

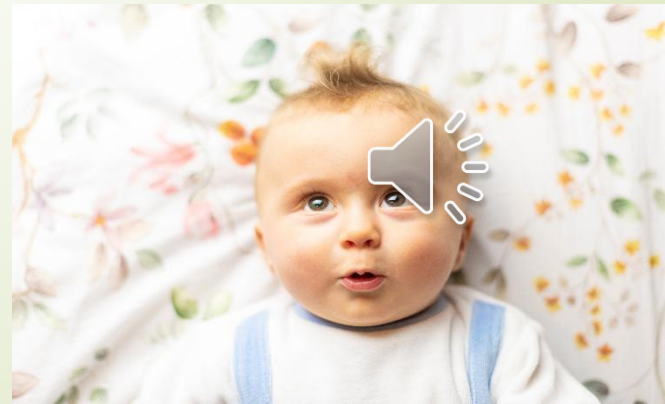


Neonatal and Pediatric Disorders Bootcamp

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Learning Objectives

- Discuss the clinical findings, radiographic abnormalities, and treatment of patients with:
 - Infant/Neonatal respiratory distress syndrome
 - Transient tachypnea of the newborn
 - Meconium aspiration syndrome
 - Bronchopulmonary dysplasia
 - Apnea of prematurity
 - Persistent pulmonary hypertension of the newborn
 - Congenital diaphragmatic hernia.



Learning Objectives (Cont.)

- Identify the anatomic defects, clinical presentation and treatment associated with congenital cardiac defects including Tetralogy of Fallot and ventricular septal defect.
- Describe types and associated conditions for abdominal wall defects.
- Define the epidemiologic factors associated with increased risk for sudden infant death syndrome.
- Identify the respiratory problems associated with gastroesophageal reflux disease.
- State the clinical findings commonly observed in patients with bronchiolitis.
- Describe the clinical features and treatment of children with epiglottitis, croup and cystic fibrosis.





Lung Parenchymal Diseases

- Respiratory Distress Syndrome
- Transient Tachypnea of the Newborn
- Meconium Aspiration Syndrome
- Bronchopulmonary Dysplasia



Infant/Neonatal Respiratory Distress Syndrome

- Neonatal respiratory distress syndrome (RDS) affects approximately 40,000 infants each year in the United State
 - Occurs in greater than 80% in neonates less than 28 weeks
- Major factors in the pathophysiology:
 - Surfactant deficiency
 - Underdeveloped alveolar units and capillaries
 - Reduced lung compliance and alveolar surface area
 - Poor gas exchange
 - Presence of a ductus arteriosus



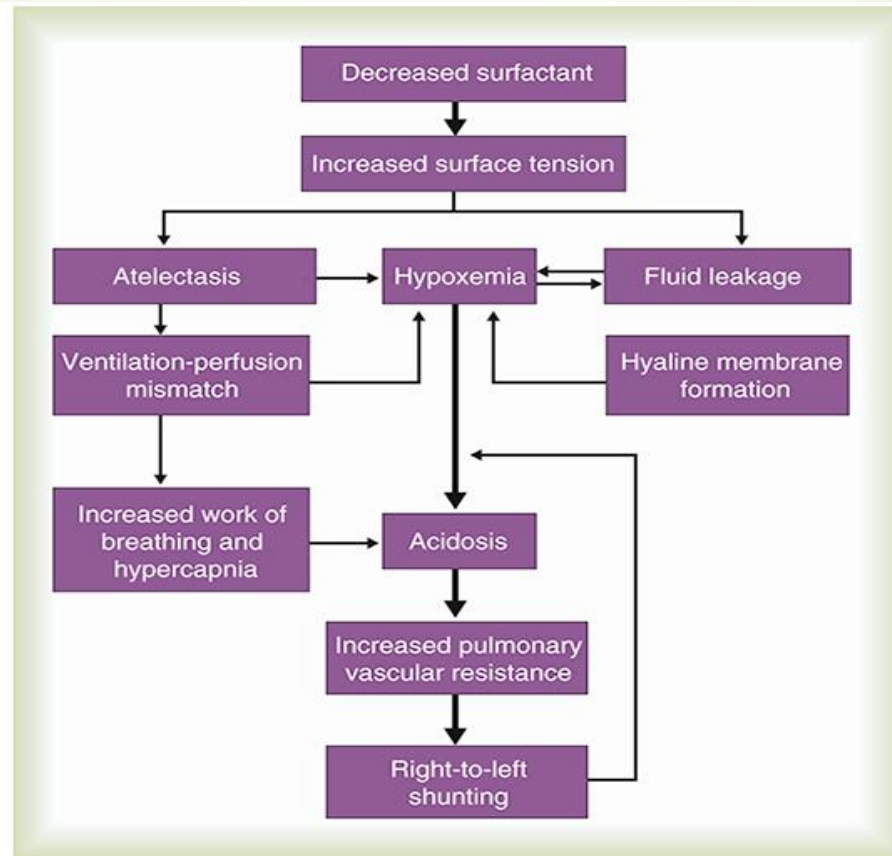


Respiratory Distress Syndrome (Cont.)

