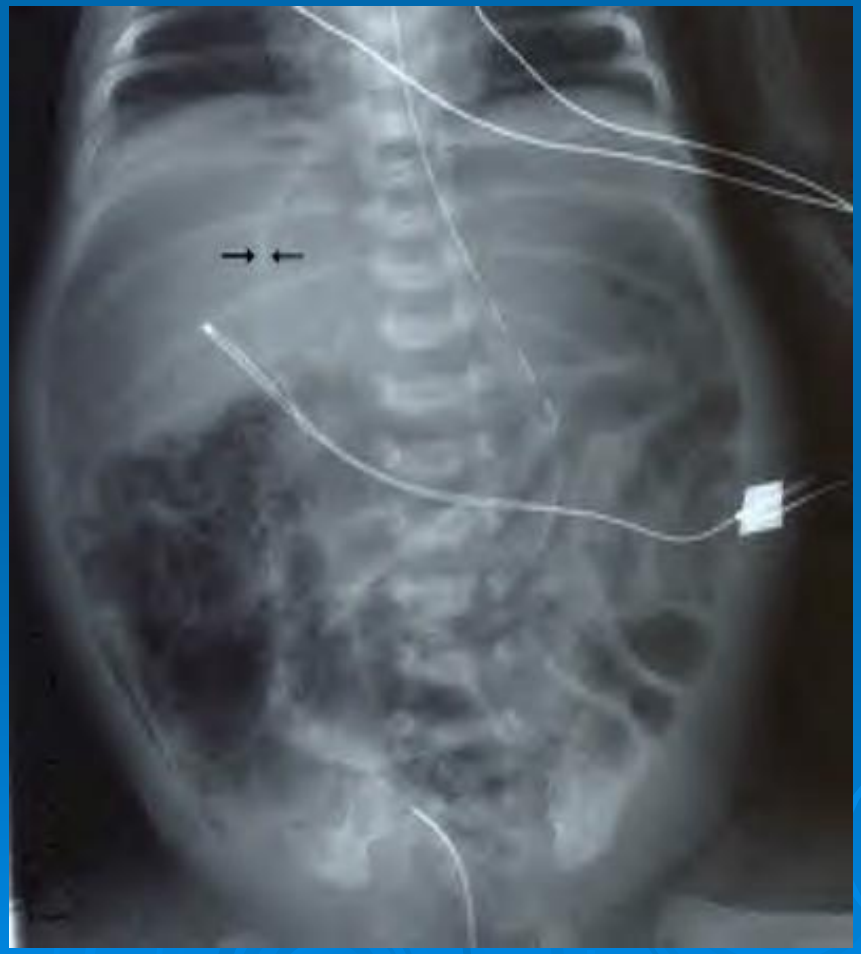
- Overall survival ~ 80%, improving in LBW
- In pts w/perforation, 65% perioperative mortality, no perf--30% mortality
- 25% of Survivors develop stricture
- 6% pts have recurrent NEC
- Postop NEC--Myelomeningocele, Gastroschisis--45-65% mortality

