Top 3 Referrals to Pediatric Pulmonology
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At the conclusion of this presentation, the participant will be able to

- Recognize the clinical presentation of common chronic pulmonary conditions.
- Differentiate diagnoses requiring urgent vs. non-urgent referral.
- Order appropriate diagnostic tests prior to the referral to maximize the benefit of the consultation.

#1. Recurrent Wheezing

What is a wheeze?

- Oscillation of opposing airway walls in narrowed airway
- Continuous "musical" sound
  - 100-5000 Hz
  - > 80 ms in duration
- Expiratory or biphasic
- Requires sufficient flow
  - no wheeze = bad

ABP content specifications

- Wheezing
  - Plan the appropriate clinical and diagnostic evaluation of wheezing of various etiologies
  - Plan appropriate management for wheezing of various etiologies

Which of the following is true?

A. Prevalence of wheezing is 30% in infants < 1 year of age.
B. There is significant agreement between parent-reported “wheeze” and provider documented wheeze.
C. The most likely diagnosis in children with recurrent wheezing is asthma.
Wheezing is a primary presenting symptom in which of the following?

A. Cystic Fibrosis
B. Primary Ciliary Dyskinesia
C. Interstitial Lung Disease
D. Vocal Cord Dysfunction
E. Airway malacia

Case # 1

2-year old with wheezing

- Multiple choice audience response questions.

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Recurrent Preschool Wheeze

- Epidemiology
  - 10% prevalence and rising
- Signs and Symptoms
  - Cough/wheeze/SOB
  - Often seen with eczema and allergic rhinitis
  - Cough is dry and tends to be worst at night and early AM; may worsen with activity and emotional upset

Asthma

- Diagnosis
  - Without spirometry, the dx of asthma is clinical; therapeutic trial of ICS is appropriate; no response suggests an alternate diagnosis and therapy should be stopped
- Treatment
  - ICS are preventative treatment of choice
  - Step up/down therapies as needed

Difficult to Treat Asthma

- Asthma which requires treatment with guidelines suggested medications for GINA steps 4–5 asthma (high dose ICS and LABA or leukotriene modifier/thesophylline) for the previous year or systemic CS for ≥50% of the previous year to prevent it from becoming “uncontrolled” or which remains “uncontrolled” despite therapy.

Uncontrolled asthma defined as at least one of the following:
1) Poor symptom control: ACT <20 (or “not well controlled” by NAEPP/GINA guidelines).
2) Frequent severe exacerbations: two or more bursts of systemic CS (>3 days each) in the previous year.
3) Serious exacerbations: at least one hospitalization, ICU stay or mechanical ventilation in the previous year.
4) Airflow limitation: after appropriate bronchodilator withhold FEV1 <80% predicted (in the face of reduced FEV1/FVC defined as less than the lower limit of normal).
5) Controlled asthma that worsens on tapering of these high doses of ICS or systemic CS (or additional biologics).

Which of the following suggest a diagnosis other than asthma?

A. Poor response to asthma medications.
B. Wheezing since birth.
C. Wheezing with feeding or vomiting.
D. History of choking and coughing.
E. Poor weight gain.
F. A, B, C
G. A and E
H. All of the above

Case #3

- 6 year old with shortness of breath and reported wheezing
Multiple choice audience response questions.

Which of the following additional symptoms is most concerning for chronic lung disease?

- The illness started with high grade fever for a few days.
- It gets worse when he is at grandmothers house (who smokes).
- Cough worsens when he is anxious, improves with distraction.
- He takes frequent breaks to catch his breath while playing with his peers.

Case # 4

4 year old girl with noisy breathing for 2 years

Multiple choice audience response questions.

