IT’S NOT ALWAYS ASTHMA: RESPIRATORY DISTRESS IN PEDIATRICS

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CONFLICT OF INTEREST DISCLOSURE

- I have no financial relationships, commercial interests/funding or other relevant conflicts of interest to disclose pertaining to today’s presentation
OBJECTIVES

- Recognize respiratory presentations that mimic asthma
- Identify how to assess asthma medication adherence
- Describe presentation of vocal cord dysfunction in pediatrics
- Evaluate children for sleep-disordered breathing
CASE 1

- 16-year-old female with **difficult-to-control asthma**
  - Endorses doing well since her last visit 2 months ago
  - Denies any current daytime or nighttime symptoms
  - Admits to using her **controller medication only 2-3 times/week**
  - **4 ED visits and 2 hospitalizations in the last year** due to acute asthma exacerbations
  - States that she is asymptomatic between episodes

- You perform spirometry to assess her lung function and note a normal FEV1, normal FVC and **low FEV1/FVC ratio**
CASE 1: ASTHMA

- Asthma is a chronic disease
  - Characterized by hyper-responsive airways and airway inflammation
  - Presenting with coughing, wheezing and breathlessness

- Acute exacerbations of asthma occur virtually in all asthmatic children

- Severity of exacerbations is variable and based on:
  - Severity of asthma
  - Exposures to specific triggers
  - Optimization and adherence to asthma management
CASE 1: ASTHMA ACTION PLAN

- Home exacerbation management is frequently successful
  - Acute management should be part of every Asthma Action Plan
  - When symptoms do not respond to outpatient management, evaluation by a physician or other healthcare provider is essential

- Most children with asthma exacerbations can be successfully treated with increased medication without additional diagnostic studies

- Frequent need for systemic steroids (>2/year) and/or hospitalizations for acute management indicate poorly-controlled asthma
  - Need to consider stepping-up preventative therapies, identifying comorbidities worsening asthma or in this patient, consider lack of adherence
### CASE 1: SPIROMETRY

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![Spirometry Graph](image-url)
CASE 1: SPIROMETRY IN ASTHMA

- Majority of children with asthma have normal spirometry at baseline
  - Normal spirometry has values above lower limit of normal (LLN) for all major spirometric parameters including FEV1, FVC, and FEV1/FVC ratio

- In severe or poorly-controlled asthma, the most common finding in spirometry is an obstructive ventilatory defect defined as an FEV1/FVC below LLN
  - FEV1 is sometimes low
  - Most common pattern in children with severe asthma: slightly low FEV1/FVC in absence of any other spirometric abnormalities
CASE 1: ASTHMA EXACERBATION

- Most common spirometric pattern in an acute exacerbation is an obstructive ventilatory defect with a low FEV1.

- Several abnormalities in arterial blood gases (ABG) depending on clinical picture and degree of airway obstruction:
  - Initially increased respiratory rate, consequent decreased PaCO2 and increased pH (respiratory alkalosis).
  - Respiratory muscle fatigue and air trapping develop, PaCO2 increases with normalization of PaCO2 and pH.
    - During this period, examination is essential in identifying impending respiratory failure.
  - Course progresses, abnormally high PaCO2 with a low pH (respiratory acidosis).
  - PaO2 is usually decreased throughout course of an acute asthma exacerbation until treatment is started.

- In most, there is improvement of all spirometric measures and ABG during resolution.
**CASE 1: PREDICTORS OF MORTALITY IN CHILDREN WITH ASTHMA**

- Prevalence and severity of childhood asthma have increased, particularly developing countries
  - Mortality from asthma has decreased over last decade
  - Rate of asthma deaths for children is approximately 0.03/1,000 individuals in the U.S.

- Risk factors associated with increased odds of fatal asthma in children include:
  - Adolescent age
  - Male
  - History of near-fatal asthma in past or poorly-controlled asthma
  - African American or Puerto Rican heritage
  - Low socioeconomic status
  - Poor perception of symptoms
  - Children with behavioral or psychological comorbidities
CASE 1: CLINICAL RESOURCES TO OPTIMIZE ADHERENCE

- **Detailed asthma education**
  - Review inhaled technique and use with aerochamber
  - Breath-activated medications are difficult for children < 6 yrs or those with developmental delays
  - Develop a clear, preferably-color Asthma Action Plan
  - Laminated treatment tools and picture checklists

- **Confirm medications** being utilized
  - Clarify medications at home with laminated medication tools
  - Contact pharmacy to confirm refills are being pick-up appropriately
  - Encourage patients/families to bring in medications
    - Count puffs in inhalers, pills in bottles
What is the most common spirometric finding in children with severe or poorly-controlled asthma? (Clue: even when FEV1 is normal.)
LOW FEV1/FVC is the hallmark of obstruction
CASE 1: QUESTION 2

- The initial arterial blood gas abnormality with an acute asthma exacerbation is ________________.
CASE 1: ANSWER 2

The initial arterial blood gas abnormality with an acute asthma exacerbation is **respiratory alkalosis**.

