The Annual General Pediatric Review & Self Assessment

Nicklaus Children's Hospital

NEONATOLOGY

Magaly Diaz-Barbosa, MD

Chief, Division of Neonatology

Medical Director, NICU

Nicklaus Children's Hospital

Miami, Florida

Nicklaus Children's Hospital

Disclosure of Relevant Relationship

Dr. Diaz-Barbosa has not had (in the past 24 months) any relevant conflicts of interest or relevant financial relationship with the manufacturers of products or services that will be discussed in this CME activity or in his presentation.

Dr. Diaz-Barbosa will support this presentation and clinical recommendations with the “best available evidence” from medical literature.

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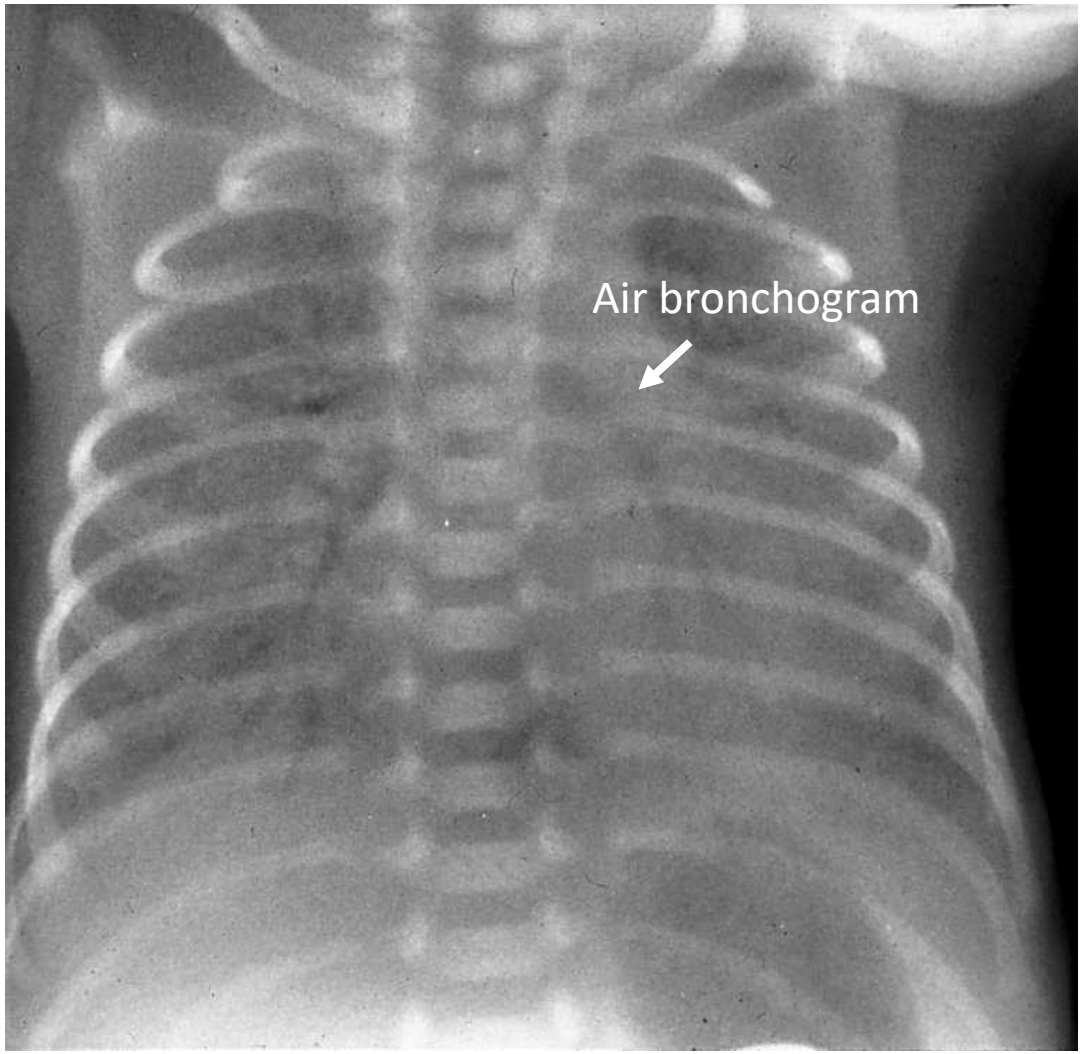
Diseases of the Newborn

- Respiratory
- Cardiology
- Neurology
- Infectious Disease
- Gastroenterology
- Ophthalmology
- Renal

Respiratory Distress Syndrome (RDS)

Incidence	• **>90% <28 wks GA** • **<5% >36 wks GA**
Etiology	• **Surfactant deficiency**: decreased production and secretion • Decreased lung compliance • Unstable alveoli->atelectasis • lung injury: inflammation and PE
Findings	tachypnea, grunting, flaring, retractions, cyanosis, apnea, increased WOB, hypoxia
Xray	**Reticogranular pattern**, ground glass appearance (diffuse microatelectasis) **air bronchograms**, low lung volumes
Management	**Surfactant** ,supportive care
Complications	Air leaks, BPD

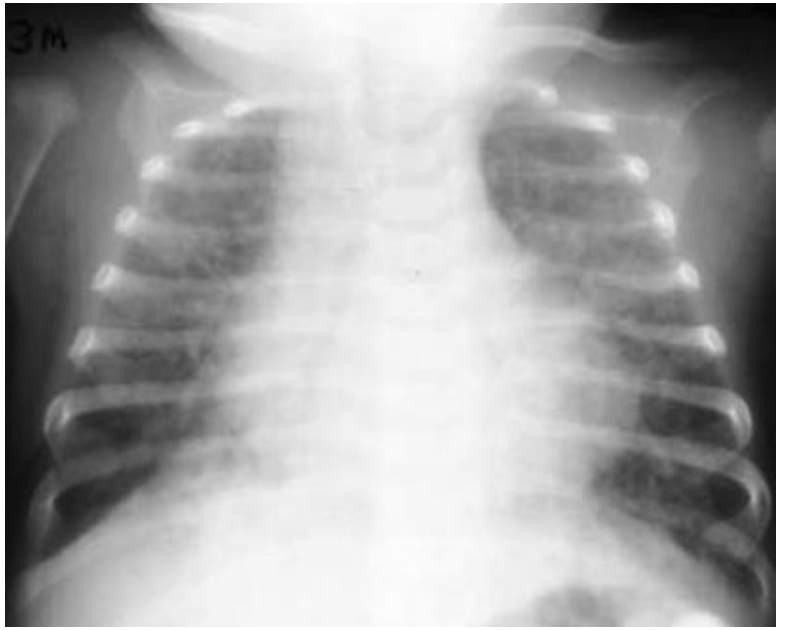


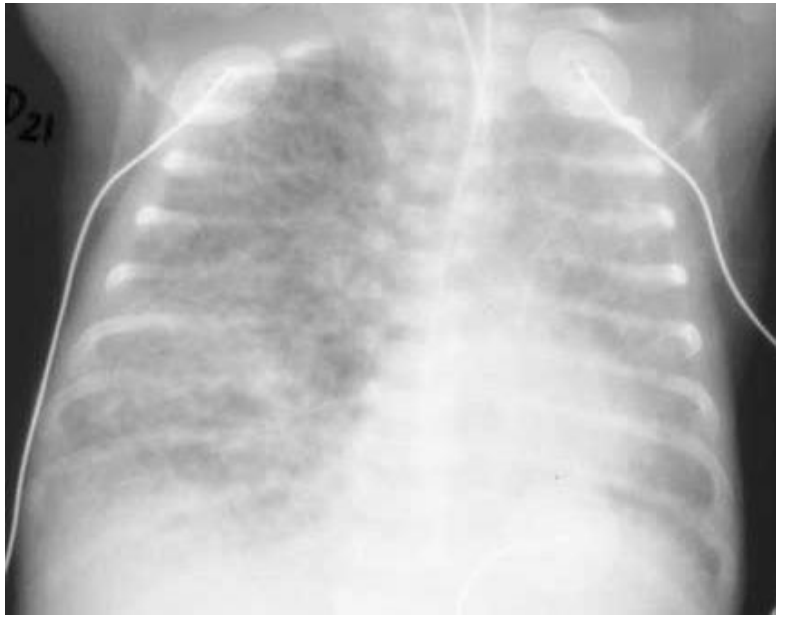
Surfactant

Function	• **Decreases surface tension** • Facilitates alveolar expansion, improves alveolar stability decreasing alveolar collapse • Increases pulmonary compliance
Production/Storage	**Type II Pneumocytes** starting at 20 wks Mature levels > 35 weeks gestation
Composition	• 90 % Lipids 70 % Phospholipids Dipalmitoylphosphatidylcholine (DPPC) • 10% Surfactant proteins (SP-A, SP-B, SP-C and SP-D)
Surfactant replacement	given as prophylaxis or rescue treatment, reduces the incidence of RDS, air leaks, and mortality in preterm infants with RDS

Broncopulmonary Dysplasia (BPD)

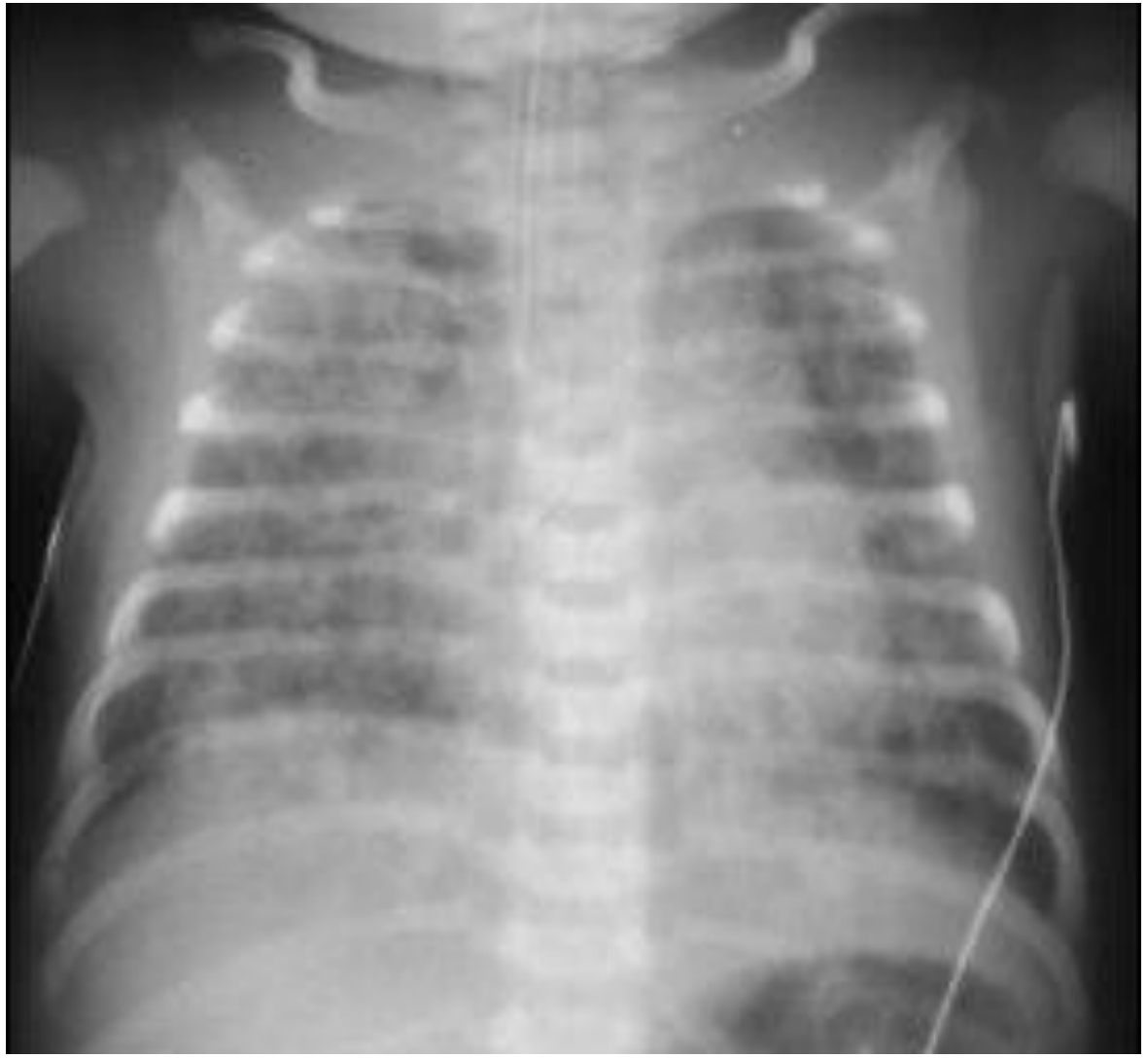
Characteristics	Chronic respiratory insufficiency **Disruption of lung development and injury** Associated with **barotrauma, oxygen toxicity and inflammation**
Definition (<32wks GA)	Need for O2 at 36 wks PCA Severity: • Mild • Moderate • Severe Based on X ray findings, intervention and FiO2
X-ray findings	Diffuse haziness to coarse interstitial densities
Treatment	Gentle ventilation strategies rapid wean of FiO2, permissive hypercapnia, stringent fluid management, adequate nutrition, reduction of airway resistance, treat PHTN if present





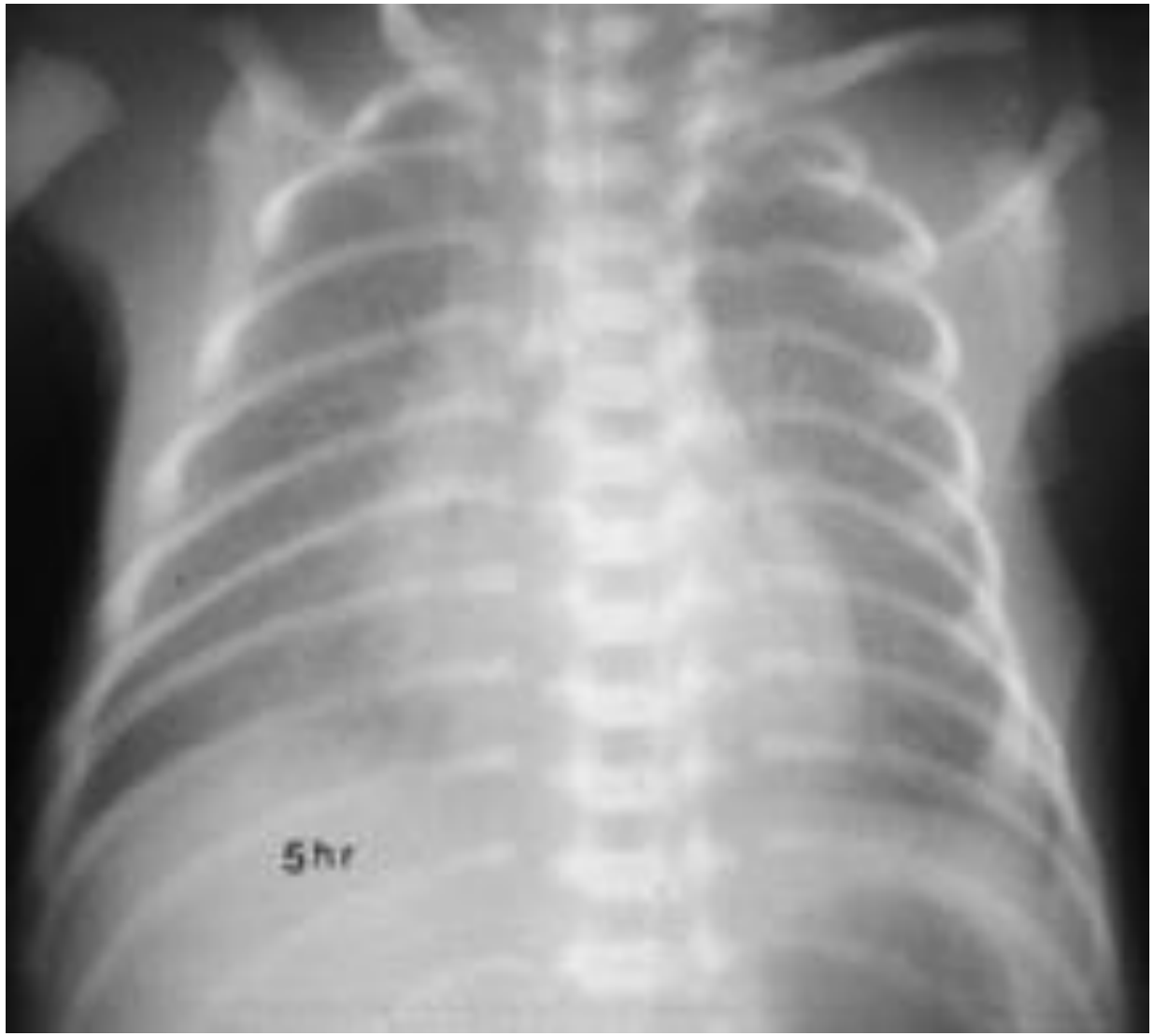
Meconium Aspiration Syndrome (MAS)

Risk factors	• **Postmaturity,** IUGR • Vaginal breech delivery , C/S delivery • Fetal distress (NRFHR) • Low Apgar scores/need for resuscitation • Maternal fever, infection
Incidence	• 0.1 to 0.4 percent of live births
Pathology	• Lung injury: chemical pneumonitis • Complete or partial airway obstruction • Inactivation of surfactant • Vasoconstriction of pulmonary vessels, at risk for **PPHN (hypoxemia/acidosis)**
Clinical Findings	SGA/Post term, meconium staining on PE, barrel shaped chest, grunting, nasal flaring, retractions, cyanosis
X-ray	Patchy infiltrates, hyperinflation
Management	Supportive care



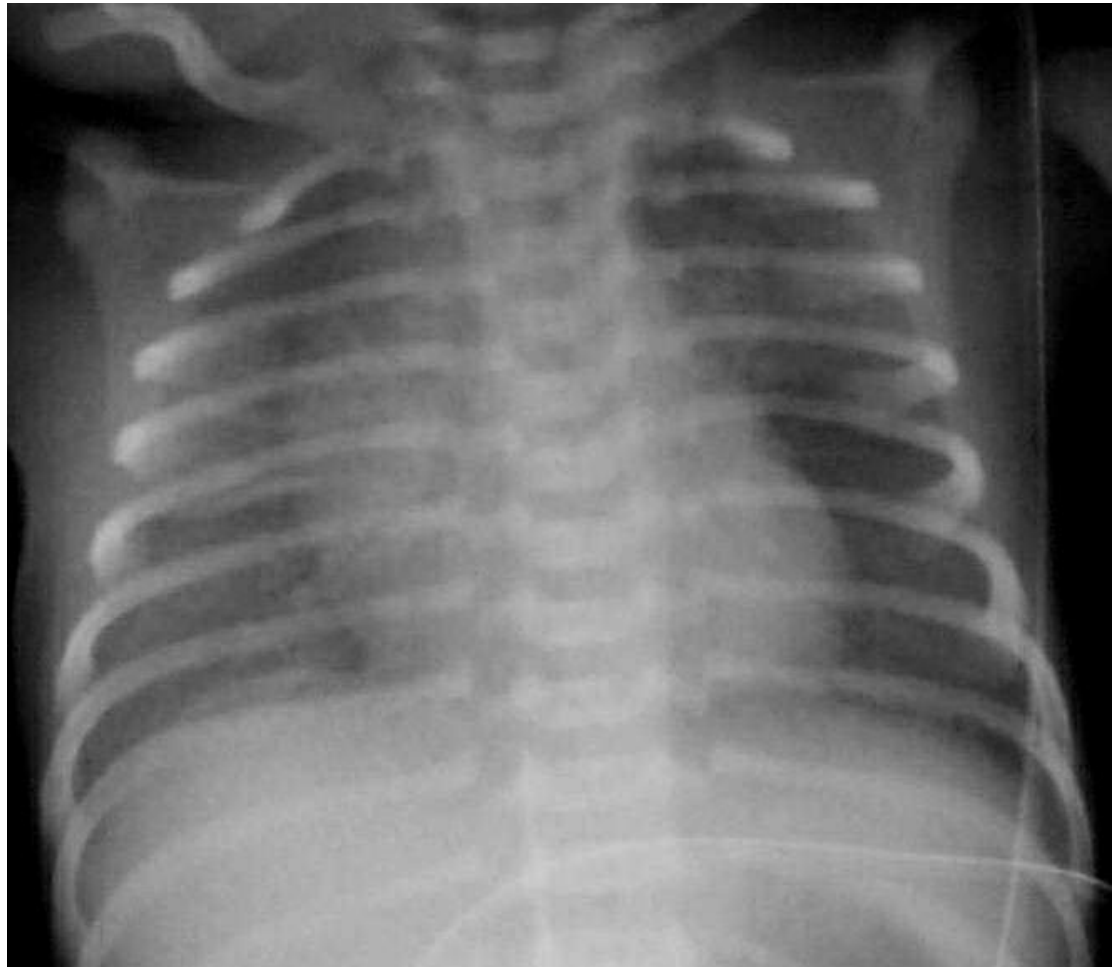
Pneumonia

Transmission	• **Transplacental** • Aspiration of infected amniotic fluid (before, during or after birth) • Vertical - mother • Horizontal -direct contact or environment
Etiology	• **Early (≤7D)**: GBS, E.Coli, Klebsiella species, Listeria, Enterococcus • **Late (≥7D)**: above plus Staph aureus, Pseudomonas, fungal, chlamydia
Findings	Presents within 48 h of age respiratory distress, apnea
X-ray	May be normal initially, hazy, reticulogranular, +/-pleural effusion GBS may mimic RDS Listeria may mimic MAS
Treatment	Antibiotics, supportive care



