Approach to the Wheezing Child

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Children's Hospital

The Problem: “Wheezing”

- Very common
- Multiple sounds and descriptions
- Broad differential

Educational Goals

- List causes of wheezing in children
- Outline a strategy for the initial assessment and treatment of a “typical” child with wheezing
- Outline a strategy for addressing sub-optimal responders
- Recognize wheezing child “red flags”

Uncommon Causes of Childhood Wheezing

- Large airway obstruction (congenital)
  - Vascular ring
  - Tracheomalacia
  - Tracheal stenosis
- Large airway obstruction (acquired)
  - Foreign body
  - Mediastinal mass
  - Endobronchial tumor
- Abnormal GI - airway anatomy
### Uncommon Causes of Childhood Wheezing

- Persistent airway infection states
  - Cystic fibrosis
  - Immunoglobulin deficiency
  - Dysmotile cilia syndromes
- Cardiac failure

### Common Causes of Childhood Wheezing

- Recurrent
  - ASTHMA, ASTHMA, ASTHMA
  - “Infantile asthma” & post-RSV wheezing
  - Asthma complicated by persistent triggers
  - Aspiration disorders (Infants)
  - Vocal Cord Dysfunction (Older child)

### Common Causes of Childhood Wheezing

- Single episode
  - VIRAL
  - “First event” asthma presentation

### Goals of Initial Evaluation

- Screen for “red flags”
- Assess for symptom pattern c/w asthma
- Screen for alternate clinical problems which might cause wheeze, or make asthma control difficult
### Tools for Evaluation: a Good History

<table>
<thead>
<tr>
<th>Description of onset</th>
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<tbody>
<tr>
<td>Present since birth</td>
</tr>
<tr>
<td>Onset in infancy</td>
</tr>
<tr>
<td>Onset in later childhood</td>
</tr>
</tbody>
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<table>
<thead>
<tr>
<th>Description of wheeze</th>
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<tbody>
<tr>
<td>Inspiratory v.s. expiratory</td>
</tr>
<tr>
<td>Intermittent v.s. daily</td>
</tr>
<tr>
<td>Associated symptoms</td>
</tr>
<tr>
<td>Rattling</td>
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<tr>
<td>Cough</td>
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<tr>
<td>Retractions</td>
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<table>
<thead>
<tr>
<th>Triggers</th>
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<tbody>
<tr>
<td>URI’s, cold air, exercise, allergen exposure, smoke exposure</td>
</tr>
<tr>
<td>Eating, lying down</td>
</tr>
<tr>
<td>“No apparent reason”</td>
</tr>
<tr>
<td>It never changes</td>
</tr>
<tr>
<td>Began following choking episode</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Past Medical History</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonatal history</td>
</tr>
<tr>
<td>State of birth (CF screening?)</td>
</tr>
<tr>
<td>Major health issues</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Family history</th>
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<tbody>
<tr>
<td>Social history (smoke exposure, pets)</td>
</tr>
</tbody>
</table>
Tools for Evaluation: a Good History

- ROS
  - GERD markers/ feeding difficulty
  - Chronic nasal discharge
  - Eczema
  - Poor growth
  - Chronic diarrhea

Tools for Evaluation: a Good Physical Exam

- Presence of:
  - Non-physiologic heart murmurs
  - Liver enlargement
  - Clubbing

Tools for Evaluation: a Good Physical Exam

- General growth/health
- Upper respiratory infection signs
- Upper respiratory allergy signs
- Pulmonary exam
  - High pitched versus rhonchorous sounds
  - Variability of sound
  - Location of sound

*Persistent unilateral location

Initial Testing: CXR

- Typical: normal, peribronchial thickening, hyperinflation
- Atypical: Asymmetric inflation, large heart, dense or extensive infiltrates
**Initial Testing: PFT’s**

- Obstructive changes on initial spirometry that improve following a bronchodilator establishes an asthma diagnosis
- Spirometry can also help estimate asthma severity

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**PFT Testing Limitations**

- Most asthmatics develop symptoms prior to age 5 years.
- Most cannot perform quality spirometry until age 6 years.
- Childhood asthmatics usually have normal spirometry when “well”

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**Initial Assessment**

- **Red Flags:**
  - Initial symptoms associated with choking episode
  - Poor growth, clubbing, chronic diarrhea
  - Findings suggestive of heart failure
  - Monotonous wheezing, truly “present from birth”
  - Atypical CXR

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**Initial Approach:**

**Trial of Asthma Therapy**

- Bronchodilators*
- Oral steroids*
- (Controller therapy)

* Failure to respond to combination suggests problem is NOT asthma
What about Controller Therapy Trial?

