Approach to the Wheezing Child

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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

- Description of onset
  - ✓ Present since birth
  - ✓ Onset in infancy
  - ✓ Onset in later childhood

Tools for Evaluation: a Good History

- Triggers
  - ✓ URI's, cold air, exercise, allergen exposure, smoke exposure
  - ✓ Eating, lying down
  - ✓ "No apparent reason"
  - ✓ It never changes
  - ✓ Began following choking episode

Tools for Evaluation: a Good History

- Description of wheeze
  - ✓ Inspiratory v.s. expiratory
  - ✓ Intermittent v.s. daily
- Associated symptoms
  - ✓ Rattling
  - ✓ Cough
  - ✓ Retractions

Tools for Evaluation: a Good History

- Past Medical History
  - ✓ Neonatal history
  - ✓ State of birth (CF screening?)
  - ✓ Major health issues
- Family history
- Social history (smoke exposure, pets)

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

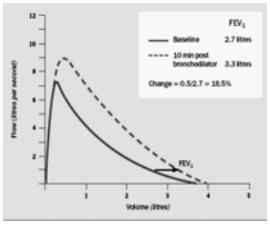
- General growth/health
- Upper respiratory infection signs
- Upper respiratory allergy signs
- Pulmonary exam
  - ✓ High pitched versus rhonchorous sounds
  - ✓ Variability of sound
  - ✓ Location of sound*

*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

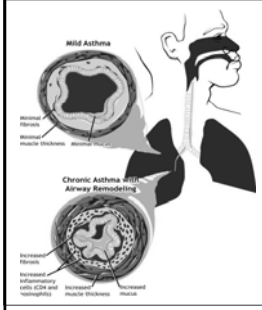
Initial Assessment Red Flags:

- Initial symptoms associated with choking episode
- Poor growth, clubbing, chronic diarrhea
- Findings suggestive of heart failure
- Monotonous wheezing, truly "present from birth"
- Atypical CXR

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"

Initial Approach: Trial of Asthma Therapy



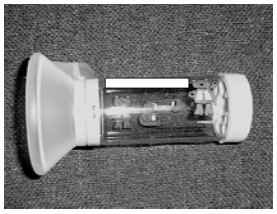
- Bronchodilators*
- Oral steroids*
- (Controller therapy)

* 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**

ICS Delivery in Young Children



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**

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

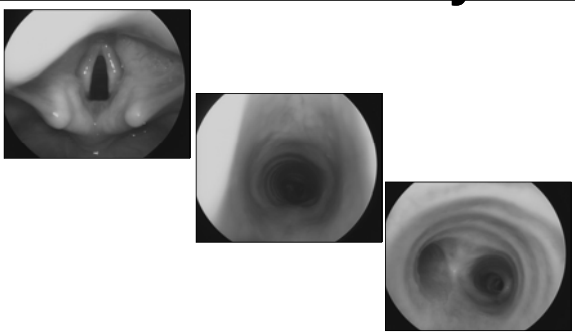
Nonbronchospasm Related Wheezing

- 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

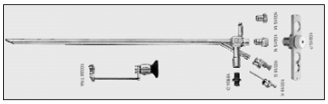
- **Anatomic:**
  - ✓ **Primary Tracheomalacia**
  - ✓ **Secondary Tracheomalacia**
  - ✓ **Tracheal stenosis**
  - ✓ **Tracheal masses**
  - ✓ **Bronchomalacia**
- **Neurologic:**
  - ✓ **Vocal cord paralysis**
  - ✓ **Vocal cord dysfunction**
- **Other:**
  - ✓ **Airway Foreign Body**

Normal Airway



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)

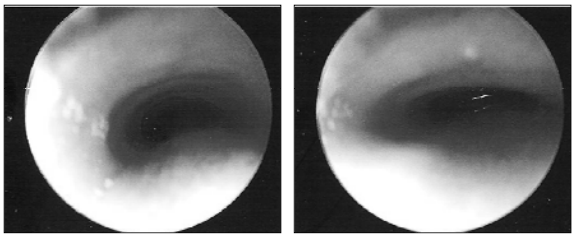
C. Boney thorax abnormality (i.e. pectus excavatum)