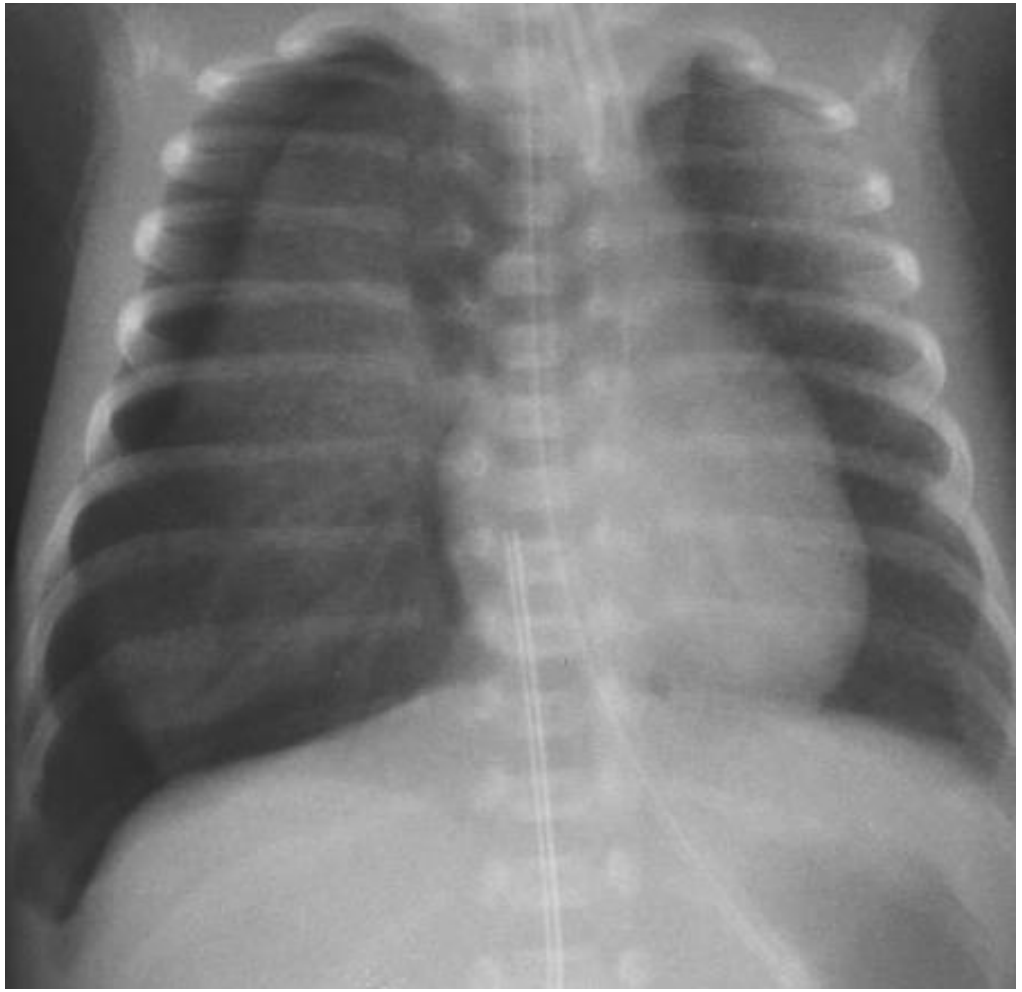
Transient Tachypnea of the Newborn (TTN)

Characteristics	• Relatively **benign** • **Self-limited** disease • Full term, C/S delivery
Etiology	Pulmonary Edema due to d**elayed absorption and clearance of fetal lung fluid** by lymphatics. Excess fluid decreases lung compliance
Findings	Tachypnea after birth or within two hrs, grunting, retractions, increased WOB
X-ray	Prominent vascular markings, interlobar fluid accumulation (fluid in fissure), may have diffuse parenchymal infiltrates
Treatment	Supportive care, supplemental O2 Symptoms can last few hours to two days



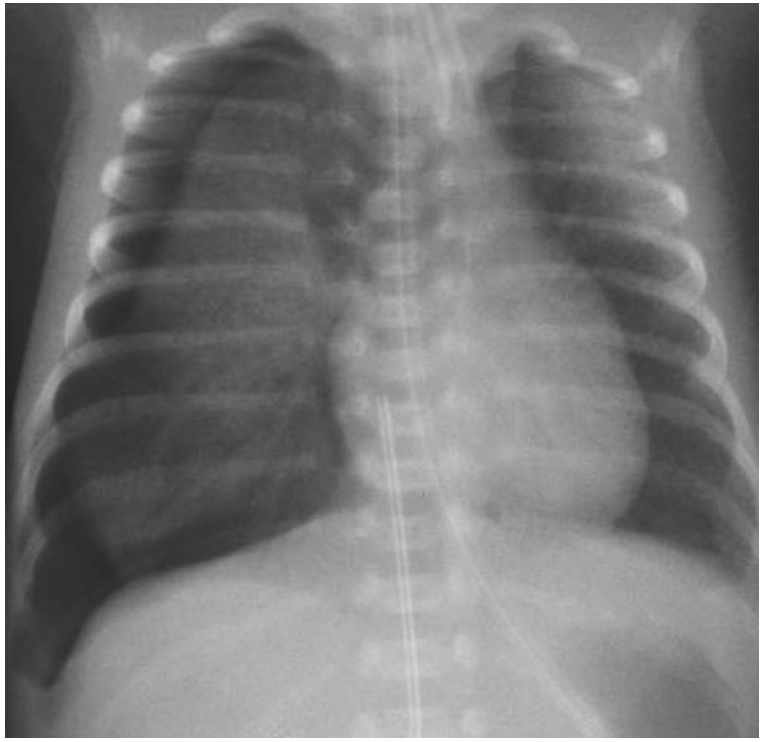
Pneumothorax

Etiology	• Air leak of air from lungs into the pleural space • Spontaneous -up to 1-2% healthy infants • Increased risk- lung disease, **mechanical ventilation, MAS,**
Clinical findings	• Tachypnea, grunting, flaring, retractions, cyanosis • Diminished or absent breath sounds on affected side • Asymmetric chest ,displaced PMI
X-ray findings	Decreased lung volume mediastinal shift
Treatment	Observation, needle aspiration, chest tube insertion

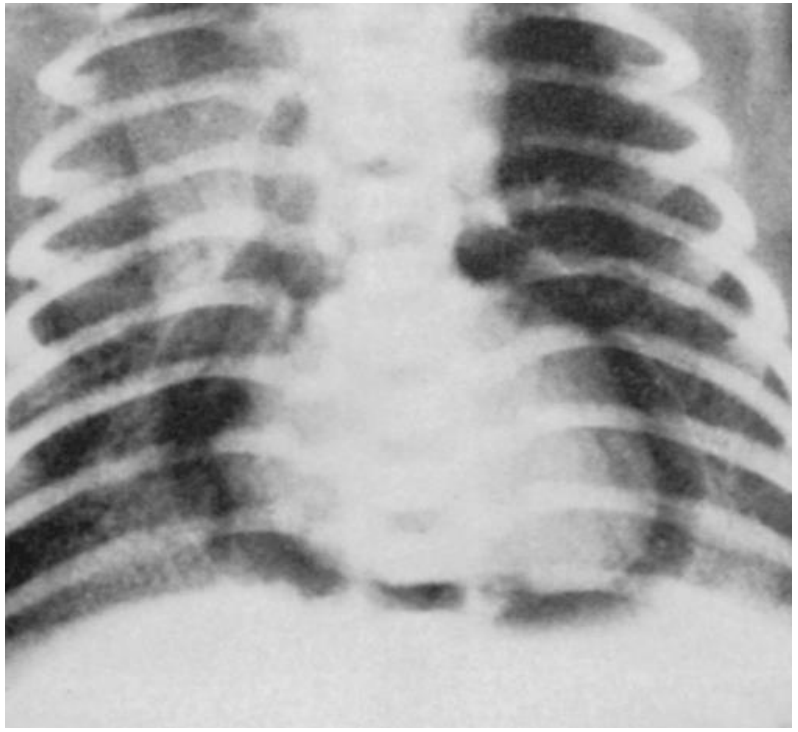


Air Leaks

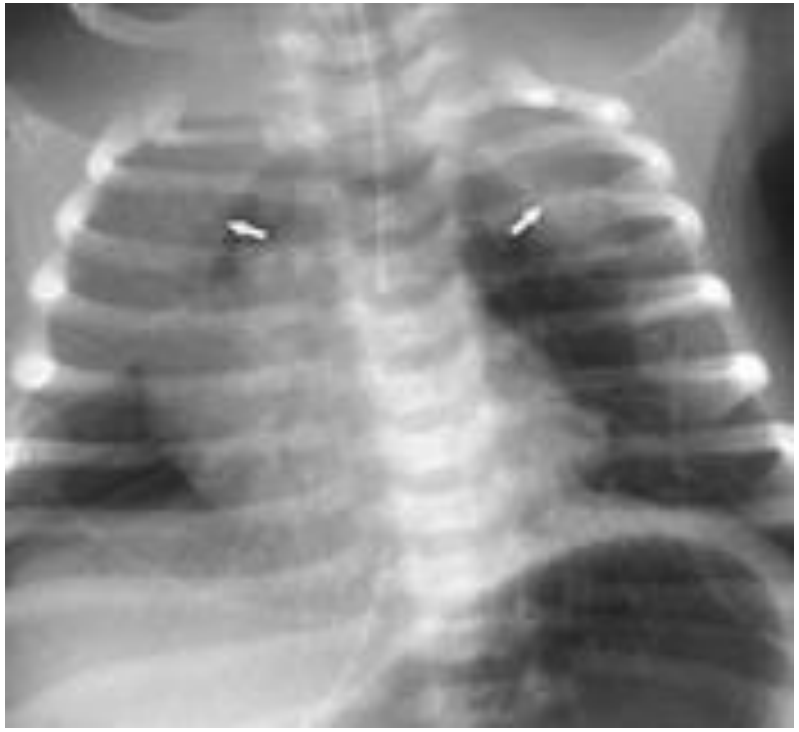
Complication of mechanical ventilation, MAS



Pneumothorax



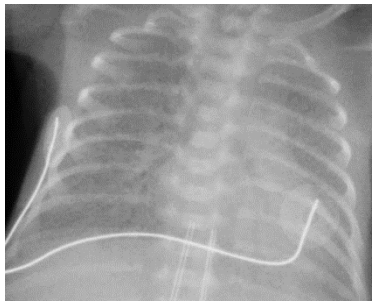
Pneumopericardium



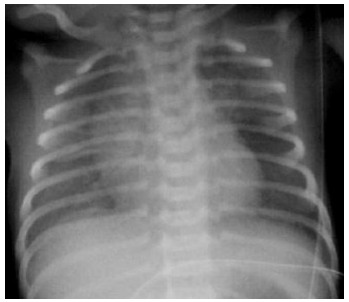
Pneumomediastinum

Respiratory Diseases

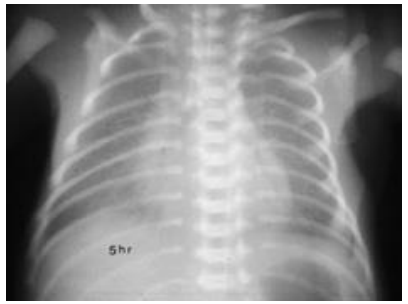
Disease	Cause	X-ray Description
RDS	Surfactant deficiency	Reticulogranular (ground glass), air broncograms
TTN	Fluid retention	Fluid in minor fissure
Pneumonia	Bacteria	Hazy, reticulogranular
MAS	Meconium	Patchy infiltrates
Air Leak	Alveolar rupture	Air dissection



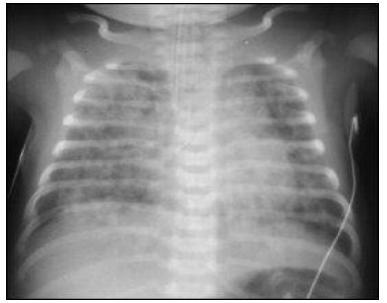
RDS



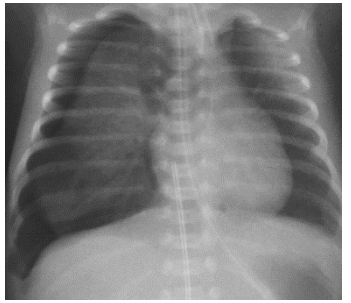
TTN



Pneumonia



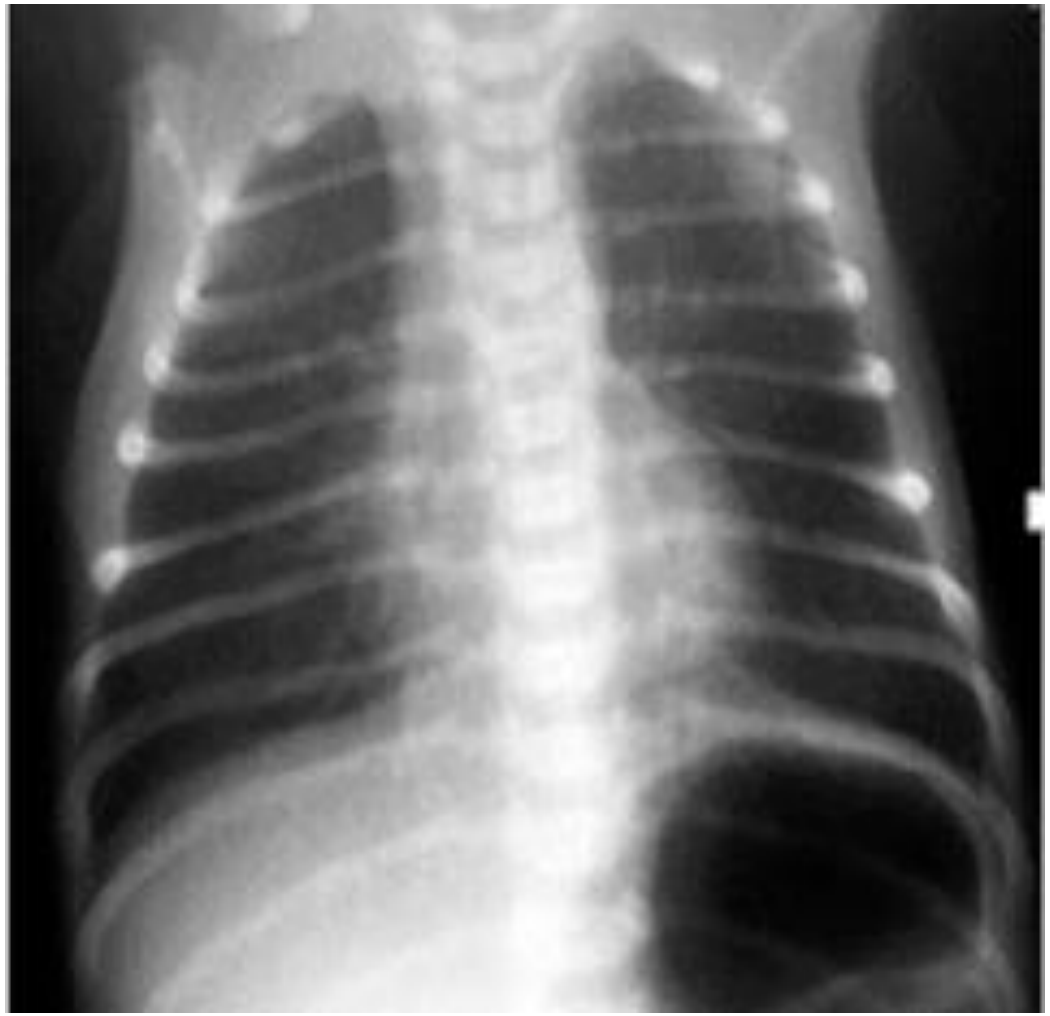
MAS



Air Leak

Persistent Pulmonary Hypertension (PPHN)

Etiology	Transitional circulation: Mean pulmonary pressure ↓, PBF ↑,PVR ↓ If **PVR remains elevated-> PPHN-Rt -> L shunt**
Types	• Idiopathic • Secondary to MAS, sepsis, PNA, CDH, BPD,
Findings	• Cyanosis and signs of respiratory distress.
Diagnosis	Cardiac ECHO ↑Rt ventricular pressure ↓function **Preductal-Post ductal O2 Sat difference at least 10%, OI - Severity of hypoxemia**
X-ray Findings	May be normal, or findings of associated pulmonary condition, pulmonary blood flow may appear normal or reduced.
Treatment	O2, HFOV, iNO, sildenafil, sedation, hemodynamic support, ECMO (for severe respiratory failure)

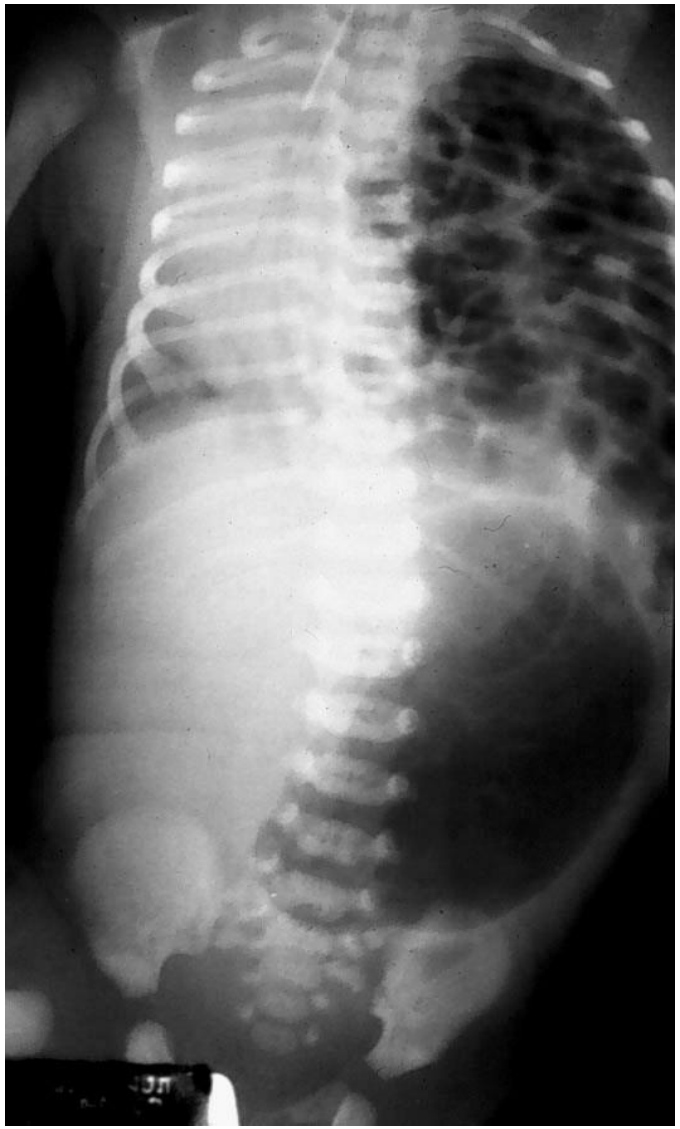


Developmental Abnormalities

- Esophageal Atresia/TE fistula (TEF)
- Congenital diaphragmatic hernia (CDH)
- Congenital pulmonary airway malformation (CPAM)
- Pulmonary sequestration