Pneumatosis

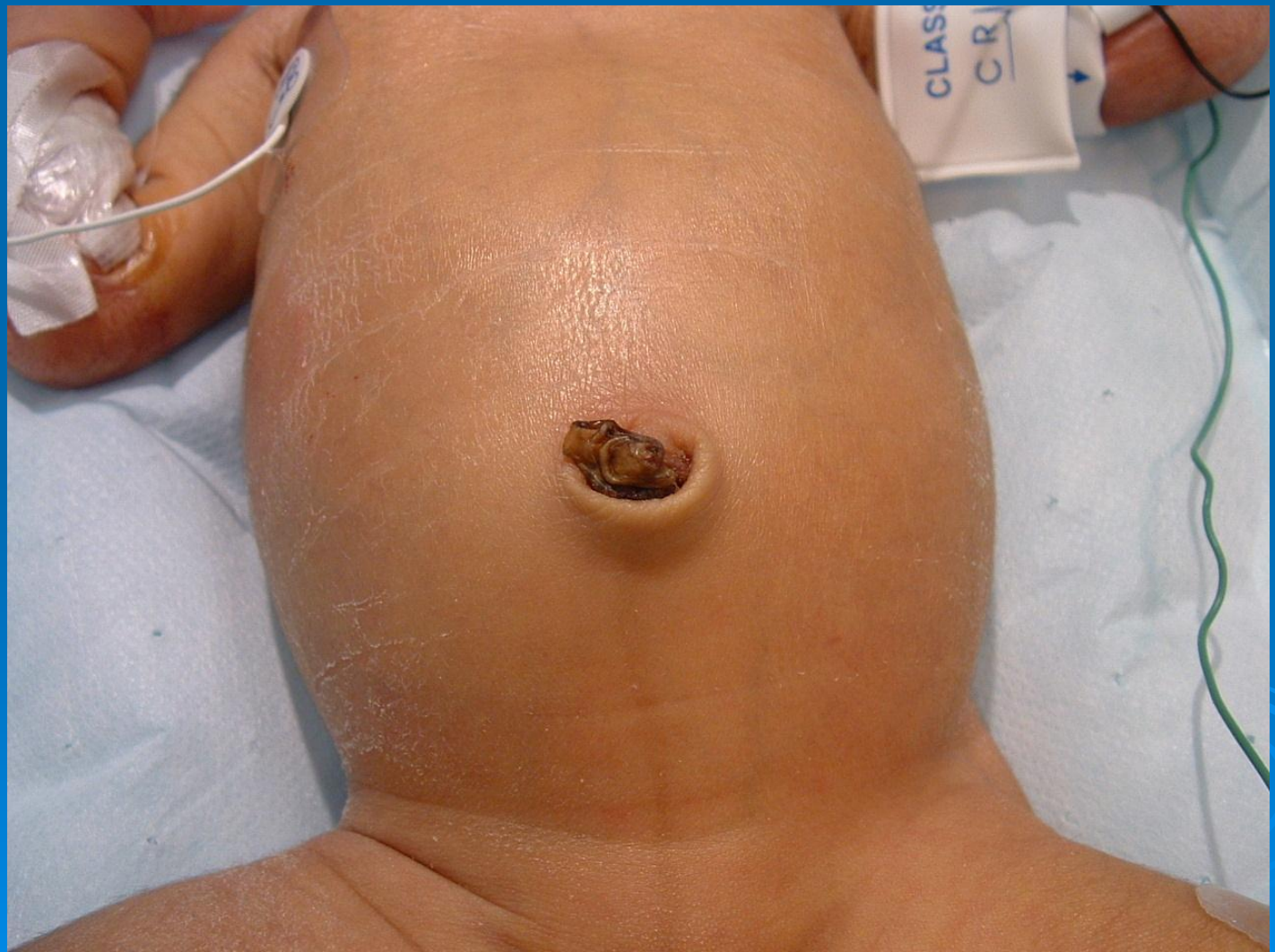




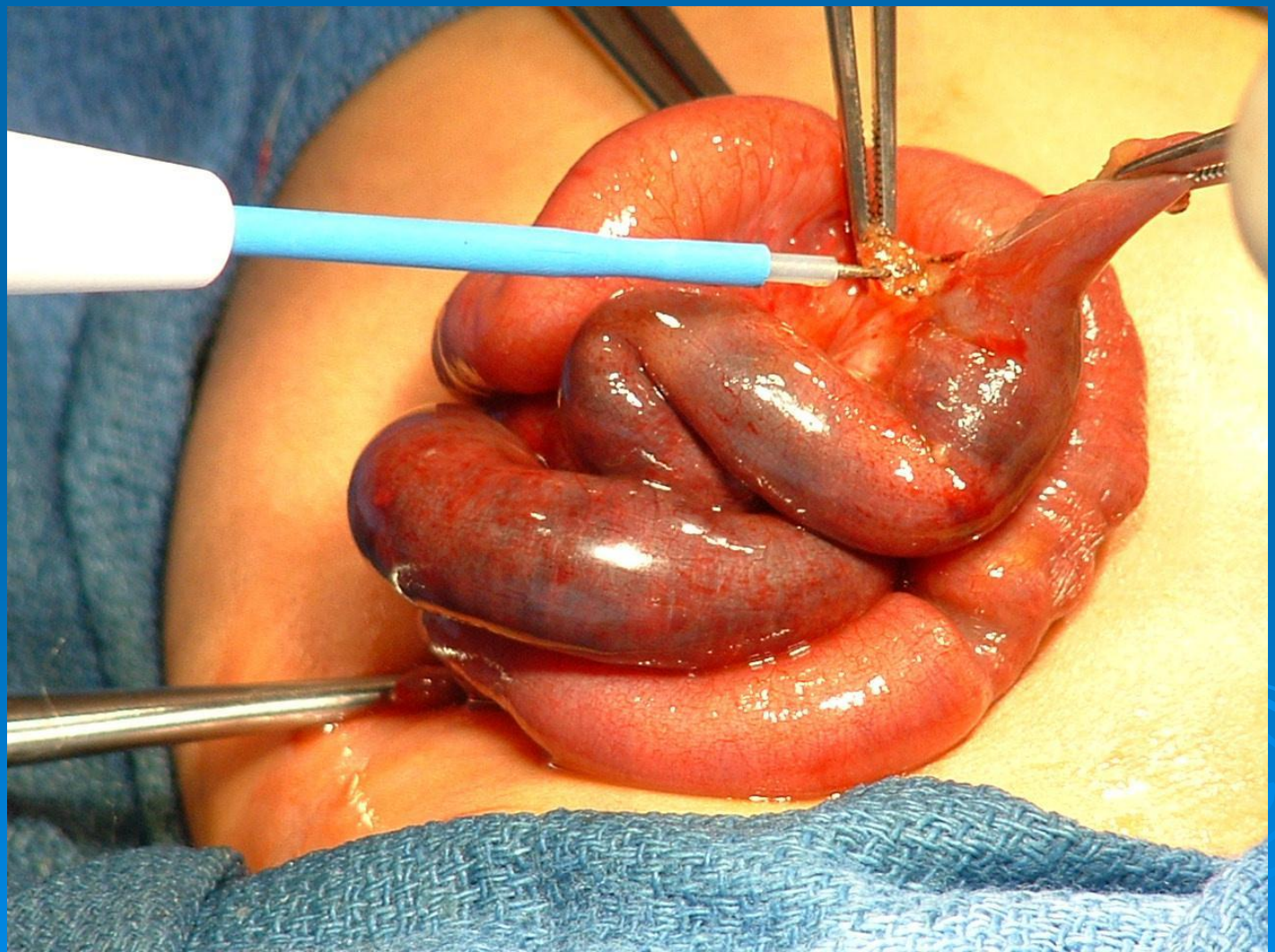
Pneumoperitoneum



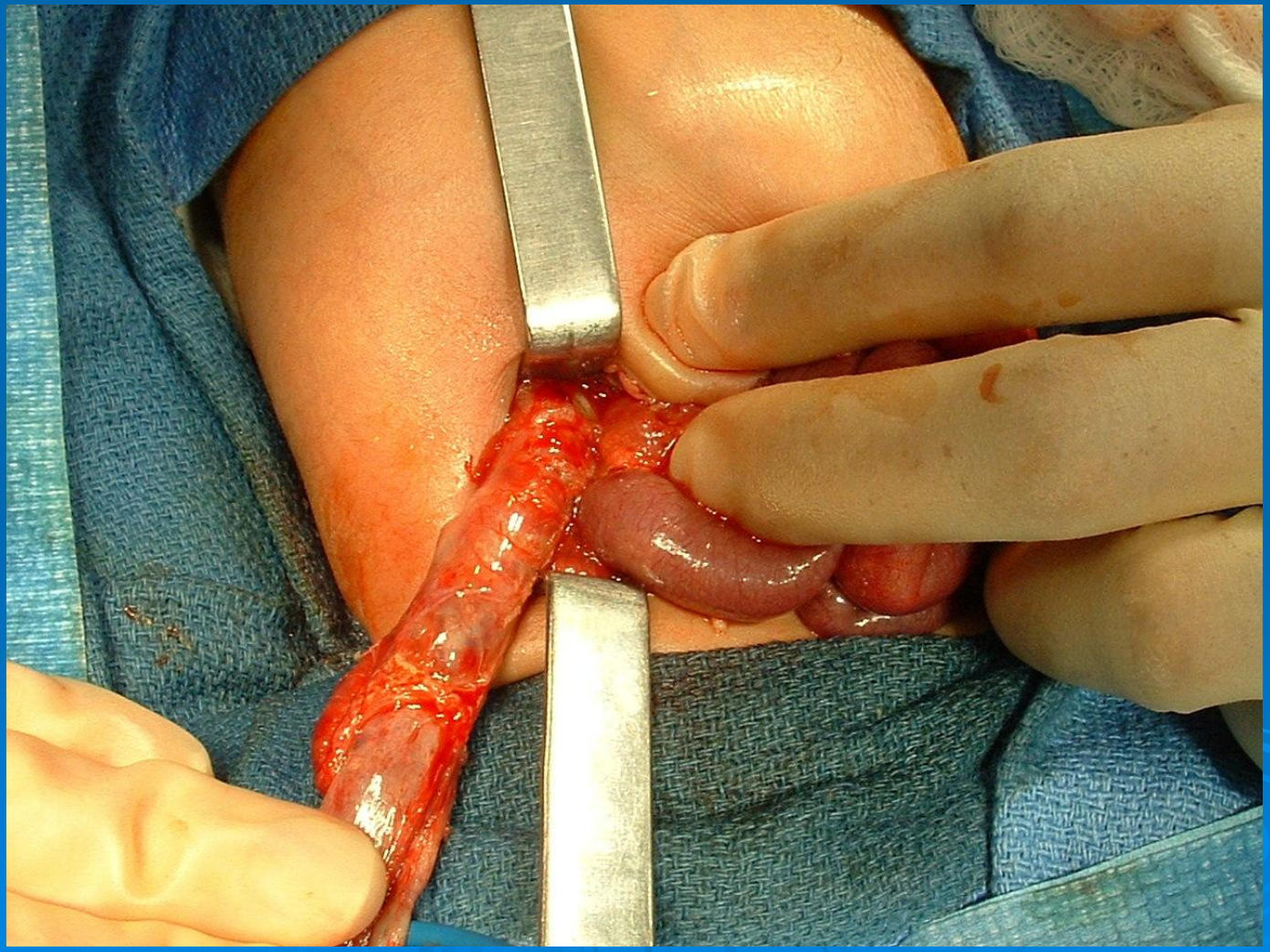
NEC--Abd Distension/Erythema



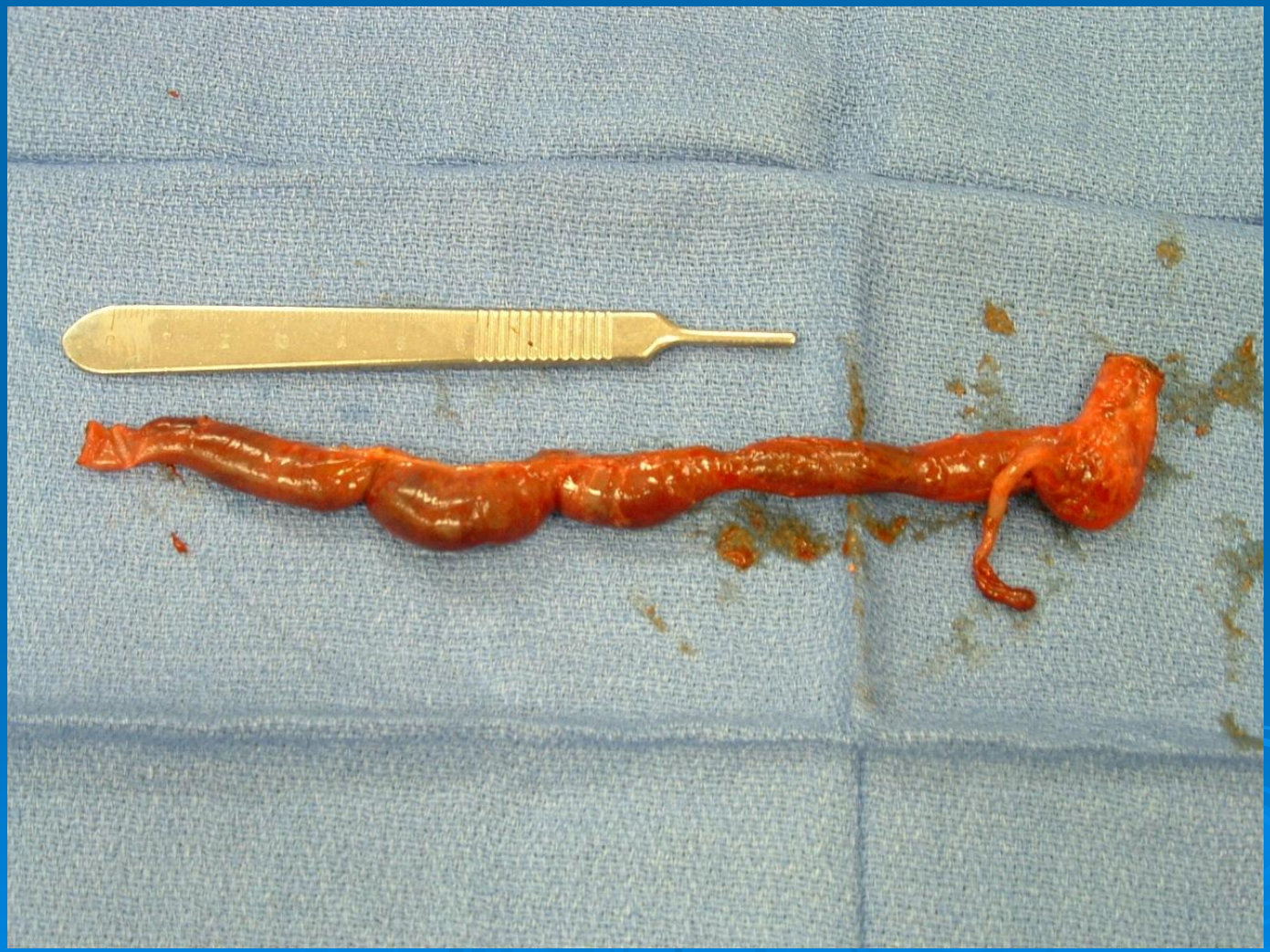
Necrotic Segment Ileum



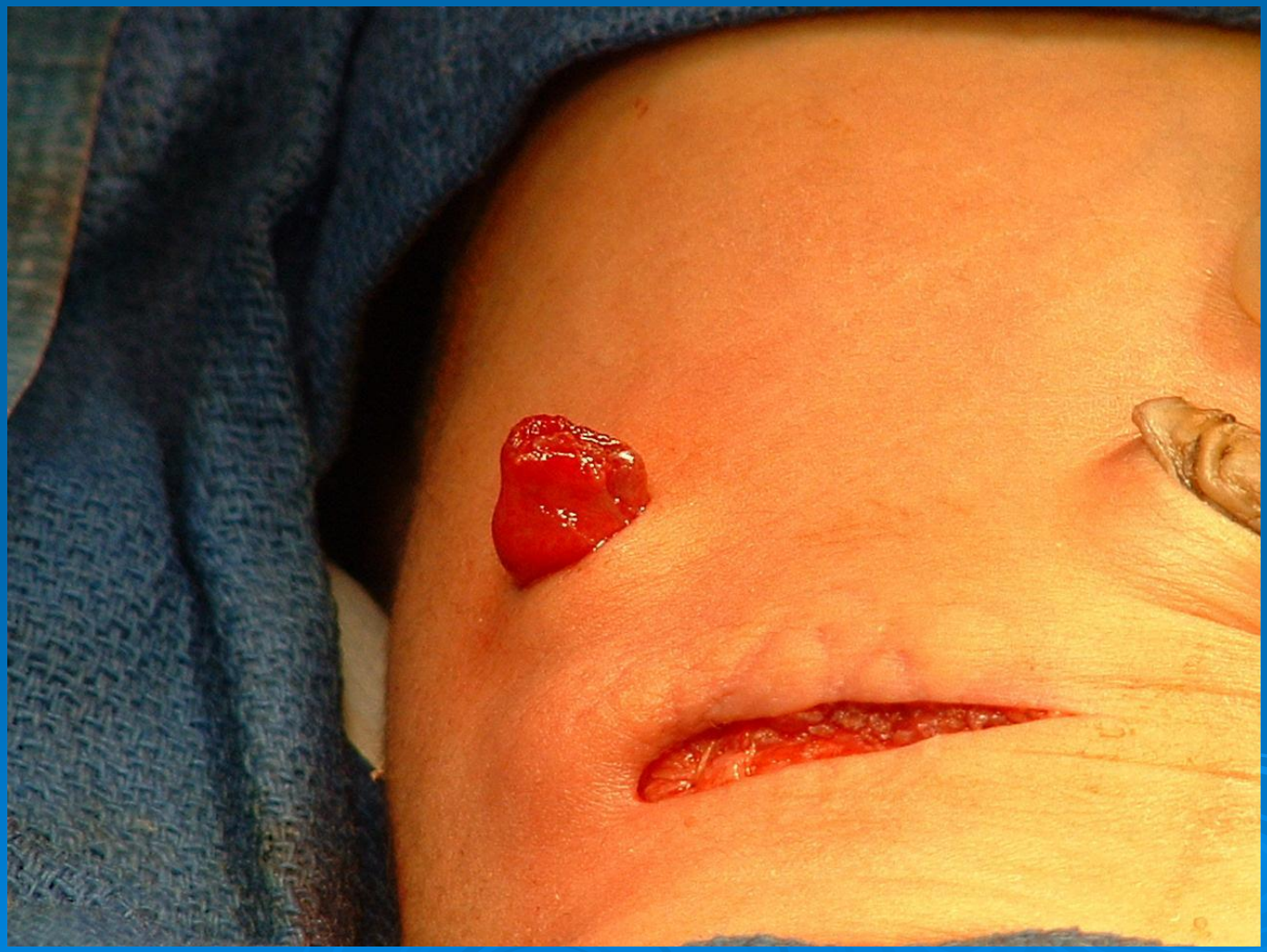
Resection



Specimen--Ileocecectomy



Ileostomy



Common Disorders

- NEC
- Duodenal Atresia
- Small Bowel Atresia
- Malrotation
- Hirschsprung's

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