D. Dyschondroplasia

Tracheomalacia and Tracheobronchomalacia

- Flaciddity 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

Primary Tracheomalacia Diagnosis- Bronchoscopy

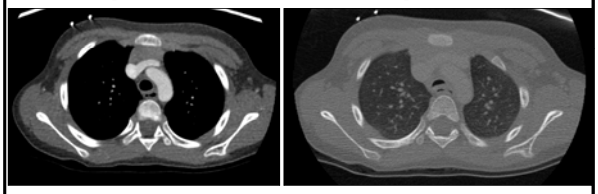


Tracheomalacia

- 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- CT Chest

Inspiratory Expiratory



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

I. Aberrant innominate artery
II. Vascular Rings
  A. Double aortic arch
  B. Right aortic arch with left ligamentum arteriosum
III. Pulmonary artery sling
IV. Aberrant right subclavian artery
V. Congenital cardiac defects

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%

1. McLaughlin R, Wetmore R, Tavill M, Gaynor J, Spray T. Vascular Anomalies Causing Symptomatic Tracheobronchial Compression. *Laryngoscope*. 109:312-319, February 1999.
2. Gormley P, Colreavy M, Patil N, Woods A. Congenital vascular anomalies and persistent respiratory symptoms in children. *International Journal of Pediatric Otorhinolaryngology*. 51: 23-21, 1999.

Reflex Apnea

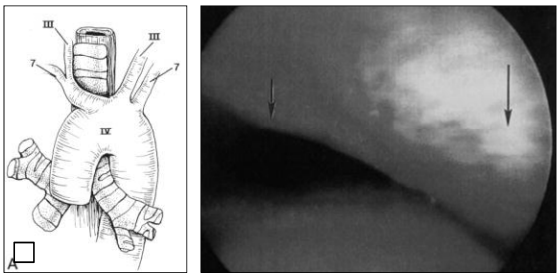
- **"Dying Spells" or Acute Life Threatening Events (ALTEs)**
- **Proposed that these events are reflex respiratory arrests due to irritation of the compressed trachea**
- **May occur when food bolus passes through esophagus, causing the posterior membranous trachea to bulge forward and narrow the airway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

Fearon B, Shortreed R. Tracheobronchial compression by congenital cardiovascular anomalies in childhood: syndrome of apnoea. *Annals of Otology Rhinology, and Laryngology*. 72: 949-969, 1963.

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



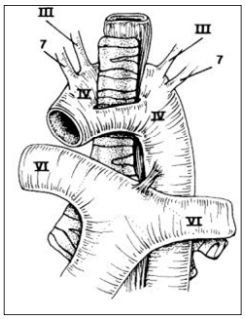
McLaughlin R, Wetmore R, Tavill M, Gaynor J, Spray T. Vascular Anomalies Causing Symptomatic Tracheobronchial Compression. *Laryngoscope*. 109:312-319, February 1999.

Aberrant Innominate Artery Treatment

- **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**



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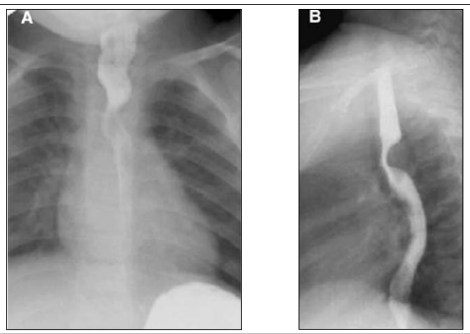
Double Aortic Arch



Hernanz-Schulman, M. Vascular Rings: A Practical Approach to Imaging Diagnosis. *Pediatric Radiology*. 35: 961-979, 2005.

