Respiratory Disorders

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Objectives

- Identify and distinguish causes of severe upper airway infections in children
- Discuss the presentation and treatment of a child with cystic fibrosis
- Know the typical presentation and treatment of pneumothoraces
- Distinguish foreign body aspiration from asthma
Case 1:

A 3 year old boy is seen for chronic wet-sounding cough. Growth has been poor. There has been partial improvement with antibiotics. Albuterol does not help.

- What is your next intervention?
- What is the likely diagnosis?
- How will this child’s condition be treated?
- What other organ systems are involved in his disease process?
Cystic Fibrosis

- Autosomal recessive condition
- CFTR (cystic fibrosis transmembrane conductance regulator) dysfunction
  - More than 2000 mutations identified
- Principal manifestations
  - Recurrent respiratory tract infections
  - Failure to thrive/Malnutrition
  - Meconium ileus (15-20% of cases)
- Infants may present with severe malnutrition, low sodium/chloride, hypoproteinemina
Variable Presentation

- Recurrent pneumonia
- Meconium ileus
- Failure to thrive
- Rectal prolapse
- Malnutrition
- Steatorrhea
- Cholelithiasis
- Pneumothorax
- Nasal polyposis
- Clubbing
- Hemoptysis
- Chronic sinusitis
- Pancreatitis
- Diabetes
- Cirrhosis
- Intussusception
- ABPA
- Infertility
- CBAVD
- Hypertrophic osteoarthropathy
- Newborn screening
CF Diagnosis

- NBS programs for CF have been approved in 50 states
- More than 60% of new diagnosed by NBS
- Elevation of immunoreactive trypsinogen (IRT) as a marker of pancreatic injury
- Sweat chloride testing is gold standard
- CFTR mutation analysis indicated for all
CF: Extrapulmonary

- Exocrine pancreatic insufficiency
  - Steatorrhea, rectal prolapse, vitamin deficiency
- Distal intestinal obstruction syndrome
  - Meconium ileus equivalent
- Hepatobiliary disease
  - May present with prolonged jaundice
- CF related diabetes
  - Increasing incidence with age
- Infertility
  - 98% of males with obstructive aspermia
Exocrine Pancreatic Insufficiency (PI)

- Obstruction of pancreatic ducts by inspissated secretions
- Damage to acinar cells, autodigestion
- 60% of CF infants have PI at birth
- Eventually 85-90% develop PI
- Leads to malabsorption of fat, protein, decreased bicarbonate secretion
Diagnosis of Exocrine Pancreatic Insufficiency

- Congenital bowel obstruction
- Steatorrhea/large stools
- Failure to thrive
- Rectal prolapse
- Diagnosis confirmed with
  - Fecal elastase or coefficient of fat absorption
Nutritional Management

- Fat soluble vitamins (A, D, E, K)
- 1/8 tsp of salt daily for infants
- Pancreatic enzyme replacement
  - 1000-2500 units lipase/kg/meal
- Adequate calories/high fat diet
  - 1.5X RDA for calories, may need oral or enteral supplementation
Better Nutrition = Better Lung Function

Median FEV₁ Percent Predicted vs. Median BMI Percentile for Children 6 to 19 Years in 2013

Goal: 50th percentile

Males
Females
CF Hepatobiliary Disease

- Infants may have conjugated hyperbilirubinemia or transaminitis
  - Complicated by malnutrition, prolonged TPN
- 25-30% develop biliary tract involvement
- Dilation of ducts with periportal fibrosis
- May develop portal hypertension and hepatic failure (very infrequent)
CF: Pulmonary

- **Early**
  - CXR may be normal or show bronchial wall thickening
  - BAL fluid may show increased neutrophilia even in infancy

- **Late**
  - Bronchial dilation leading to bronchiectasis, mucous plugging
  - Marked neutrophilic inflammation
CF Bronchiectasis
Pulmonary Disease in CF

- **Goal**: prevent bronchiectasis/structural lung disease and preserve function
- **Airway clearance/exercise**
- **Abnormal mucus properties**
  - DNase, hypertonic saline
- **Inflammation**
  - High dose ibuprofen, azithromycin
- **Antibiotics**: oral, inhaled, parenteral
Airway Clearance

Flutter

Acapella

Manual

Vest

Frequencer

IPV

And EXERCISE!
CFTR Modulators

- 2012: ivacaftor (Kalydeco) for patients with at least 1 CFTR gating mutation (&R117H): ↑lung function/weight and fewer exacerbations
- 2015: lumicaftor/ivacaftor (Orkambi) for patients >12 yrs & homozygous for F508del (50% of CF pop.)
- 2016 Orkambi approved for 6-11 yrs
Case 1

- The same 3 year old boy with chronic wet cough, sweat chloride < 10 mmol/L.
- What other diagnoses are in your differential?
Non-CF Bronchiectasis

- Pneumonia (usually unilobar)
- Chronic aspiration
- Foreign body (retained)
- Immunodeficiency
- Primary ciliary dyskinesia
Primary Ciliary Dyskinesia

- Consider with unexplained neonatal respiratory distress/hypoxemia
- Recurrent/chronic purulent otitis
- Chronic nasal congestion/rhinorrhea
- Heterotaxy, 50% with dextrocardia
- Diagnosis: clinical features, ciliary biopsy, genetic testing, nasal nitric oxide (low)
Primary Ciliary Dyskinesia

What’s Wrong?
Case 2: Barking child

The father of a 2 year old previously healthy boy calls you after his child awoke with a dry, harsh sounding cough.