Congenital Diaphragmatic Hernia (CDH)

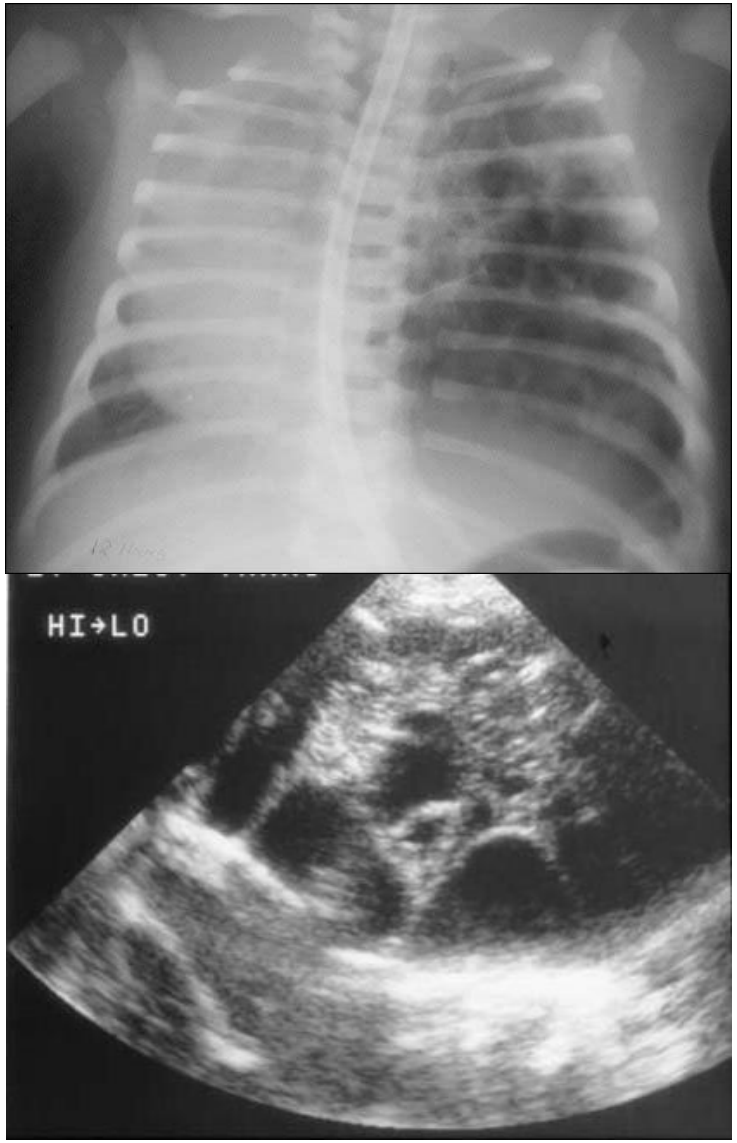
Etiology	Herniation of abdominal organs into thoracic cavity through a defect in the diaphragm. Left sided 80-85%
Clinical Findings	• Respiratory distress and cyanosis at birth with decreased breath sounds on ipsilateral chest • Bowel sounds in affected chest • **Scaphoid abdomen** • Shifted heart sounds • Hypoxemia, hypercapnia, respiratory acidosis, hypoperfusion and hypoxia
Associations	**Hypoplastic lungs, PPHN** Chromosomal abnormalities, CHD and NTD.
Post Natal Management	• **Intubation in DR** • Decompression (low constant suction) • Supportive care/NPO • Mechanical ventilation –HFOV • Nitric Oxide (iNO) • ECMO if indicated



Lt sided

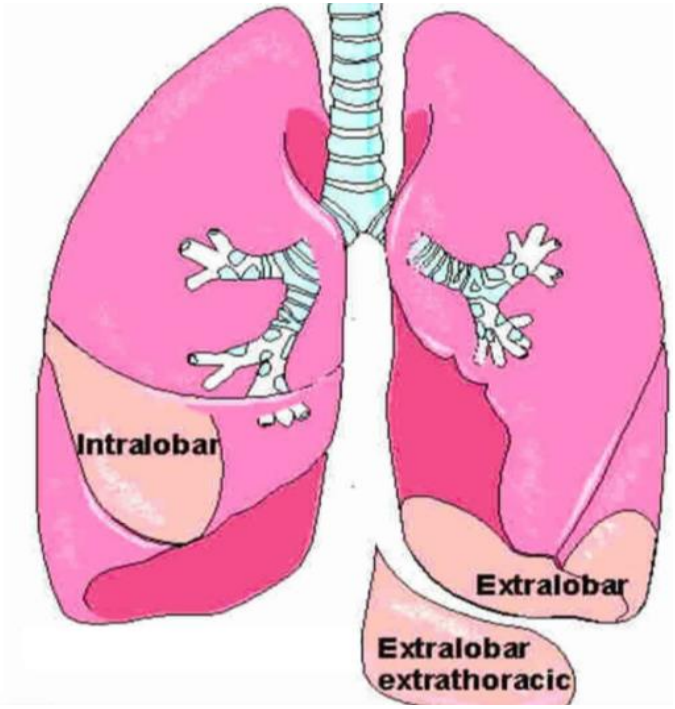
Congenital Pulmonary Airway Malformation (CPAM)

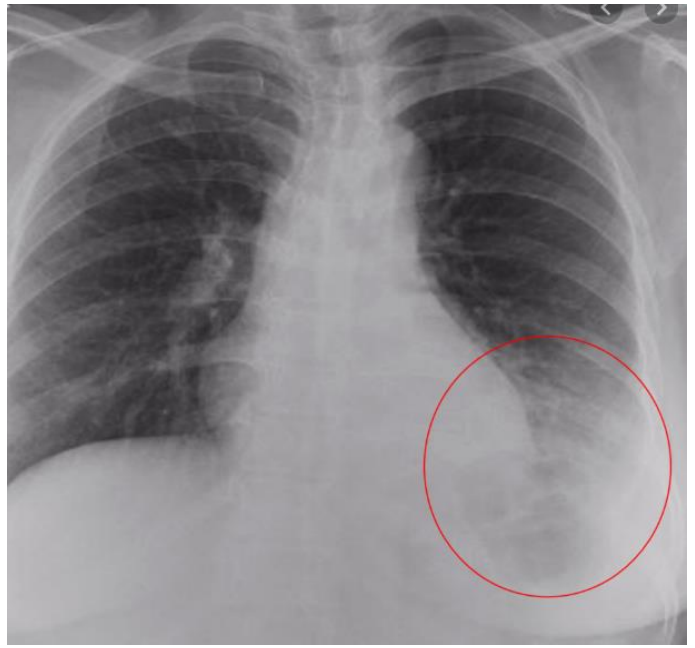
Etiology	Result from abnormalities of branching morphogenesis of the lung. Hamartomatous lesions comprised of cystic and adenomatous elements arising from tracheal, bronchial, bronchiolar, or alveolar tissue. Connected to tracheobronchial tree with blood supply from pulmonary circulation
Clinical findings	25% have symptoms at birth, including mild or severe respiratory distress **Type 1** 60-70%-large cysts 2-10 cm) **Type 2** 15-20% -cysts 0.5-2 cm- 60% associated other anomalies (pulmonary,, cardiac, renal, CNS, bone, Gi) **Type 4** high malignant potential
X-ray findings	Multiple cysts that increase in size with time, compression of surrounding lung tissue
Treatment	Supportive care, complete resection usually recommended d Due to risk of complications



Bronchopulmonary Sequestration

Etiology	Congenital nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its arterial blood supply from the systemic circulation
Types	**Extralobar**: contained within its own visceral pleural envelope 25% (most common in infants M>F) **Intralobar**: contained within the lung 75%
Clinical findings	May be asymptomatic. Can present with polyhydramnios, fetal hydrops, pleural effusion, pulmonary mass lesion. respiratory distress, feeding difficulties
Diagnosis	Prenatal Dx: Doppler US Post natal: CT, angiography (CT or MR)
Treatment	Supportive care, endovascular embolization and coiling, resection



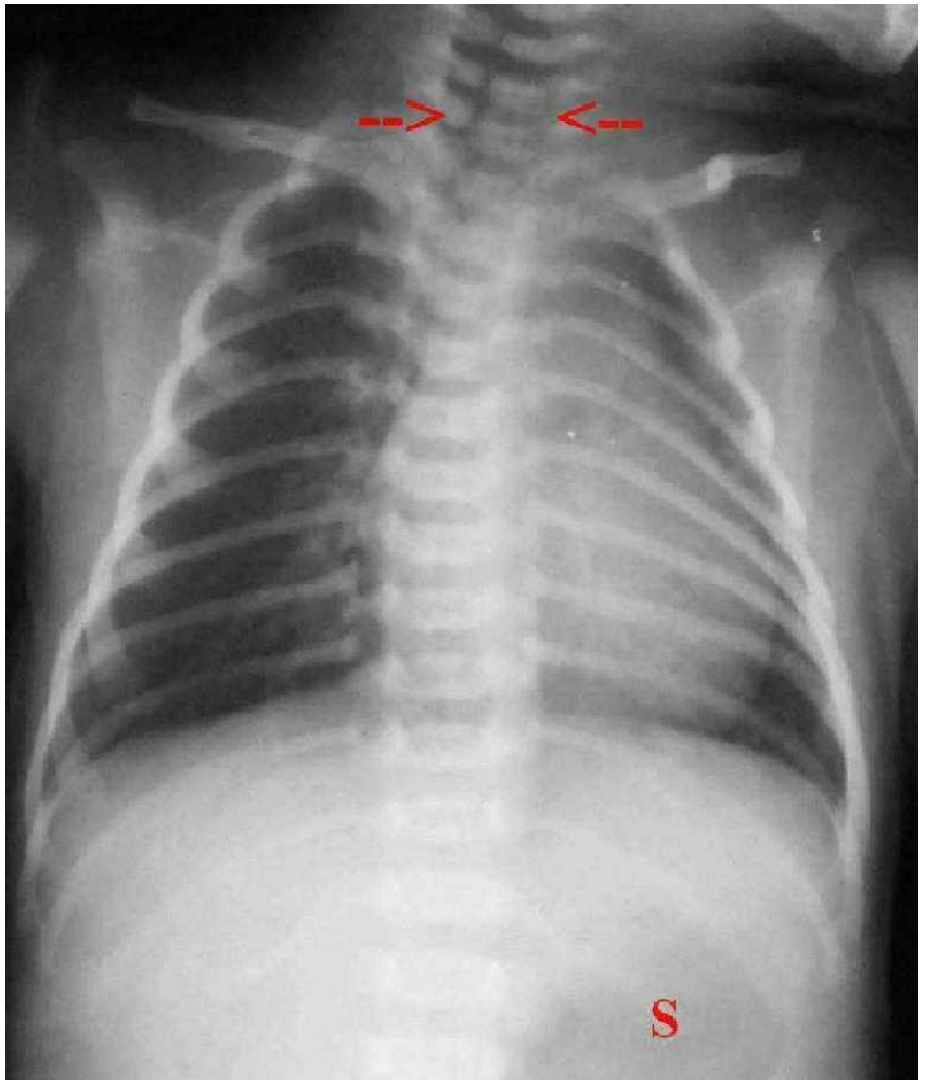


CPAM vs Broncopulmonary Sequestation

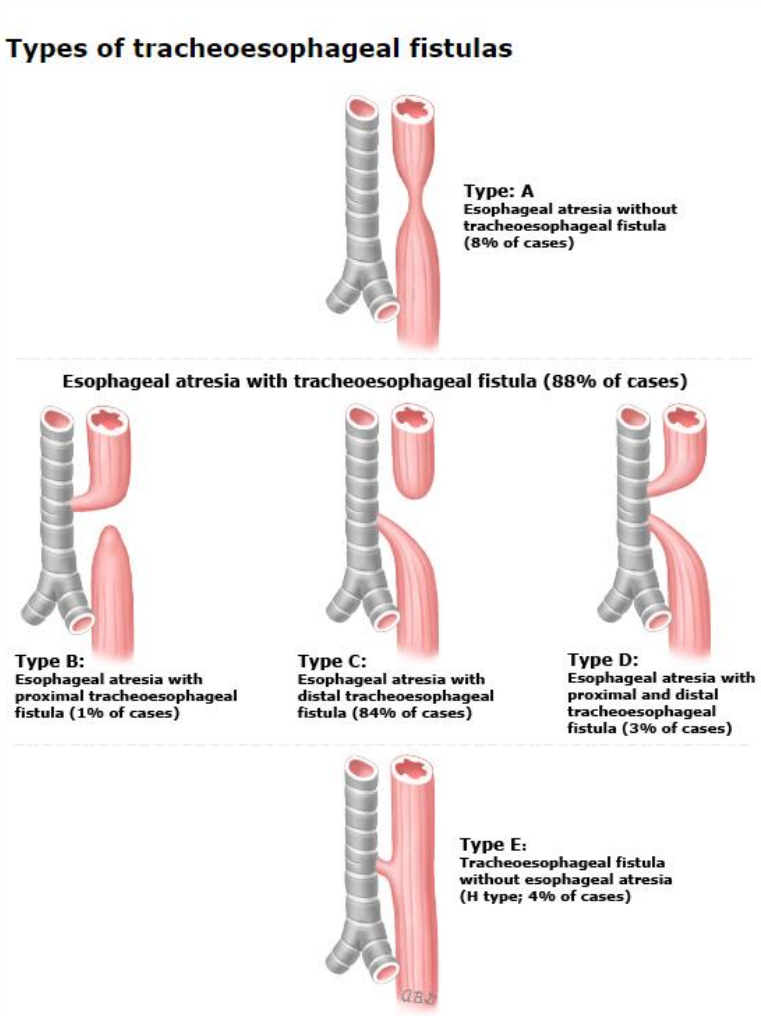
	Congenital Pulmonary Airway Malformation	Bronchopulmonary Sequestration
Classification	Microcystic and macrocystic	Intralobar and extralobar
Connection to tracheobronchial tree	**Yes**	**No**
Systemic blood supply	**No**	**yes**
Associated malformation	Common	Less common
Location	Either lower lobe	Left lower lobe

Tracheoesophageal Defects

Incidence	1 in 3500 to 1 in 4500 live births
Clinical Findings	• Polyhydramnios • **Inability to insert NG tube** • Intermittent cyanosis due to pooling of oral secretions • VACTERL association
X-ray findings	Air delineating esophageal pouch or coiled NG tube, stomach bubble if EA with fistula present
Treatment	Primary anastomosis, fistula ligation



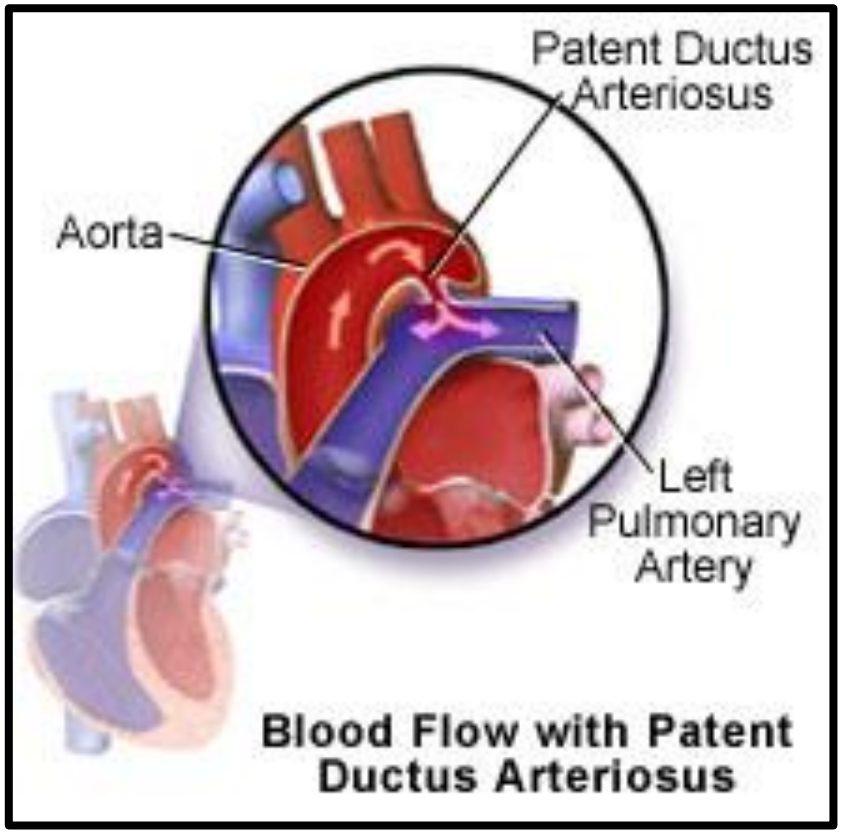
EA with TEF



Tracheoesophageal fistula types classified according to the scheme developed by EC Vogt[1] in 1929, as modified by Gross[2] in 1953.

