Case Presentation #1

- A 5 year old male presents to your office with a chronic cough
  - Cough is productive, increased at night, recurrent
  - Worse with exercise and with upper respiratory infections
  - Growth has been normal
  - Chest xray findings are normal except for mild hyperinflation

Differential Diagnosis: Which is the MOST likely diagnosis?

A. Sinusitis
B. Asthma
C. Gastroesophageal reflux disease
D. Tuberculosis
E. Cystic Fibrosis
F. Psychogenic cough
Asthma: Overview

- Chronic inflammatory disease of the airway
- Affects 20 million people in the US (9 million children)
- More than 70% also have allergies
- Prevalence has increased by almost 40% in all ages in the past decade.
- Typically develops in childhood - 50% before 3 years of age, and the majority before 8 years of age.
- 470,000 hospitalizations per year

Pathogenesis

- Airway inflammation also contributes to airflow limitation, which includes:
  - Bronchoconstriction
  - Edema
  - Chronic mucus plugging
  - Airway wall remodeling
- All this leads to bronchial obstruction
All of the following are asthma Risk Factors EXCEPT:

A. Inner city minority  
B. Low birth weight  
C. Cigarette smoking  
D. Family history of asthma  
E. History of atopy (allergies, eczema)  
F. Living on a farm

**History**

- Asthma is primarily a **clinical** diagnosis
- **History**
  - Cough
  - Wheezing
  - Shortness of breath, particularly with exercise
  - Chest pain or tightness
  - “Difficulty catching my breath”
  - Vomiting, particularly mucus
Physical Exam

- Wheezing
- Crackles in the lung
- Muscle retractions
- Often can be normal

Pulmonary Function Testing

- Determines
  - Degree of airway obstruction
  - Other lung disorders
Flow Volume Loops

Other Flow-Volume Loops

Diagnostic Studies

- Chest x-ray
- Immunoglobulins
  - Identify allergic components
  - Rule out associated immunodeficiencies
- Skin testing/RAST testing for allergies
- Sweat test to rule out Cystic Fibrosis
Treatment

- Bronchodilators
  - Short-acting
- Leukotriene modifiers
- Inhaled corticosteroids
- Combination therapy (inhaled steroid + long-acting bronchodilator)
- Systemic steroids (acute exacerbation)
- Methyloxanthines (Theophylline)
- IgE blocker (Omalizumab (Xolair))

Treatment—Inhaled Steroids

- Inhaled corticosteroids are standard of care for all categories except for mild intermittent asthma
  - Long term prevention of symptoms; suppression, control and reversal of inflammation.
  - Block late reaction to allergen
  - Reduce airway hyper-responsiveness
  - Inhibit inflammatory cell migration and activation
  - Increase B2 receptor affinity

Inhaled Steroids

- Budesonide (Pulmicort®)
- Fluticasone propionate (Flovent®)
- Mometasone (Asmanex®)
- Beclomethasone (Qvar®)
- Ciclesonide (Alvesco®)
- Flunisolide (Aerospan®)
- Fluticasone furoate (Arnuity Ellipta®)
Combination Therapy

- Fluticasone + Salmeterol (Advair®)
- Budesonide + Formoterol (Symbicort®)
- Mometasone + Formoterol (Dulera®)
- Fluticasone + Salmeterol (AirDuo Respliclick®)

All of the following are side effects of inhaled steroids EXCEPT:

A. Cough
B. Hoarse voice
C. Palpitations
D. Oral thrush.
E. Adrenal suppression
F. Growth suppression
G. Osteoporosis

Asthma and Exercise

- Exercise can trigger asthma
- Symptoms are worse with cold, dry air
- However, exercise helps lungs function better and prevents obesity
- As long as asthma is well-controlled and a short-acting bronchodilator (rescue medicine) is used beforehand, children with asthma should be able to do sports
- Pulmonary function testing best first test; then exercise testing.
Case # 2

- A 4-month-old infant boy is brought to the Emergency Room because of lethargy.

