**Neonatal Pulmonary Conditions**

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**Learning Objectives:**
- Explain the etiologies, pathophysiology and management of common neonatal conditions.
- Explain the pathogenesis, prevention and management of iatrogenic neonatal conditions.

**Neonatal asphyxia**

- **AKA-** asphyxia neonatorium
- **Definition**- failure of an infant to cry or breathe well after delivery.
- **Neonatal asphyxia may be a continuation of fetal asphyxia**

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**Neonatal asphyxia**
- Management - resuscitation
  - Dry & warm the patient
  - Oxygen - room air not effective*
  - Ventilation
  - NaHCO₃
  - Glucose


**Respiratory distress syndrome**
- Disease of pulmonary immaturity
  - Severity associated with low birth weight
  - Immature lung deficient in surfactant
  - Immature lung has insufficient alveoli for gas exchange
  - Lungs usually mature at 35 weeks

**Respiratory distress syndrome**
- Prediction - lab fetal lung profile
  - Principal constituent of surfactant - lecithin
  - Low lecithin levels ==> early gestation
  - After 35 weeks L:S > 2 ==> OK surfactant
  - Infants of diabetic mothers (IDM) - require phosphatidyl glycerol (PG), a more sensitive test

FYI - Link to more information about fetal lung profile
http://www.mayomedicallaboratories.com/test-catalog/Clinical-and-Interpretive/8929

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**Surfactant activity**
- Atelectasis
  - Compliance
  - WO
  - RR
  - PaCO₂
  - PaO₂
  - VQ defects
  - Metabolic acidemia
  - Combined acidemia
  - Pulmonary vasoconstriction
  - PFO & R = L anatomic shunt

- Surfactant deficiency
  - Alveolar necrosis

Capillary damage
RDS Manifestations
- Hypoxemia, hypercapnia, acidemia
- Tachypnea
- High Silverman respiratory distress score:
  - asynchrony of upper & lower chest
  - retractions- xiphoid, lower chest
  - nasal flaring
  - grunting
- Fine crackles

Click to see Downes and Silverman scoring systems
http://members.tripod.com/puffnicu/rd.html

RDS Chest Radiograph
- massive atelectasis
- "ground glass"
- air bronchograms

Click to see chest Xray of RDS with air bronchograms

RDS treatment- break the cycle
- >28 wks- CPAP*
- <28 wks- intubate & ventilate
- Continuous mechanical ventilation
  - Gentle ventilation (permissive hypercapnia)
  - PCIRV- first used on neonates
  - High-frequency oscillation


RDS Treatment
- Exogenous surfactant
  - treatment
  - prophylaxis for at-risk infants
  - preparations:
    - curosurf (pork lung)
    - beractant (bovine lung)
    - calfactant (calf lung)
    - lucinactant (Surfaxin)- synthetic-instilled or aerosolized

RDS- Prevention
- Accurate prediction, index of suspicion
  - Stop labor- terbutaline, Ritudrine
  - Administer steroids to mom to speed fetal lung maturation.
  - Cocaine and tobacco use also speed lung maturation

Click for consensus on steroids for fetal lung maturation

Meconium aspiration syndrome
- Aspiration of fetal stool
- Predisposing factors
  - fetal asphyxia
  - post-maturity
  - oligohydramnios- deficient amniotic fluid
Meconium aspiration syndrome

Onset
- May occur prior to labor
- Asphyxia ==> relaxation of anal sphincter
- Increased peristalsis
- Fetal gasping
- Asphyxia continues in extrauterine state

Complications
- RDS
- Extraneous air; e.g., pneumothorax
- Hypoxemia ==> encephalopathy
- Persistent pulmonary hypertension
- Pneumonia

Types of obstruction
- Check valve- air entry-- no exit ==> distal hyperinflation (emphysema)
- Stop valve- no air movement ==> distal atelectasis