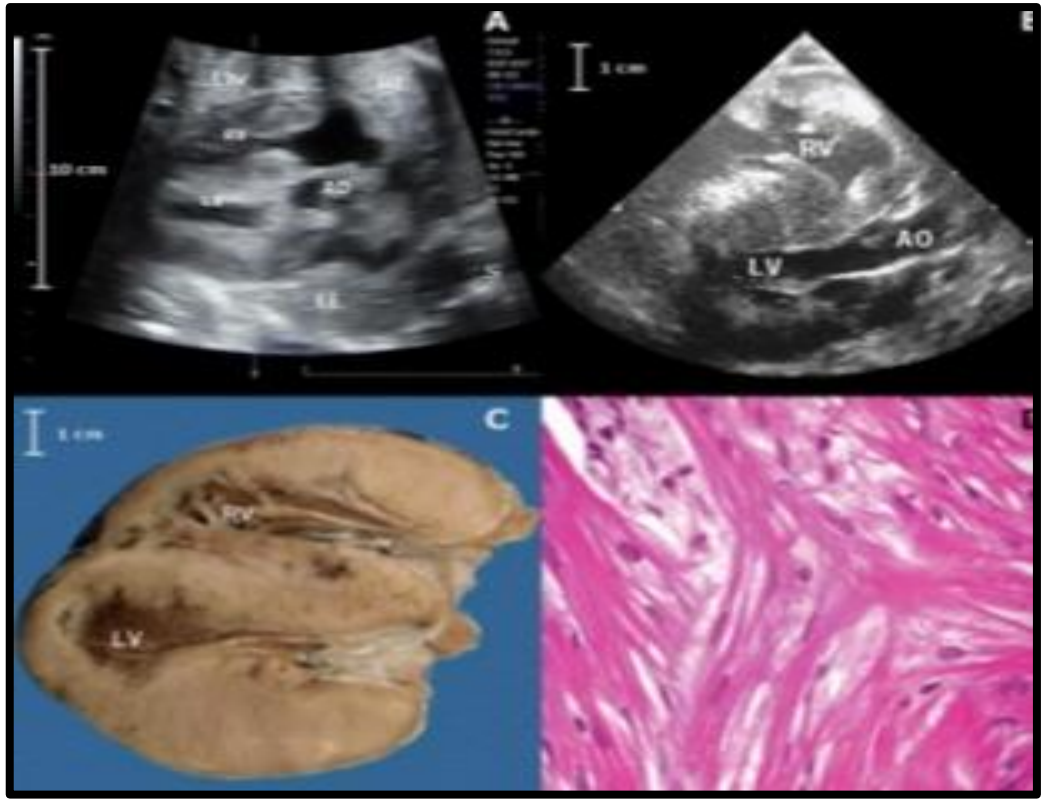
Patent Ductus Arteriosus

Clinical findings	bounding pulses, wide pulse pressure, prominent apical impulse, hyperdynamic precordium, murmur, worsening respiratory status
Diagnosis	Cardiac ECHO
Management	• Observation • Fluid restriction • Meds: Indomethacin/Ibuprofen/ Acetaminophen • Surgical: Ligation or Catheter based interventions



Hypertrophic Cardiomyopathy

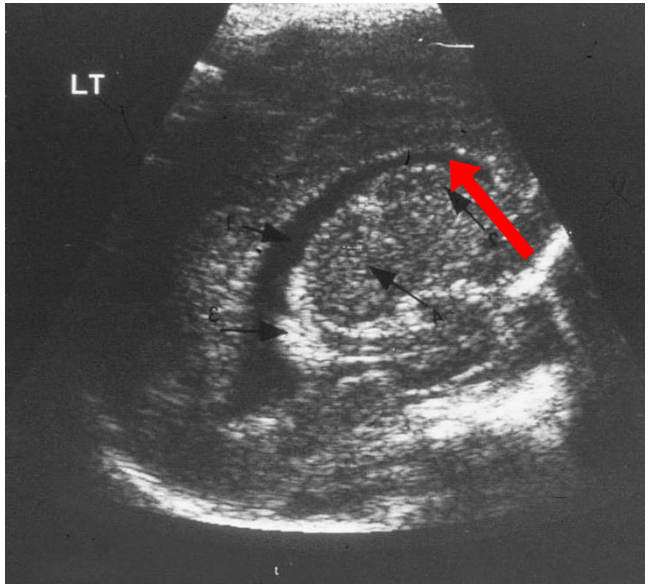
Associations	**IDM**
Presentation	SEM with or without CHF
Diagnosis	Asymmetric thickening of the intraventricular septum and or the ventricular walls
Course	Most often resolves by 6-12 months If severely affected may have signs of LV outflow obstruction and CHF
Treatment	Supportive



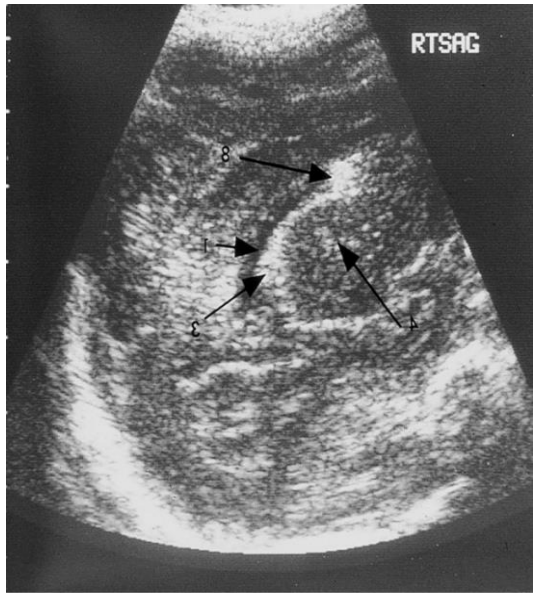
Intraventricular Hemorrhage

Incidence	~20 % BW less than 1500gms
Clinical Findings	Most asymptomatic or abnormal neurologic status, Sz, apnea, bradycardia, increasing support, anemia, full fontanel
Classification	• **Grade I Geminal matrix bleed** • **Grade II Intraventricular without ventricular dilatation** • **Grade III Intraventricular with ventricular dilatation** • **Grade IV Periventricular hemorrhagic infarction**
Diagnosis	Head U/S
Management	Management: supportive care, treat Sz if present,
Complications	Post hemorrhagic ventricular dilation, Periventricular Leukomalacia, White matter injury

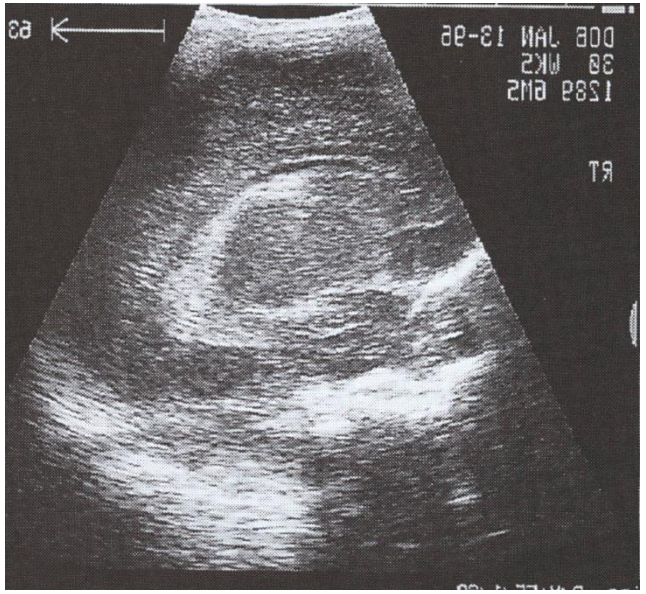
Intraventricular Hemorrhage



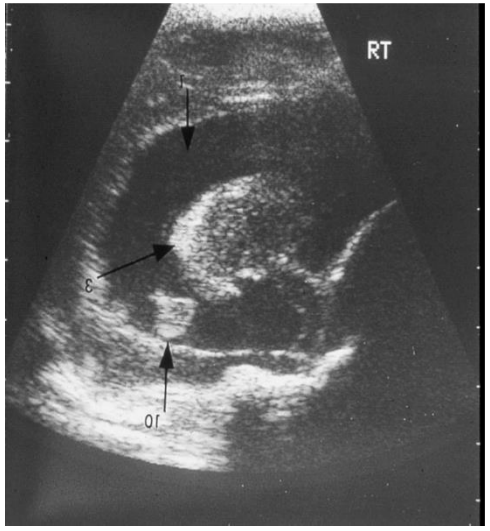
Normal



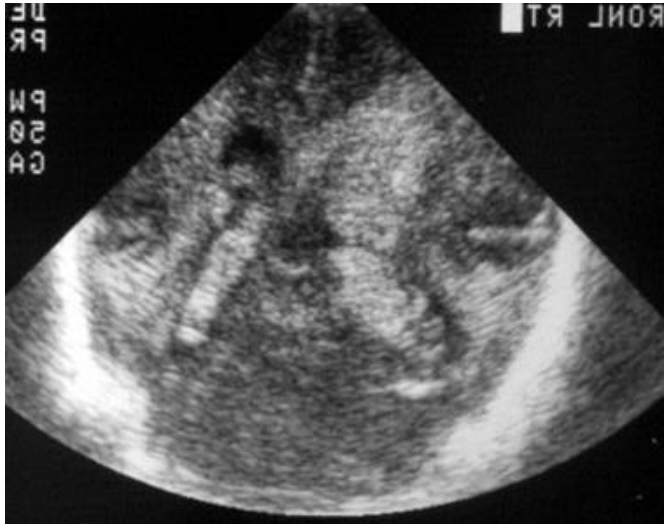
Grade I



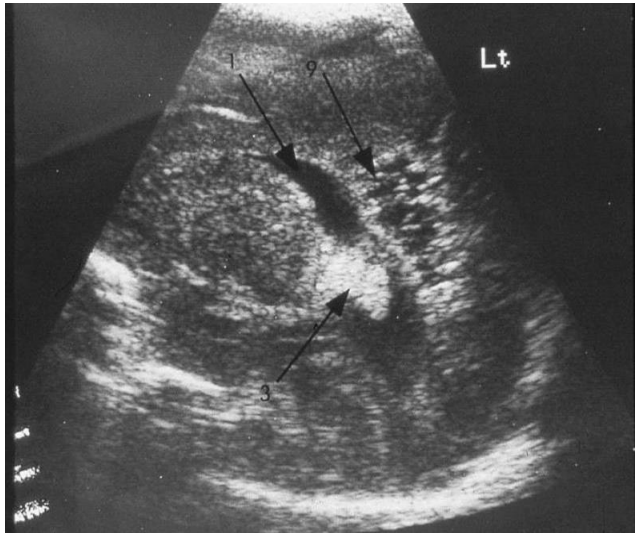
Grade II



Grade III



Grade IV



PVL

Neonatal Seizures

Clinical Findings	May be subtle or present with clonic or tonic movement of one or more extremities
Etiology	**HIE (most common cause if Sz in first 24 hrs)** Other: Hypocalcemia, hypoglycemia, drug withdrawal, causes of cerebral injury, Pyridoxine deficiency
Evaluation	EEG, MRI, metabolic screen, LP
Treatment	Anticonvulsants (phenobarbital, keppra), supportive care, treat underlying condition

Hypoxic Ischemic Encephalopathy (HIE)

Etiology	**Significant hypoxic event in utero or at delivery**
Findings	• Acidosis (pH <7, BD ≥12 mmol/l) • Apgar score <5 at 5 and 10 min • Neurologic findings (Sz, coma, hypotonia) • Acute brain injury on MRI or MRS • Multiple organ involvement (kidney, lungs, intestine, heart and brain
Severity	Sarnat staging
Management	Adequate resuscitation, supportive care, **hypothermia** within 6hrs for 72 hrs

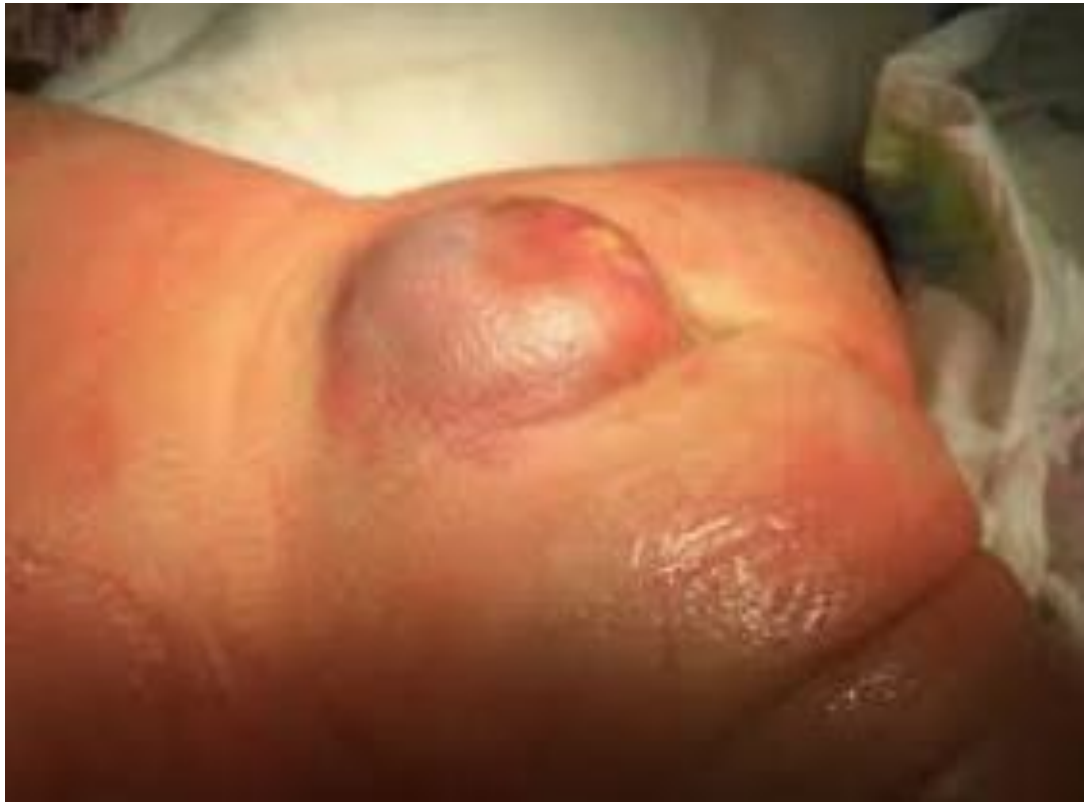
HIE Clinical Staging (Sarnat Staging)

Factor	Stage I (mild)	Stage II (Moderate)	Stage III (severe)
Duration	**<24hrs**	**2-14 days**	**Hours to weeks**
Level of consciousness	**Hyperalertness, irritability**	**Lethargy**	**Deep stupor or coma**
Muscle tone	**Normal or increased**	**Hypotonia**	**Flaccidity**
Pupils	**Dilated, reactive**	**Constricted, reactive**	**Variable, fixed**
Respirations	**Regular**	**Periodic**	**Apnea**
Seizures	**None**	**Common (70%)**	**Uncommon**
EEG	Normal	Low voltage/Sz activity	Periodic or isoelectric
Risk of death	<1	5%	>60%
Risk of handicap	<1%	20%	>70%

Sarnat staging bolded

Neural Tube Defects

Etiology	•Multifactorial •Abnormal embryological formation of the neural tube during the early weeks of pregnancy
Prevalence	• USA 5.5-6.5 per 10,000 live births
Detection	• Elevated AFP • Prenatal U/S
Types	• Anencephaly (lethal) • Encephalocele (most occipital) • Spina Bifida Oculta (intact spinal cord) • Meningocele (herniation of meninges) • Myelomenigocele (herniation of spinal cord tissue)
Prevention	**Folic acid**
Treatment	Multidisciplinary care ***Surgical closure soon after birth**



Neonatal Infections

- Clinical syndrome characterized by systemic signs of infection frequently accompanied by bacteremia
  - Positive blood culture confirms sepsis
  - Negative defined as “clinical sepsis”

Early Onset Sepsis (EOS) (< 72hrs)	Acquired vertically from the mother, related to perinatal risk factor and to maternal flora
Late Onset Sepsis (LOS) (≥ 72 hrs)	Vertical transmission, resulting in initial neonatal colonization that evolves into later infection •Horizontal transmission from contact with care providers or environmental sources

Sepsis Calculator

Early Onset Sepsis in Infants >35 wks

Risk calculator predictors:	• Incidence of Early Onset Sepsis
	• Gestational age
	• Highest maternal antepartum temperature
	• ROM (hrs)
	• Maternal GBS status (positive, negative or unknown)
	• Type of intrapartum antibiotics
	  ○ Broad spectrum antibiotics >4 hrs PTD
	  ○ Broad spectrum antibiotics 2-3.9 hrs PTD
	  ○ GBS specific antibiotics > 2 hrs PRD
	  ○ No antibiotics or any antibiotics < 2hrs PTD

Predictor	Scenario
Incidence of Early-Onset Sepsis	
Gestational age	weeks days
Highest maternal antepartum temperature	Fahrenheit
ROM (Hours)	
Maternal GBS status	○ Negative ○ Positive ○ Unknown
Type of intrapartum antibiotics	○ Broad spectrum antibiotics > 4 hrs prior to birth ○ Broad spectrum antibiotics 2-3.9 hrs prior to birth ○ GBS specific antibiotics > 2 hrs prior to birth ○ No antibiotics or any antibiotics < 2 hrs prior to birth

Calculate » Clear

Risk per 1000/births	
EOS Risk @ Birth	

EOS Risk after Clinical Exam	Risk per 1000/births	Clinical Recommendation	Vitals
Well Appearing			
Equivocal			
Clinical Illness			

Classification of Infant's Clinical Presentation Clinical Illness Equivocal Well Appearing

Neonatal Early-Onset Sepsis Calculator

Neonatal Sepsis

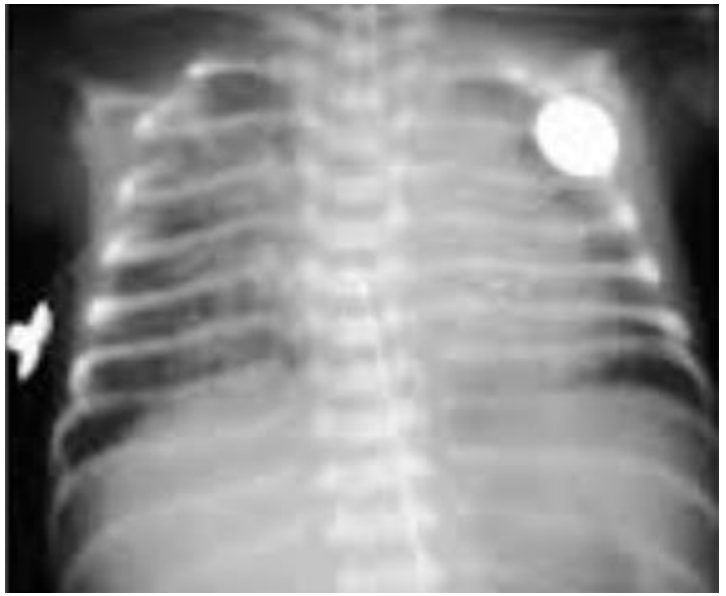
	Early	Late
Etiology	• GBS • E.Coli • Listeria	• **Home:** GBS,E. Coli, Listeria, Gram negatives, Herpes • **Hospital:** Staph Epi,Gram negatives (Klebsiella, Enterobacter, Pseudomonas, Serratia) • Fungal (Candida)
Risk Factors	Maternal: • GBS colonization • Chorioamnionitis • Amniotic fluid infection • PROM >18 hrs Infant: • Prematurity	• Intravascular catheters • Reservoir, VP shunts • Urinary catheters
Treatment	Ampicillin & Gentamicin	Coverage for suspected organisms

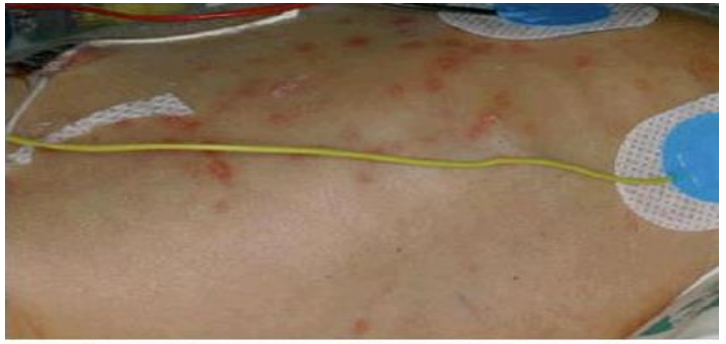
Group B Strep Sepsis

Presentation	Early	Late
Transmission	Vertical	Horizontal
Prevention (Abx prophylaxis)	**Yes**	**No**
Presentation	Respiratory distress **Pneumonia** Mostly presents in the first 24 hrs	**Meningitis** Can also cause focal infection in bone and joints

Listeria Monocytogenes Infection

Etiology	Listeria Monocytogenes
History	• Maternal ingestion of unpasteurized milk, soft cheeses and undercooked poultry • Presents with PTL, **maternal flu-like symptoms**, GI symptoms and fever • Meconium stained fluid at delivery
Presentation	• **Early onset**: Pneumonia, septicemia and rash ”granulomatosis infantisepticum” • **Late onset:** Meningitis
X-ray	Patchy infiltrates (resembles MAS)
Treatment	Ampicillin





(Reprinted with permission from Heras PC, Garcia-Patos V, Palacio L *Actas Dermosifiliogr.* 2006;97:59-61)