1. What other history is important?
2. What conditions are you concerned about?
3. What management, if any, is indicated?
Croup

- **Infectious** (Viral) laryngotracheobronchitis
- Fever
- URI symptoms
- Barking cough
- Inspiratory stridor
- Hoarseness

- **Non-infectious** “spasmodic”
- Afebrile
- No other signs of illness
- Recurrent disorder
- Acute nocturnal onset
- Rapid resolution
Laryngotracheobronchitis

- Viral etiologies: parainfluenza type 1>2,3, RSV, influenza A & B, metapneumovirus
- Very common (1-6% of kids under 6 yrs)
- Rarely requires hospitalization for impending respiratory failure
- Usually resolves in 3-5 days
Croup Differential Diagnosis

- Bacterial tracheitis
- Epiglottitis
- Retropharyngeal abscess
- Peritonsillar abscess
- Foreign body aspiration
- Caustic ingestion
- Subglottic stenosis
- Vocal cord paralysis
- Anaphylaxis
- Tumor
- Hemangioma
- Diphtheria
LTB Management

- Minimize child anxiety
- Symptom control/prevent hospitalization
- Racemic epinephrine or L-epinephrine
  - 0.5 ml of 2.25% solution in NS (racemic)
  - 5 ml of 1:1000 (5 mg) L-epinephrine
- Systemic steroids
  - Dexamethasone 0.6 mg/kg x 1 (oral or IM)
- Nebulized steroids: Budesonide 2 mg
Croup

- AP and lateral x-rays may show subglottic narrowing: “steeple sign”
- Laboratory studies unhelpful
Croup
Case 2a

2 year old boy is brought to the ED with T of 40°C, toxic appearance, “brassy” cough, marked dyspnea, hoarseness, fails to respond to racemic epinephrine.

1. Now what are you worried about?
2. How should you proceed?
Emergent Airway Infections

- **Bacterial Tracheitis**
  - Rapid progression
  - Often evolves from croup (biphasic illness)
  - *S. aureus, S. pyogenes*

- **Supraglottitis (epiglottitis)**
  - Seen rarely in young children since Hib vaccine
  - Now seen more in older children with nontypeable *H. influenzae, S. pyogenes*
Bacterial Tracheitis

- Assemble airway team
- Prepare for intubation/bronchoscopy in a controlled setting (OR)
- Prompt coverage with parenteral antibiotics (cover S.aureus)
- May require extended period of mechanical ventilation
Bacterial Tracheitis
Supraglottitis

- Maintain child in upright position in parent’s arms or lap
- Assemble airway team
- If concern for loss of airway, then proceed with endoscopy/intubation in controlled setting rather than obtaining neck films
- Quick resolution following intubation, antibiotics, steroids
Supraglottitis

Smalhout B  *The Suffocating Child*
1980
Supraglottitis
Case 3: “His asthma isn’t getting any better”

18 month old boy presents with a 3 day history of cough and wheeze. No antecedent URI or fever. He has diffuse wheezing which responds poorly to albuterol.

- What further evaluation is indicated?
- What are causes of chronic wheezing in children?
Foreign Body Aspiration

- More common in toddlers
- 50% of children with foreign body aspiration have no witnessed history
- Symptoms include sudden onset of cough, choking, wheezing or shortness of breath
- Usually no signs of infection (unless symptoms have persisted for weeks)
Foreign Body Aspiration

- Physical examination
  - May be normal
  - Unilateral wheezing or decreased aeration
  - Asymmetric thorax
  - +/- signs of respiratory distress
    - More likely with laryngeal foreign body
Evaluation of FB Aspiration

- Chest radiographs may demonstrate differential aeration
- Inspiratory/expiratory films can be performed in cooperative children
- Bilateral decubitus films in infants
- Airway fluoroscopy can show air trapping
- Bronchoscopy with any clinical suspicion
Foreign Body Aspiration
Foreign Bodies
Foreign Bodies
FB Aspiration Complications

- Severe airway inflammation
- Scarring/granulation tissue
- Recurrent pneumonia
- Bronchiectasis
- Hemoptysis
- Bronchopleural fistula
Differential: Chronic Wheeze

- Asthma
- Tracheobronchomalacia
- Foreign body aspiration
- Oral or gastric chronic aspiration
- Vascular ring
- Hemangioma
- Congenital malformation (CPAM)
Normal Trachea     Tracheomalacia
Primary Tracheomalacia

- Common
- Symptoms include stridor, wheeze, harsh cough, recurrent infections, reflex apnea
- Abnormal cartilage to membrane ratio
Case 4: My chest hurts...

15 year old presents with a 12 hour history of sudden left sided upper chest/shoulder pain, that began during basketball practice.

- What is the usual presentation of a pneumothorax?
- What is the appropriate management of pneumothoraces?
Pneumothorax

- Spontaneous
  - Apical blebs
  - More common in young, tall, thin males
- Secondary
  - Underlying pulmonary disease (asthma, cystic fibrosis, pneumonia)
  - Trauma
Pneumothorax

- **Symptoms:**
  - Sudden onset
  - Chest pain, worse with inspiration (pleuritic)
  - Shortness of breath

- **Signs:**
  - Tachypnea
  - Decreased aeration
  - Hypoxemia
  - Tension pneumothorax: unilateral absent BS, tracheal deviation, bradycardia, ↓BP
Pneumothorax Management

- Depends on severity
- Supplemental oxygen (FiO2 1.0)
- For worsening or persistent symptoms, large air collection or underlying disease
  - Thoracentesis
  - Chest tube drainage
  - Bleb resection
  - Pleurodesis
- High recurrence rate (>20%): ipsilateral and contralateral
QUESTIONS ???