Meconium aspiration syndrome

Complications
- RDS
- Extraneous air; e.g., pneumothorax
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- Persistent pulmonary hypertension
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Stop valve obstruction

Check valve obstruction

Meconium ==> inflammation ==> pneumonitis ==> pneumonia

Hypoxia ==> pulmonary vasoconstriction ==> PPHN
**MAS- Manifestations**
- Meconium-stained amniotic fluid and neonate
- Hypoxemia
- Tachypnea, gasping
- Hyperexpansion ==> bulging intercostal spaces
- Rhonchi, crackles

**MAS- Manifestations**
- Chest radiograph
  - mixed densities, hyperexpansion
  - commonly associated with pneumothorax

Click to see article with chest radiographs of MAS
http://www.emedicine.com/radio/TOPIC426.HTM

**MAS- treatment**
- All newborn mouths suctioned with bulb syringe before thorax is delivered
- Thin meconium- may require no further treatment

**MAS- treatment**
- Thick meconium
  - Avoid PPV, because it worsens condition
  - Laryngoscopy- meconium below cords
  - Intubate
  - Deep, tracheal suctioning with meconium aspirator

**MAS- Treatment**
- Surfactant lavage
  - Survanta
  - Surfaxin
- Mechanical ventilation
  - Lung protective strategies
  - HFOV
- Extracorporeal membrane oxygenation (ECMO)

**MAS- management**
- NICU care
  - CPT- NOT
  - Antibiotics for pneumonia
  - Suctioning
  - Treatment for PPHN
Persistent pulmonary hypertension

- **Etiologies**
  - hypoxemia
  - congenital - abnormal vasoactivity

- **Associated conditions**
  - RDS
  - MAS
  - Asphyxia
  - Diaphragmatic hernia

Pre and post-ductal blood flow

- **Left-right shunt- absence of PPHN**
  - Normal pre-ductal PaO2 (right arm, head)
  - Left arm PaO2?
  - Increased mixed venous PO2

- **Right-left shunt with PPHN**
  - Decreased post-ductal PaO2

Persistent pulmonary hypertension

- **Manifestation- severe, refractory hypoxemia (right-left shunt)**

- **Diagnosis**
  - Echocardiography shows shunt
  - Hyperoxia test- 100% O2 shows shunt
  - Pre & post-ductal test >20 mm Hg difference
  - Hyperoxia-hyperventilation
  - PaCO2 < 25 ==> 100 mm Hg increase in PaO2

Persistent pulmonary hypertension

- **Treatment**
  - Hyperventilation- conventional
  - High frequency ventilation
  - Medications
    - tolazoline
    - nitric oxide
    - prostacyclin (Flolan)
  - ECMO- non-responders
**Transient tachypnea of the newborn (TTN)**
- **Etiology & onset- term newborns**
  - Failure to expel fetal lung fluid ==> cesarean-section delivery
  - Signs appear at birth, or shortly afterward

**TTN- Pathophysiology**
- Retained fetal lung fluid ==> 
  - Obstruction (mild)
  - Mild hypoxemia, hypercapnia, acidemia

**TTN- Manifestations**
- Tachypnea
- Grunting
- Cyanosis
- Differentiate from:
  - Pneumonia
  - CHF (cardiomegaly)

**TTN- CXR**
- Generalized overexpansion
- Hilar streaking

Click for radiograph of TTN
http://radiopaedia.org/images/218523

**TTN- Management**
- Oxygen
- Supportive care
- Usually clears within days
- Albuterol aerosol - reduces edema*

*Reference

**Apnea**
**Apnea and periodic breathing**

- Periodic breathing
  - apnea < 15 sec.
  - no bradycardia, cyanosis
  - reverse spontaneously

- True apnea (primary)
  - Associated with prematurity & BW < 1800g
  - duration >15 sec.
  - Associated with bradycardia, cyanosis
  - Reversible with stimulation of patient