  **Physical Examination**
  
  Afebrile  HR 160 bpm   RR 50 breaths/min  SaO2: 98% on RA
  Weight: 3.2 kg
  
  **GENERAL** : Very thin, appearing to be *malnourished*;
  Lethargic but arousable
  **HEENT** : dry mucous membranes
  **CHEST** : equal breath sounds
  **ABDOMEN** : distended; no organomegaly
  **SKIN** : decreased turgor and elasticity
  **NEUROLOGIC** : poor muscle tone; poor suck

Case # 2 (Continued)

- **PMHx**: Born at term; No problems at birth.
  Hospitalized at 1 month of age for pneumonia:
  Chronic cough; Frequent diarrhea
- **Immunizations**: UTD
- **Social Hx**: Lives with parents
- **FHx**: Cousin with recurrent respiratory infections.
- **Nutrition & Growth**: breast fed; used to have good appetite but it got progressively worse; poor weight gain in the beginning; actual *weight loss* lately

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**SWEAT TEST**

- **Sweat Chloride**: 78.12 mmol/L

Normal under 6mos <30 mmol/L
Normal over 6mos <40mmol/L
Borderline 30-60 mmol/L
Abnormal >60 mmol/L
*In infants anything >30 should be repeated and worked up*
OVERVIEW OF CYSTIC FIBROSIS

- Most common fatal inherited disorder in Caucasians

- **Genetics:** Autosomal-recessive genetic disease caused by mutations in chromosome 7. The CF gene codes for a protein called the CF Transmembrane Regulator (CFTR).

  There are approximately 1,700 known mutations; however 75% of the patients are homozygous for the Δ508 mutation.

  Genetic testing for the 30 most frequent mutations is sensitive for the genotype of up to 90% of Americans.

- **Incidence:** varies significantly among racial groups
  - Caucasians: ~1/377-3500 live births
  - African-Americans: ~1/17,000 live births (US)
  - Asians: ~1/90,000 live births (Hawaii)

Cystic Fibrosis Gene

<table>
<thead>
<tr>
<th>Chromosome 7</th>
<th>Sequence of nucleotides in CFTR gene</th>
<th>Amino acid sequence of CFTR protein</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Isoleucine 506</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Isoleucine 507</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Phenylalanine 508</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Glycine 509</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Valine 510</td>
</tr>
</tbody>
</table>

Deleted in many patients with cystic fibrosis


Pathophysiology of CF

- The CFTR controls the Cl conductance in the apical epithelial cells (via the cAMP).

- The epithelial cells are unable to secrete salt and water on the airway surface.

- Thus, they can not hydrate secretions that in turn become **viscous and elastic** and difficult to be cleared by the mucociliary mechanisms.

- Similar events may take place in the pancreatic and biliary ducts as well as in the vas deferens.

- Because the sweat glands absorb chloride, salt is not retrieved from the primary sweat as it is transported to the skin surface and as a result its sodium and chloride levels are elevated.
The genetic defect underlying CF disrupts the functioning of several organs by causing ducts or other tubes to become clogged, usually by thick, sticky mucus or other secretions.

Organs Affected By CF
- Airways
- Liver
- Pancreas
- Sinuses
- Small intestine
- Reproductive tract
- Skin


All of the following are manifestations of CF:
1. Cough (productive)
2. Bulky, greasy stools with droplets of fat
3. Diabetes
4. Meconium ileus
5. Constipation
6. Azoospermia
7. Biliary cirrhosis
8. Pancreatitis

Potential Pulmonary Treatments in Cystic Fibrosis
- Airway clearance
- Antibiotics
- Corticosteroids
- Ibuprofen, other anti-inflammatory agents
- Dornase Alfa
- Hypertonic saline (HTS)
- Genetic or Protein Correctors
**CF: Newborn Screening**

- Assessment of Immunoreactive trypsinogen (IRT)
- Confirmation of positive IRT by CF gene mutation analysis
- Confirmation of results with a sweat test
- Now present in all 50 states in the US

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**Presenting Features of CF**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persistent respiratory symptoms</td>
<td>50%</td>
</tr>
<tr>
<td>Failure to thrive</td>
<td>43%</td>
</tr>
<tr>
<td>Abnormal stools</td>
<td>35%</td>
</tr>
<tr>
<td>Meconium ileus, intestinal obstruction</td>
<td>19%</td>
</tr>
<tr>
<td>Family history</td>
<td>17%</td>
</tr>
<tr>
<td>Hyponatremia, acid-base abnormality</td>
<td>5%</td>
</tr>
<tr>
<td>Rectal prolapse</td>
<td>3%</td>
</tr>
<tr>
<td>Nasal polyps, chronic sinusitis</td>
<td>2%</td>
</tr>
<tr>
<td>Hepatobiliary disease</td>
<td>1%</td>
</tr>
</tbody>
</table>

