Respiratory Disorders 1 (or Pulmonary Potpourri)

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Objectives

- Identify causes of stridor in children
- Formulate a differential diagnosis for noisy breathing
- Understand indications for further evaluation of children with suspected airway or pulmonary malformations
Lecture Content

- Noisy Breathing & Stridor
- Laryngeal Disorders
- Vascular Airway Compression
- Pulmonary Malformations
Noisy Breathing

- Stridor
  - Inspiratory, often high pitched noise
  - Heard best over face, neck
- Wheeze
  - Primarily expiratory
  - Monophonic and transmitted
Stridor- By Age

- Neonatal: congenital and acquired
- Toddlers: infectious vs. non-infectious
  - Croup
  - Laryngeal foreign body
  - Vascular compression
- School Aged/Adolescent
  - Vocal cord dysfunction
  - Supraglottitis
Neonatal Stridor

- Laryngomalacia
- Vocal cord paresis
- Subglottic stenosis (congenital or acquired)
- Subglottic/tracheal cysts/web
- Hemangiomas
- Vascular compression of the airways
Laryngeal Anatomy
The Floppy Airway

- Laryngomalacia—usually inspiratory stridor heard best over the face/neck
- Tracheobronchomalacia—primarily expiratory monophonic wheeze
- Noise may be accentuated in the supine position, with activity or illness
The Floppy Airway

- “Raspy” breathing
- Often relieved with position changes
- No response to bronchodilator
- Resolution by 18-24 months in most cases
Severe Laryngomalacia
Laryngomalacia
Vocal Cord Paresis

- **Unilateral**
  - Hoarse cry
  - Inspiratory stridor
  - More common on left (recurrent laryngeal)
  - Birth trauma
  - Surgical- PDA ligation

- **Bilateral**
  - Weak cry
  - May have stridor
  - Aspiration
  - Traumatic intubation
  - Chiari II malformation
  - Prolonged NGT
Anterior Commissure Web
Interarytenoid Web
Severe Subglottic Stenosis

Congenital
Post-intubation Stenosis

Grade III Stenosis
Neonatal Stridor: Who Needs a Scope?

- Feeding difficulties
- Poor weight gain
- Apnea
- Cyanosis
- Progressively increasing work of breathing
- Small minority of cases require surgery
Vascular Rings

- Double aortic arch
  - Encircles trachea and esophagus
- Right sided arch
  - With aberrant lt. subclavian or mirror image branching
- Aberrant right subclavian artery
- Anomalous innominate artery
- Pulmonary artery sling
Vascular Ring: Presentation

- Persistent or increasing stridor/wheeze
- Feeding intolerance (choking/gagging)
- Also may present with
  - Apnea/cyanotic spells
  - Tachypnea/respiratory distress/exertional sx
  - Failure to thrive
  - Recurrent pneumonia
  - Dysphagia
Double Aortic Arch
Innominate Artery Compression
Right Aortic Arch
Double Aortic Arch
Double Aortic Arch
Congenital Pulmonary Malformations

- CPAMs are often detected by prenatal ultrasound
- Presence of large CPAM and fetal hydrops is ominous
- Even regressing lesions should be evaluated after birth
CPAM Presenting Symptoms

- Unexplained neonatal respiratory distress
- Noisy breathing
  - Persistent wheeze/stridor
- Recurrent pneumonia/infections
- Swallowing problems
- Persistent infiltrate on radiographs
Congenital Thoracic Malformations

- Bronchogenic cyst
  - Foregut cyst
  - Duplication cyst
- Lobar overinflation
- Cystic adenomatoid malformation
- Bronchopulmonary sequestration
Bronchogenic Cyst

- Abnormal budding that occurs at various points during gestation
- Early = mediastinal cysts
- Late = pulmonary cysts
- Size of cyst often related to presentation
Bronchogenic Cyst

- Mediastinal cysts may cause severe respiratory distress
- May contain multiple cell types (esophageal and respiratory origin)
- Secondary infection may develop
- Surgery is usually indicated
DOL #1
Fetal ultrasound showed left CLL
Noisy breathing since birth
Cyst contrast study repeated during bronchoscopy without ETT in place.
Large Hyperlucent Lung

- Lobar “emphysema”- actually overinflation
- LUL>RML>RUL>lower lobes
- Ball valve mechanism due to abnormal bronchus
- Occurs following airway development
- Surgery often indicated
Lobar Overinflation
Lobar Overinflation
Lobar Overinflation
Congenital Cystic Adenomatoid Malformation

- Communication with bronchial tree
- Usually unilobar (no particular location)
- Mix of cysts with adenomatous tissue
- 5 pathologic types
CCAMs

- Surgical resection indicated
- Potential for malignant transformation
Sequestrations

- Non-functioning lung tissue
- Supplied by systemic circulation
- Diagnose by CT/MRI (feeding vessel)
- Lower lobes most common
- Two types:
  - Intralobar
  - Extralobar
Sequestrations

- **Intralobar**
  - More common
  - Supplied by pulmonary & systemic circulation
  - Late presentation (30% are < 10yrs)
  - Recurrent pneumonia

- **Extralobar**
  - Rare
  - More common in males
  - Systemic arterial and venous supply
  - Neonatal presentation
  - Other anomalies
1st day of life

Age 3 weeks