Double Aortic Arch

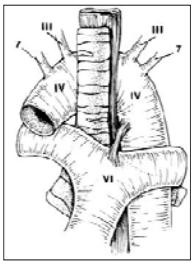
- **Symptoms begin by age 3 months**
  - ✓ **Stridor, worse with feeding**
  - ✓ **Dysphagia, especially to solids**
  - ✓ **Cyanosis**
  - ✓ **Recurrent respiratory infections**
- **Bronchoscopy shows "teardrop" appearance of tracheal lumen**
- **CT with contrast will show ring and which limb of aorta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

Double Aortic Arch Treatment

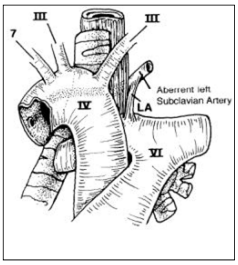
- **Surgical treatment requires division of nondominant arch with or without aortopexy**
- **Tracheomalacia can persist after surgical correction**

Right-Sided Aortic Arch

With left descending aorta and left ligamentum arteriosum



With aberrant left subclavian artery and left ligamentum arteriosum

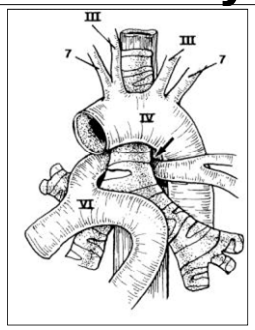


McLaughlin R, Wetmore R, Tavill M, Gaynor J, Spray T. Vascular Anomalies Causing Symptomatic Tracheobronchial Compression. *Laryngoscope.* 109:312-319, February 1999.

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

Pulmonary Artery Sling



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

McLaughlin R, Wetmore R, Tavill M, Gaynor J, Spray T. Vascular Anomalies Causing Symptomatic Tracheobronchial Compression. *Laryngoscope.* 109:312-319, February 1999.

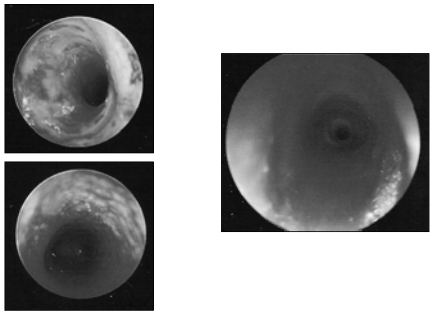
Tracheal Stenosis

- 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

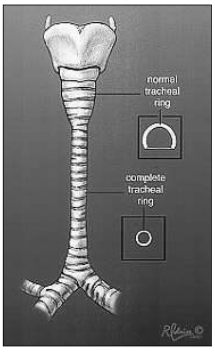
Tracheal Stenosis

- 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

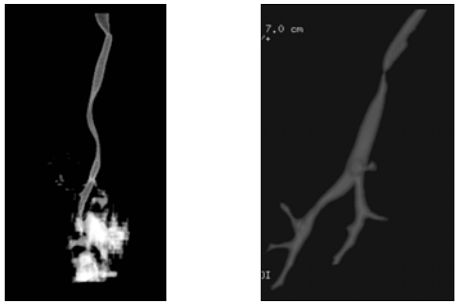


Complete Tracheal Rings



- Often associated with cardiovascular and other extrathoracic anomalies
  - ✓ Pulmonary artery sling
  - ✓ VATER/ VACTERL
  - ✓ ASD/ VSD
  - ✓ TAVPR

Complete Tracheal Rings Radiography



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

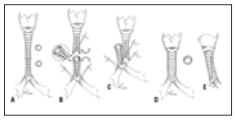
Tracheal Masses

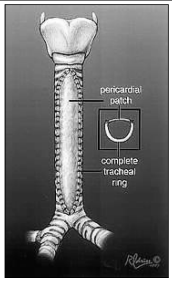
- Primary tracheal and bronchial tumors are rare
  - ✓ 64-90% are benign
  - ✓ Benign tumors more commonly in the proximal trachea
  - ✓ Malignant tumors more commonly in the distal trachea
- Symptoms:
  - ✓ Wheezing
  - ✓ Stridor
  - ✓ Cough
  - ✓ Dyspnea
  - ✓ Hemoptysis

Desai D, Holinger L, Gonzalez-Crussi F. Tracheal neoplasms in children. *Annals of Otology, Rhinology, and Laryngology*. 1998: 790, 107.