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**All the following support a Dx of CF except:**

1. Typical clinical features (e.g. cough, FTT)
2. A positive newborn screening testing
3. 2 sweat chloride concentrations of 20 and 24 mEq/L
4. Identification of 2 CF mutations
5. Abnormal nasal potential difference
Common Respiratory Pathogens in CF

- Staph Aureus
- MRSA
- Non-typable Haemophilus Influenza
- Pseudomonas Aeruginosa
- Burkholderia cepacia

Also:
- Candida
- Aspergillus Fumigatus
- Nontuberculous Mycobacteria

Signs and Symptoms of a Pulmonary Exacerbation in CF

- Increased frequency and duration of cough or increased pulmonary symptoms
- Increased sputum production and change in appearance
- Appearance of rhonchi and crackles
- Decline in indices of pulmonary function
- Weight loss
- New infiltrate on Chest X-ray

Case Study #3

BG “A” is an ex-24 week preemie with chronic lung disease of the newborn, a history of a patent ductus arteriosus (PDA), and apnea of prematurity, who is now preparing to be discharged home from the NICU. She is now 4 months of age (41 weeks gestational age). She still has occasional apneic episodes, mostly occurring with feeds, with desats to the 80s and bradycardia. Baseline oxygen saturations are normal.
Apnea of Infancy

Unexplained episode of cessation of breathing for 20 seconds or longer, or a shorter respiratory pause associated with bradycardia, cyanosis, pallor, and/or marked hypotonia

- Called apnea of prematurity when present in an infant younger than 37 weeks gestational age.
- Usually ceases by 37 weeks postmenstrual age, but may persist for several weeks beyond term.
- Extreme episodes usually cease at 43 weeks postconceptional age.

Apparent Life-Threatening Event (ALTE)

- Episode in an infant that is frightening to the observer and is characterized by some combination of:
  - Apnea (central or occasionally obstructive)
  - Color change
  - Unresponsiveness
  - Change in muscle tone, choking, or gagging

SIDS

Sudden death of an infant under 1 year* of age that remains unexplained after a thorough investigation, including autopsy, examination of the death scene, and review of the clinical history.

*Risk much lower >6mos of age
Risk Factors for SIDS

- Sleeping in prone position
- Co-sleeping
- Smoking
- Low socioeconomic status
- Overheating
- Young parents

*Apnea appears to resolve at a postnatal age before which most SIDS deaths occur and apnea is not a predictor or a precursor to SIDS

Prematurity

- Preterm infants at greater risk of extreme apnea episodes
- Risk decreases with time, ceasing at approximately 43 weeks postmenstrual age
- In infants with recurrent, significant apnea, monitoring may be considered

AAP Recommendations

- Home monitors should not be prescribed to prevent SIDS
- Home monitors may be warranted for premature infants who are at high risk of recurrent episodes of apnea, bradycardia, and hypoxemia after hospital discharge.
- However, the use of home monitors should be limited to approximately 43 weeks postmenstrual age or after the cessation of extreme episodes, whichever comes last
AAP Recommendations

- Parents should be advised that home monitoring has not been proven to prevent SIDS
- Pediatricians should continue to promote proven practices that decrease the risk of SIDS—supine sleep position, safe sleeping environments, and elimination of prenatal and postnatal exposure to tobacco smoke


Obstructive Sleep Apnea

Disorder of breathing during sleep characterized by prolonged partial upper airway obstruction and/or intermittent complete obstruction (obstructive apnea) that disrupts normal ventilation during sleep and normal sleep patterns


Airway Obstruction during Sleep

- Combination of structural and neuromuscular factors
- Dynamic process
- Site of airway collapse in children most often at level of the adenoid
All of the following are risk factors for obstructive sleep apnea EXCEPT:

A. Adenotonsillar hypertrophy
B. Obesity
C. Craniofacial anomalies
D. Gastroesophageal reflux disease
E. Neuromuscular disorders

Prevalence of OSAS

- Children of all ages
- Most common in preschool-aged children (age at which tonsils and adenoids are the largest in relation to the underlying airway size)
- Estimated prevalence rates of approximately 2%