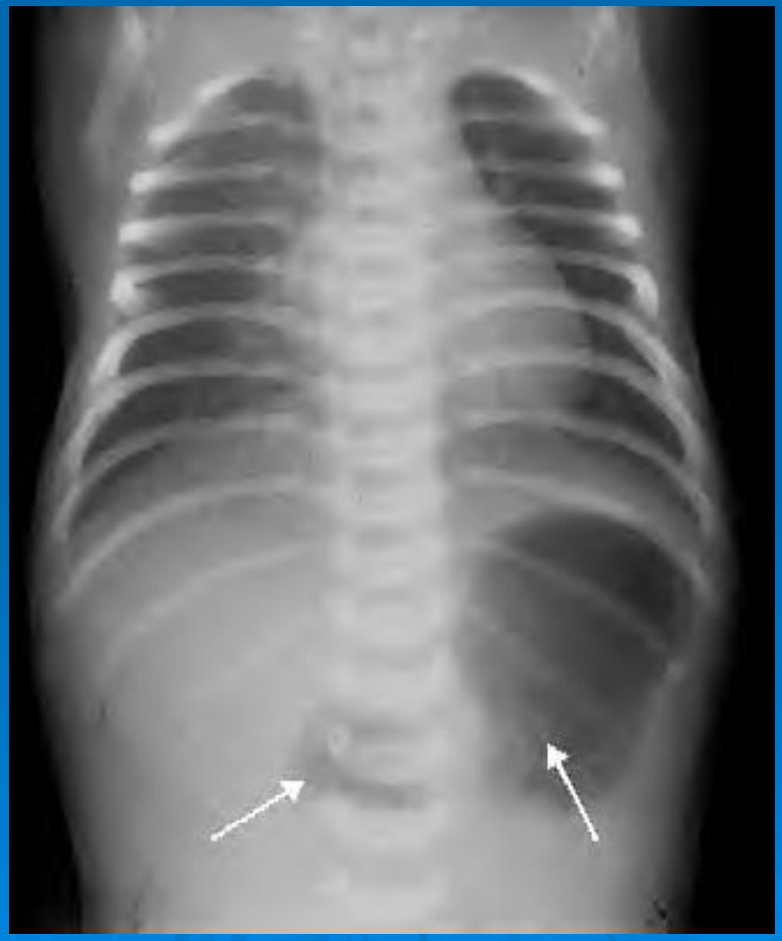
Duodenal Atresia Con't

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

Duodenal Atresia Diagnosis

- Radiographs
  - “Double-Bubble”
  - Pyloric dimple sign
  - Absence of “beak” sign seen in pyloric obstruction
- Workup of potential associated anomalies
  - ECHO, abd US, possible VCUG

“Double Bubble”



Duodenal Atresia Treatment

- Nasogastric decompression, hydration
- Surgery
  - Double diamond duodenoduodenostomy