- Lung prematurity, and particularly surfactant deficiency, sets up following sequelae:
 - Alveolar instability and collapse, leads to increased WOB
 - Simultaneously increased surface tension draws fluid into alveoli
 - Combined, these cause impaired oxygenation with hypoxemia and acidosis, which lead to increased PVR
 - Increases right-to-left shunting through PDA
 - Worsens acidosis, hypoxemia, and surfactant production, which establishes downward spiral



Neonatal Respiratory Distress Syndrome



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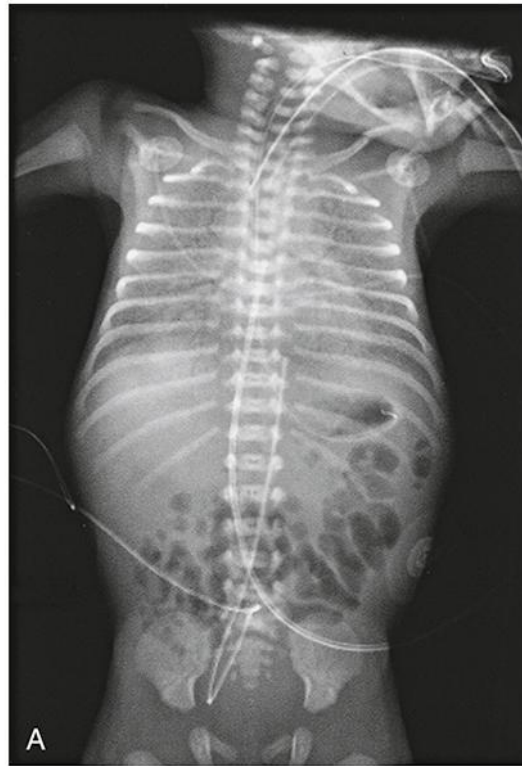
Respiratory Distress Syndrome (Cont.)

- Clinical manifestations
 - Tachypnea occurs soon after birth
 - Followed by worsening retractions, paradoxical breathing, and grunting
 - Nasal flaring may be seen
 - Chest auscultation: fine inspiratory crackles
 - Diagnosis is made from chest radiography
 - Diffuse, hazy reticulogranular densities, air bronchograms, and low lung volumes are typical



CXR Neonatal Respiratory Distress Syndrome

Diffuse hazy appearance with low lung volumes and air bronchograms that extend into the periphery.



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Respiratory Distress Syndrome (Cont.)

➤ Treatment

- CPAP, surfactant replacement therapy, and mechanical ventilation with conventional or high frequency ventilation (HFV)
- HFNC, NIMV, a trial of nasal CPAP is indicated unless the infant's condition is severe (high work of breathing, FiO_2 greater than 0.4, and/or severe apnea)
- Nasal CPAP is indicated (4 to 6 cm H_2O)
- Mechanical ventilation with PEEP should be initiated if gas exchange does not improve with CPAP or if the patient's frequent apnea that does not respond to stimulation



Respiratory Distress Syndrome (Cont.)

➤ Treatment

➤ INSURE

➤ Intubate, SURfactant, and Extubation to CPAP

- HFNC has been shown to be as clinically effective as nasal CPAP but with fewer nasal injuries following extubation from mechanical ventilation in premature infants
- Surfactant replacement therapy also is used as both a rescue treatment (in infants who already have RDS) and a prophylactic therapy



Transient Tachypnea of the Newborn (TTN)

- Often called Type II RDS
- Term babies with no specific predisposing factors
- Probably occurs due to delayed clearance of fetal lung fluid
 - Thoracic squeeze expels about two thirds of fluid in birth canal
 - Cesarean section avoids this squeeze
 - Lymphatics' absorption accounts for final third



Transient Tachypnea of the Newborn (Cont.)

- Clinical manifestations
 - Tachypnea
 - pH, and PaCO₂ are usually normal
 - Features of chest radiograph:
 - May look like pneumonia
 - Hyperinflation
 - Pleural effusions
 - Perihilar streaking (lymphatic engorgement)



CXR-TTN – Overinflation, increased interstitial streaky markings and fluid in the intralobar fissure.



Transient Tachypnea of the Newborn (Cont.)

➤ Treatment

- Responds well to low FiO_2 with oxygen hood or nasal cannula
- May benefit from CPAP if higher FiO_2 needed
- May use antibiotics (TTN hard to tell from pneumonia)
- Frequent changes in infant's position may help speed lung fluid clearance
- Most treatments have rapid response (24 to 48 hours)



