Acute Respiratory Distress in Infants and Children

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Overview

- Etiologies of respiratory distress
- Common radiographic presentations
- Cases presenting through the Emergency Department
3-month-old previously healthy female

Mild congestion and decreased oral intake for 24 hrs.

Transferred to ED after being seen in local urgent care
- Tachypnea with grunting
- Hypoxic
- Afebrile
- Large cardiopericardial silhouette
- Fissural thickening
- Ill defined pulmonary vasculature
- Lower lobe atelectasis (left > right)
Heart Failure

- **Volume overload**
  - L to R shunts
  - Valvular insufficiency
  - Systemic AV communication

- **Pressure overload**
  - Outflow or inflow obstruction

- **Disorders affecting the inotropic state**
  - Myocarditis, electrolyte disturbances, hypoxia, acidosis, cardiomyopathies, coronary artery lesions, metabolic derangements, septic shock

- **Alterations in the chronotropic state**
  - SVT, VT, complete heart block
Age is critical

Drop in pulmonary vascular resistance (falls in the first 3 months of life)
- Ventricular septal defect (VSD)
- Patent ductus arteriosus (PDA)
- Anomalous left coronary artery from the pulmonary artery (ALCAPA)
VSD

- **Isolated VSD**
  - 2-6/1,000 live births
  - 20% CHD
    - Second to BAV

- **Small VSDs**
  - Asymptomatic
  - Excellent long term prognosis

- **Moderate/Large VSD**
  - Trial of medical therapy
  - Uncontrolled HF
    - Surgical repair
PDA

- Functionally closed within 48 hrs. after birth
  - Most authorities consider patent ductus abnormal > 3 months
- Term infants
  - 1 in 2,000 births
- Hemodynamic impact of PDA determined by magnitude of shunting
  - Flow resistance of the ductus arteriosus
    - Length
    - Narrowest diameter
    - Overall shape and configuration
ALCAPA

- Anomalous left coronary artery from the pulmonary artery
  - AKA Bland-Garland-White syndrome
- 1 in 300,000 live births
- Usually isolated
Spectrum of pathophysiologic changes that take place after birth in patients with ALCAPA

Peña E et al. Radiographics 2009;29:553-565
ALCAPA
ALCAPA
Structurally Normal Heart

- Primary cardiomyopathy
  - Dilated
  - Hypertrophic
  - Restrictive
- Secondary
  - Arrhythmogenic
  - Ischemic
  - Toxic
  - Infiltrative
  - Infectious
Cardiomyopathy (LVNC)
Back to our patient

- **ECHO**
  - Severely dilated LV
  - Severely depressed LV systolic performance
    - EF 15%
  - No VSD/PDA
  - Normal coronaries
- **Cardiac catheterization**
  - Endomyocardial biopsy
    - Active myocarditis
Myocarditis

- Inflammation of cardiac muscle
- Myocellular damage → heart failure
- Causes
  - Infectious (most common cause in pediatrics)
    - Viral (enterovirus, adenovirus, parvovirus, HHV-6)
  - Toxic
  - Autoimmune
- Mean age of Dx 9.2 years
  - Bimodal distribution
    - Between 6 and 12 months of age (infancy)
    - 16 years of age (adolescence)
Worsening despite inotropic support
Case 2

- 10-month-old boy with recurrent respiratory distress
- Seen in the ED with admissions at the ages of 5, 7, 8, and now at 10 months
- Born after full term gestation and had received all routine immunizations
- No pets in the house
- Father, maternal grandmother, paternal grandfather, and sibling with asthma
Case 2

- With each admission retractions, cough, and tachypnea with fine crackles were described on physical exam with O2 sat between 85-90% on room air
- Each time, his condition improved with supplemental oxygen, prednisolone, nebulizer treatments, and antibiotics
- d/c home after 5 to 7 days following each admission
- Symmetric hyperinflation
- Parahilar opacities
Reactive airway disease

- Not unusual in infants and can be seen in up to 19% of children
- Many names including transient early wheeze
- Underlying cause small airways of infants
  - With infection airway diameter $\downarrow = \text{wheeze}$
- Gradually resolves with growth of airways
- Hyperinflation and wheeze are characteristic
- Persistent fine crackles and prolonged hypoxemia are not consistent with RAD
DDx