  - Con’t prolonged NG decompression, sometimes more than 2 weeks needed

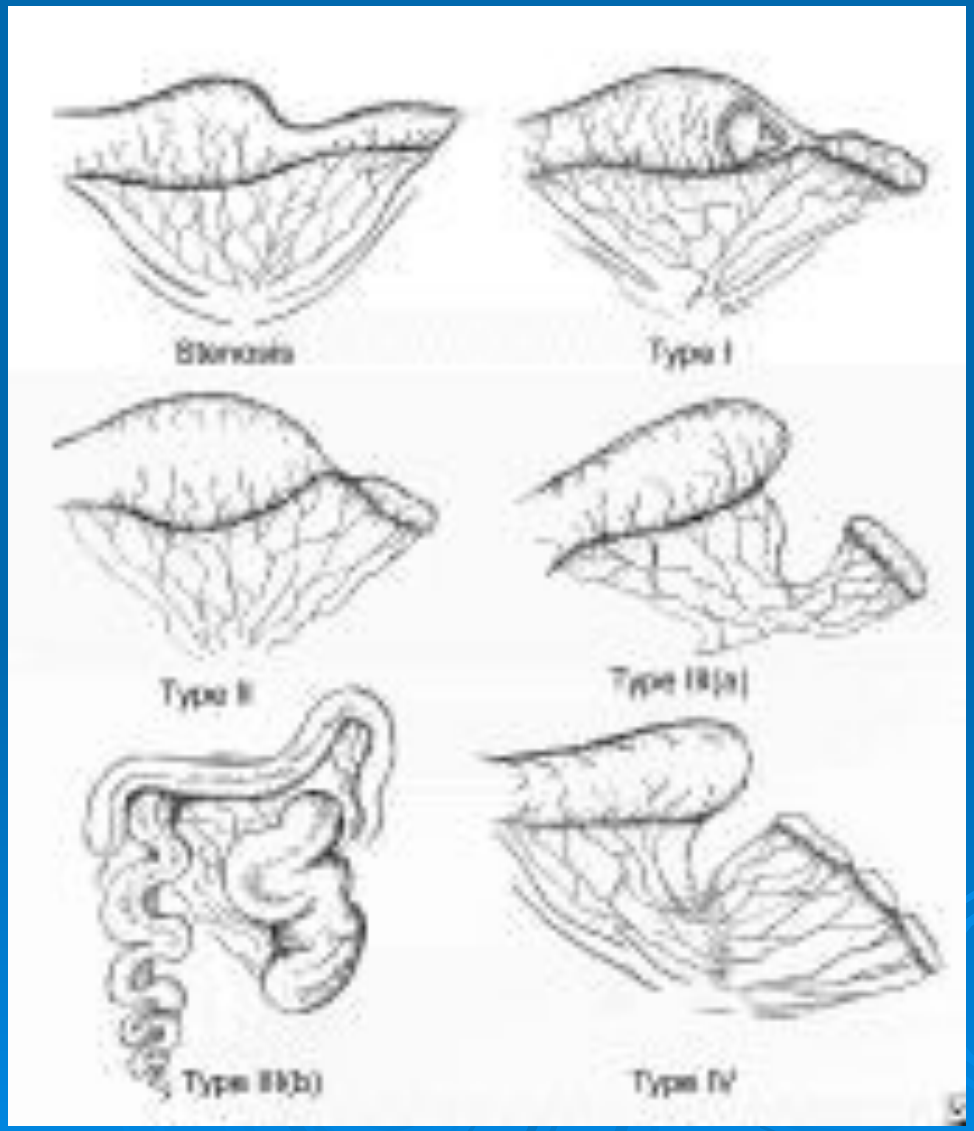
Common Disorders

- NEC
- Duodenal Atresia
- Small Bowel Atresia
- Malrotation
- Hirschsprung's

Small Bowel Atresia

- Jejunal is most common, about 1 per 2,000 live births
- Atresia due to in-utero occlusion of all or part of the blood supply to the bowel
- Classification--Types I-IV
- Presents w/bilious emesis, abd distension, failure to pass meconium (70%)