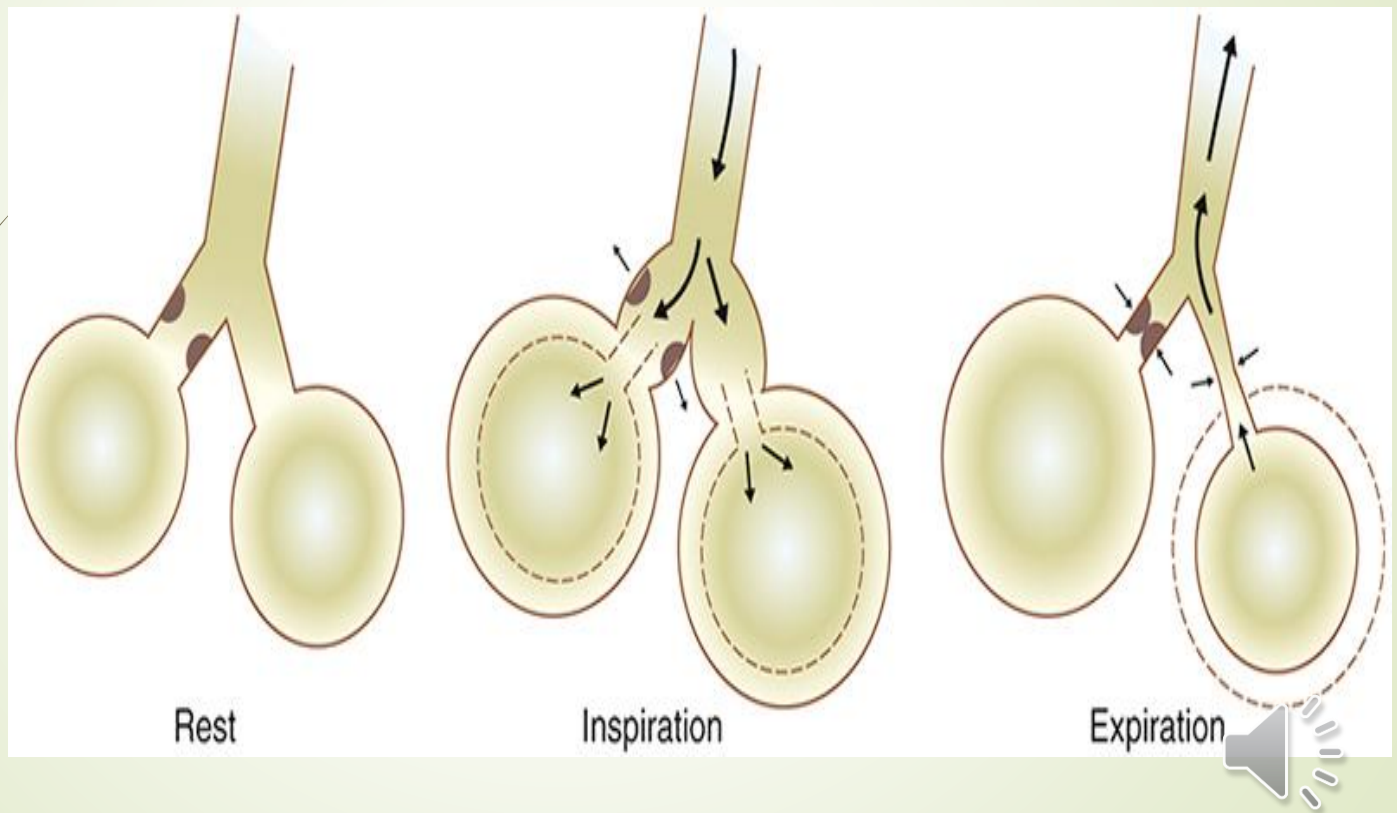
Meconium Aspiration Syndrome (MAS)

- Disorder of term or near-term infants, associated with perinatal depression and asphyxia
- Pathophysiology
 - Depression and asphyxia lead to passing of meconium
 - MAS involves three primary problems:
 - Pulmonary obstruction due to meconium plugging
 - Lung damage due to chemical injury
 - Persistent pulmonary hypertension (PPHN)
 - Chemical effects include inflammatory responses, cytokine and chemokine activations, complement activation, phospholipase A2 activation



MAS - Mechanical Disruption to Ventilation

Air goes in but exhalation is inhibited



Meconium Aspiration Syndrome (MAS) (Cont.)

- Clinical manifestations
 - Gasping respirations, tachypnea, grunting, retractions
 - Chest radiograph
 - Irregular pulmonary densities (atelectasis)
 - Hyperlucent areas (hyperinflation)
 - ABGs: hypoxemia, mixed acidosis due to PPHN



CXR-MAS - Diffuse patchy areas of atelectasis and emphysema



Meconium Aspiration Syndrome (MAS)

► Treatment

- It is no longer recommended that vigorous infants with meconium-stained fluid be intubated and suctioned
 - Traditionally, an ETT has been inserted immediately in severely respiratory-compromised (non-vigorous) infants with thick meconium, and suction applied directly to the ETT
- If attempted intubation is prolonged and unsuccessful, bag-mask ventilation should be considered, particularly if there is persistent bradycardia
- The use of positive pressure ventilation is preferred to minimize any delay in initiating ventilation
- If the infant's condition worsens as evidenced by nasal flaring, retractions, increased RR, and/or cyanosis, the use of NCPAP, HFNC or mechanical ventilation may be needed
- CPAP is indicated if the primary problem is hypoxemia
- INO if PPHN





Bronchopulmonary Dysplasia (BPD)

➤ Background

- A complex disease that is characterized by lung injury/inflammation from oxygen and/or positive pressure
- May cause altered lung development, pulmonary emphysema, and pulmonary hypertension
- Historically
 - Defined by the requirement for supplemental oxygen at 28 days in infants born at below 32 weeks gestation
- BPD is divided into 3 severity grades (mild, moderate, or severe) based on respiratory support needs at 36 weeks of postmenstrual age





Bronchopulmonary Dysplasia (BPD) (Cont.)

- ▶ Pathophysiology
 - ▶ Atelectrauma is result and cause of lung injury
 - ▶ Volutrauma injures airways and lung parenchyma
 - ▶ Above two synergistically increase lung injury, necessitate oxygen therapy (leads to oxygen toxicity)
 - ▶ Immaturity, genetics, malnutrition, oxygen toxicity, hypoxia, and MV all contribute to BPD development