High Risk Populations

- Obesity
- Down syndrome
- Prader Willi syndrome
- Neuromuscular disease
- Craniofacial anomalies
Symptoms

- Habitual nightly snoring
- Disturbed sleep
- Daytime neurobehavioral problems
  - Think about it with ADHD and midfacial anomalies
- Daytime sleepiness may occur, but is uncommon in young children

All are consequences of obstructive sleep apnea in children EXCEPT:

A. Cor pulmonale
B. Failure to thrive
C. Seizures
D. Attention problems
E. Hyperactivity

Case Presentation #4

- Six year old female presents to the ER after a one week history of nasal congestion and mild cough. Two days ago, she developed high fevers, chills, and increased cough.
- Upon arrival in the ER, she is ill-appearing, tachypneic, and febrile.
- PE: Rales are appreciated on exam over right posterior lung fields.
Case Presentation #4

PMHx: No prior pneumonia or wheezing
FHx: +Asthma (brother)
ALL: NKDA
IMM: Missing part of primary series; no recent ppd done.
SHx: No recent travel out of the country.
Laboratory: WBC 35,000

Radiographic Findings

Definition: Pneumonia
An inflammation of the lung parenchyma
Which is the MOST likely causative organism in this patient?

1. Group B strep
2. Streptococcus pneumoniae
3. Tuberculosis
4. Mycoplasma
5. Legionella

Background
- More than 2 million children die annually of pneumonia worldwide
- Mortality rare in the developed world
- In U.S., 35-40 episodes of community-acquired pneumonia /1,000 children per year
- Respiratory viruses most common cause of pneumonia during the first years of life
- Risk factors: daycare, cigarette smoking

Pathophysiology of Bacterial Pneumonia
- Most common event disturbing lung defense mechanisms is a viral infection
- Alters properties of normal lung secretions
- Inhibits phagocytosis
- Modifies normal bacterial flora
- Often precedes development of a bacterial pneumonia by a few days
High Risk Populations

- Agammaglobulinemia
- CF
- Cleft palate
- Congenital bronchiectasis
- Ciliary dyskinesis
- TEF
- Immunodeficiency
- Neutropenia
- Deficient gag reflex
- Trauma
- Anesthesia
- Dental work
- Aspiration

Organisms

- Neonates
  - E. coli
  - Group B strep
  - H. influenzae
  - S. pneumoniae
  - Listeria
  - Anaerobes

- Infants
  - S. pneumoniae
  - S. aureus
  - H. Influenzae

Organisms

- Preschool age
  - S. pneumoniae
  - Moraxella
  - H. Influenzae
  - Neisseria meningitidis

- School age and adolescent
  - S. pneumoniae
  - S. aureus
  - Mycoplasma
  - C.pneumoniae (TWAR)
Clinical Sxs

- Shaking chills
- High Fever
- Cough
- Chest pain
- Mild URI sx
- Decreased appetite
- Abrupt onset high fever
- Rusty-colored sputum
- Respiratory distress
- Cyanosis

*Pattern more variable in infants and young children and PE often unrevealing

Physical Exam

- Retractions
- Dullness to percussion
- Tubular breath sounds
- Rales
- Diminished tactile and vocal fremitus
- Decreased breath sounds

Laboratory Findings

- Leukocytosis with left shift
- WBC <5,000/mm³ poor prognosis
- Bacteremia on blood culture
- Positive sputum culture

Severity of community acquired pneumonia in infants and children

<table>
<thead>
<tr>
<th>Clinical features of mild pneumonia</th>
<th>Clinical features of severe pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Temperature &lt;38.5°C (101.3°F)</td>
<td>Temperature ≥38.5°C (101.3°F)</td>
</tr>
<tr>
<td>Mild or absent respiratory distress</td>
<td>Moderate to severe respiratory distress</td>
</tr>
<tr>
<td>Mild or absent retractions</td>
<td>RR &gt;70 breaths/minute for infants; RR &gt;50 breaths/minute for older children</td>
</tr>
<tr>
<td>No grunting</td>
<td>Grunting</td>
</tr>
<tr>
<td>No nasal flaring</td>
<td>Nasal flaring</td>
</tr>
<tr>
<td>No apnea</td>
<td>Apnea</td>
</tr>
<tr>
<td>Mild shortness of breath</td>
<td>Significant shortness of breath</td>
</tr>
</tbody>
</table>