- Congenital heart disease
  - Clinically confined to the respiratory system
    - Normal auscultatory exam
- Immunodeficiency
  - No true documented infections
  - No history of recurrent/incompletely cleared
    - Otitis
    - Sinusitis
    - Pharyngitis
    - Conjunctivitis
  - Normal growth
Diffuse/Interstitial Lung Disease

- In pediatric patients, a diagnosis of ILD is considered when the child has (chILD syndrome):
  - Respiratory symptoms (cough✓, rapid breathing✓, exercise intolerance)
  - Signs (resting tachypnea✓, adventitious sounds✓, retractions✓, clubbing, FTT)
  - Hypoxemia✓
  - Diffuse abnormalities on CXR or CT
- 75% cases are manifested before 1 year of age
Pediatric Diffuse Lung Disease

- Disorders more prevalent in infancy
  - Diffuse developmental disorders
  - Alveolar growth abnormalities
  - Pulmonary interstitial glycogenosis (PIG)*
  - Neuroendocrine cell hyperplasia of infancy (NEHI)*
  - Surfactant dysfunction disorders

*Specific conditions of undefined etiology
Diffuse Developmental disorders

- Acinar dysplasia
- Congenital alveolar dysplasia
- Alveolar capillary dysplasia with misalignment of pulmonary veins

- Term infants present in the immediate neonatal period
  - Rapid progressive respiratory failure
  - Severe pulmonary HTN
  - Universally fatal (1 case of successful lung transplant)
Alveolar growth abnormalities

- Most common cause of diffuse lung disease in infancy
- Impaired alveolarization
  - Lobular simplification
    - Fewer and larger alveoli
    - Deficient septation and vascularization
Alveolar growth abnormalities

- **Prenatal**
  - Reduced in utero thoracic space
    - CDH, oligohydramnios, CPAM/sequestration, skeletal dysplasia

- **Postnatal**
  - Chronic neonatal lung disease/"new" BPD
    - Extremely premature neonates
      - Arrested or delayed alveolar and pulmonary vascular development

- CHD, genetic disorders (trisomy 21, filamin A X-linked gene), primary idiopathic
Pulmonary interstitial glycogenosis

- Unknown etiology
  - ? Reactive feature that associates with conditions related to lung development and injury*
- Glycogen-laden mesenchymal cells in alveolar interstitium
  - Patchy or diffuse
- Particularly common in alveolar growth abnormalities
  - Patchy PIG present in 40% of lung biopsies having lung growth abnormalities*
  - Most common in neonates (immediately or soon after birth) and very young infants

*Deutsch GH, Young LR (2010) Pediatric Radiology 40

PIG with alveolar growth abnormality

PIG in a patient with aortic coarctation
Surfactant dysfunction disorders

- Genetic mutations in proteins involved in surfactant metabolism
  - ABCA3
  - Surfactant protein B (SP-B)
  - Surfactant protein C (SP-C)
  - GM-CSF-Rα
  - Thyroid transcription factor 1 (TTF-1)
  - Solute carrier 747
Surfactant dysfunction disorders

- **Autosomal recessive mutations in SP-B**
  - Severe respiratory distress within hours of birth
  - Unresponsive to exogenous surfactant or ECHMO
  - Fatal within a few months without lung transplant

- **Autosomal recessive ABCA3 and inherited/spontaneous autosomal dominant SP-C mutations**
  - Variable expression
    - Acute severe respiratory distress to chronic diffuse lung disease in children or adults
Surfactant protein C mutation

ABCA3 mutation
NEHI

- ↑# pulmonary neuroendocrine cells
  - Within bronchiolar epithelium
  - Bombesin stain
  - Lack of inflammatory cells
- Usually presents in infancy
  - Tachypnea
  - Hypoxemia
  - Crackles
  - ↑ Symptoms with viral respiratory infections
- Infant PFT
  - Significant air trapping and airway obstruction
Treatment

- Supportive
  - O2 supplementation
  - Nutritional support
- Bronchodilators/Corticosteroids
  - Only beneficial during treatment of superimposed viral infxn
- No reported deaths
Radiography
- Hyperinflation
  - Resembles RAD/bronchiolitis

CT
- Air trapping
- Geographic ground-glass opacities
  - Middle lobe
  - Lingula
  - Paramediastinal

Characteristic symptoms, CT findings, and infant PFT for NEHI
- Forego lung biopsy
Case 3

- 5-year-old fully immunized otherwise healthy girl
  - Fever
  - Tachypnea (RR 60s)
  - Hypoxia (Sp02 85% RA)
Differential Diagnosis