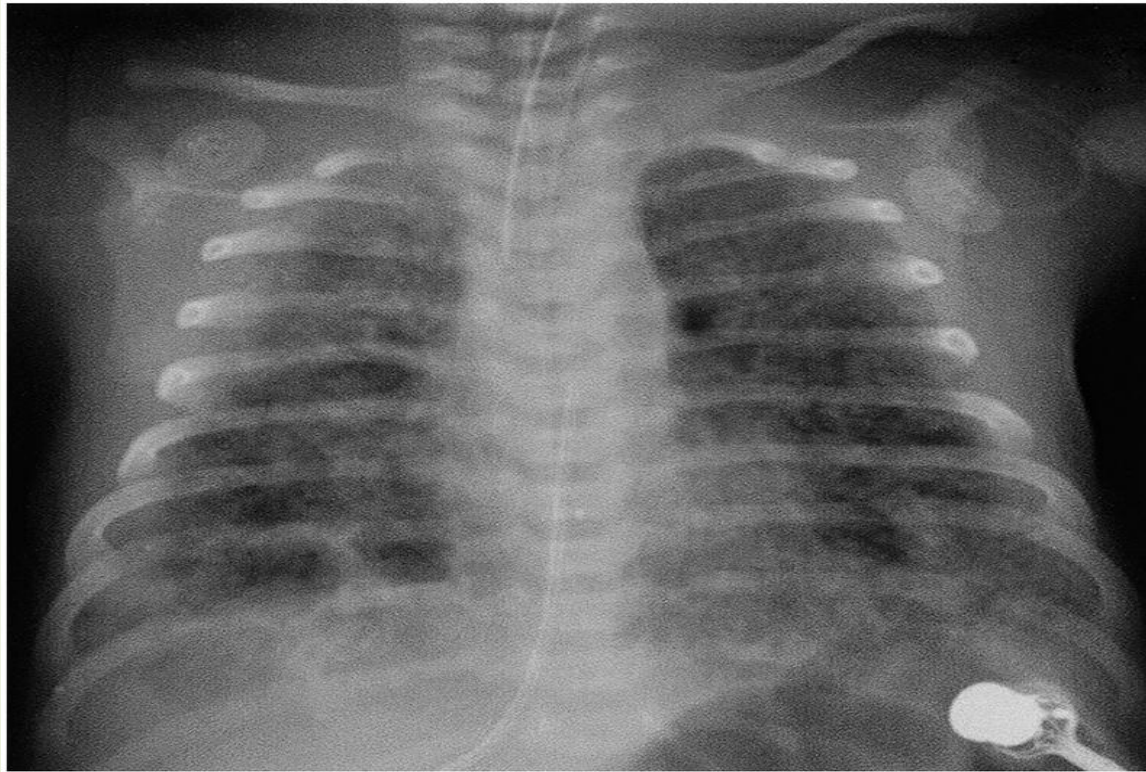


Bronchopulmonary Dysplasia (BPD) (Cont.)

- Clinical manifestations
 - Progressive respiratory distress at 2-3 weeks of life
 - If did not require O₂ and MV before, will now
 - Chest radiograph shows diffuse areas of atelectasis, emphysema, and fibrosis
 - May develop floppy upper airways (tracheobronchomalacia) due to prolonged ventilation of stiff lungs through relatively compliant airways
 - ABGs show varying hypoxemia and hypercapnia



CXR-BPD – Severe BPD as evidenced by scarring, atelectasis, emphysema and cysts



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Bronchopulmonary Dysplasia (BPD) (Cont.)

➤ Treatment

- Prevention of atelectrauma and volutrauma is KEY
- Establish optimal FRC in delivery room
- Early use of surfactant
- Minimize lung damage: monitor carefully and use lowest oxygen and MV settings possible
- Diuretics to treat pulmonary edema
- Antibiotics for pulmonary infections
- Chest physiotherapy for retained secretions
- Bronchodilator therapy to decrease R_{aw}
- Steroids used cautiously



Control of Breathing: Apnea of Prematurity

- Common in premature infants
- Apnea is abnormal if:
 - Lasts longer than 15 seconds
 - Causes cyanosis, pallor, hypotonia, or bradycardia
- Central apnea: no respiratory effort during apnea
- Obstructive apnea: effort but no flow
- Mixed apnea: combination of central and obstructive apnea



Control of Breathing: Apnea of Prematurity (Cont.)

- Cause
 - Premature infants have immature control of respiratory drive in response to O₂ and carbon dioxide
- Diagnosis –
 - Pneumocardigram-Infant Sleep Study
 - Home apnea monitoring
- Treatment
 - Continuous monitoring of HR and RR
 - Continuous noninvasive monitoring of oxygenation
 - Methylxanthines
 - “Back to Sleep”
 - CPAP





Sudden Infant Death Syndrome (SIDS)

- Leading cause of death in infants less than 1 year of age
- Prevention of SIDS
 - Identification of infant at risk
 - Train family in apnea monitoring and CPR
 - Place patient in supine or side-lying position
 - Reduce soft objects in sleeping environment—No more crib bumpers!!!!





Sudden Infant Death Syndrome (Cont.)

- Age less than 6 months (peak between 1 month and 3 months)
- Winter season
- Asleep at night
- Mild illness in week before death
- History of apparent life-threatening event
- Prone sleep position
- Mother smokes cigarettes



Pulmonary Vascular Disease

- Persistent pulmonary hypertension of the newborn (PPHN)
 - Prolonged postpartum fetal circulation due to increased PVR
 - Pathophysiology
 - Pulmonary blood flow is low due to right-to-left shunt through patent foramen ovale or ductus arteriosus
 - Three types of PPHN
 1. Vascular spasm: triggered by hypoxemia or pain
 2. Increased muscle wall thickness: chronic
 3. Decreased cross-sectional area of vasculature



Pulmonary Vascular Disease (Cont.)

- Persistent pulmonary hypertension of the newborn
 - Clinical manifestations
 - Suspect when rapidly changing SpO_2 , hypoxemia worse than indicated on chest radiograph
 - Detected by performing preductal and postductal SpO_2
 - Preductal SpO_2 greater than 5% postductal SpO_2



Pulmonary Vascular Disease (Cont.)

- Persistent pulmonary hypertension of the newborn
 - Treatment
 - Correct underlying problem
 - Hypoxemia with oxygen, surfactant for RDS
 - May require intubation, MV, and sedation
 - Inhaled nitric oxide (INO) therapy-
 - **PPHN is the only approved indication for INO!!!**
 - Often requires: paralysis, HFV, worst cases ECMO





Congenital Abnormalities Affecting Respiration

- Airway Diseases
- Lung Malformations
- Congenital Diaphragmatic Hernia
- Abdominal Wall Abnormalities
- Neuromuscular Control



Airway Diseases

- ▶ Three mechanisms

1. Internal obstruction

- ▶ Laryngomalacia, tracheomalacia, laryngeal webs, tracheal stenosis, and hemangiomas

- ▶ Stridor, gas-trapping, wheezing, accessory muscles

2. External obstruction (symptoms similar to internal)

- ▶ Neck or thoracic mass, vascular rings



Airway Diseases (Cont.)