- Secondary apnea
  - Sequel to untreated primary apnea
  - Irreversible with stimulation
  - Immature respiratory centers
  - Blunted response to CO2
  - Hypoxemia depresses ventilation

- Treatment
  - mild stimulation
  - oxygen (23-25%)
  - xanthines- theophylline, caffeine
  - CPAP
  - ventilation for secondary apnea

**Etiologic factors in apnea**

- Prematurity- immature chemoreceptors
- Anatomic obstruction
  - cranial malformations
  - airway anomalies
- CNS lesions
- Hyper/hypo-thermia
- Abdominal distension; e.g., necrotizing enterocolitis (NEC)
Iatrogenic Conditions

Bronchopulmonary dysplasia (BPD)
- Definition: infant requires supplemental O2 at one week still O2 dependent at 28 D has BPD
- progressive chronic lung disease in infants following periods of mechanical ventilation

BPD- Etiologic Factors
- Pulmonary immaturity-- lung vulnerable to insult
- FIO2 > .21 x time
- Endotracheal tube
- Positive pressure ventilation
- PDA-increases pulmonary blood flow
  ==> reduced CL ==> need for increased PIP

BPD- Etiologic Factors
- BPD can be predicted in very low birthweight by a small thymus gland on the chest radiograph*
  - identifies impaired immune function
  - BPD may not be preventable

FYI - Link to function of thymus gland
http://www.intech.mnsu.edu/angelamonson/DH319/Short%20Papers/thymus_gland_1.htm


BPD- clinical course
- Stage I- precipitating dx
  - refractory hypoxemia
  - increased level of ventilatory support
  - pulmonary inflammation
- Stage II
  - diffuse atelectasis- opaque CXR
  - pulmonary edema- common
  - PDA- common

BPD- clinical course
- Stage III
  - some improvement
  - pulmonary cystic changes
  - continued O2 dependence
  - hypercapnia- common
  - airway hyperreactivity
**BPD- clinical course**

- Stage IV-- chronic
  - cardiomegaly
  - hyperexpansion, interspersed with fibrosis
  - bronchiolar smooth muscle hypertrophy
  - tracheotomy often required

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**BPD- manifestations**

- history of ventilation with O2
- chronic hypoxemia
- chronic hypercapnia
- wheezing
- hyperinflation
- reversible (usually) airway obstruction
- cardiomegaly

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**BPD- manifestations**

- chest radiograph

Click to see chest radiograph of BPD

Click to see chest radiograph of 15 YO with BPD
http://www.hawaii.edu/medicine/pediatrics/pemxray/v3c02.html

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**BPD- management**

- Minimal, gentle ventilation
- Cautious O2 therapy
  - too much- apnea
  - too little- cyanosis, apnea

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**BPD- management**

- Bronchodilators-- if reversible
  - clinical signs
  - pre-post pulmonary mechanics: $R_{AW}$, $C_{DYN}$, flow-volume curve
  - MDI- albuterol and/or Atrovent
- Inhaled steroids- NOT
- Prevent infection

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**BPD- management**

- Furosemide (Lasix)
- Digoxin
- Theophylline
- Caffeine
- Nutrition- development of new alveoli
- Home care- for stage IV patients
BPD Prevention

- Gentle ventilation
- Avoid fluid overload, CHF
- Antioxidants??
- Exogenous surfactant*


Extraneous Air Syndromes

- air, where air should not be.

Pathogenesis

- PPV ruptures alveoli
- air enters perivascular sheath
- alveolar perivascularature (PIE)
- reduced CL
- need for increased PIP
- extraneous air

Extraneous Air Syndromes

- Pathogenesis

PIE Manifestations

- Progressive hypoxemia
- Decreasing compliance
- increased PIP required for ventilation
- Diagnosed by CXR

Extraneous Air Syndromes

- Affected sites
  - Pleura-- blebs, pneumothoraces
  - Subcutaneous tissue
  - Mediastinum-- pneumomediastinum
  - Intravascular, intracardiac spaces-- air embolus
  - Pericardium
  - Cerebral vessels- cerebral air embolism