- Age
  - Neonatal or beyond
- Clinical symptoms
- Position of mediastinal structures
Ipsilateral mediastinal displacement
  - Lung collapse
    - Severe respiratory symptoms
    - DDx
      - Endobronchial foreign body
      - Mucus plug (asthma, surgery)
      - Endobronchial tumor
      - Extrinsic bronchial compression
  - Radiography
    - Interrupted bronchus
    - +- pneumomediastinum/subcutaneous gas
Aspirated chicken obstructing left main bronchus
Mucus plug
Endobronchial tumor
Endobronchial tumor

Mucoepidermoid carcinoma
Extrinsic compression
Necrotic left hilar adenopathy → Mycobacterium Avium
Extrinsic compression

Necrotic left hilar adenopathy → Mycobacterium Avium
Contralateral mediastinal displacement

- **DDx**
  - Isolated pleural effusion
  - Congenital malformations (secondary infection)
  - Pneumonia with pleural effusion
  - Tumor

- **Imaging**
  - US extremely useful
    - Detect and characterize pleural effusion
    - Characterize consolidation
    - Potentially differentiate tumor from consolidation
Pleural Effusion with Atelectais
Tumor

Pleuropulmonary Blastoma
Tumor

T-Cell Lymphoma
Our patient
Our patient
Necrotizing pneumonia
Uncommon but increasing complication of community acquired pneumonia

*Staphlococcus aureus, Streptococcus pneumoniae* most common

Lung necrosis and cavitation
  - Microangiopathic changes
    - Thrombosis of intrapulmonary blood vessels
Ultrasound

- US similar to CT in detection of lung necrosis and lung abscess*
- Increased lung water in inflammation
  - Renders parenchyma visible
    - Hepatization
  - Dynamic air bronchogram
- Altered echogenicity and vascularity
  - Necrosis

*Kurian J et al. AJR Dec 2009
Case 4

- 15-year-old male with sudden onset sharp left chest pain
- SOB at rest and with minimal exertion with no improvement overnight prior to arrival at ED
Case 4
Spontaneous Pneumothorax

- Primary
  - No underlying lung disease that would predispose to air leak
    - Asthenic body habitus
    - Increased transpulmonary pressure
      - Valsalva maneuver
      - Diving/military flying
Spontaneous Pneumothorax

- Secondary
  - Complication of underlying lung disease
    - Asthma
    - CF
    - Necrotizing pneumonia
    - ILD
Langerhans Cell Histiocytosis
Etiology
- Mechanical stress highest at lung apices
  - Larger alveoli
  - More negative intrapleural pressures
    - Alveolar rupture forming air collection beneath visceral pleura (bleb)
- Rupture of apical subpleural bleb → PTX
- Pleural porosity
  - Replacement of mesothelial cells in the pleura by porous inflammatory cell layer → air leakage
- Resolved left PTX with chest tube
- 3 week f/u no symptoms
  - Repeat radiograph showing bi-apical irregularity
Returns to ED 3 months later

New sudden onset right chest pain and SOB

- Pain started after coughing
- Substantial residual PTX when placed to water seal
- CT chest to “evaluate parenchyma”
Limited Chest CT
Limited Chest CT

Right greater than left apical “blebs” and left apical “lines”
Ipsilateral apical blebs identified in 28% of children (13-19 years) with PSP
- Published adult series varies from 56% to 88%
78% of the imaged events with blebs ipsilateral to the PTX also showed contralateral blebs
100% of the identified lesions in the apical subpleural regions
- Limited CT confined to upper zones → ↓ radiation exposure

“Apical Lines”

- Linear opacities seen at the apices of the lungs on the cephalad most axial images
- 56% of pts with PSP and 28% of age and gender matched controls
  - ? Normal variant
  - Should not be confused with apical blebs

Goals
- Treat acute episode
- Prevent recurrence

Recurrence
- Following first episode between 50-60%
  - Children tx with O2 or chest tube
- Higher than rates reported in adults

Currently no evidence to support the use of CT and subsequent surgical intervention in first episodes of PSP*

Clinical presentation of CHD is often age dependent...don’t forget ALCAPA

Keep DLD in the back of your mind when seeing infants with repeat “viral/RAD”

Ultrasound is your friend...especially with thoracic infections

- More commonly being used for lung evaluation
  - PTX
    - Alveolar/interstitial syndromes

Limit chest CT to apices in primary spontaneous PTX