Intestinal Atresia Classification



Small Bowel Atresia Con't

- Associated Anomalies
  - other atresias
  - Hirschsprung's
  - Biliary atresia
  - polysplenia syndrome (situs inversus, cardiac anomalies, atresias)
  - CF (10%)

Atresia--Diagnosis and Treatment

- Plain films show dilated loops small bowel
- Contrast enema shows small unused colon
- UGI/SBFT shows failure of contrast to pass beyond atretic point
- Treatment is surgical
  - tapered primary anastamosis
  - check for other atresias/associated anomalies

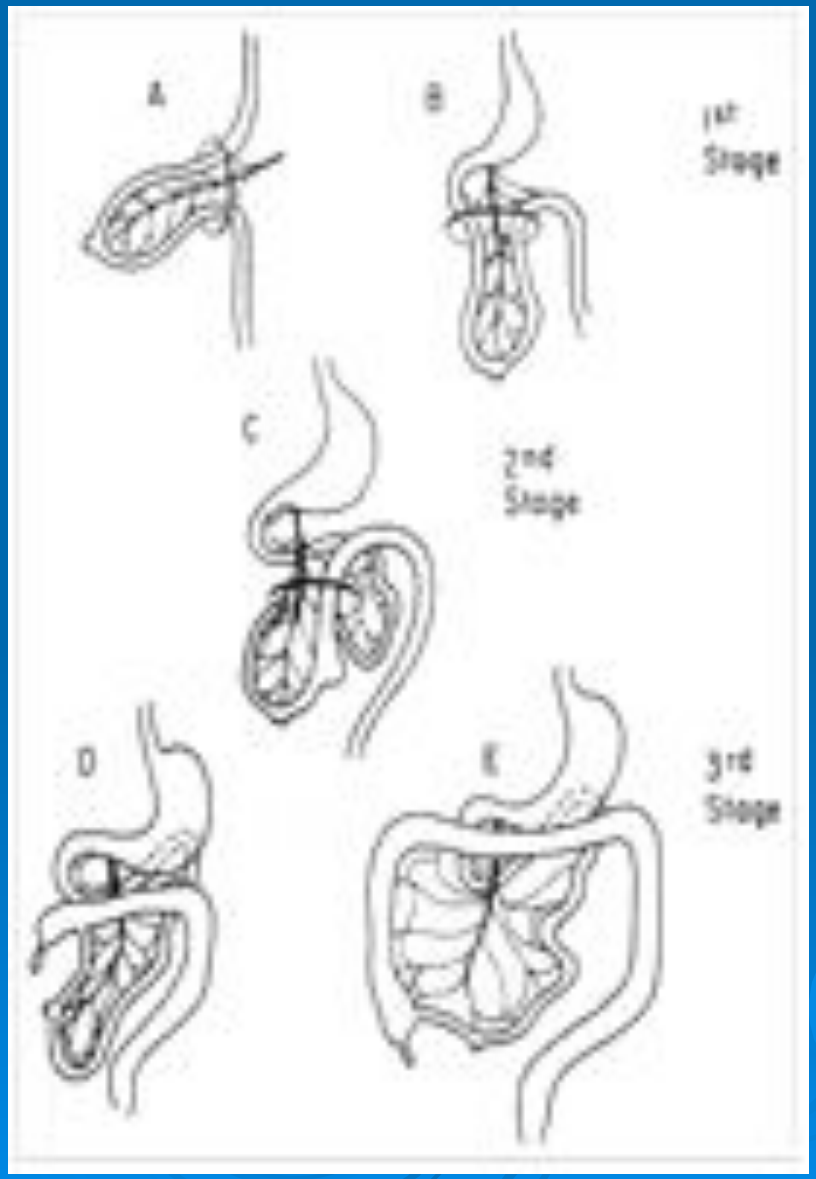
Common Disorders

- NEC
- Duodenal Atresia
- Small Bowel Atresia
- Malrotation/Volvulus
- Hirschsprung's

Malrotation

- 1 per 6,000 live births
- can be asymptomatic throughout life
- Usually presents in first 6 months of life
- 18% children w/short gut had malrotation with volvulus
- Etiology
  - physiologic umbilical hernia--4th wk gestation
  - Reduction of hernia 10th - 12th wks of gestation