Congenital Anomalies

- Epidemiology
  - Rare cause but includes wide variety of pathology (cystic, rings, malacia, TEF, laryngeal cleft, etc.
- Signs and Symptoms
  - Newborn: dyspnea, cyanosis, resp distress
  - Later life: cough, noisy breathing, recurrent pneumonia, dysphagia
  - Asymmetric breath sounds, wheezes, crackles
- Diagnosis
  - Cysts often diagnosed in utero
  - Chest Imaging – CXR, CT, MRI, UGI, bronchoscopy, swallow function, etc.
- Treatment
  - Generally surgical

Vascular Ring

- Epidemiology
  - DAA, RAA, PA Sling, aberrant subclavian
- Signs and Symptoms
  - Cough, wheezes, stridor, dysphagia, dyspnea
- Diagnosis
  - Esophagram, bronchoscopy, MRA, CT Angio, echocardiogram
- Treatment
  - CT surgery, trach/vent, aortopexy
Which of the following is false?

A. GERD and swallowing dysfunction can present as chronic wheezing.
B. Children with aspiration syndromes can be misdiagnosed for difficult to treat asthma.
C. Difficult to treat asthma can be associated with poorly controlled GERD.
D. It is necessary to have symptoms of coughing with feeding.

Recurrent wheeze red flags

1. Symptoms since birth
2. Respiratory symptoms associated with feeds or vomiting
3. Recurrent productive cough
4. Failure to thrive
5. Failure therapeutic trial with conventional therapy
6. Underlying neuromuscular or neurodevelopmental disease
7. Additional clinical findings such as digital clubbing, murmur, focal lung sounds.

#2. Chronic Cough

ABP Specifications

- Cough
  - Plan the appropriate clinical and diagnostic evaluation of cough of various etiologies.
  - Plan effective screening evaluation of chronic cough.
  - Plan appropriate management for cough of various etiologies.
Case # 5

- 5 year old boy with cough for 2 weeks

Multiple choice audience response questions.

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Which of the following is true regarding acute cough?

- Duration is < 4 weeks.
- Majority are viral related.
- Testing is typically indicated.
- Foreign body does not present as acute cough.

Diagnostic Algorithm

<table>
<thead>
<tr>
<th>Cough &gt; 4 weeks with Indicators of Specific Cough?</th>
<th>CXR and Spirometry</th>
<th>Specific Testing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>No</td>
<td></td>
<td>No</td>
</tr>
</tbody>
</table>

Diagnostic Cough

Cough Type

- Barking or brassy
- Honking
- Paroxysmal
- Wet

Underlying Disease

- Croup, mediated habit
- Pertussis
- Pharyngitis
- Plastic or cartilaginous subglottic stenosis

Specific Cough Indicators or Abnormal CXR or Spirometry

- Reversibility of airway obstruction
- Comprehensive assessment (suggest pulmonology referral)

Reassure & Reassess

- Yes
- No

Resolution + Success

- Yes & Yes
- No & No

Persistent Non-specific Cough

- Referral
- EmpiriTherapy
- Specific Cough
- Resolution + Failure

Specific Cough

- Dry & Wet
- Persistent / Referral

Reassess & Reassess
### Differential Diagnosis by Age*

<table>
<thead>
<tr>
<th>Infant</th>
<th>Preschool</th>
<th>School Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>GERD/Aspiration*</td>
<td>Sinusitis/UACS</td>
<td>Asthma</td>
</tr>
<tr>
<td>Infection*</td>
<td>Asthma*</td>
<td>UACS/sinusitis*</td>
</tr>
<tr>
<td>Asthma/RAD</td>
<td>Infection*</td>
<td>Suppurative Lung Ds</td>
</tr>
<tr>
<td>Cystic Fibrosis*</td>
<td>Suppurative Lung Ds*</td>
<td>Infection*</td>
</tr>
<tr>
<td>Congenital Anomaly*</td>
<td>GERD/Aspiration</td>
<td>Smoking*</td>
</tr>
<tr>
<td></td>
<td>Foreign Body*</td>
<td>Habit or Tic*</td>
</tr>
</tbody>
</table>

### Case # 6
- 5 year old boy with acute onset of cough
- Aspirated Foreign Body

### Epidemiology
- Peak between 1 & 2 year of age
- $\varnothing : \varnothing \approx 2:1$
- Food items most common
- 70% witnessed; 50% delayed diagnosis