Normal color
Normal mental status
Noncynotic (oxygen saturation ≥90 percent in room air)
Normal feeding (infants); no vomiting
Normal heart rate
Capillary refill <2 seconds

Cyanosis
Altered mental status
Hypoxic (oxygen saturation <90 percent in room air or at sea level)
Not feeding (infants) or signs of dehydration (older children)
Tachypnea
Capillary refill ≥2 seconds
Complications

- Empyema—pus in the pleural space
- Pleural effusion
- Pericarditis
- Meningitis
- Osteomyelitis
- Metastatic abscesses

*Antibiotic therapy has reduced spread of infection
Pre-antibiotic era mortality rate high in infants

Pleural Effusion

- Decision to hospitalize based on severity of the illness and home environment
- Can often treat simple pneumonia as an outpatient
- Patients with empyema or pleural effusion should be hospitalized
- Oxygen
- Thoracentesis
- Chest tube drainage
- Decortication
Empiric Therapy In Infants

- Neonates
  - Parenteral antibiotics
  - Ampicillin and Gentamicin or Amikacin
  - Treat as rule out sepsis
- Infants 1 to 6 months
  - Should use parenteral initially
  - Ceftriaxone if less than one year of age and more severe pneumonia
  - Once stabilized, can give Augmentin for total of 10 day course

Empiric Therapy 6 months to 5 years

Typical bacterial

- Amoxicillin 90 mg/kg per day in 2 or 3 divided doses (MAX 4 g/day), OR
- Amoxicillin-clavulanate 90 mg/kg per day of the amoxicillin component in 2 or 3 divided doses (MAX 4 g/day amoxicillin component), OR
- For patients with non type 1 hypersensitivity to penicillins:
  - Ceftriaxone 14 mg/kg per day in 2 divided doses (MAX 400 mg/day), OR
  - For patients with type 1 hypersensitivity to penicillins:
    - Levofloxacin\(^{*}\) 16 to 20 mg/kg per day in 2 divided doses (MAX 750 mg/day), OR
    - Cephalexin 30 to 40 mg/kg per day in 3 or 4 divided doses (MAX 1.6 g/day), OR
    - Erythromycin 30 to 50 mg/kg per day in 4 divided doses (MAX 2 g/day as base, 3.2 g/day as ethylsuccinate), OR
    - Azithromycin 10 mg/kg on day 1 followed by 5 mg/kg daily for 4 more days (MAX 500 mg on day 1 and 250 mg thereafter), OR
    - Clarithromycin 15 mg/kg per day in 2 divided doses (MAX 1 g/day), OR
    - In communities with a high rate of pneumococcal resistance to penicillin:
      - Levofloxacin\(^{*}\) 16 to 20 mg/kg per day in 2 divided doses (MAX 750 mg/day), OR
      - Linezolid 60 mg/kg per day in 3 divided doses (MAX 1800 mg/day)

- For patients ≥18 years:
  - Levofloxacin 8 to 10 mg/kg once daily for children 5 to 16 years (MAX 500 mg/day); 500 mg once per day for children ≥16 years, OR
  - Moxifloxacin 400 mg once per day

In the United States, fluoroquinolones (eg, levofloxacin and moxifloxacin) are approved by the Food and Drug Administration for community acquired pneumonia for patients ≥18 years of age. However, they may be used in younger children if other antibiotics are inappropriate (eg, due to hypersensitivity or local antimicrobial resistance patterns).

Empiric therapy > 5 years

Mycoplasma pneumoniae or Chlamydophila pneumoniae

- Erythromycin 40 to 50 mg/kg per day in 4 divided doses (MAX 2 g/day as base, 3.2 g/day as ethylsuccinate), OR
- Azithromycin 10 mg/kg on day 1 followed by 5 mg/kg daily for 4 more days (MAX 500 mg on day 1 and 250 mg thereafter), OR
- Clarithromycin 15 mg/kg per day in 2 divided doses (MAX 1 g/day), OR
- Doxycycline 4 mg/kg per day in 2 divided doses (MAX 200 mg/day), OR
- For patients ≥18 years:
  - Levofloxacin 8 to 10 mg/kg once daily for children 5 to 16 years (MAX 500 mg/day); 500 mg once per day for children ≥16 years, OR
  - Moxifloxacin 400 mg once per day