- Clear response/improvement to controller therapy can help establish a diagnosis of asthma in a child with typical symptoms

Controller Therapy Trial Limitations

A failure of improvement may indicate:
- Medication chosen was not “potent” enough
- Montelukast trials
- Low dose ICS trials
- Medication was not used long enough
- Medication was not actually inhaled

ICS Delivery in Young Children

Asthma Therapy Responders: Ongoing Management

- Establish clear plan for response to acute symptoms
- If symptoms are frequent (or repeatedly severe) begin controller therapy
- Titrate controller therapy according to disease severity/level of control
### Approach to Initial Responders Who are in Poor Control

- Review adequacy of controller dosing
- Review medication usage
  - Technique problems
  - Compliance problems
- In older child, obtain PFT’s

### Advanced Approach: Infants

- Trial of GERD Therapy (BID PPI)
- Videoswallow study
- UGI?
- Sweat test
- Allergy testing

### Approach to Ongoing Poor Control, and “Partial Responders”

- Advanced Approach: Look for and address problems that persistently irritate airways:
  - Smoke exposure
  - Infants: dysphagia, GERD, food allergy
  - Older children: Allergen exposure, chronic sinusitis, GERD

### Advanced Approach: Older Child

- Trial of GERD Therapy (up to BID PPI)
- Prolonged antibiotics for sinus disease
- Sinus CT
- Allergy testing
- Pulmonary function testing
Treatment Red Flags – Consider Referral

- Failure to respond to beta-agonist and oral steroid trial
- Failure to achieve control despite:
  - Adequate controller therapy
  - Addressing common complicating disorders
- Persistently abnormal lung function studies

Approach to the Wheezing Child

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Summary

- Wheezing is common in childhood
- Recurrent wheezing is usually due to asthma
- Initial approach establishes whether the history, and therapeutic response, is consistent with asthma
- If asthma therapy is unsuccessful, look for common inflammation-contributing co-morbidities
- Watch for red flags - if found, consider referral

Nonbronchospasm Related Wheezing

- When symptoms are atypical, consider other causes for persistent wheezing:
  - Wheezing that is poorly responsive to medical treatment
  - Wheezing that returns after withdrawal of medications
  - Unilateral wheezing
  - Barky/ Croupy cough, especially in a young infant
  - Symptoms that are worse during eating
  - Reflex apnea
  - Recurrent pneumonias or infections
### Nonbronchospasm Related Wheezing

<table>
<thead>
<tr>
<th>Anatomic:</th>
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<tbody>
<tr>
<td>✓ Primary Tracheomalacia</td>
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<tr>
<td>✓ Secondary Tracheomalacia</td>
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<tr>
<td>✓ Tracheal stenosis</td>
</tr>
<tr>
<td>✓ Tracheal masses</td>
</tr>
<tr>
<td>✓ Bronchomalacia</td>
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<tr>
<th>Neurologic:</th>
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<tr>
<td>✓ Vocal cord paralysis</td>
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<tr>
<td>✓ Vocal cord dysfunction</td>
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<tr>
<th>Other:</th>
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<tr>
<td>✓ Airway Foreign Body</td>
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### Normal Airway

![Normal Airway](image)

### Diagnostic Evaluation

- Nasopharyngoscopy allows evaluation to the level of the larynx and limited evaluation of the subglottic airway
- Rigid bronchoscopy allows evaluation of the tracheal and bronchial airways
- Imaging may be useful in some cases
  ✓ 3-D Reconstructions

### Tracheomalacia

- Classification system described by Benjamin in 1984:
  I. Primary tracheomalacia
  II. Secondary tracheomalacia
    A. Tracheoesophageal fistula and esophageal atresia
    B. External compression (vascular/cardiac/neoplastic)
    C. Boney thorax abnormality (i.e. pectus excavatum)
    D. Dyschondroplasia
### Tracheomalacia and Tracheobronchomalacia