### Signs and Symptoms
- Cough, wheeze, stridor, distress, dysphagia, drooling
- Wheeze or chest asymmetry

### Diagnosis
- Chest imaging
- Insp/Exp
- Lateral Decubitus
- Bronchoscopy

### Treatment
- Rigid bronchoscopy

### Case # 7
- 4 year old with cough for 1 year
  - Productive
  - Multiple antibiotic courses for “pneumonia”
  - Cough resolves between episodes

### Protracted Bacterial Bronchitis

#### Epidemiology
- 25-40% of chronic wet cough in children
- Generally < 5 years of age
- Organisms: H. influenzae, S. pneumoniae, M. catharralis

#### Signs and Symptoms
- Chronic wet cough
- Crackles
- Wheeze

#### Diagnosis
- Symptoms: chronic wet cough, response to antibiotics, no other identifiable cause
- Bronchoscopy
- Treatment
- 2-4 weeks oral antibiotics
- Airway clearance
- Systemic steroids

#### Multiple choice audience response questions.
- ______________________
- ______________________
- ______________________
- ______________________
- ______________________
Case # 8

- 13 year old boy with seasonal allergies and chronic dry cough.

UACS

- Epidemiology
  - AR common (20%-40%)
  - Sinusitis less common (10%)
- Signs and Symptoms
  - Allergic shiners, allergic salute, or allergic crease
  - Associated nasal and ophthalmic symptoms
  - Sinusitis with fever, facial pain, and swelling
- Diagnosis
  - AR is primarily clinical
  - Sinusitis also clinical with use of CT
- Treatment
  - AR: long-acting antihistamines & nasal steroids
  - Sinusitis: antibiotics until symptom-free for 7 days

Habit or Tic Cough

- Epidemiology
  - Up to 10% of all chronic cough (most common 6-16 years of age)
- Signs & Symptoms
  - Honking cough
  - Attention getting
  - Most noticeable at school or with audience
  - Extreme anxiety and irritation from parents
  - La Belle Indifference from child
- Often preceded by or associated with URI, UACS, asthma, VCD
- Diagnosis
  - Classic “diagnosable” cough
  - Child does NOT cough when asleep
- Treatment
  - Pharmacological
  - Psychosocial
  - Speech pathology

Summary

- Chronic cough requires careful, systematic evaluation
- CXR and Spirometry
- Specific additional tests for “specific” cough symptoms
- Wet cough warrants further investigation for bronchiectasis or PBB
- Therapy for chronic cough should be etiology-based i.e. specific therapy for diagnosed “specific” cough
- “Non-specific” chronic cough may resolve spontaneously but warrants reevaluation
- A trial of ICS may be appropriate
- Ineffective medications should be discontinued
- OTC cough medicine and antihistamines are not appropriate for non-specific cough in children

Chronic cough red flags

1. Duration > 4 weeks
2. Hemoptysis
3. Feeding difficulty
4. Failure to thrive
5. Symptoms since birth
6. Hypoxemia
7. Digital clubbing

#3. Recurrent Pneumonia
ABP content specifications

1. Plan appropriate management of the different types of pneumonia
   Identify the major acute and chronic complications of pneumonia
2. Recognize the clinical features of pneumonias of various etiologies
   and the associated sequelae
3. Plan the appropriate diagnostic evaluation for pneumonias of
   various etiologies

Pneumonia Definitions

- Pneumonitis vs Pneumonia
  - Pathophysiologic – lobar, broncho, interstitial, miliary
  - Epidemiologic – CAP, VAP, Atypical

<table>
<thead>
<tr>
<th>Organization</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>WHO</td>
<td>Cough or difficulty breathing, Tachypnea</td>
</tr>
<tr>
<td>BTS</td>
<td>Persistent or repetitive fever &gt;38.5°C together with chest recession (retractions) and an increased respiratory rate</td>
</tr>
<tr>
<td>IDSA</td>
<td>Presence of signs and symptoms of pneumonia in a previously healthy child caused by an infection that has been acquired outside the hospital</td>
</tr>
</tbody>
</table>