Extraneous Air Syndromes

- Pathogenesis

PIE- Manifestations

- Progressive hypoxemia
- Decreasing compliance
- increased PIP required for ventilation
- Diagnosed by CXR
PIE - Manifestations
➤ CXR

Click to see article with radiographs of PIE
http://www.emedicine.com/ped/topic2596.htm#Multimedia1

PIE - Management
➤ Minimize PIP
➤ HFOV
➤ Intubate unaffected side
➤ Affected side positioned down
➤ Intermittent 100% O₂

Retinopathy of Prematurity (ROP)
➤ Retinal disease - vision impairment
➤ Etiology
✓ ROP is due to immaturity of retina
✓ Increased incidence of ROP is due to increased survival of infants
BW < 1 Kg

Retinopathy of Prematurity (ROP)
➤ Prevention - maintain PaO₂ < 100
➤ Patients who were never on O₂ have developed ROP
➤ Management
✓ Laser surgery
✓ Intravitreal bevacizumab (Avastin)
- Prevents vascular proliferation

Retinopathy of Prematurity (ROP)
➤ Pathology with hyperoxia
PaO₂ > 100 ==> retinal vasoconstriction ==> necrosis of vessels ==> proliferation of new vessels ==> extend into vitreous humor ==> hemorrhage ==> scar ==> retinal detachment ==> blindness

Retinopathy of Prematurity (ROP)
➤ Etiology
✓ ROP may not be preventable
✓ 20-40% neonates with birth weight < 1 kg have ROP
✓ Aggravated by hyperoxia

FYI - Click for article on Bevacizumab (Avastin) and ROP
http://www.presstv.ir/detail/166016.html
Airway, Esophageal & Abdominal Anomalies

Choanal atresia
- obstructed nasopharynx - may be soft tissue or bone
- Infant is obligate nose breather ==> severe distress
- Bag-mask ventilation produces temporary recovery

Choanal atresia
- Diagnosis - inability to pass catheter through nares to pharynx
- Treatment
  - temporary oropharyngeal airway
  - intubation
  - surgical correction

Tracheoesophageal (TE) fistulae
- Esophageal atresia without TE fistula

Tracheoesophageal (TE) fistulae
- Esophageal atresia with proximal fistula

Tracheoesophageal (TE) fistulae
- Esophageal atresia with distal fistula (most common)
Tracheoesophageal (TE) fistulae

- Esophageal atresia with proximal and distal fistulae

Tracheoesophageal (TE) fistulae

- Tracheoesophageal fistula without esophageal atresia (H-type)

TE Fistula- Manifestations

- Depends on type
- Accumulation of oral secretions
- Regurgitation of feedings
- Respiratory distress

TE Fistula

- Complications
  - Aspiration
  - Distension/rupture of stomach with positive-pressure breathing
  - Failure to feed

TE Fistula

- Management
  - Hold oral feedings
  - Surgical correction- atresias may recur

TE Fistula

- Associated anomalies
  - vertebral
  - anal
  - cardiac
  - tracheal
  - esophageal
  - renal
  - limb
# Miscellaneous Airway Anomalies
- Vocal cord paralysis
- Laryngeal web
- Vascular ring (around trachea)
- Pierre Robin syndrome
  - micrognathia
  - glossoptosis
- Treacher-Collins syndrome

# Congenital Diaphragmatic Hernia (CDH)
- **Etiology & onset**
  - Etiology-- incomplete embryonic development of diaphragm.
  - Distress at birth or shortly thereafter
  - Some are not diagnosed until childhood

## Congenital Diaphragmatic Hernia
- **Pathophysiology & course**
  - Abdominal contents compress lung
    - Left-sided (most common)-- intestines
    - Right-sided-- liver

  - Suspected in presence of polyhydramnios
  - Detected in utero by ultrasound
  - Severity depends on how much lung is compressed
  - Lung on affected side may be hypoplastic