- Flaccidity of the tracheal/bronchial cartilage leading to collapse of the airway
  - Collapse occurs during expiration
  - More pronounced with increased airflow
  - May be primary or secondary
- Cartilaginous to membranous ratio may be significantly decreased
- Incidence of primary tracheomalacia is about 1 in 2100

### Tracheomalacia

- **Symptoms:**
  - Tracheal wheeze/Expiratory wheeze or stridor
  - Harsh, barking cough/“Brassy” cough
  - Failure to thrive
  - Increasing respiratory distress with growth
  - Transmitted vibration through the back
  - Difficulty clearing secretions
  - Symptoms exacerbated with viral infections

### Primary Tracheomalacia

- **Diagnosis:** Bronchoscopy

### Primary Tracheomalacia

- **Diagnosis:** CT Chest
### Primary Tracheomalacia Treatment

- Supportive care
  - Disease resolves between ages 2-5 years
- Medical treatment
  - Only to help manage concomittant issues
- CPAP/ BiPAP
- Tracheotomy
- Airway stenting

### Secondary Tracheomalacia Vascular Compression

<table>
<thead>
<tr>
<th>I. Aberrant innominate artery</th>
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<tbody>
<tr>
<td>II. Vascular Rings</td>
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<tr>
<td>A. Double aortic arch</td>
</tr>
<tr>
<td>B. Right aortic arch with left ligamentum arteriosum</td>
</tr>
<tr>
<td>III. Pulmonary artery sling</td>
</tr>
<tr>
<td>IV. Aberrant right subclavian artery</td>
</tr>
<tr>
<td>V. Congenital cardiac defects</td>
</tr>
</tbody>
</table>

### Secondary Tracheomalacia Tracheoesophageal Fistula

- TEF occurs in 1 in 3000 to 5000 live births
- Often involves trachea and both main bronchi
- Symptoms are the same as for primary tracheomalacia
- Associated tracheomalacia often remains after repair of TEF due to cartilage deficiency in the area

### Secondary Tracheomalacia Vascular Compression

- Symptoms:
  - Tracheal wheezing or stridor 100%
  - Chronic cough 14-75%
  - Recurrent pneumonia 47-56%
  - Dysphagia 14-25%
  - Reflex apnea 30-70%
  - Failure to thrive 11%

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Reflex Apnea

- “Dying Spells” or Acute Life Threatening Events (ALTEs)
- Proposed that these events are reflex respiratory arrests due to irritation of the compressed trachea
- May occur when food bolus passes through esophagus, causing the posterior membranous trachea to bulge forward and narrow the airway further


Aberrant Innominate Artery

- Leftward origin of the innominate artery (brachiocephalic trunk) is common variant of normal anatomy
- Bronchoscopic findings:
  - Pulsatile anterior compression of the trachea, worse on the right side of the trachea
  - Right radial pulse diminishes with anterior pressure by bronchoscope


Aberrant Innominate Artery

- Expectant Management
  - Humidification, Supplemental oxygen, treatment of infection
- Surgical Management- Aortopexy
  - Absolute Indication: Reflex apnea
  - Relative Indications: Repeated infection with poor medical response, Exercise intolerance
- Tracheomalacia may still be present after compression is relieved
Double Aortic Arch

- Bifurcation of the ascending aorta that surrounds the trachea and esophagus, then rejoins to form the descending aorta


Double Aortic Arch

- Symptoms begin by age 3 months
  - Stridor, worse with feeding
  - Dysphagia, especially to solids
  - Cyanosis
  - Recurrent respiratory infections
- Bronchoscopy shows “teardrop” appearance of tracheal lumen
- CT with contrast will show ring and which limb of aorta is dominant


Double Aortic Arch

- Surgical treatment requires division of nondominant arch with or without aortopexy
- Tracheomalacia can persist after surgical correction

Right-Sided Aortic Arch

With left descending aorta and left ligamentum arteriosum

With aberrant left subclavian artery and left ligamentum arteriosum

Pulmonary Artery Sling

• Occurs because of an anomalous origin of the left pulmonary artery

Pulmonary Artery Sling

• Typically produce early and severe symptoms
• Half of patients with pulmonary artery sling have associated tracheobronchial malformations
  ✓ Most common: Complete tracheal rings
• Treatment:
  ✓ Surgical division and reimplantation of the anomalous left pulmonary artery
  ✓ Correction of associated tracheal anomalies