CXR Conundrum

- Pneumonia is significantly over-diagnosed based on clinical impression
- Pneumonia is significantly over-diagnosed based on minimal CXR abnormalities
- A CXR should
  - Confirm clinical impression of real disease
  - Characterize potential etiology with respect to a focal, diffuse, or interstitial process
  - Identify complications of pneumonia
  - Provide a baseline for future comparison

Recurrent & Nonresolving Pneumonia Definitions

RECURRENT
- 2 episodes of real pneumonia within a 1 year period
- 3 episodes of real pneumonia ever
- Must document resolution between episodes

NON-RESOLVING
- Unimproved or worsening radiographic finding for more than 1 month

Case # 9

- 4-year old with pneumonia

Multiple choice audience response questions.

- ________________________________
- ________________________________
- ________________________________
- ________________________________
- ________________________________
Uni-Focal

- Intraluminal Obstruction
  - Foreign Body
  - Tumor
- Extraluminal Obstruction
  - Lymph Nodes
  - Infectious: TB, fungus, other
  - Non-infectious: malignancy, sarcoidosis, other
- Cardiovascular Structures
- Congenital anomalies
  - Tracheal bronchus, bronchial stenosis or malacia, bronchiectasis, cystic

Case # 10

- 5-month old with recurrent pneumonia

Multiple choice audience response questions.

Multi-Focal

- Asthma with or without RML Syndrome
- Aspiration
  - Impaired swallow (CNS, neuromuscular, anatomic), Esophageal obstruction or dysmotility
- Mucociliary Dysfunction
  - CF, PCD, PBB, non-CF bronchiectasis, other
- Immune Deficiency
- Structural anomalies
- Congenital heart disease
- CLD of prematurity (BPD)

Multiple choice audience response questions.

Quick Hits

<table>
<thead>
<tr>
<th>Condition</th>
<th>Evaluation</th>
<th>Prior to Referral</th>
<th>When To Refer</th>
<th>What To Send Along</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asthma, recurrent cough and wheeze</td>
<td>Chest x-ray</td>
<td>Following hospitalization especially ICU</td>
<td>Frequent ER visits</td>
<td>Complicating conditions such as FTT, prematurity, BPD, pneumonia.</td>
<td>Age &lt; 2 years 2 steroid bursts per year Moderate/severe persistent asthma Chief concern List of treatments in response Respiratory history since birth Chest x-ray images on CD Workup may include: Spirometry (&gt; 5 years) Chest x-ray Blood work RAST, IgE</td>
</tr>
<tr>
<td>Bronchopulmonary dysplasia, chronic lung disease</td>
<td>Chest x-ray, Oxygen saturation</td>
<td></td>
<td>Unstable respiratory status Poor growth Rehospitalization after discharge from NICU Unable to wean oxygen Complicating conditions i.e. tracheostomy or pulmonary hypertension</td>
<td>Chief concern NICU discharge summary Current oxygen requirement List of medications Growth chart Chest x-ray images on CD Workup may include: Chest x-ray Serum electrolytes Let gas</td>
<td></td>
</tr>
<tr>
<td>Cystic fibrosis</td>
<td>Chest x-ray, Sweat chloride (at CF accredited center)</td>
<td>Positive newborn screen</td>
<td>Positive sweat testing</td>
<td>Cough, wheeze, malabsorption or failure to thrive with a family history of CF</td>
<td>Sweat chloride results Genotyping results if available Chest x-ray Growth chart Spirometry DNA analysis Vitamin D levels, LFT, electrolytes Sputum her throat culture KS x-ray</td>
</tr>
<tr>
<td>Chronic cough, recurrent pneumonia, noisy breathing, tachypnea, chest pain</td>
<td>Chest x-ray</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pneumonia</td>
<td>Chest x-ray</td>
<td>RECURRENT</td>
<td>2 episodes of real pneumonia within a 1 year period</td>
<td>Unimproved or worsening radiographic finding for more than 1 month</td>
<td>Chest X Ray images Growth chart Summary of episodes and antibiotics used</td>
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