Congenital Infections

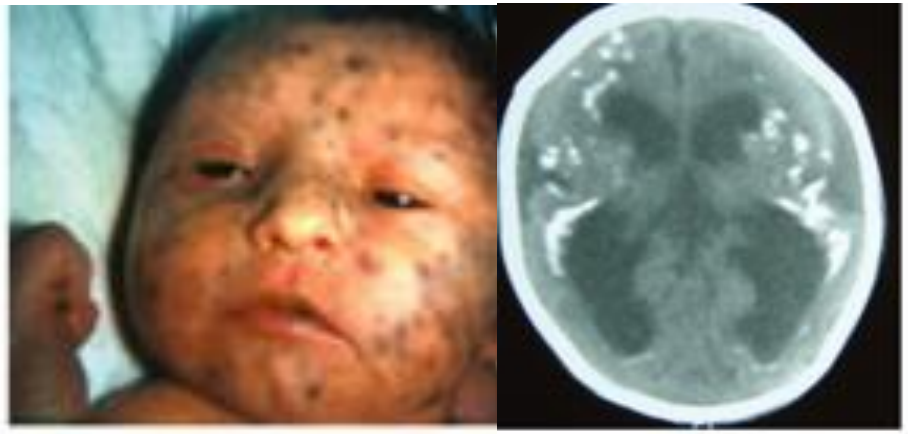
Disease	Findings	Treatment
CMV	IUGR, microcephaly, CNS calcifications (periventricular), chorioretinitis neurosensory hearing loss, HSM jaundice,rash, diffuse petechiae blueberry muffin, pneumonia, MR	Ganciclovir
Congenital Rubella	Classical findings: • **Sensorineural deafness** • **Cataracts** • **Cardiac defect** (**PDA**, PA stenosis,CoA) • **IUGR**	No effective treatment/isolate Maternal vaccination
Neonatal HSV Infection	Disseminated, Encephalitis or Localized	Acyclovir
HIV Infection	Potential growth restriction, does not manifest in newborn period C/S delivery (prior ROM)) Avoidance of breast feeding	Antiretroviral prophylaxis PCP prophylaxis-Bactrim

Congenital Infections

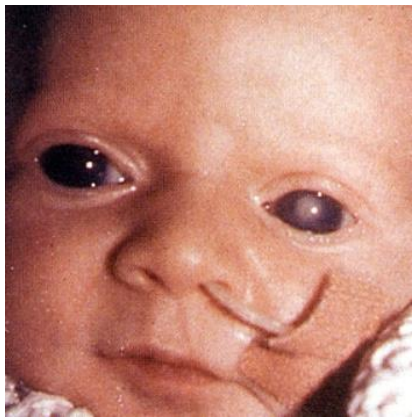
Disease	Findings	Treatment
Toxoplasmosis	**Retinitis, CNS calcifications**, seizures HSM, jaundice	Pyrimethamine, sulfadiazine, folinic acid
Syphyllis	PT, Hydrops, IUGR, HSM, hem anemia, jaundice, **rash palm and soles, periosteal changes,** neurosensory hearing loss, rhinorrhea	Penicillin
Varicella	Eye, CNS damge, **skin scarring** **Limb hypoplasia**	Acyclovir VZIG- if 5days PTD to 2 days after delivery, isolate
Parvovirus	Anemia, **nonimmune hydrops**, ascitis, cardiomegaly	Supportive care

Congenital Infections

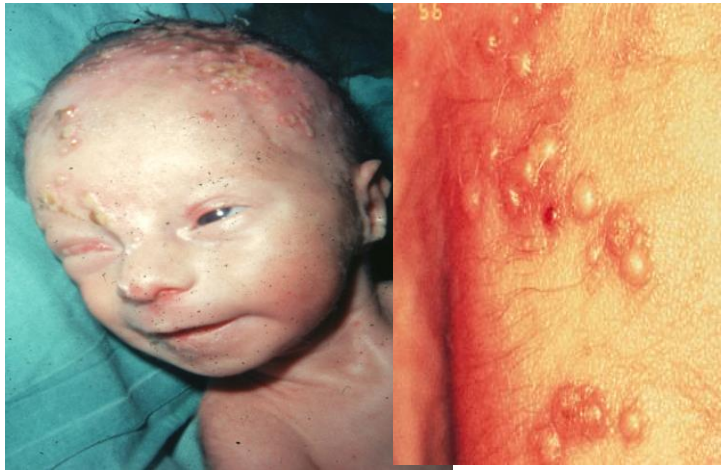
TOXOPLASMOSIS



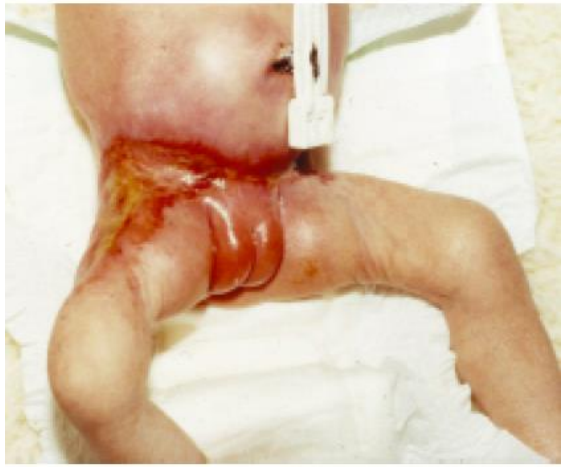
CMV



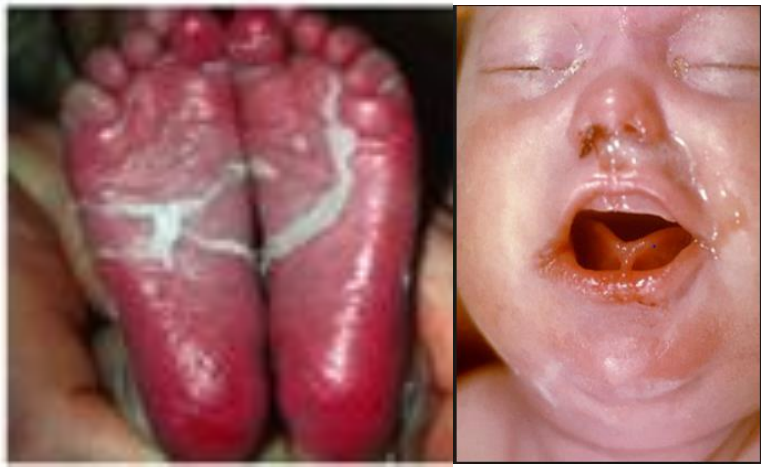
RUBELLA



HERPES



VARICELLA



SYPHILLIS

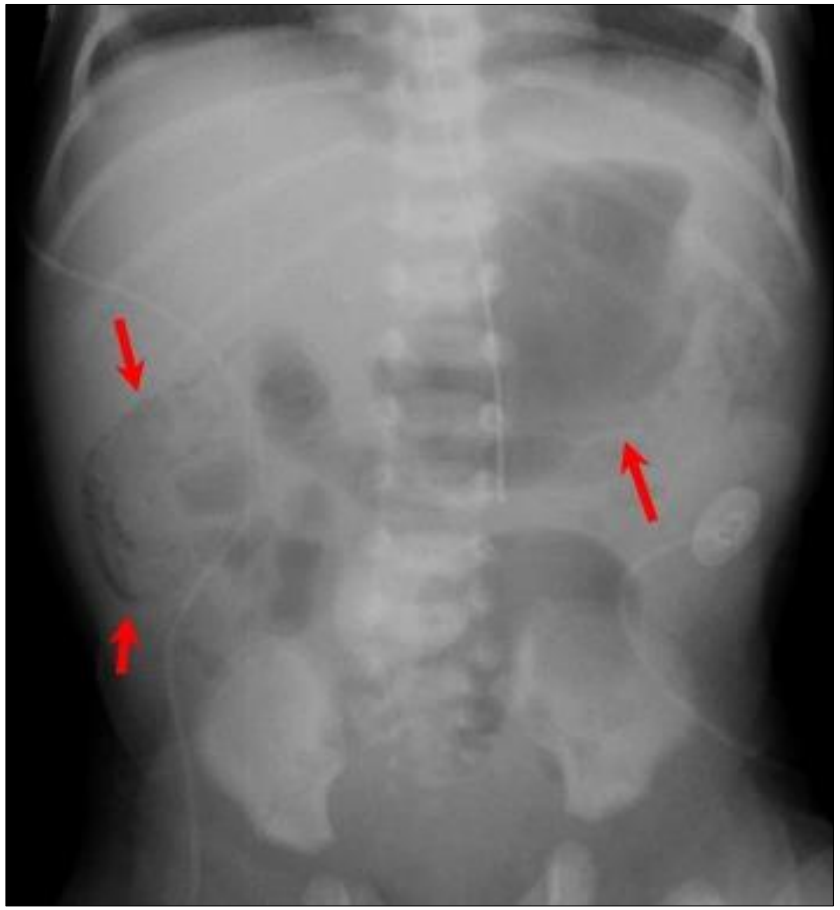
Neonatology at a Glance ,2nd edition 2011

Conjuntivitis

Type	Onset	Characteristics	Treatment
Chemical	**Within 24 hr of exposure**	After Silver Nitrate, (presently used <1%)	Resolves after 48 hrs
Acute Purulent	**24-48 hrs or later**	Staph Aureus, GBS,H Influenza, Strep Pn, Pseudomonas	Topic antibiotic Pseudomonas + IV Antibiotics
Gonorrhea	**2-5 days**	Copious purulent, bilateral-emergency eval/Rx – May lead to ulceration/perforation if untreated	Third generation cephalosporin
Chlamydia	**5-14 days**	Watery discharge that becomes purulent. Often associated with pneumonia	Erythromycin 14d
Herpes simplex	**4 d-3 wks**	Most frequent viral etiology, may lead to keratitis, chorioretinitis Assess for systemic herpes	Acyclovir

Necrotizing Enterocolitis (NEC)

Risk Factors	• Prematurity • Intestinal ischemia • Feedings (formula>BM, rapid increases), Abnormal bacterial colonization
Clinical Findings	abdominal distention, gastric aspirates, heme + or bloody stools
Xray	**Pneumatosis intestinalis,** portal air, free air, fixed loop
Management	NPO (bowel rest), NG suction, IVF, antibiotics, serial KUB's. Surgery if perforation
Complications	Short bowel syndrome, strictures



NEC: Bell Classification

Stage	Clinical Findings	X-ray Findings
I. Suspect	Abdominal distension, poor feeding, vomiting	Ileus
2. Definite	Above + gastrointestinal bleeding	Pneumatosis intestinalis
3. Advanced	Above + clinical deterioration, shock	Portal venous gas, perforation

Spontaneous Intestinal Perforation (SIP)

At risk	**ELBW infants most affected**
Comparison to NEC	• May be indistinguishable from NEC • Less frequent with • Better prognosis • Infants smaller and more PT
Clinical findings	• Obvious clinical signs of bowel perforation are infrequent • May present with dramatic abdominal distention often associated with bluish discoloration of abdominal wall
Associations	postnatal steroids use, systemic candidiasis fetal or neonatal hypoxia, **Indomethacin use**, PDA, IVH
Treatment	Peritoneal drainage or exploratory lap

Short Bowel Syndrome

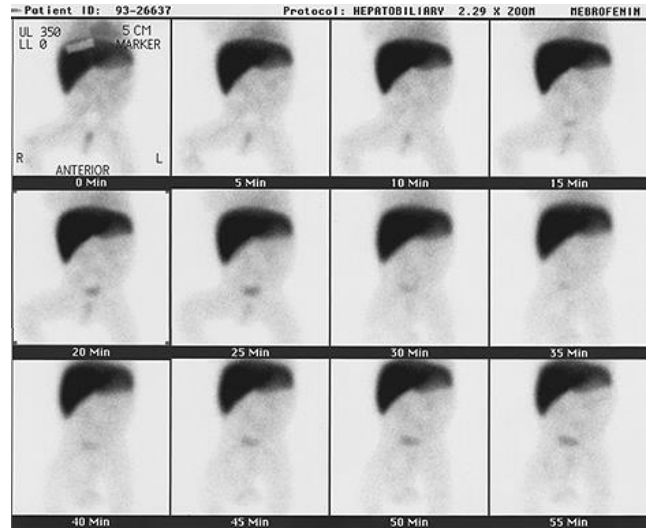
Findings	Malabsorption and malnutrition as a result of bowel shortening
Causes	Extensive resection of bowel • **NEC** *** most common, approximately 50% • Jejunal or ileal atresia • Midgut volvulus • Extensive Hirschsprung's disease • Omphalocele or gastroschisis
Prognosis	Depends on length of remaining gut Site of intestinal loss (Adaptive process greater in ileum than jejunum),presence or absence of ileocecal valve
Treatment	• Nutritional support (diet adjustments, vitamins and mineral supplementation) • GT feeds/Parental Nutrition • Medications • Surgical intervention

Direct Hyperbilirubinemia

Hepatocellular disorders	• Neonatal idiopathic hepatitis • Infection (TORCH, syphilis, systemic infections) • Prolonged hyperalimentation • Intestinal obstruction • Metabolic disorders
Ductal disorders	• Biliary Atresia • Alagille syndrome • Choledocal cyst
Management	Treat underlying condition Ursodiol Biliary Atresia-Surgery

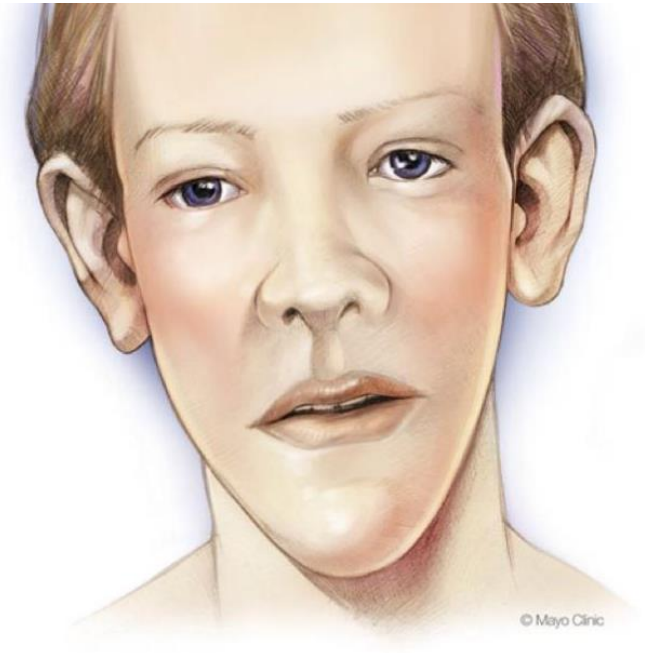
Biliary Atresia

Etiology	Obstruction of bile flow in the extrahepatic bile duct system
Clinical findings	• Jaundice between 2nd and 6th week of life • **Acholic stools** (clay or white colored) • Hepatomegaly • Portal hypertension
Associated anomalies	Cardiovascular, polysplenia, malrotation, situs inversus, intestinal atresia
Diagnosis	Elevated Direct Bilirubin Gallbladder US Hepatobiliary scan
Treatment	Kasai Procedure: critical age for surgery: less than 10-12 weeks

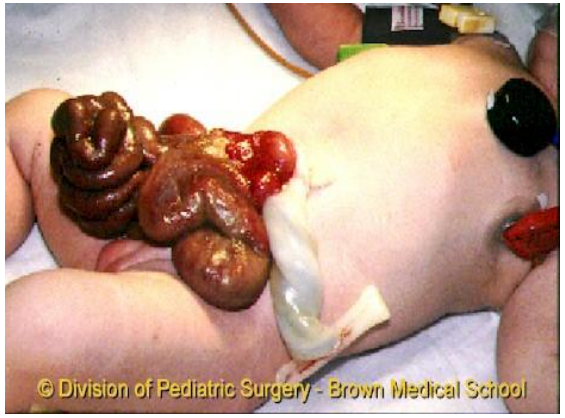


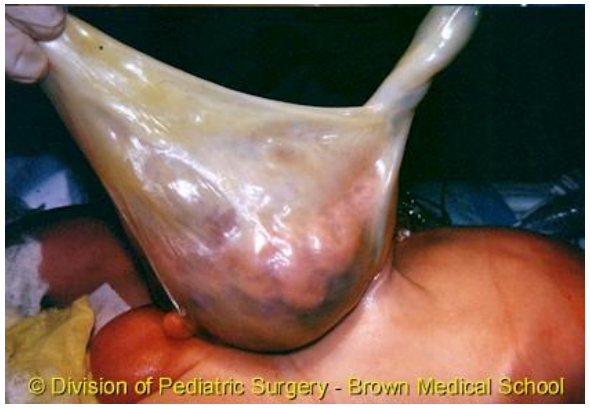
Alagille Syndrome

Inheritance	Autosomal dominant disorder with variable phenotype
Findings	Bile duct paucity, cholestatic liver disease, eye, cardiac (PPS), renal, and skeletal malformations, CNS vasculopathy
Characteristic facial appearance	Broad forehead, deep set eyes, pointed chin, elongated nose with bulbous tip



Abdominal Wall Defects

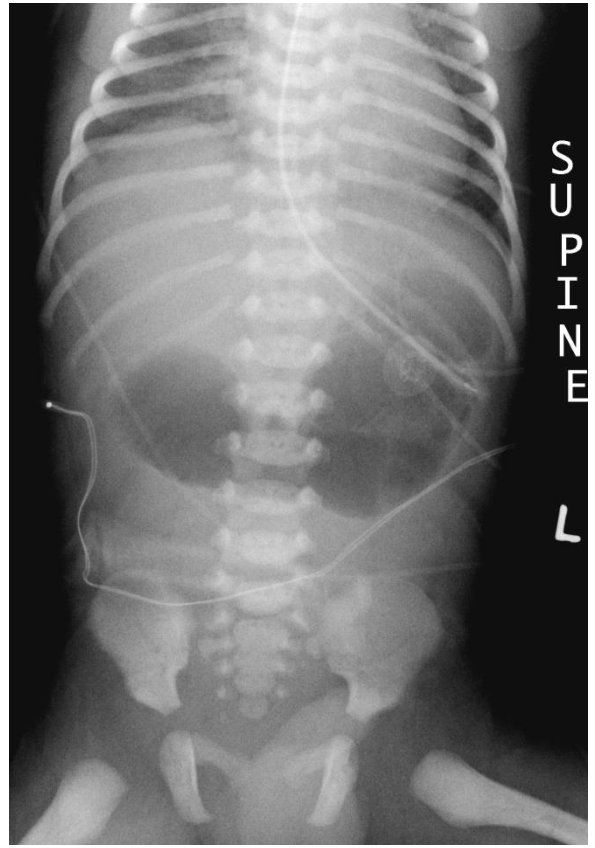




Bowel Herniation	Gastroschisis	Omphalocele
Location	**To Rt of umbilicus**	**In the umbilicus**
Findings	**Extruding organs**	**Covering sac/umbilical vessels at apex**
Bowel appearance	Matted, edematous	Pink, normal
GI anomalies	Atresias	Volvulus (rare)
Non GI anomalies	rare	**common**
Syndrome association	none	Beckwith-Weiderman, CHD, T13, T18, GI and urinary tract anomalies