3. Airway disruptions

➤ Tracheoesophageal fistula (TEF)

➤ Five types, all manifest as difficulty swallowing, frothing at mouth, and choking

- Esophageal atresia with proximal fistula

- Esophageal atresia with distal fistula

- Esophageal atresia with both proximal and distal fistula

- Esophageal atresia without either fistula

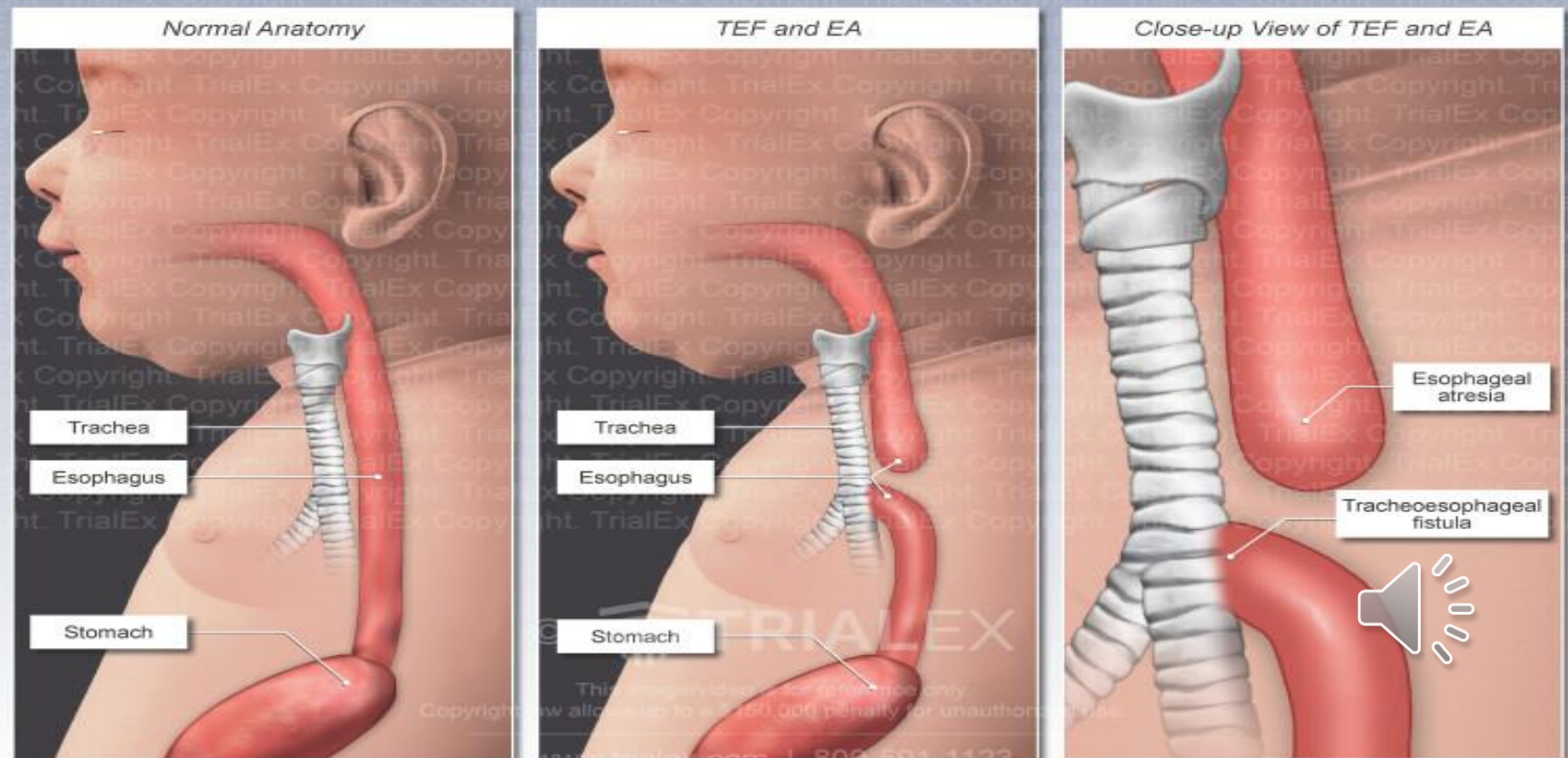
- Intact esophagus with H fistula

➤ Often associated with other congenital abnormalities



T/E Fistula & Esophageal Atresia

Tracheoesophageal fistula (TEF) and Esophageal Atresia (EA)



Congenital Diaphragmatic Hernia

- Two types:
 1. Bochdalek hernia
 - Lateral and posterior defect, usually on the left
 2. Morgagni hernia
 - Medial and anterior, may be on either side
- Combination of lung hypoplasia and abnormal development
- Hernias that occur in the right hemidiaphragm may be less severe
 - Liver can block the defect decrease the volume of abdominal contents that can enter the thorax





Congenital Diaphragmatic Hernia (Cont.)

- Physical findings:
 - Scaphoid abdomen
 - Decreased breath sounds
 - Displaced heart sounds
 - Severe cyanosis
- Manifests as severe respiratory distress
- Diagnosis established with chest radiography



Congenital Diaphragmatic Hernia (Cont.)