In the United States, fluoroquinolones (eg, levofloxacin and moxifloxacin) are approved by the Food and Drug Administration for community acquired pneumonia for patients ≥18 years of age. However, they may be used in younger children if other antibiotics are inappropriate (eg, due to hypersensitivity or local antimicrobial resistance patterns).
Empiric therapy > 5 years Typical Bacterial

- Amoxicillin 90 mg/kg per day in 2 or 3 divided doses (MAX 4 g/day), OR
- For patients with non type 1 hypersensitivity to penicillins:
  - Cefdinir 14 mg/kg per day in 2 divided doses (MAX 600 mg/day), OR
  - Cefpodoxime 10 mg/kg per day in 2 divided doses (MAX 400 mg/day), OR
- For patients with type 1 hypersensitivity to penicillins:
  - Levofolexacin 8 to 10 mg/kg once daily for children 5 to 16 years (MAX 750 mg/day); 750 mg once daily for children ≤16 years
  - Clindamycin 30 to 40 mg/kg per day in 3 or 4 divided doses (MAX 1.8 g/day), OR
- For patients with non type 1 hypersensitivity to penicillins:
  - Erythromycin 40 to 50 mg/kg per day in 4 divided doses (MAX 2 g/day as base, 3.2 g/day as ethyl succinate), OR
- Azithromycin 10 mg/kg on day 1 followed by 5 mg/kg daily for 4 more days (MAX 500 mg on day 1 and 250 mg thereafter), OR
- Clarithromycin 15 mg/kg per day in 2 divided doses (MAX 1 g/day), OR
- In communities with a high rate of pneumococcal resistance to penicillin:
  - Levofolexacin 8 to 10 mg/kg once daily for children 5 to 16 years (MAX 750 mg/day); 750 mg once daily for children ≤16 years, OR
  - Linezolid 30 mg/kg per day divided in 3 doses (MAX 1800 mg/day) for children <12 years; 20 mg/kg per day divided in 2 doses (MAX 1200 mg/day) for children ≥12 years

Follow-Up

- Most children have normal x-rays by 2-3 months after acute infection*
- 20% with residual changes 3-4 weeks after infection
- Children with persistent symptoms should have follow-up x-rays to rule out such things as foreign body, congenital malformations, or TB

Case #5

A 2-month-old infant boy is brought to the Emergency Room because of persistent cough and difficulty in breathing.

On examination the infant has audible stridor, a harsh, "honking" cough, and suprasternal and subcostal chest wall retractions

He has URI symptoms and a low-grade fever and is nontoxic appearing
Overview

- Stridor is a harsh, high-pitched predominantly inspiratory sound produced by partial obstruction of the airway, resulting in turbulent airflow.
- It is associated with variable degrees of difficulty in breathing
- Usually associated with suprasternal retractions, and when severe with intercostal, substernal and subcostal as well.

Sites & Sounds of Airway Obstruction

All of the following are potential causes of stridor in a 2-month-old infant EXCEPT:

1. Infectious
2. Trauma
3. Laryngomalacia
4. Airway hemangioma(s)
5. FB aspiration
6. Vascular ring
Case #5: Neonatal History

- Cyanosis and respiratory distress developed during the first 24 hours of life
- Cardiac echocardiogram revealed congenital cyanotic heart disease necessitating a Blalock-Taussig shunt
- He was intubated and mechanically ventilated until 10 days of life.

Which is the least likely cause for his stridor:

1. Subglottic stenosis
2. Vocal Cord Paralysis
3. Pulmonary artery sling
4. Idiopathic laryngomalacia
5. Vascular ring

What would be the least useful test in determining the cause of the stridor:

1. Lateral neck x-rays
2. CT scan of the neck and chest
3. Barium swallow
4. Bedside flexible laryngoscopy
5. Flexible fiberoptic bronchoscopy
Causes of Stridor in Infants & Children According to Site of Obstruction & Age

Nasopharynx
- Choanal atresia
- Thyroglossal cyst
- Macroglossia
- Hypertrophic tonsils
- Retropharyngeal or peritonsillar abscess