Complete Tracheal Rings Treatment

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





Grillo H, Wright C, Vlahakes G, MacGillivray T. Management of congenital tracheal stenosis by means of slide tracheoplasty or resection and reconstruction. *Journal of Thoracic and Cardiovascular Surgery*. 123: 145-152, 2002.

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

- Vocal Cord Paralysis
  - ✓ Can be congenital or acquired
  - ✓ Unilateral or Bilateral
- Vocal Cord Dysfunction
  - ✓ Paradoxical vocal cord movement
  - ✓ Paradoxical vocal cord dysfunction

Vocal Cord Paralysis Etiology

- Congenital
  - ✓ Hydrocephalus
  - ✓ Arnold-Chiari Malformation
  - ✓ Myasthenia Gravis
  - ✓ Cardiovascular anomalies
- Acquired
  - ✓ Trauma
  - ✓ Infectious
  - ✓ Supranuclear lesions (i.e. MS)
  - ✓ Iatrogenic
  - ✓ Idiopathic

Vocal Cord Paralysis

- Accounts for 10% of congenital laryngeal lesions
- Symptoms
  - ✓ Weak or abnormal cry
  - ✓ Stridor or wheezing
  - ✓ Dysphagia
  - ✓ Chronic cough or cough during eating

Vocal Cord Dysfunction

- The larynx exhibits paradoxical vocal cord motion, with vocal cord adduction during inspiration
- Symptoms:
  - ✓ Wheezing
  - ✓ Dyspnea
  - ✓ Cough
  - ✓ Throat tightness
  - ✓ Shortness of breath
  - ✓ Exercise intolerance
- Common in asthmatics

Vocal Cord Dysfunction Etiology

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

Vocal Cord Dysfunction 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

1. Powell DM, Karanfilov BI, Beechler KB, Treole K, Trudeau MD, Forrest AL. Paradoxical vocal cord dysfunction in juveniles. *Archives of Otolaryngology, Head and Neck Surgery*, 126: 29-34, 2000.
2. Landwehr LP, Wood PP, Blager FB, Milgram H. Vocal cord dysfunction mimicking exercise-induced bronchospasm in adolescents. *Pediatrics*, 98: 971-974, 1996.

Vocal Cord Dysfunction

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

Vocal Cord Dysfunction 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

Vocal Cord Dysfunction Treatment

- **Acute Phase:**
  - ✓ **Heliox**
  - ✓ **CPAP**
  - ✓ **Anxiolytic medications**
  - ✓ **General anesthesia**
- **Long-Term Management:**
  - ✓ **Education**
  - ✓ **Avoidance of known triggers**
  - ✓ **Breathing for vocal cord dysfunction**
  - ✓ **Relaxation of muscles of neck, shoulder, and chest**
  - ✓ **Psychotherapy**

Airway Foreign Body

- **Acute Phase- Choking or gagging episode**
- **Asymptomatic Interval- Foreign body becomes lodged and airway reflexes become fatigued**
- **Complications- Erosion, obstruction or infection occurs resulting in symptoms**

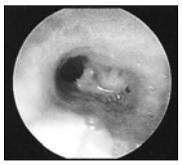
Airway Foreign Body

- **Presentation is variable**
  - ✓ **There is usually a choking or gagging event, however this may not be recognized or reported (present in about 80% of cases)**
  - ✓ **Other symptoms include cough, wheezing, recurrent croup**
  - ✓ **Symptoms may respond to "asthma" treatment, but recur after medications are stopped**
- **85% of patients are younger than 5 years of age**

Airway Foreign Body Diagnosis

- **Radiography**
  - ✓ **CXR may be normal, show radio-opaque foreign body, or hyperinflation**
  - ✓ **Inspiratory and Expiratory views or Decubitus views**
  - ✓ **CT with fine (<3 mm) cuts**
- **Bronchoscopy**
  - ✓ **Gold standard for diagnosis**
  - ✓ **High clinical suspicion with normal imaging does *not* rule out foreign body**

Airway Foreign Body



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

Airway Foreign Body