■ Treatment

- Initially intubation, paralysis, MV, continuous gastric insufflation
- Repair delayed for PVR to fall due to adequate V_A
- Severe cases may require HFV and ECMO





Abdominal Wall Abnormalities

- Affect breathing because infants are abdominal breathers
- If severe may require MV
- One of most common is omphalocele
 - An abdominal wall defect that involves the insertion of the umbilical cord
 - Gastroschisis is an abdominal wall defect that is completely separate from the insertion of the umbilical cord





Congenital Heart Disease

■ Two large categories:

1. Cyanotic Heart Disease

■ Blood shunts from right to left, bypassing lungs

2. Acyanotic Heart Disease

■ Blood shunts left to right causing congestive heart failure



Cyanotic Congenital Heart Defects

- Tetralogy of Fallot:
 - Pulmonary stenosis, VSD, dextroposition aorta, RVH
 - Timing of surgical repair dependent on case
 - Chest radiograph shows “boot-shaped” heart
- Transposition of great arteries
 - Manifests as moderate to severe cyanosis at birth
 - Emergency atrial septostomy relieves distress
 - Perform arterial switch operation at 2-3 weeks of life
 - Chest radiograph shows “egg-shaped” heart



Acyanotic Congenital Heart Defects

- Ventricular septal defect (VSD):
 - Most common
 - Results in left-to-right shunt and CHF
 - Appears 6-8 weeks as PVR falls
- Atrial septal defect (ASD):
 - Typically of little importance
 - Most common
 - Incomplete closure of foramen ovale



Acyanotic Congenital Heart Defects (Cont.)

- Patent ductus arteriosus (PDA)
 - Typically occurs in premature infants
 - Normally closes 5 to 7 days after birth of term infants
 - May act as right-to-left or left-to-right shunt depending on SVR and PVR
 - Treatment is either pharmacologic (indomethacin) or surgical (ligation)

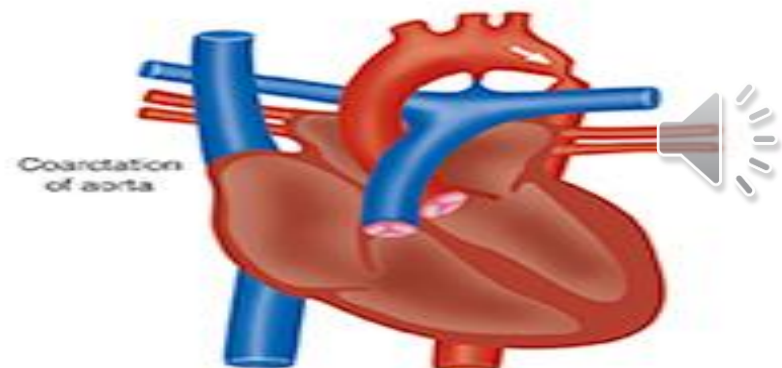
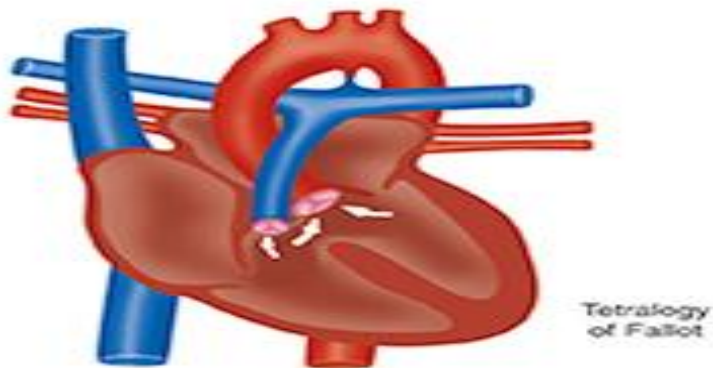
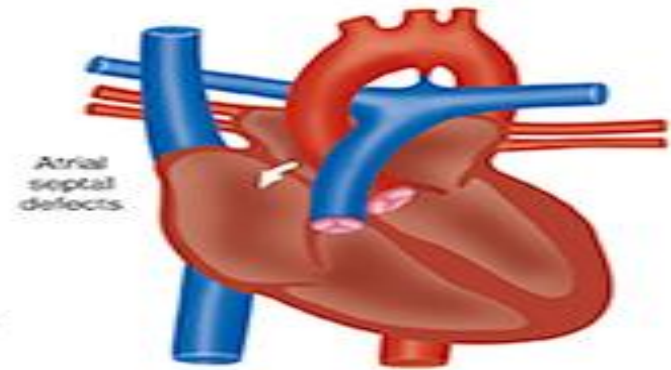
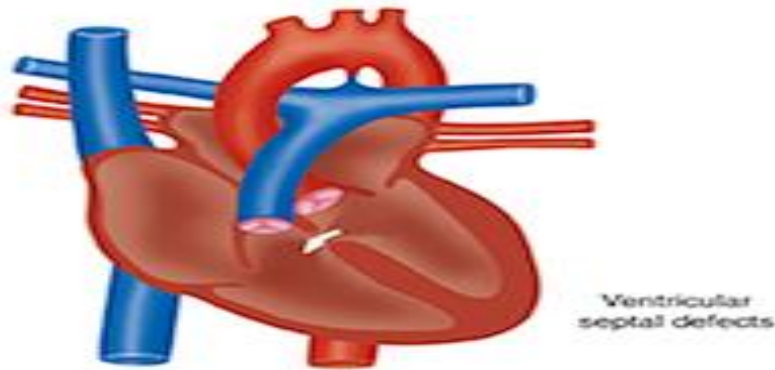
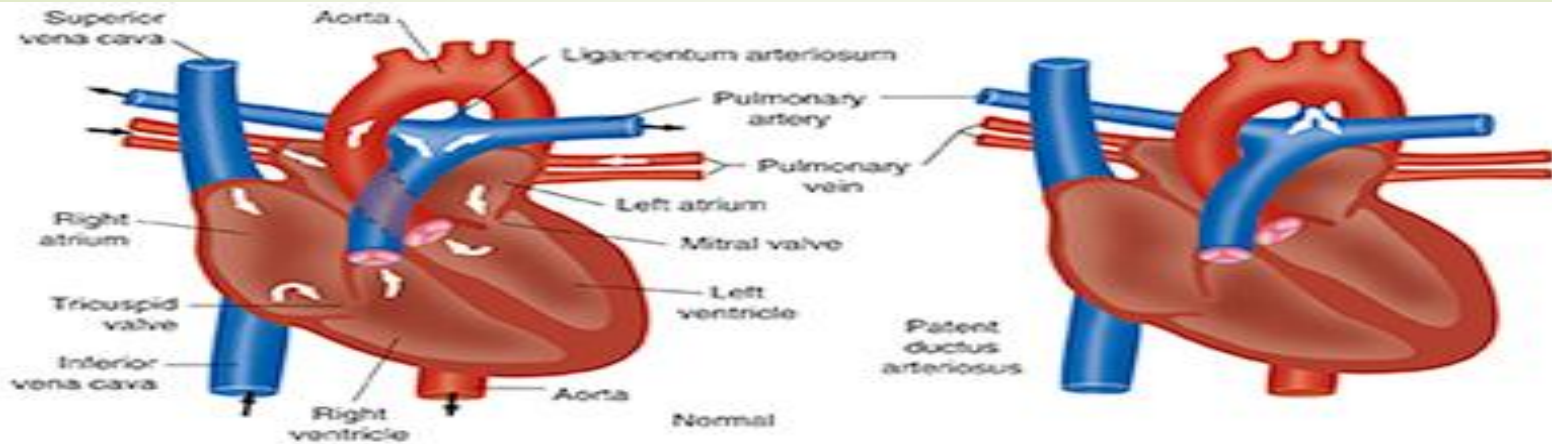