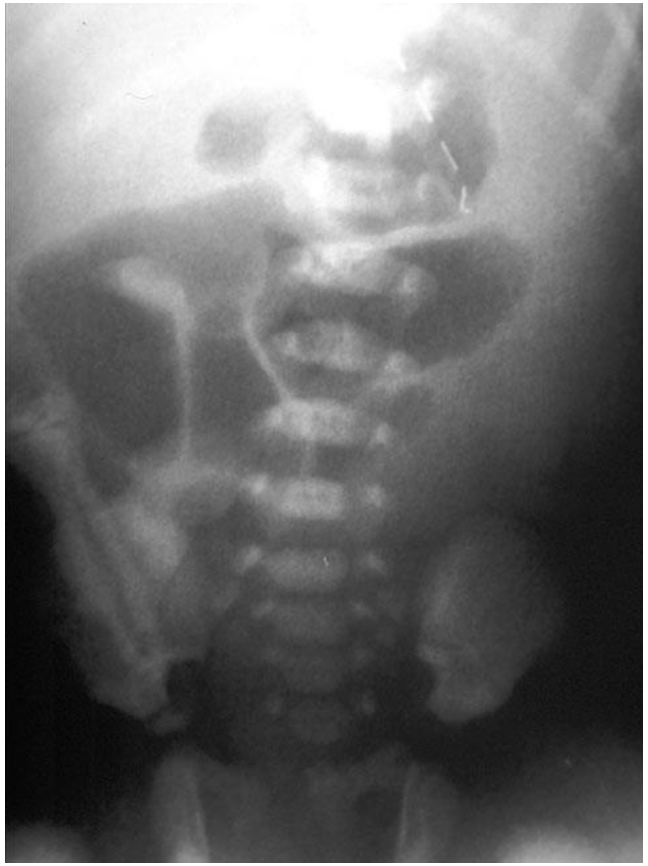
Duodenal Atresia

Incidence	1 per 10,000 live births
Etiology	Failure of recanalization of GI tract during first trimester
Associations	Isolated or associated with congenital heart disease or renal anomalies 30% of affected infants have **Down syndrome**
Clinical findings	Bilious vomiting, abdominal distension, signs of bowel obstruction
Diagnosis	X-ray **double bubble** UGI- confirms atresia
Treatment:	Resection/reanstomosis (duodenoduodenostomy)

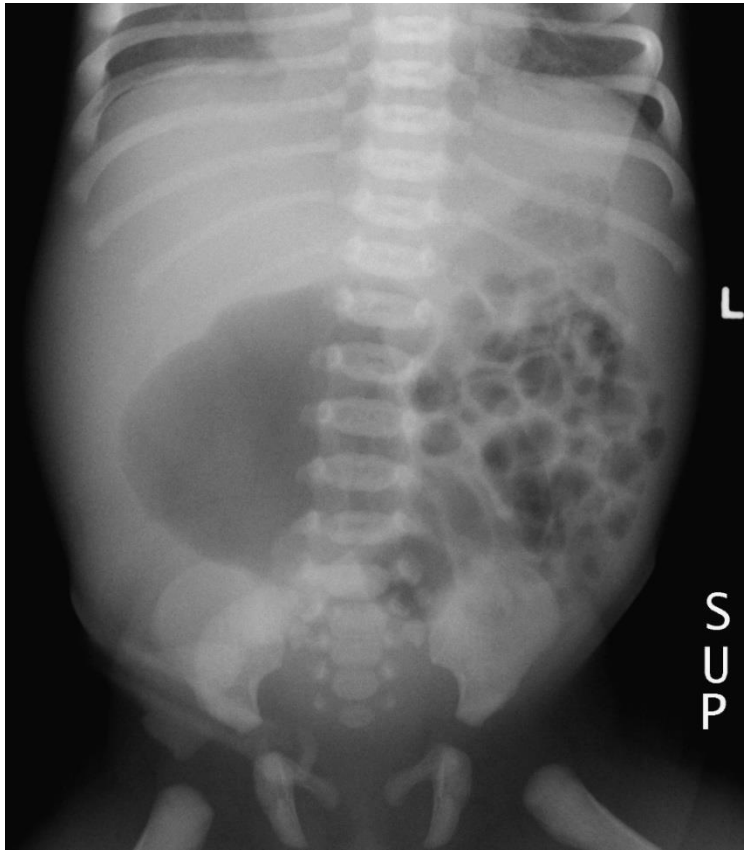


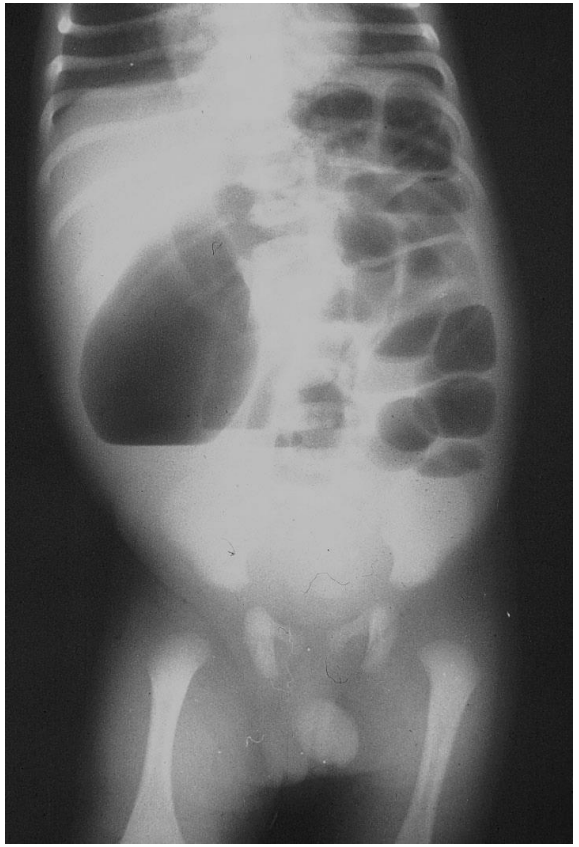
(double bubble sign)

Jejunal Atresia



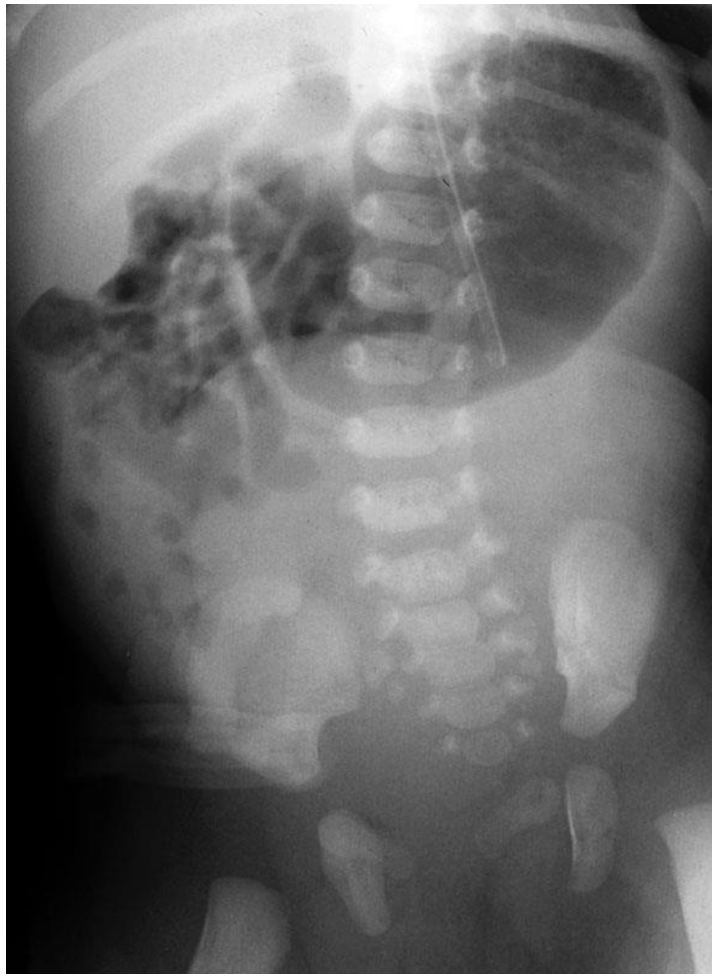
Ileal Atresia





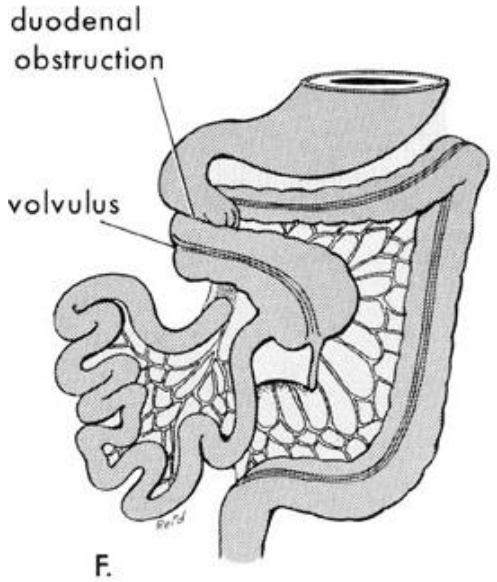
Malrotation

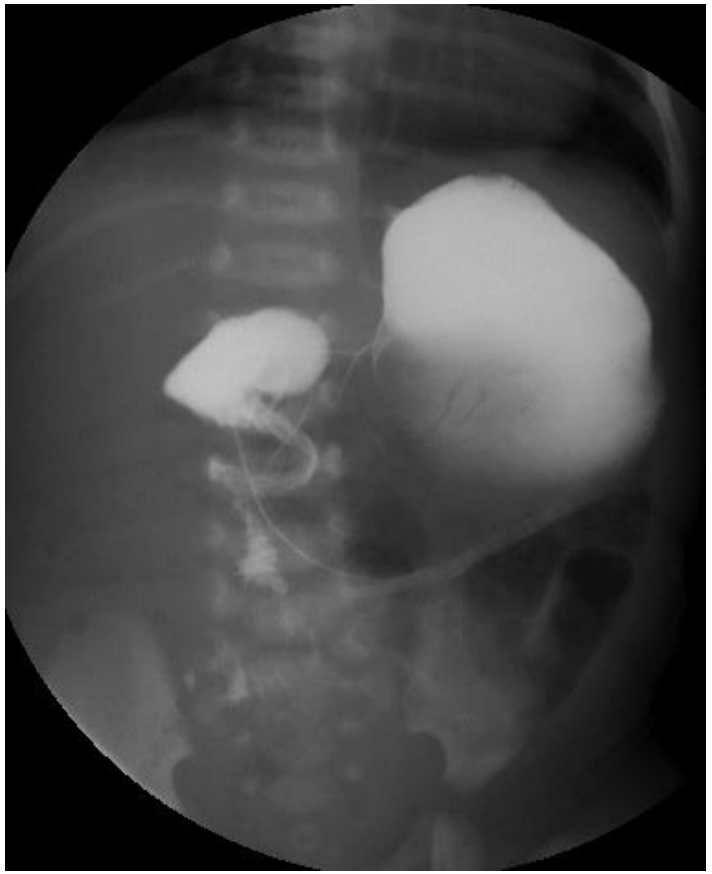
Etiology	Due to incomplete rotation and fixation of the embryonic intestine as it returns to the fetal abdominal cavity from its embryonic extracoelomic position
Findings	Distal duodenum fails to cross to the left of the vertebral bodies to join the jejunum at the ligament of Treitz
Associations	DA, small intestinal atresia, gastroschisis, omphalocele and CDH, cardiac and renal malformations
Clinical findings	Presents with bile stained emesis and distension and obstruction may be intermittent
Diagnosis	**UGI**
Treatment	Ladd's procedure



Midgut Volvulus

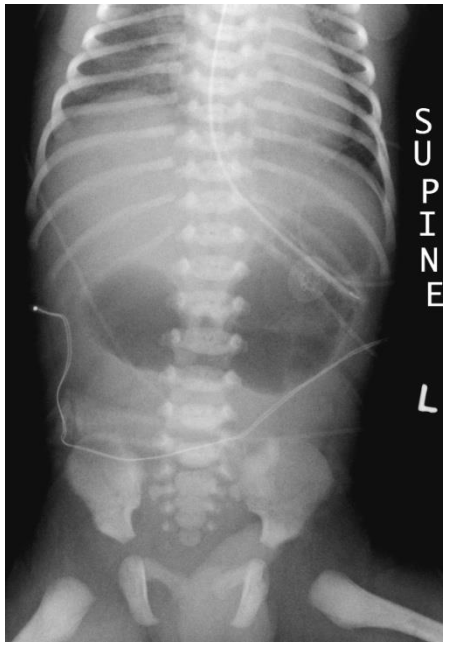
Etiology	Strangulation obstruction with occlusion of blood flow to the gut
Clinical Findings	Variable depending on: • Degree of volvulus (i.e. number of twists of the bowel) • Duration of volvulus, and viability of the bowel involved Bilious emesis in previously well newborn, abdominal distention, abdominal mass, electrolyte imbalance, shock, lethargy, death
Diagnosis	UGI
Treatment	**SURGICAL EMERGENCY!!**



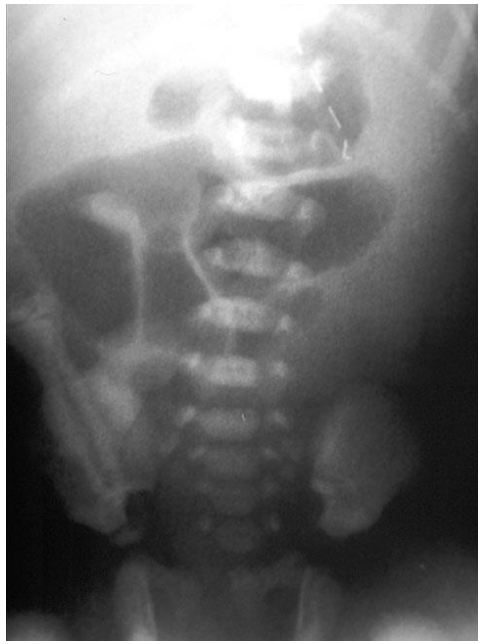


Proximal Bowel Obstruction

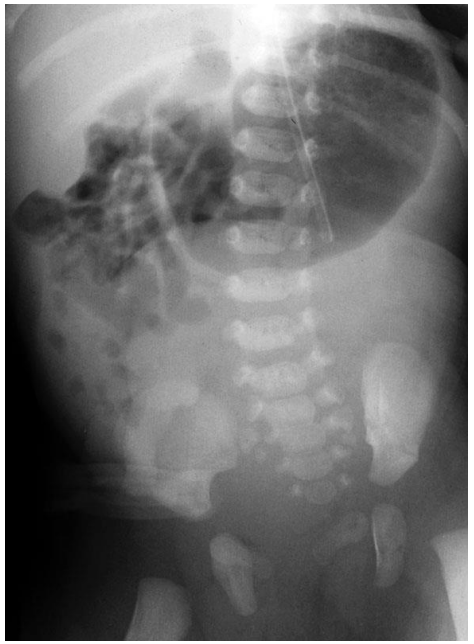
- Bilious vomiting- Bowel obstruction
- Abdominal distension- Bowel atresias, malrotation
- **Diagnosis- UGI**



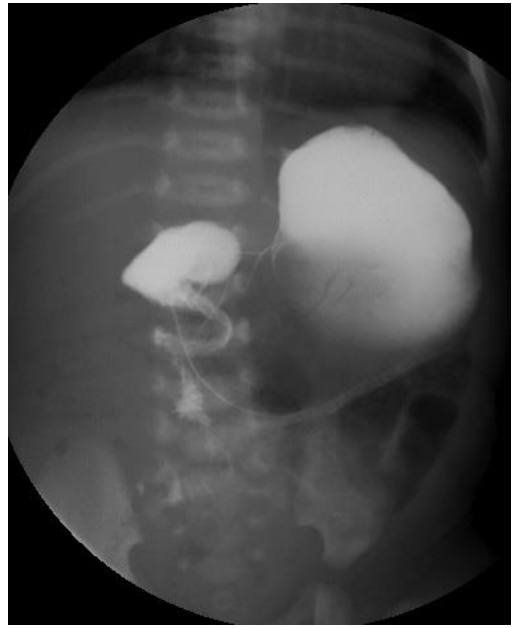
Duodenal Atresia



Jejunal Atresia



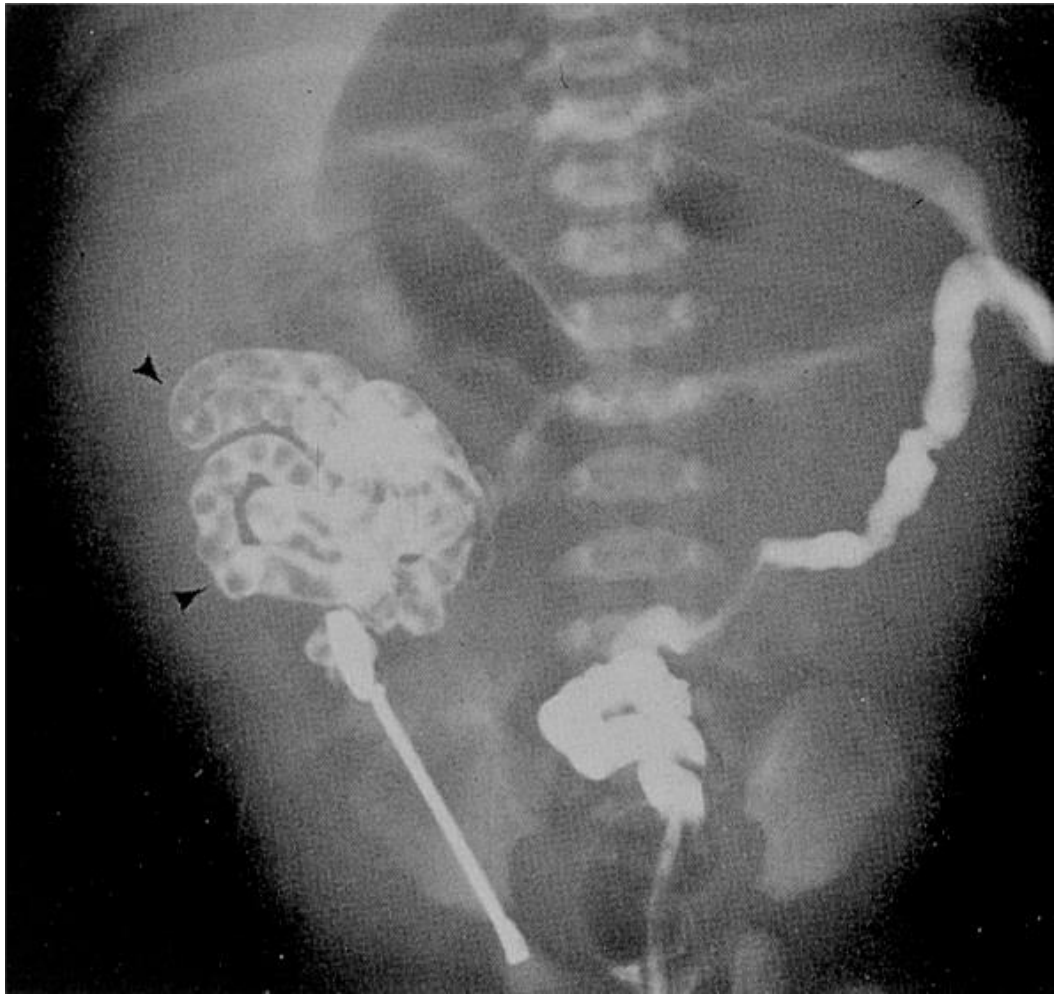
Malrotation



Midgut Volvulus

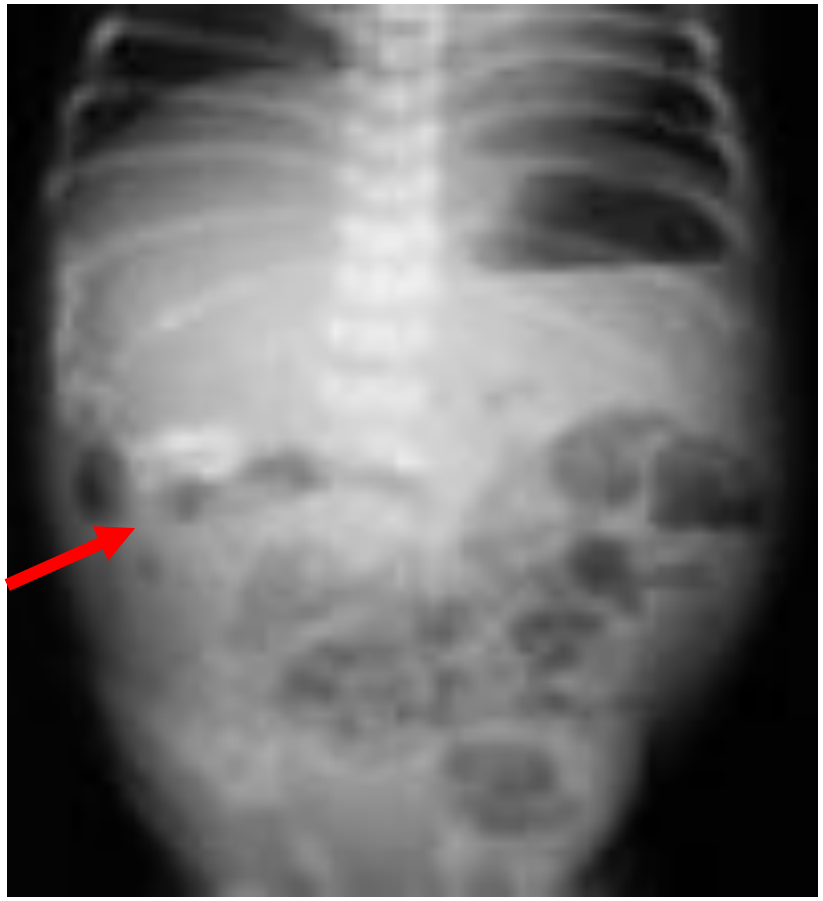
Meconium Ileus

Incidence	~10% of infants with CF presenet with MI at birth. **>80-90 % cases with MI have CF**
Etiology	obstruction of the small intestine at the level of the terminal ileum with inspissated meconium. .→ may lead to perforation
Presentation	• Bile stained emesis • Abdominal distension secondary to obstruction from impaction of thick meconium meconium filled loops have a doughy feel
Radiologic findings	Small bowel obstruction with numerous, variably sized air filled loops. “Soap bubble” appearance in the right lower quadrant
Management	A contrast enema will demonstrate a microcolon and can be therapeutic