Tracheal Stenosis

• Complete tracheal rings
  ✓ Normal trachea has membranous posterior wall
  ✓ Complete cartilaginous rings result in narrowing of trachea to varying degree
  ✓ May involve single ring to entire tracheal length
• Tracheal cartilaginous sleeve

### Tracheal Stenosis

- Symptoms may be present at birth or start after acute illness or other insult
  - Wheezing or stridor
  - Cough
  - Recurrent croup
  - Recurrent respiratory infections
  - Difficulty clearing secretions

### Complete Tracheal Rings

#### Endoscopy

![Endoscopy Images]

#### Radiography

![Radiography Images]
Complete Tracheal Rings Treatment

- Surgical management is indicated for:
  - Significant respiratory symptoms
  - ALTEs
  - Need for intubation
  - Repeated respiratory infection
  - Failure to wean from ventilatory support
- Operative management is typically pursued when stenotic segment is less than 40% of normal tracheal diameter

Tracheal Masses

- Primary tracheal and bronchial tumors are rare
  - 64-90% are benign
  - Benign tumors more commonly in the proximal trachea
  - Malignant tumors more commonly in the distal trachea
- Symptoms:
  - Wheezing
  - Stridor
  - Cough
  - Dyspnea
  - Hemoptysis

Treatment Options

- Patch tracheoplasty
- Cartilage
- Pericardium
- Tracheal sleeve resection
- Slide tracheoplasty


Complete Tracheal Rings Treatment

- Treatment Options
  - Patch tracheoplasty
  - Cartilage
  - Pericardium
  - Tracheal sleeve resection
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Complete Tracheal Rings Treatment

- Treatment Options
  - Patch tracheoplasty
  - Cartilage
  - Pericardium
  - Tracheal sleeve resection
  - Slide tracheoplasty


Tracheal and Bronchial Masses

- Benign:
  - Papilloma
  - Fibroma
  - Hemangioma
  - Hamartoma

- Malignant:
  - Malignant fibrous histiocytoma
  - Mucoepidermoid carcinoma
  - Adenoid cystic carcinoma
  - Rhabdomyosarcoma
  - Squamous cell carcinoma
  - Bronchogenic carcinoma

### Neurologic Causes of Wheezing

<table>
<thead>
<tr>
<th>• Vocal Cord Paralysis</th>
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<tbody>
<tr>
<td>✓ Can be congenital or acquired</td>
</tr>
<tr>
<td>✓ Unilateral or Bilateral</td>
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<table>
<thead>
<tr>
<th>• Vocal Cord Dysfunction</th>
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</thead>
<tbody>
<tr>
<td>✓ Paradoxical vocal cord movement</td>
</tr>
<tr>
<td>✓ Paradoxical vocal cord dysfunction</td>
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### Vocal Cord Paralysis

<table>
<thead>
<tr>
<th>• Accounts for 10% of congenital laryngeal lesions</th>
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<table>
<thead>
<tr>
<th>• Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ Weak or abnormal cry</td>
</tr>
<tr>
<td>✓ Stridor or wheezing</td>
</tr>
<tr>
<td>✓ Dysphagia</td>
</tr>
<tr>
<td>✓ Chronic cough or cough during eating</td>
</tr>
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</table>

### Vocal Cord Dysfunction

<table>
<thead>
<tr>
<th>• The larynx exhibits paradoxical vocal cord motion, with vocal cord adduction during inspiration</th>
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</thead>
</table>

<table>
<thead>
<tr>
<th>• Symptoms:</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ Wheezing</td>
</tr>
<tr>
<td>✓ Dyspnea</td>
</tr>
<tr>
<td>✓ Cough</td>
</tr>
<tr>
<td>✓ Throat tightness</td>
</tr>
<tr>
<td>✓ Shortness of breath</td>
</tr>
<tr>
<td>✓ Exercise intolerance</td>
</tr>
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<table>
<thead>
<tr>
<th>• Common in asthmatics</th>
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### Vocal Cord Paralysis Etiology