Larynx
- Laryngomalacia
- Laryngeal web, cyst or laryngocele
- Viral Croup
- Spasmodic croup
- Epiglottitis
- Vocal cord paralysis
- Laryngeal stenosis
- Cystic hygroma
- Laryngeal papilloma
- Angioneurotic edema
- Laryngospasm
- Vocal Cord Dysfunction

Trachea
- Subglottic stenosis
- Hemangioma
- Foreign body
- Tracheomalacia
- Bacterial tracheitis
- External compression
Laryngomalacia

- Arises as a dilatation of the saccule of the laryngeal ventricle
- Stridor can present at birth

Laryngocele

- Arises as a dilatation of the saccule of the laryngeal ventricle
- Stridor can present at birth

Laryngeal Cyst
Vocal Cord Paralysis

Subglottic Hemangioma

- Female: male is 2:1
- Usually a submucosal lesion
- No color change or bluish discoloration
- Frequently associated with hemangiomas elsewhere on the body
- Stridor biphasic, increased with crying or Valsalva

Vascular Ring
Acute Laryngotracheobronchitis (Croup)

Etiology

Parainfluenza virus 1 (also 2 & 3)
- Respiratory Syncytial Virus
- Rhinovirus
- Influenza virus A (and less often B)
- Adenovirus

Croup: Epidemiology

- Season: fall and early winter
- Gender: more common in boys
- Onset of symptoms: mostly at night
- Duration: from hours to several days

Recurrent (Spasmodic) Croup

- Affects about 6% of children
- Not associated with obvious infection
- Abrupt onset, usually during sleep
- Barking cough, hoarseness, stridor
- Usually resolves within hours
- May be a hypersensitivity reaction
- Associated with airway hyperreactivity
**Epiglottitis**

- Life-threatening infection
- Incidence is 10-40 cases per million people in the United States.
- Since 1985, with the widespread vaccination against *Haemophilus influenzae* type b (or *Hib*), which was the most common organism related to epiglottitis, the overall incidence of the disease has dropped dramatically.
- Epiglottitis typically peaks in children aged 2-4 years.
- Various organisms that can cause epiglottitis include *Streptococcus pneumoniae*, *Haemophilus parainfluenzae*, varicella-zoster, *herpes simplex virus* type 1, and *Staphylococcus aureus*.

**Epiglottitis: Symptoms**

- The most common symptoms include:
  - Sore throat
  - Muffling or changes in the voice,
  - Difficulty speaking
  - High fever
  - Dysphagia
  - Drooling
  - Respiratory distress

---

*Lateral X-Ray of Epiglottitis showing the enlarged epiglottis. This is also known as the thumbs-up sign.*
CASE #6

- PMH: unremarkable until onset of above symptoms; Normal growth until 1 year of age; no weight gain for past 3-4 months
- FHx: Significant for asthma in his 5-year-old sister.

Physical Examination

VS: T 37.3°C; HR 140 bpm  RR 42 breaths/min
SaO₂: 91% on RA  Wt: 10 kg (25th)  
**General**: well nourished but thin child; tachypneic but not in distress

**Chest**: symmetric with mild intercostal retractions; equal but somewhat decreased breath sounds bilaterally; scattered fine crackles

**Extremities**: mild (1+) clubbing

**Chest X-ray**: increased interstitial markings

Case #6

Differential Diagnosis

A. Cystic Fibrosis
B. Dysmorphic Cilia Syndrome
C. Interstitial Lung Disease
D. Immunodeficiency
E. Asthma
Children’s Interstitial Lung Disease (chILD)

- Heterogenous group of disorders of rare and diffuse lung diseases that produce considerable morbidity and mortality

Epidemiology of Pediatric ILD

- Prevalence: estimates range from 0.36/100,000 up to ~90/100,000
- Affects slightly more males (1.4:1)
- Affects mostly Caucasians (88%)
- Affected siblings in about 10% of cases
- Infants and young children more affected

ILD: Symptoms & Signs

**SYMPTOMS**
- Cough: 78%
- Tachypnea/Dyspnea: 76%
- Failure to thrive: 37%
- Fever: 20%

**SIGNS**
- Crackles: 44%
- Cyanosis: 28%
- Clubbing: 13%
ILD : Disorders in Infancy

1. Diffuse developmental disorders
   1. Congenital alveolar dysplasia
2. Growth abnormalities
   1. Pulmonary hypoplasia, chronic neonatal lung disease of prematurity, congenital heart dz
3. Pulmonary interstitial glycogenosis (PIG)
4. Neuroendocrine cell hyperplasia of infancy (NEHI)
5. Surfactant dysfunction disorders