Acyanotic Congenital Heart Defects (Cont.)

- Left ventricular outflow obstructions
 - Includes aortic stenosis, coarctation of aorta, and hypoplastic left heart syndrome
 - Treatment: Depends on severity
 - Initially prostaglandin E1 may be administered
 - In severe cases:
 - Mechanical ventilation is required
 - Surgical procedures
 - Stenosis and coarctation: Resection or graft
 - Hypoplastic left heart: Norwood procedure or transplantation



Congenital Heart Defects





Pediatric Respiratory Disorders

- Gastroesophageal Reflux Disease (GERD)
- Bronchiolitis
- Croup
- Epiglottitis
- Cystic Fibrosis



Gastroesophageal Reflux Disease (GERD)

- Associated respiratory problems include:
 - Reactive airways disease
 - Aspiration pneumonia
 - Laryngospasm
 - Stridor
 - Chronic cough
 - Choking
 - Apnea
- Diagnosed with esophageal pH testing, upper gastrointestinal contrast studies, and gastric scintiscanning
- Once diagnosed, medical and surgical management is required



Bronchiolitis

- Caused by respiratory syncytial virus (RSV)
- Clinical manifestations
 - Usually follows URTI
 - Slight fever and cough worsen to dyspnea and tachypnea
 - Inspiratory and expiratory wheezing may develop
 - Radiograph shows hyperinflation and consolidation
- Prophylaxis: passive immunization became available for RSV
- Treatment:
 - Relief of airway obstruction and hypoxemia
 - MV with prolonged expiration used in most severe cases



Croup

- Also called laryngotracheobronchitis
- **Viral infection** resulting in subglottic swelling
 - Most common cause of obstruction in 6-month- to 6-year-olds
- Clinical manifestations
 - Follows 2-3 days of nasal congestions, fever, cough
 - Progressive inspiratory and expiratory stridor
 - Barking cough
 - Progression to dyspnea, cyanosis, and exhaustion
 - Chest radiograph shows classic “steeple sign”





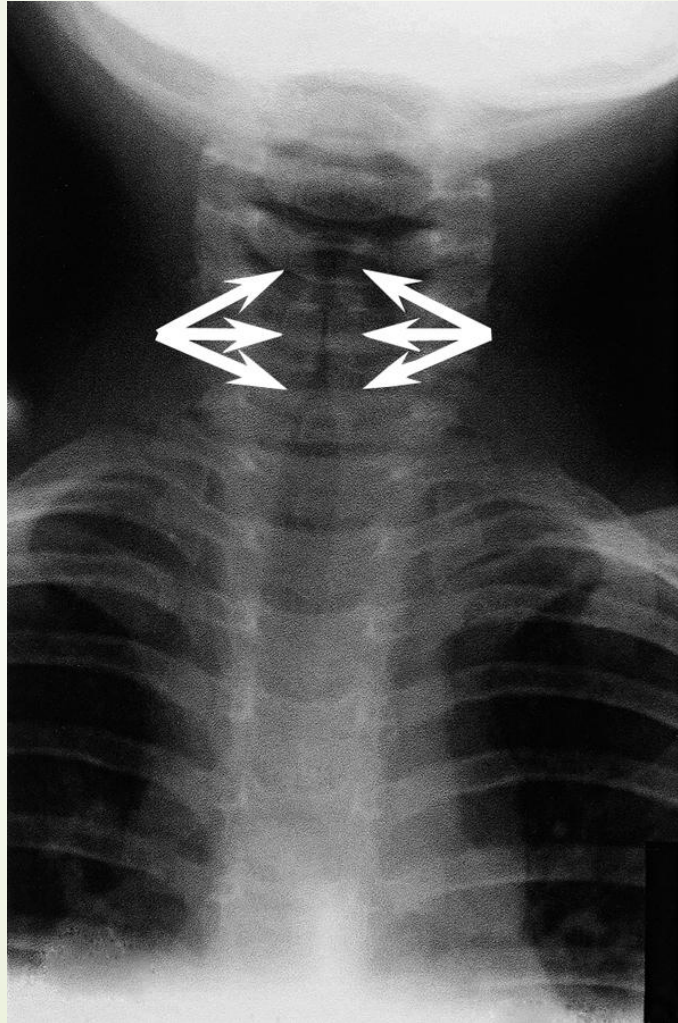
Croup (Cont.)