<table>
<thead>
<tr>
<th>• Congenital</th>
</tr>
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<tbody>
<tr>
<td>✓ Hydrocephalus</td>
</tr>
<tr>
<td>✓ Arnold-Chiari Malformation</td>
</tr>
<tr>
<td>✓ Myasthenia Gravis</td>
</tr>
<tr>
<td>✓ Cardiovascular anomalies</td>
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<table>
<thead>
<tr>
<th>• Acquired</th>
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<tbody>
<tr>
<td>✓ Trauma</td>
</tr>
<tr>
<td>✓ Infectious</td>
</tr>
<tr>
<td>✓ Supranuclear lesions (i.e. MS)</td>
</tr>
<tr>
<td>✓ Iatrogenic</td>
</tr>
<tr>
<td>✓ Idiopathic</td>
</tr>
</tbody>
</table>
Vocal Cord Dysfunction

Etiology

- Cortical injury
  ✓ Stroke
  ✓ ALS
- Brainstem compression
  ✓ Chiari malformation
- Conversion disorder
- Malingering
- Irritant-induced
  ✓ Solvents, Amonia, Smoke

Demographics

- Mean age at presentation: about 14 years
- 82-86% of patients are female
- Patients tend to be high functioning, participate in organized sports, have high level of social and life stressors
- Exercise often induces symptoms
- High association with GERD

Precipitating Factors:

- Exercise
  ✓ Especially when there is poor response to bronchodilators
- Psychological Conditions
  ✓ PTSD, Anxiety Disorder, Depression
- Irritants
- Rhinosinusitis
- GERD
- Medication use
  ✓ Phenothiazines

Diagnosis

- Can be difficult due to episodic nature
- Laryngoscopy during symptoms shows pathognomonic adduction of the vocal folds during inspiration
- Symptoms resolve when patient is distracted or asleep
- Wheezing is most prominent over the larynx and less notable in lung fields
- PFT’s are inconclusive due to high correlation with asthma

References:
### Vocal Cord Dysfunction Treatment

<table>
<thead>
<tr>
<th>• Acute Phase:</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ Heliox</td>
</tr>
<tr>
<td>✓ CPAP</td>
</tr>
<tr>
<td>✓ Anxiolytic medications</td>
</tr>
<tr>
<td>✓ General anesthesia</td>
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</tbody>
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<table>
<thead>
<tr>
<th>• Long-Term Management:</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ Education</td>
</tr>
<tr>
<td>✓ Avoidance of known triggers</td>
</tr>
<tr>
<td>✓ Breathing for vocal cord dysfunction</td>
</tr>
<tr>
<td>✓ Relaxation of muscles of neck, shoulder, and chest</td>
</tr>
<tr>
<td>✓ Psychotherapy</td>
</tr>
</tbody>
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### Airway Foreign Body

<table>
<thead>
<tr>
<th>• Acute Phase- Choking or gagging episode</th>
</tr>
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</table>

<table>
<thead>
<tr>
<th>• Asymptomatic Interval- Foreign body becomes lodged and airway reflexes become fatigued</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>• Complications- Erosion, obstruction or infection occurs resulting in symptoms</th>
</tr>
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</table>

### Airway Foreign Body

<table>
<thead>
<tr>
<th>• Presentation is variable</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ There is usually a choking or gagging event, however this may not be recognized or reported (present in about 80% of cases)</td>
</tr>
<tr>
<td>✓ Other symptoms include cough, wheezing, recurrent croup</td>
</tr>
<tr>
<td>✓ Symptoms may respond to “asthma” treatment, but recur after medications are stopped</td>
</tr>
<tr>
<td>✓ 85% of patients are younger than 5 years of age</td>
</tr>
</tbody>
</table>

### Airway Foreign Body Diagnosis

<table>
<thead>
<tr>
<th>• Radiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ CXR may be normal, show radio-opaque foreign body, or hyperinflation</td>
</tr>
<tr>
<td>✓ Inspiratory and Expiratory views or Decubitus views</td>
</tr>
<tr>
<td>✓ CT with fine (&lt;3 mm) cuts</td>
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<table>
<thead>
<tr>
<th>• Bronchoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>✓ Gold standard for diagnosis</td>
</tr>
<tr>
<td>✓ High clinical suspicion with normal imaging does not rule out foreign body</td>
</tr>
</tbody>
</table>
Conclusions

- When symptoms are atypical, consider other causes for persistent wheezing
- ALTE’s raise concern for anatomic compression of the trachea, such as vascular ring or sling
- Tracheal narrowing may not become evident until exacerbated by a viral infection
- Vocal cord dysfunction often presents like poorly controlled asthma
- Airway foreign body may present as unilateral wheezing or as asthma that gets worse after medications are stopped