ILD: Causes presenting in children over 2 years of age

- Intrinsic lung disease
  - Nonspecific interstitial pneumonia
  - Cryptogenic organizing pneumonia
  - Acute interstitial pneumonia
  - Desquamative interstitial pneumonia
  - Lymphoctic interstitial pneumonia
- Other primary disorders
  - Alveolar hemorrhage syndromes
  - Aspiration syndromes
  - Hypersensitivity pneumonitis
  - Infectious or postinfectious (bronchiolitis obliterans)
  - Pulmonary alveolar microlithiasis
  - Pulmonary alveolar proteinosis
  - Pulmonary infiltrates with eosinophilia
  - Pulmonary lymphatic disorders (lymphangiomatosis, lymphangiectasis)
- Pulmonary vascular disorders (hemangiomatosis)

ILD associated with systemic disease

1. Connective tissue diseases
2. Histiocytosis
3. Malignancies
4. Sarcoidosis
5. Neurocutaneous syndrome
6. Lipid storage diseases
7. Inborn errors of metabolism
Alveolar Hemorrhage Syndromes

- Without pulmonary capillaritis
  - With cardiac disease
    - Mitral stenosis
    - Pulmonary venoocclusive disease
    - AVM, Pulmonary HTN
  - Without cardiac disease
    - Heiner syndrome
    - Idiopathic pulmonary hemosiderosis
    - Acute idiopathic pulmonary hemorrhage of infancy

- With pulmonary capillaritis
  - Goodpasture’s syndrome
  - Idiopathic pulmonary capillaritis
  - SLE
  - Wegener’s granulomatosis
  - Henoch-Schönlein purpura

Pulmonary Alveolar Proteinosis

- Pulmonary alveolar proteinosis (PAP) is a rare lung disorder of unknown etiology characterized by alveolar filling with flocular material that stains positive using the periodic acid-Schiff (PAS) method and is derived from surfactant phospholipids and protein components.
- May be related to granulocyte-macrophage colony-stimulating factor (GM-CSF) antibodies or GM-CSF deficiency.
- Due to a defect in the homeostatic mechanism of either the production of surfactant or the clearance by alveolar macrophages and the mucociliary elevator.
- Rx: Whole lung lavage, Inhaled GM-CSF (if caused by antibodies to GM-CSF)

Surfactant Metabolism Dysfunction Disorders

- Due to surfactant mutation disorders
  - Surfactant proteins B and C
  - ABCA3
  - Mutations in thyroid transcription factor 1 (TTF1)
  - Lysinuric protein intolerance
Any child with cough and/or tachypnea lasting more than >3 months should be evaluated for possible ILD

- Most laboratory tests are rare diagnostic but they are useful to exclude other diagnoses

Which of the following is the MOST diagnostic test in this case?

1. Chest X-ray
2. Chest CT
3. Quantitative Immunoglobulins
4. Panel for collagen vascular diseases
5. Bronchoalveolar lavage
6. Sweat test
7. Lung Biopsy

ILD: Imaging Studies

- Plain chest X-rays to start
- High resolution CT (HRCT) with thin sections (1 mm) is the best modality
ILD: Diagnostic Studies

- **Pulmonary Function Tests**
  - Restrictive pattern with decreased lung volumes, decreased lung compliance and markedly decreased diffusing capacity

- **Bronchoalveolar Lavage**
  Able to confirm only a few disorders but useful to rule out others (e.g. hemorrhage)

- **Lung Biopsy**: it’s the most definitive of the studies.
  Video Assisted Thoracoscopic biopsy is becoming the method of choice

ILD: Treatment & Outcome

- Depends on classification
- Supportive care
- Long-term oxygen
- Steroids (oral and/or IV)
- Hydroxychloroquine
- Chemotherapy (Azathioprine, Methotrexate; cyclophosphamide; GM-GSF
- Lung transplantation

**OUTCOME:**
Mortality approximately 30%
Increased mortality if pulmonary HTN present

Questions?