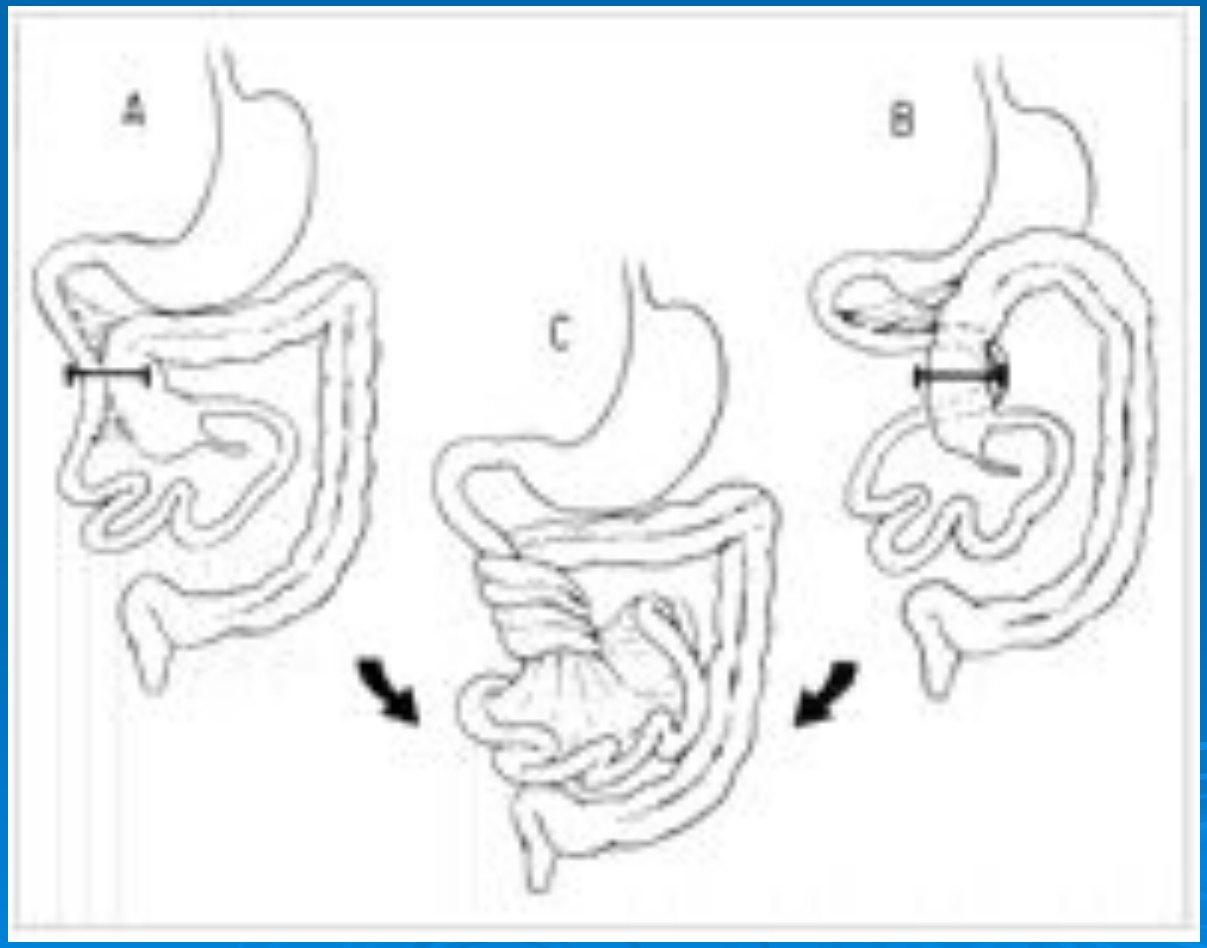
Normal Embryology



Malrotation Classification

- Nonrotation
  - when neither duodenojejunal or cecocolic limbs undergo correct rotation
- Abn Rotation of Duodenojejunal limb
  - causes Ladd's bands to form across duodenum
- Abn rotation of Cecocolic limb
  - cecum lies close to midline, narrow mesenteric base