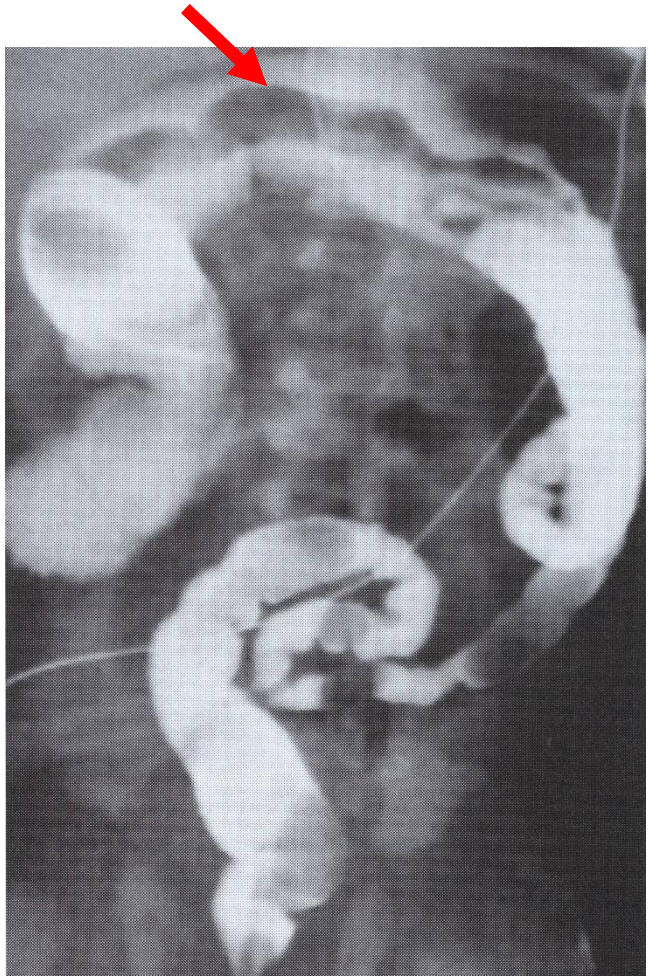
Meconium Peritonitis

Etiology	• Calcifications a results of **intrauterine intestinal perforation** with meconium spillage into the peritoneal cavity • GI perforation due to obstruction and/or volvulus
X-rays	Calcifications in abdomen
Associations	**Meconium ileus (CF)**



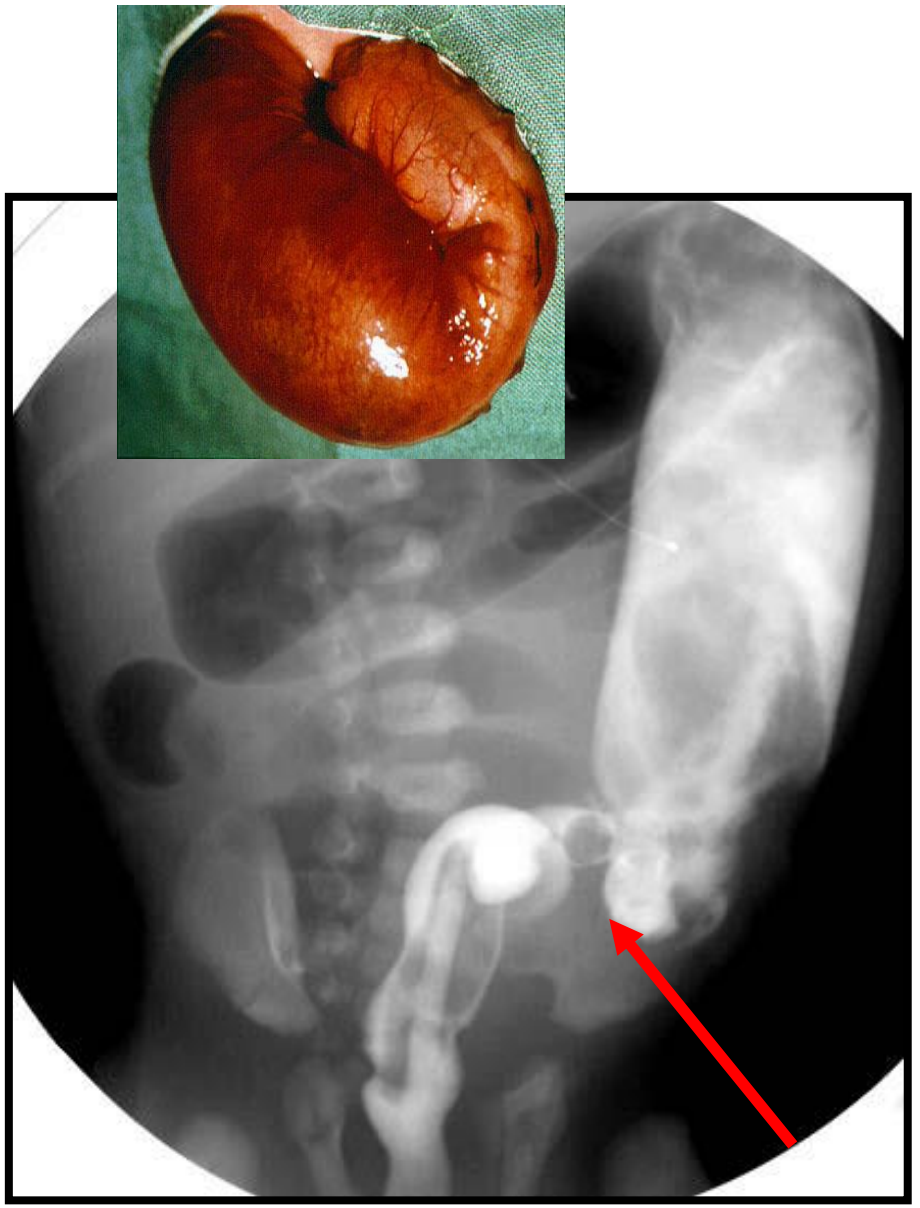
Meconium Plug

Etiology	Part of the spectrum of colonic hypomotility
Associations	PT, IDM, Mg exposure, CF
Clinical findings	Abdominal distension and delayed stool passage. Bilious vomiting may be present Typically benign course
Radiologic findings	Contrast Enema shows empty distal colon, dilated proximal bowel and a filling defect caused by the plug
Management	Abdominal decompresion **Contrast enema -dual function**: diagnosis and therapy



Hirschsprung's Disease

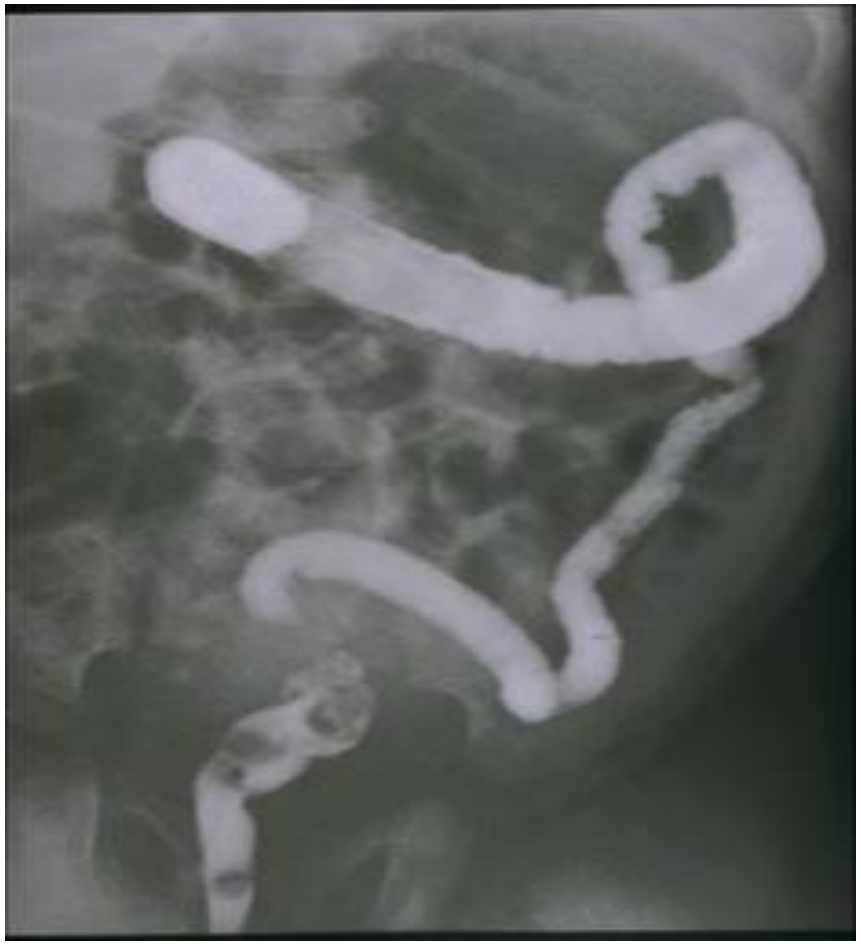
Incidence	1/5000 births
Etiology	**Absence of parasympathetic innervation** to the distal intestine due to failure of neural crest cells (precursors of enteric ganglion cells) to migrate completely during intestinal development during fetal life
Presentation	• 10-20% present in NB period • 80% affect rectosigmoid colon • Presents with distension, vomiting and delayed stool passage (>48 hrs)
Radiologic findings	Contrast enema: **transition zone**, narrowed aganglionic segment with dilated proximal segment
Diagnosis	**Rectal biopsy** (absence of ganglion cells is diagnostic
Treatment	Excision of diseased portion Primary pull-through or staged procedure



Transition zone

Small Left Colon

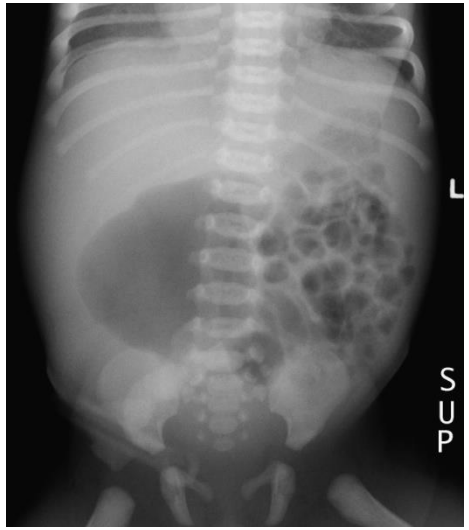
Associations	Maternal - **diabetes,** hypothyroidism, toxemia Prematurity
Etiology	• **Functional immaturity of ganglion cells** • Transient functional colonic obstruction • Primarily affects descending and rectosigmoid colon
Presentation	Abdominal distension, failure to pass meconium
Radiologic findings	Contrast enema - significant caliber reduction of sigmoid and descending colon
Treatment	• May resolve spontaneously • Contrast study usually resolves the obstruction • Most do not require surgery unless abdominal distension leads to perforation



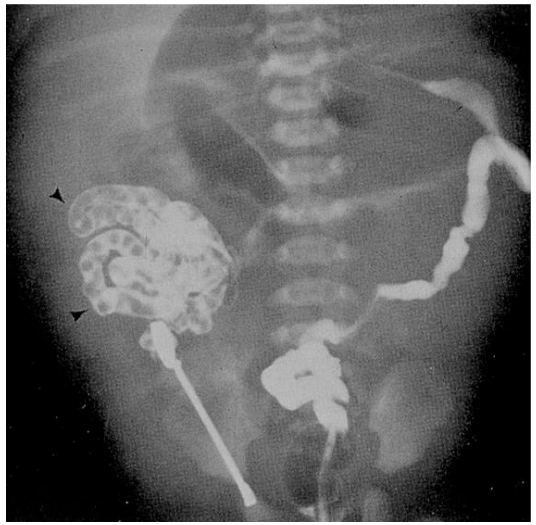
Distal Intestinal Obstruction

Delayed stool passage of meconium, distension

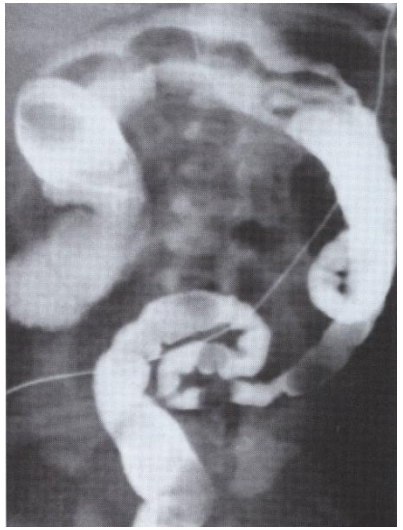
- Distal ileal atresia
- Colonic obstruction
- **Diagnosis: Contrast enema**



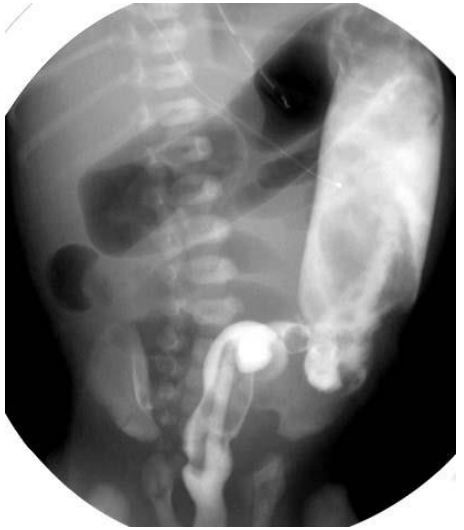
Distal Ileal Atresia



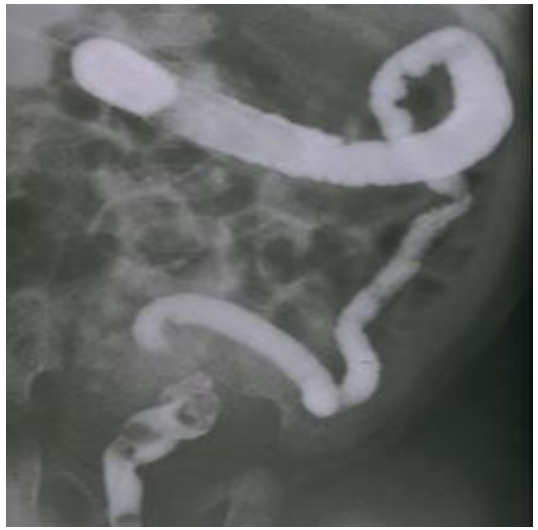
Meconium Ileus



Meconium plug



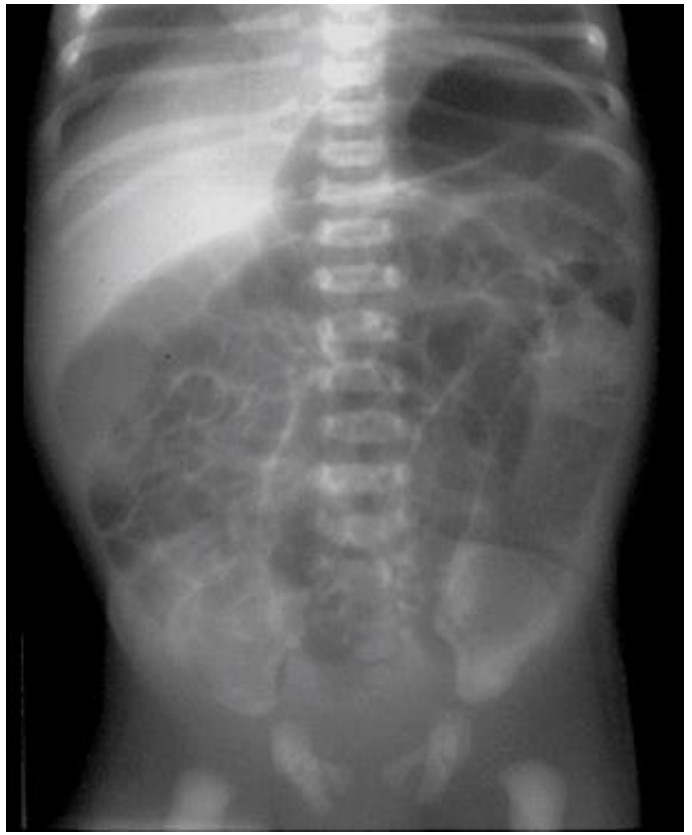
Hirschprung's



Small Left Colon

Imperforate Anus

Incidence	1/5000 births
Associations	GU, GI, Cardiac, Skeletal, CNS anomalies, VACTERL association
Types	• **High** • **Intermediate** • **Low** (usually associated with a fistula)
Diagnosis	PE, prone lateral plain abdominal film (no air in rectum)
Management	Anal dilatation, surgery (anoplasty)