  - Respiratory distress- worsens with bag-mask ventilation due to inflation of bowel
  - Absent breath sounds or presence of bowel sounds on affected side
  - Shift in point of maximal impulse
  - Need to differentiate from pneumothorax

[Click for more information and illustrations of CDH](http://www.nlm.nih.gov/medlineplus/ency/article/001135.htm)
**Congenital diaphragmatic hernia**

- Management
  - Avoid bag-mask ventilation
  - Immediate intubation, ventilation
  - Conventional ventilation
  - HFOV
  - ECMO- severe cases
  - Surgical correction

Click for radiographs of diaphragmatic hernia
http://www.mypacs.net/cases/DIAPHRAGMATIC-HERNIA-6159020.html
http://www.hawaii.edu/medicine/pediatrics/neoxray/neoxray.html

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**Omphalocele**

- protrusion of viscera, covered by peritoneum through umbilical area
- Associated with other anomalies
  - prune belly
  - congenital heart disease (TOF)

FYI - Link to index of pictures of congenital anomalies
http://library.med.utah.edu/WebPath/PEDHTML/PEDIDX.html#1

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**Gastroschisis**

- uncovered bowel protruding through abdominal wall
- Not associated with other anomalies
- Causes extrathoracic restriction
- Bowel slowly pushed into abdomen

Click to see gastroschisis
http://library.med.utah.edu/WebPath/PEDHTML/PED007.html

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**Congenital Emphysema**

- Etiology uncertain-- weak bronchial support, with dynamic compression
  ==> air trapping
- Pathophysiology-- hyperinflation of lobe
  ==> compress lung segments
  ==> V/Q defects

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**Congenital diaphragmatic hernia**

- Associated with elevated maternal serum alpha fetoprotein
- Visualized on ultrasound
- Causes extrathoracic restriction
- Repaired by pushing viscera into abdominal cavity

Click to see omphalocele
http://library.med.utah.edu/WebPath/jpeg3/PERI110.jpg
Manifestations

- Signs and symptoms may appear later
- Tachypnea - distress
- Cyanosis
- Wheezing - mistaken for asthma
- Intercostal bulging on affected side

Click to see radiograph of congenital emphysema
http://radiographics.rsna.org/cgi/content-nw/full/24/1/e17/F16B

Congenital Emphysema

- Management
  - Ventilatory support
  - Surgical resection

Summary & Review

- Neonatal asphyxia
  - hypoxemia, acidemia, hypercapnia
  - complications
- Respiratory distress syndrome
  - surfactant deficit & immature lung units
  - pathophysiologic cycle
  - treatment - surfactant

Summary & Review

- Meconium aspiration
  - initially, no PPV
  - obstruction types - non-uniform dx
- Persistent pulmonary hypertension
  - cause - hypoxemia
  - pre- post-ductal O2 differences
  - treatment - NO, Fiolan

Summary & Review

- Transient tachypnea of newborns
  - cesarean section, term babies
  - mild, self-limited
- Apnea in neonates
  - periodic breathing
  - apnea - primary and secondary

Summary & Review

- bronchopulmonary dysplasia
  - immature lung, vulnerable to injury
  - manifestations like COPD
  - resolution - growth of new lung units
- extraneous air
  - due to PPV
  - pulmonary interstitial emphysema
- retinopathy of prematurity
  - not preventable
  - maintain PaO2 less than 100 mm
Summary & Review

Airway anomalies
- choanal atresia - blocked nasal passage
- tracheoesophageal fistula-atresia
- Congenital diaphragmatic hernia
  - distress with bag-mask ventilation
  - hypoplastic lung - severity
  - may require ECMO

Omphalocele, gastroschisis
- bowel outside abdomen
- thoracic restriction with management
- Congenital lobar emphysema
  - resembles asthma, pneumothorax
  - ventilation until surgical correction

END