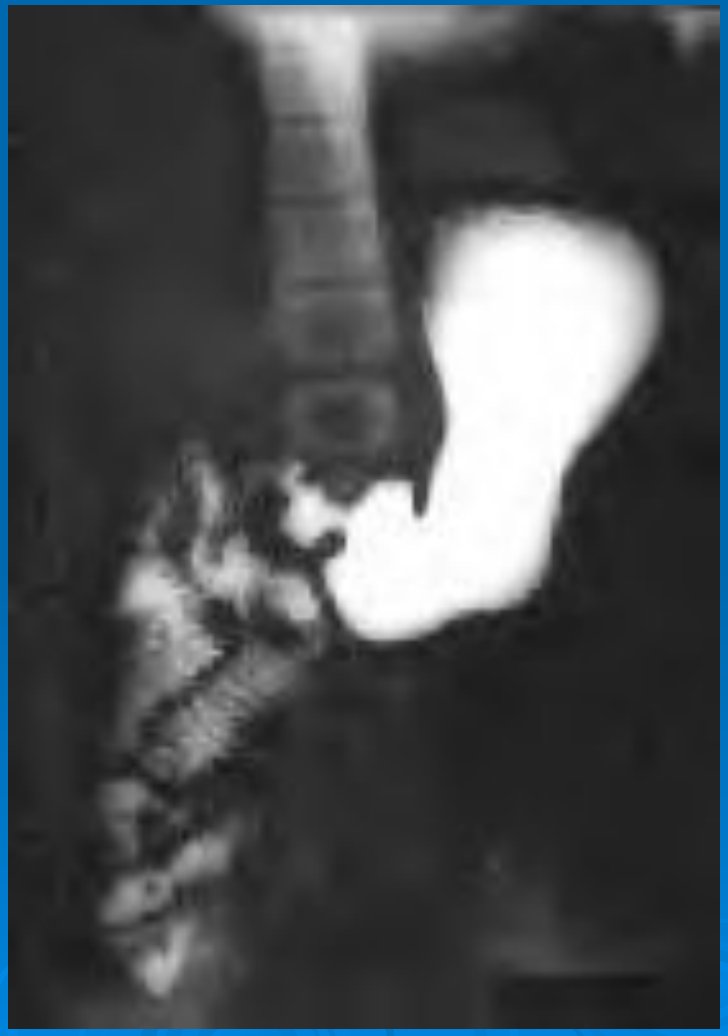
Abnormal Rotation/Fixation



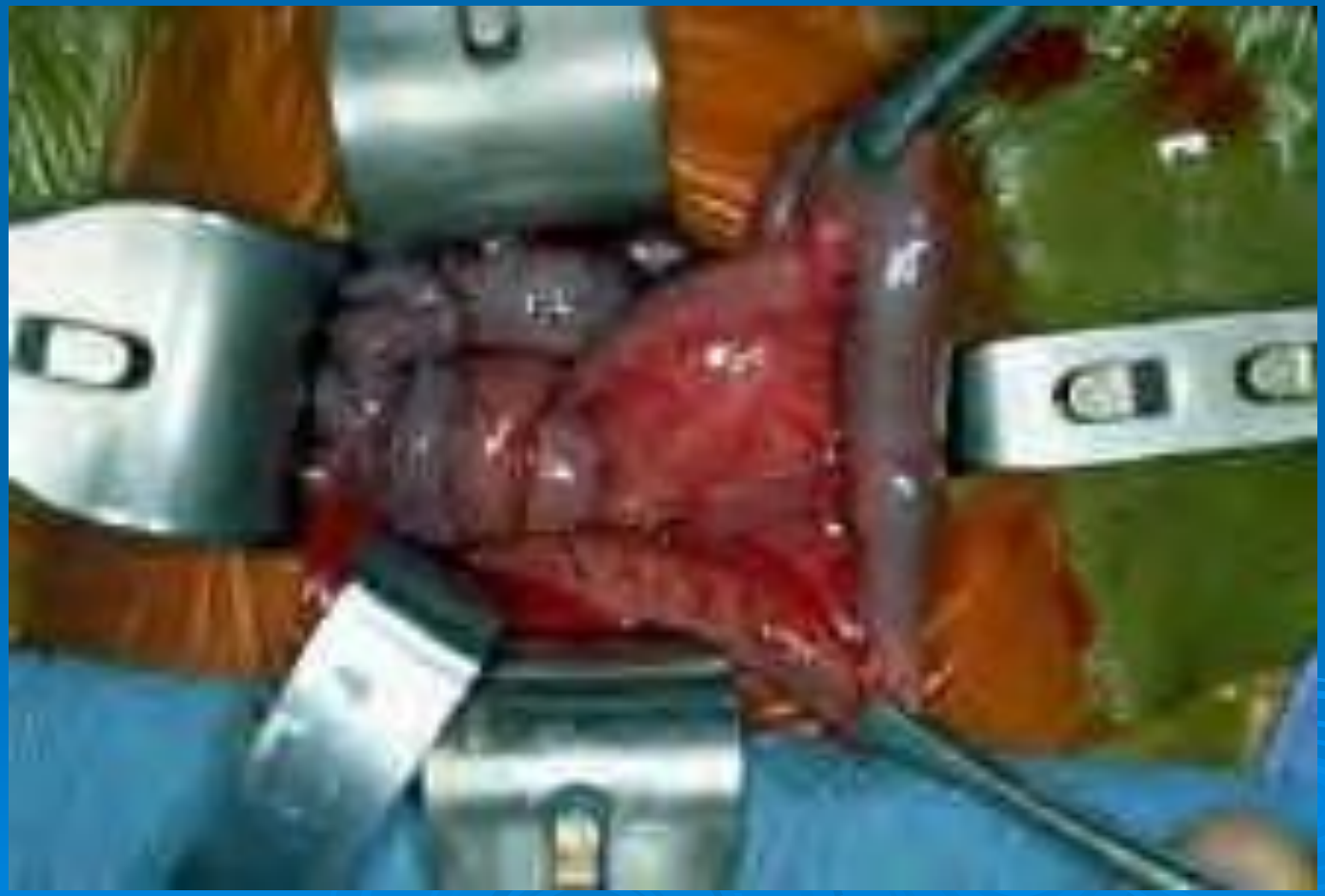
Malrotation Diagnosis

- Varying symptoms from very mild to catastrophic
- **Bilious emesis is Volvulus until proven otherwise**
- Bilious emesis, bloody diarrhea, abd distension, lethargy, shock
- UGI shows abnormal position of Duodenum
  - if Volvulus, see “bird’s beak” in duodenum

Malrotation UGI



Intraop Volvulus



Bowel Necrosis--Volvulus

Malrotation--Treatment

- Surgical--Ladd's Procedure
  - Evisceration
  - Untwisting of volvulus (counterclockwise)
  - Division of Ladd's Bands
  - Widening mesenteric base
  - Relief of Duodenal obstruction
  - Appendectomy
- Recurrence 10% after Ladd's

Common Disorders

- NEC
- Duodenal Atresia
- Small Bowel Atresia
- Malrotation
- Hirschsprung's

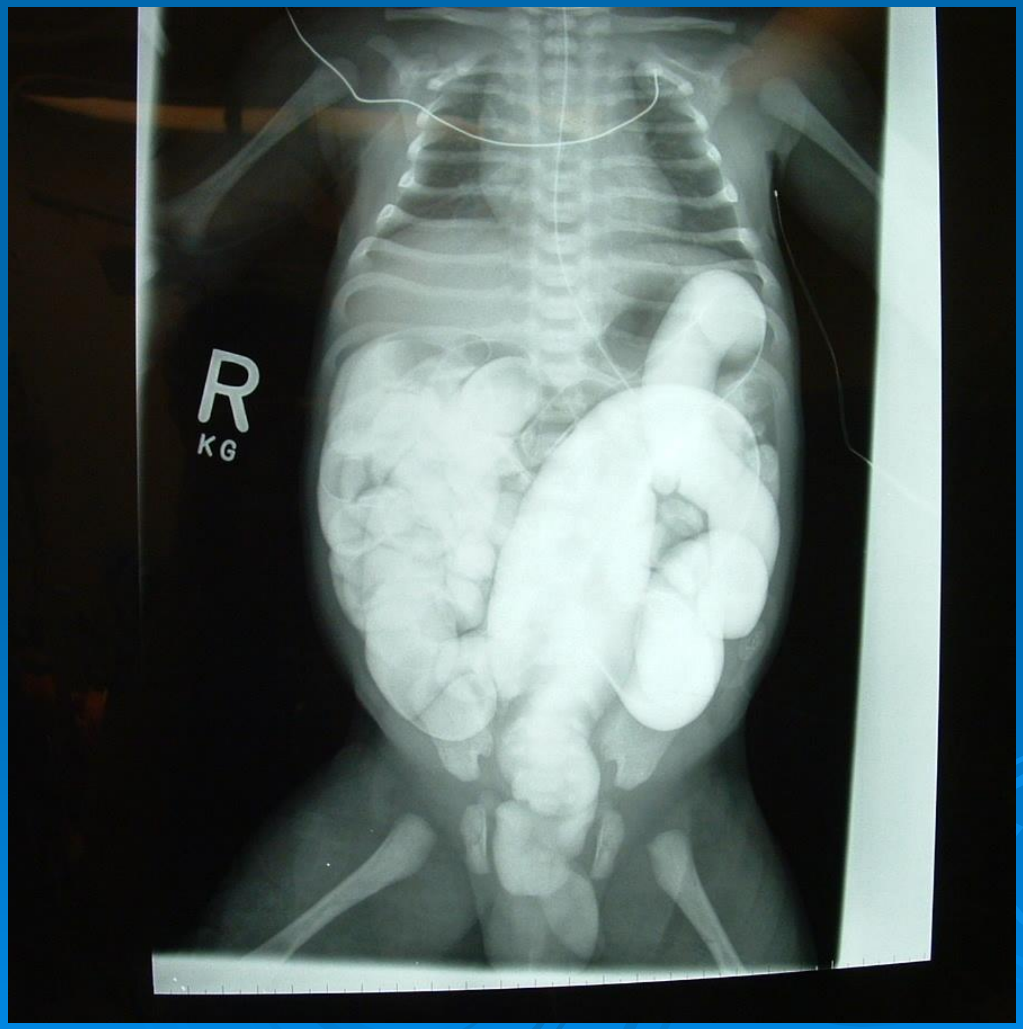
Hirschsprung's Disease

- Migratory failure of neural crest cells
- Incidence 1 in 5,000 live births, males affected 4:1 over females
- 90% of pts w/H'sprung's fail to pass meconium in first 24-48 hrs
- Abd distension, bilious emesis, obstructive enterocolitis

Hirschsprung's Diagnosis

- Barium Enema
  - Transition zone
- Anorectal Manometry
  - shows failure of reflexive relaxation
  - not very helpful in infants, young children
- Rectal Biopsy
  - Absence of Ganglion cells and hypertrophy of nerves