➤ Treatment

- Mild to moderate disease treated with cool mist therapy with or without supplemental oxygen
- Corticosteroids and epinephrine shows greatest benefit in reducing length and severity of symptoms
- Addition of budesonide may help
- Severe cases progressing despite therapy may require intubation and mechanical ventilation
- Heliox may be used for severe cases



Steeple Sign

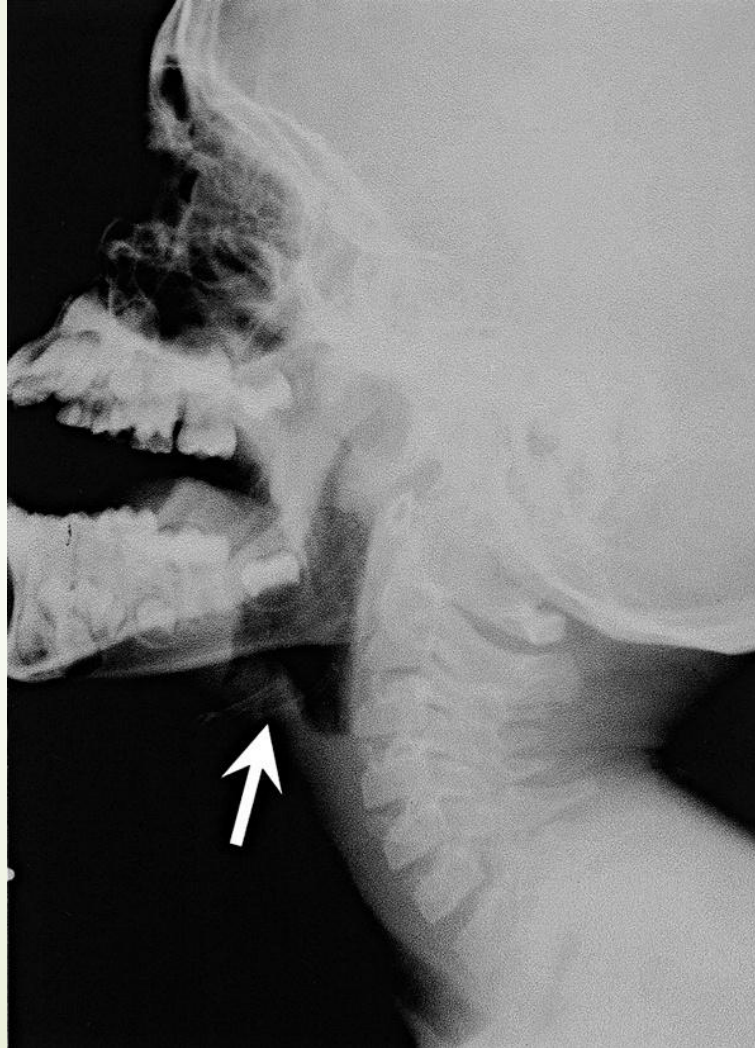


Epiglottitis

- Most commonly caused by *H. influenzae* type B
- Life-threatening infection resulting in supraglottic swelling
- Clinical manifestations
 - High fever, sore throat, stridor, labored breathing
 - NOT associated with barking cough
 - May have difficulty swallowing and muffled voice
 - Lateral neck radiograph shows “thumb sign”
- Treatment: proceed with great care
 - Elective intubation under general anesthesia
 - Corticosteroids may decrease swelling



Thumb Sign



Cystic Fibrosis

- Most common lethal genetic disease in white Americans
 - Genetic mutation of gene coding for large protein called cystic fibrosis transmembrane conductance regulator (CFTR), which affects chloride movement, particularly in exocrine glands
- Clinical manifestations
 - Most severely affected organs:
 - Sweat glands, pancreas, and lungs
 - Skin is very salty: mother notes when kissing infant
 - Sweat chloride test is diagnostic
 - Greater than 60 mEq/L indicates presence of disease
 - Pancreatic insufficiency leads to malnutrition, diarrhea, and steatorrhea
 - Lung disease is leading cause of death
 - Patients produce copious amounts of thick, mucoid sputum



Cystic Fibrosis (Cont.)

- Monitoring of progression and response to treatment
 - Routinely accomplished with spirometry and chest films
 - Cultures of sputum used to monitor airway flora and resistance
- Treatment
 - Pancreatic enzyme supplements
 - Regular chest physiotherapy
 - Strenuous exercise, external pneumatic vests, PEEP devices, or autogenic drainage may substitute for regular chest physiotherapy
 - DNase, 7% saline preserve lung function
 - Antibiotics
 - May use antiinflammatory agents and bronchodilators
 - Lung transplant





Take Home Points

- There are many cardiopulmonary disorders which are unique to infants and young pediatric patients.
- Infants respond differently to diseases and distress than older patients.
- Treatment of pediatric and neonatal respiratory disorders are important subspecialties requiring specific skills to ensure competent care.
- Expertise and knowledge to improve patient outcomes in all populations including infants and pediatric patients.
- Stay current with the latest evidence-based trends in pediatric and neonatal respiratory care.



Selected References

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