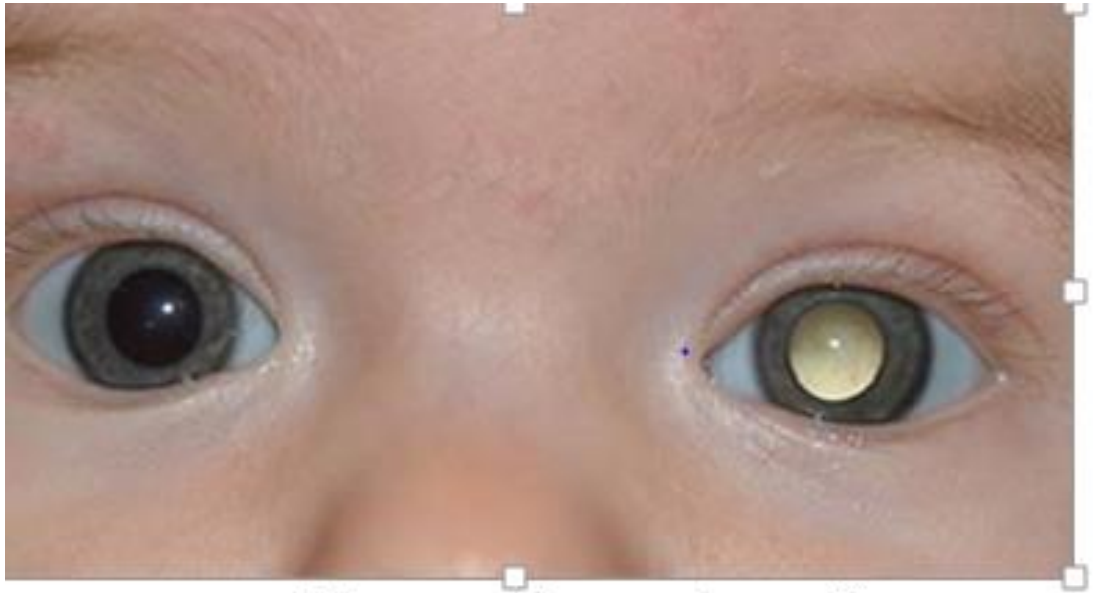
Dilated GI tract, No gas in rectum

Ophthalmology

- At birth a newborn's eyesight is between 20/200 and 20/400
- Can detect light, motion, face and large objects
- Normal infant can fixate at 6 wks
- Ability to track normally develops at 2 months
- + Red reflex should be documented at initial exam
- Dysconjugate eye movements can be present at birth

Leukoria (White Pupillary Reflex)

Causes
• Cataracts
• Coloboma
• Chorioretinitis
• Retinoblastoma
• Advanced ROP
• Retinal detachtment



Absent red reflex

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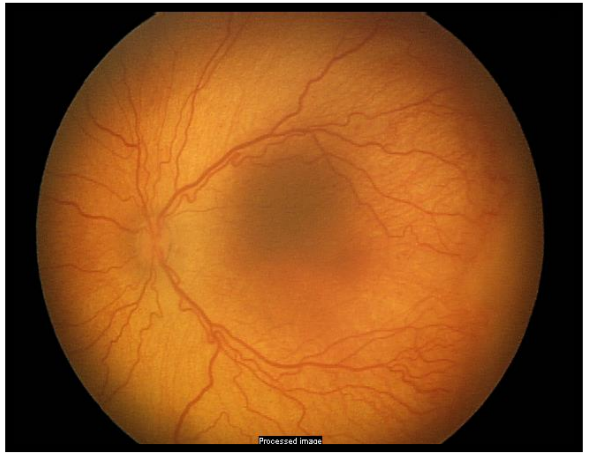
Retinopathy of Prematurity (ROP)

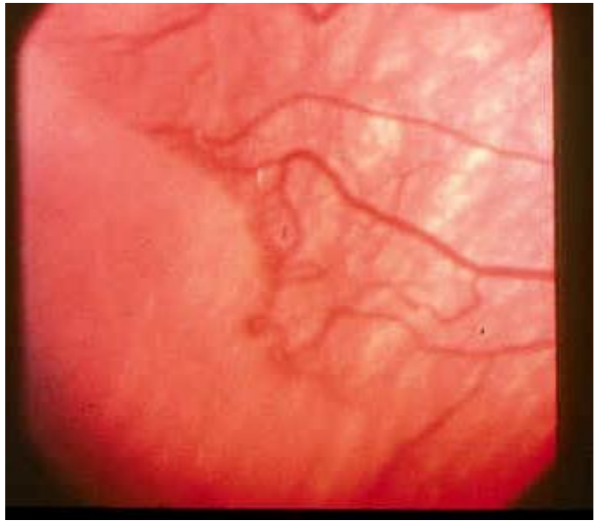
- Caused by deregulation of VEGF
- Vascular/avascular retina
- Routine screening: PT <1500 gms or <30wks

Stages	Findings
Stage 1	• Demarcation line
Stage 2	• Ridge
Stage 3	• Thicker ridge
Stage 4	• Partial detachment
Stage 5	• Retinal detachment

- Treatment: Laser therapy, Avastin (intravitreal)



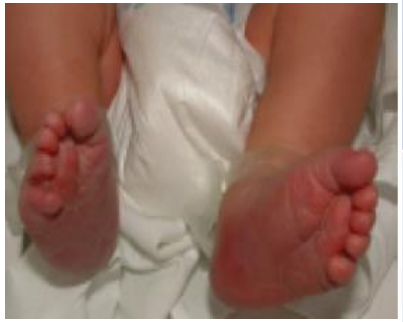
NORMAL



ROP

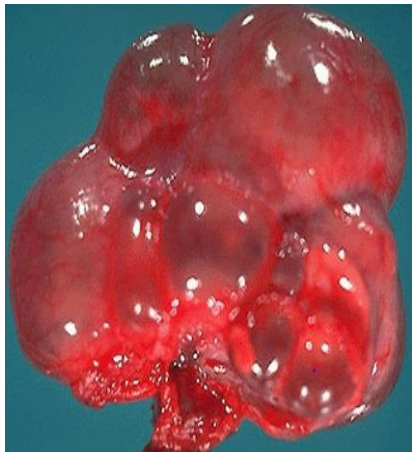
Defect of Morphogenesis

Disruption

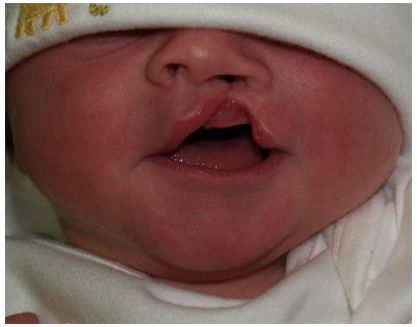


Deformation

Type	Cause	Example
Disruption	Destruction of a tissue that initially developed normally	Amniotic bands, ring-like constriction of limbs, amputation of digits
Dysplasia	Abnormal cellular organization or function in a specific tissue or organ	Multicystic renal dysplasia, hemangiomas
Deformation	Extrinsic intrauterine constraint or deformity due to neuromuscular or skeletal abnormality	Positional talipes equinovarus, torticollis
Malformation	Incomplete or abnormal progression of one or more developmental processes in early gestation	Cleft lip, palate, myelocele, CHD



Dysplasia



Malformation

CHARGE

C oloboma of the eye
H eart defects
C hoanal Atresia
R etarded growth and development
G enital and/or urinary abnormalities
E ar abnormalities

Coloboma

VATER

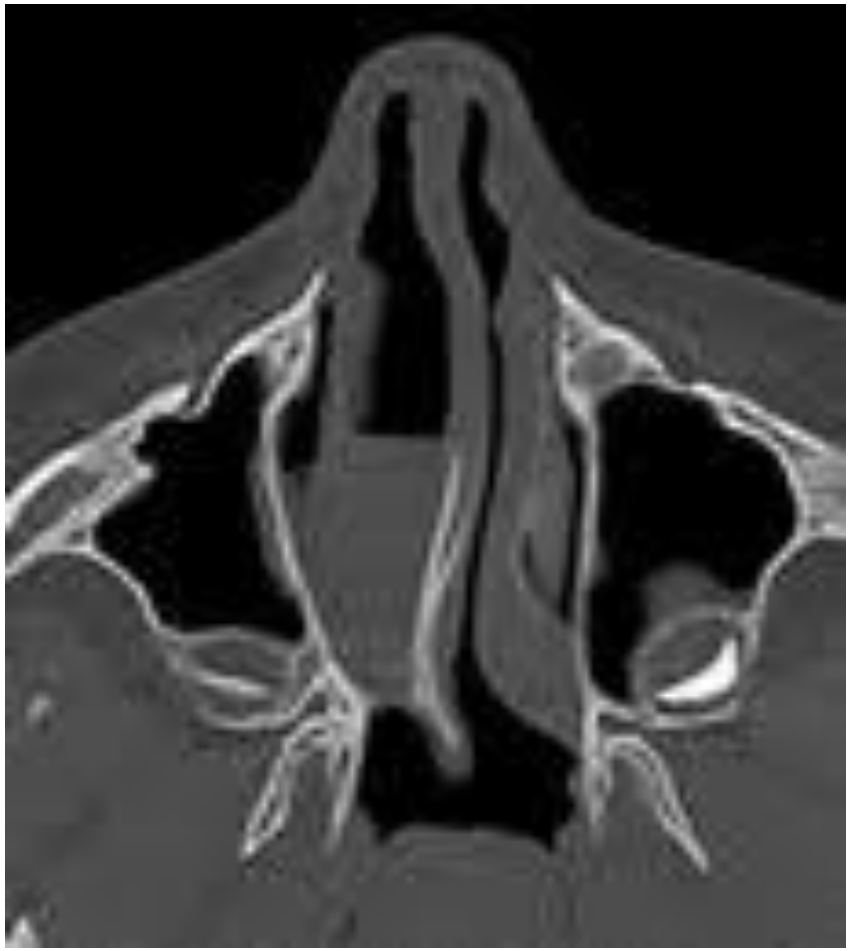
V ertebral anomalies
A norectal stenosis or atresia
T racheo **E** sophageal fistula
R enal anomalies

VACTERL

V ertebral anomalies
A norectal stenosis or atresia
C ardiac defects
T racheo **E** sophageal fistula
R enal anomalies
L imb abnormalities

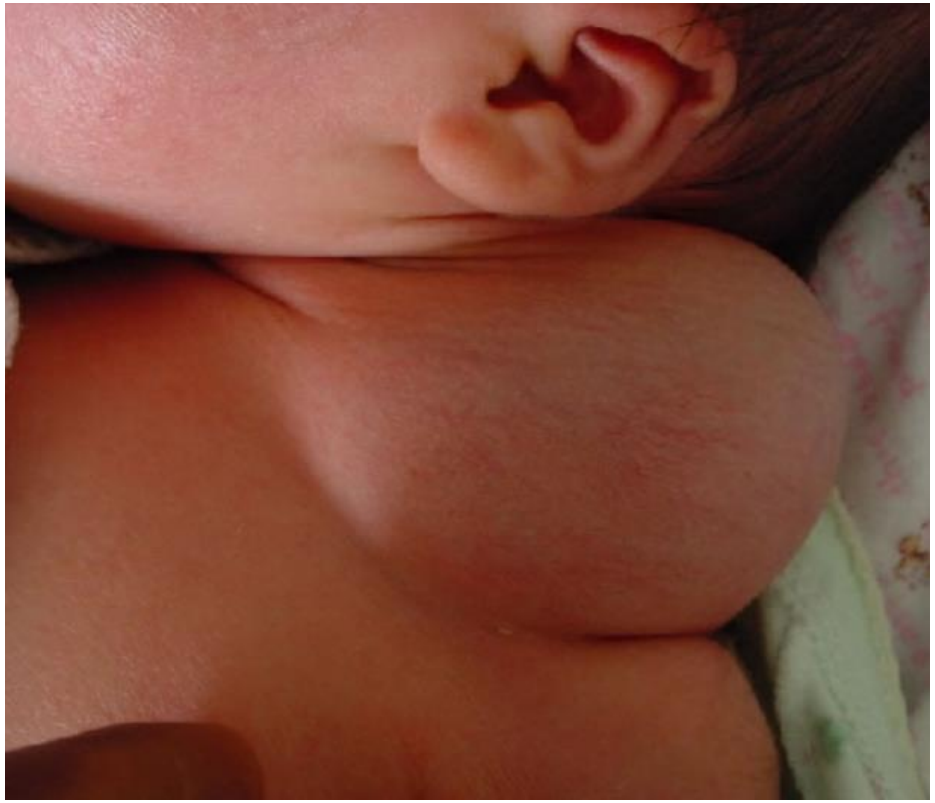
Choanal Atresia

Etiology	Blockage of back of the nasal passage (choana), usually by abnormal bony or soft tissue formed during fetal development
Incidence	Rare,1 in 7,000 births 2/3 Unilateral F>>M
Associations	CHARGE
Clinical findings	**Cyanosis** worse with feeds or pacifier which **improves with crying or opening mouth** (newborn obligatory nose breathers)
Treatment	Initial- oral airway Surgical correction, stents



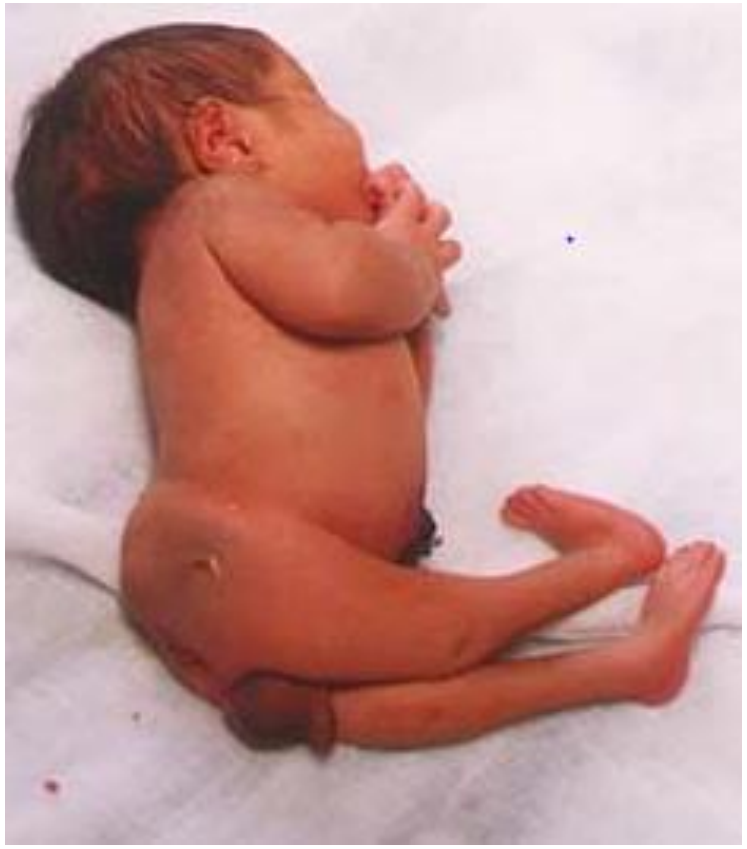
Cystic Hygroma

Etiology	Cystic lymphatic lesion due to the abnormal development of the lymphatic system leading to obstruction of lymphatic flow and sequestration of lymphatic fluid
Location	Majority located **neck** (75%), axilla (10%)
Clinical Findings	Mass is soft, compressible, painless and thick walled. Consists of numerous cyst of varying sizes, minority unilocular
Associations	T 13, T18, 21, XO,XXY



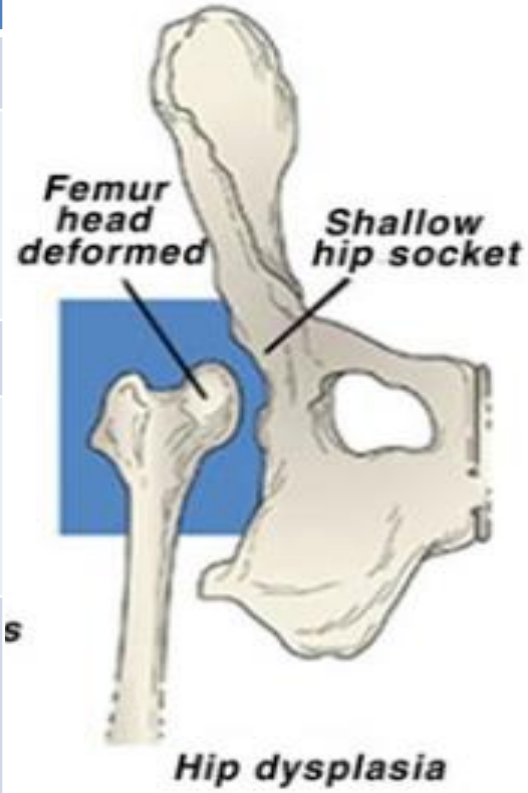
Caudal Dysplasia Syndrome

Associations	Seen in **IDM**
Clinical findings	• **Sacral or lumbar hypoplasia** • Disruption of distal spinal cord • Renal anomalies: • Double uterer • Horseshoe kidney • Neurogenic bladder • VUR • Hydronephrosis • Renal agenesis
Treatment	Multidisciplinary team approach



Developmental Dysplasia of Hip (DDH)

Incidence	1.5% of newborns, incidence of severity varies
Risk factors	• **Breech presentation** • **Female Gender** (Female/breech up to 12 % vs Males/breech 2.6 %) • Positive family history
PE findings	Barlow and Ortolani maneuvers, Asymmetrical Leg Creases
Diagnosis	**Most DDH normalize spontaneously** (60% by 1wk, 90% by 8wks) < 6month- U/S of hip, >6 months- X ray frog-leg position
Treatment	Observation and weekly re-examinations < 6months-Pavlik harness-if instability is present ➢6 months-Surgery (open or closed reduction)



Renal Disease

- UO by 24 hrs if not patient warrants evaluation
- Anuric infant:

  -Prerenal (renal, CHF)

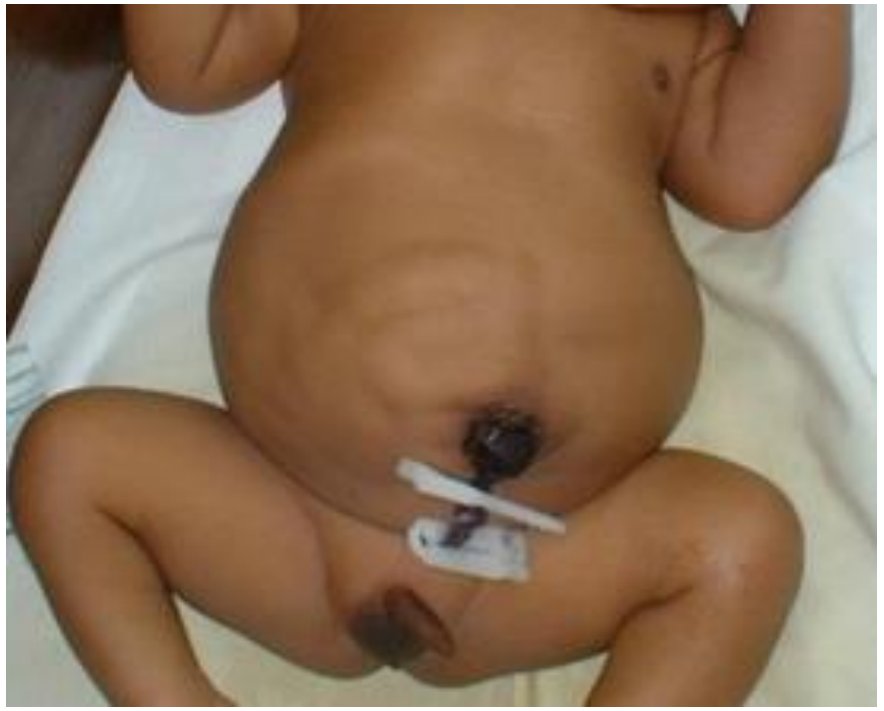
  -Renal (ATN, PCK)

  -Post renal (PUV, Prune Belly Syndrome)

	Acute Renal Failure
Etiology	Pre-renal, intrinsic, renal disease: congenital diseases (renal dysplasia, PCKD), hypoxic ischemic injury, toxic insults, vascular insults and obstructive uropathy
Presentation	HTN, proteinuria, elevated BUN/Creat
Treatment	Appropriate fluid balance, electrolyte status, acid-base status, nutrition and renal replacement therapy when appropriate

Prune Belly Syndrome

Etiology	Form of **obstructive uropathy**
Clinical findings	• Bilateral hydroureteral nephrosis • Undescended testes • Diminished or absent anterior abdominal wall musculature resulting wrinkled abdominal wall (Prune belly appearance)
Diagnosis	PE, **renal/bladder U/S**



Good luck
and
Thank you!!

Nicklaus Children's Hospital