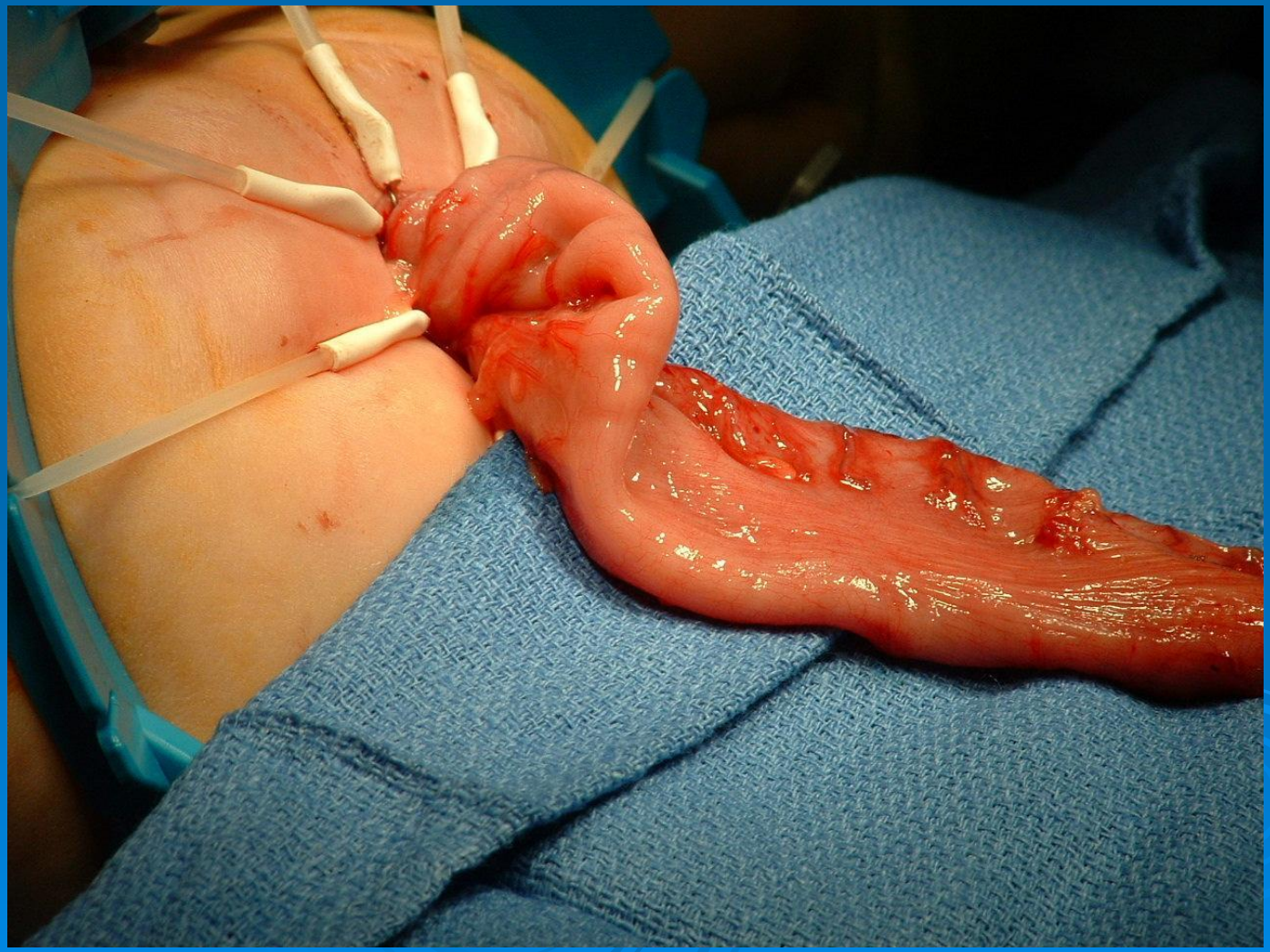
Transition Zone on BE

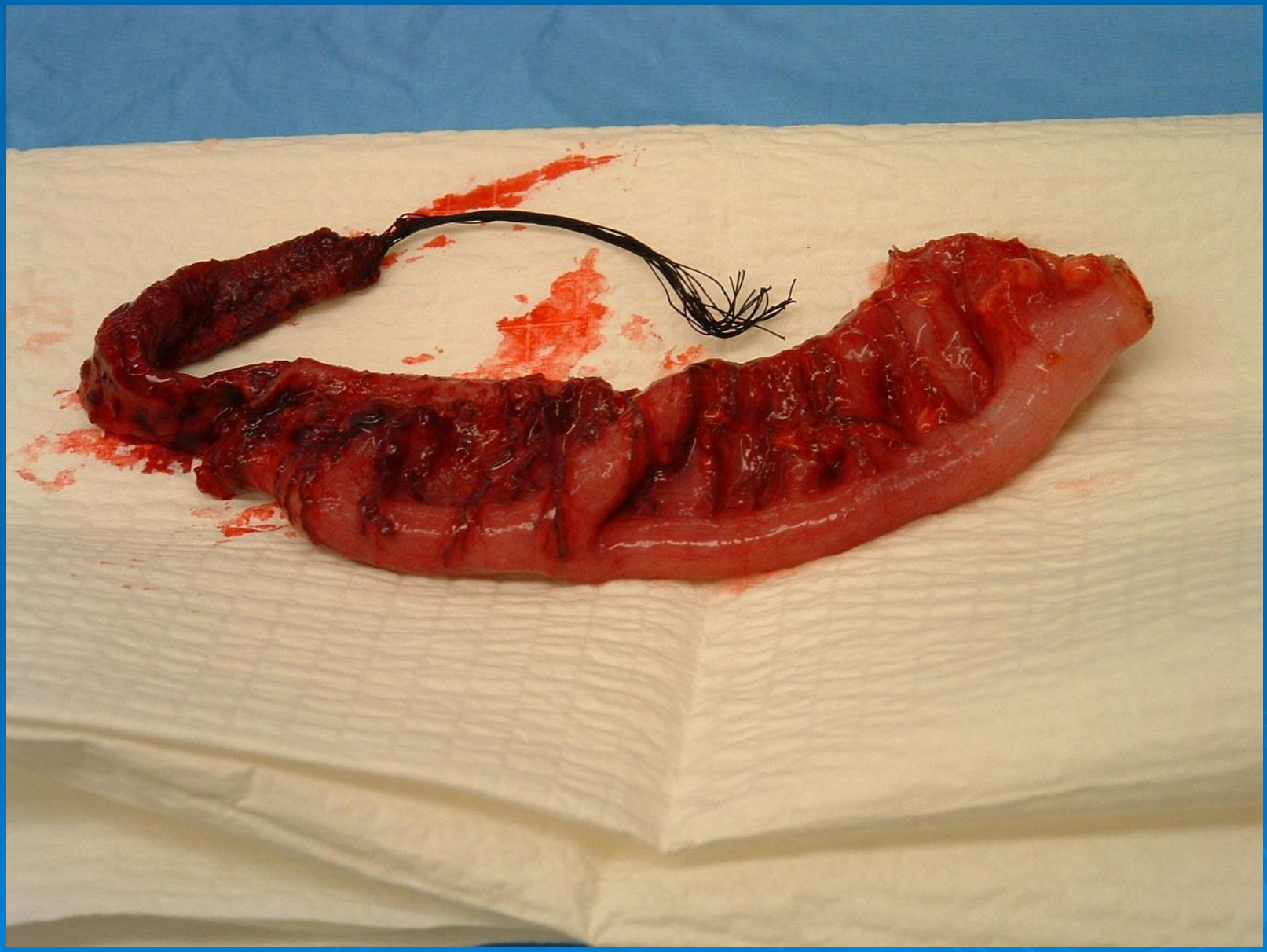


Hirschsprung's Treatment

- In neonates, can do primary pull-through--bringing normal colon down to anorectal junction
- In older infants, may need diverting colostomy first to decompress
- May need prolonged dilatations and irrigations

Pull-Through Procedure





Summary

- BILIOUS EMESIS IS VOLVULUS UNTIL PROVEN OTHERWISE
- Signs of surgical emergency
  - free air, abd wall cellulitis, fixed loop on xray, rapid distension, peritonitis, clinical deterioration
- History and plain films will guide sequence of additional studies
- Remember associated anomalies