As time progresses, ABG abnormality is respiratory acidosis.
True or False: Mortality of childhood asthma has decreased over time.
**CASE 1: ANSWER 3**

**True:** Mortality of childhood asthma has decreased over time.

- Disproportionately affects certain racial, ethnic and sociodemographic groups of children
- Know predictors of mortality in children with asthma
CASE 1: KEY POINTS

- Low FEV1/FVC is hallmark of obstruction
  - Most common spirometric finding in severe or poorly-controlled asthma, even when FEV1 is normal

- Initial ABG abnormality with an acute asthma exacerbation is respiratory alkalosis
  - As time progresses, ABG abnormality is respiratory acidosis

- Mortality of childhood asthma has decreased over time
  - Disproportionately affects certain racial, ethnic and sociodemographic groups of children
  - Know predictors of mortality in children with asthma
CASE 2

- 11 year-old female with **cough** and **wheeze** associated with viral infections
  - Born at **28 weeks of gestation** requiring 2-month neonatal intensive care unit (NICU) course prior to discharge home on supplemental oxygen
  - Required supplemental oxygen until 6 months of life
  - Now participates in competitive lacrosse and field hockey without difficulty
  - No cough or wheeze with exposure to allergens or irritants

- Chest radiograph is normal

- Spirometry follows
# CASE 2: SPIROMETRY

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![Graph showing flow-volume curve](image)
## CASE 2: SPIROMETRY

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CASE 2: BRONCHOPULMONARY DYSPLASIA

- The patient has a history of bronchopulmonary dysplasia (BPD)

- Defined by some as need for supplemental oxygen support for >28 days in children born < 32 weeks gestation (definition is debated)

- Symptoms:
  - Tachypnea at rest or with exertion: feeding, crying
  - Coughing
  - Wheezing
  - Labored breathing with retractions and nasal flaring
CASE 2: EXERCISE INTOLERANCE

- Many children who survive BPD have complaints of exercise intolerance.
- Cardiopulmonary exercise testing (CPET) is usually normal or near-normal maximum oxygen consumption.
- Long-term follow-up studies have shown normal or slightly reduced diffusing capacity and either a normal or elevated RV/TLC ratio.
- Airway resistance is normal or elevated in few long-term follow-up studies that report these resistance measurements.
CASE 2: BPD AND EARLY LUNG INJURY

- Serious lung injury in early infancy can lead to chronic lung disease (CLD) or BPD
  - Most common etiology is premature birth with respiratory distress syndrome (RDS)

- Despite management advances of prematurity, BPD incidence has not decreased
  - Probably due to increased survival of smallest premature infants
  - About 12% of births in the U.S. are preterm with a 90% survival rate
  - **BPD occurs in 10-40%** of these survivors depending on gestational age
CASE 2: BPD PRESCHOOL

- As children with BPD have been followed through childhood to early adulthood, many have **persisting physiologic abnormalities** associated with respiratory symptoms due to early lung injury.

- In **preschool children** using infant pulmonary function techniques, only about 1/3rd have a positive response to bronchodilator.
  - Can be associated with continued bronchodilator use, persistent wheezing and asthma diagnosis.

- BPD is an independent risk factor for having asthma in later childhood compared to children born at term.
CASE 2: BPD INTO ADULTHOOD

- **School-aged children with** BPD often have reduced FEV1, FEF25-75%, and elevated RV/TLC ratio
  - Few reports of diffusing capacity, reduced and normal diffusing capacity reported
  - Chest CT show abnormal peripheral airways that increase risk for chronic obstructive pulmonary disease (COPD)
  - Likely to have more respiratory symptoms than term children and greater risk for asthma

- **Physiologic abnormalities associated with** BPD may persist into adulthood
  - Young adults experience increased frequency of respiratory symptoms, suggesting that, despite ongoing lung growth, many continue to have lingering effects of early-in-life lung injury
CASE 2: QUESTION

BPD develops in ___% of very-low and extremely-low birth weight infants:

A. <10%
B. 10-20%
C. 10-40%
D. >40%
CASE 2: ANSWER

BPD develops in **10-40%** of very-low and extremely-low birth weight infants:

A. <10%

B. 10-20%

C. **10-40%**

D. >40%
CASE 2: KEY POINTS

- **Prematurity** and **BPD** are risk factors for subsequent pulmonary function abnormalities and **asthma**
- Some infants with **neonatal lung injury** (RDS, meconium aspiration, extremely low birth weight, etc.) develop BPD
- Despite advances in neonatal care, **BPD develops in 10-40% of very-low and extremely-low birth weight infants**
- BPD remains the most common complication of extreme prematurity
- **Long-term sequelae** of neonatal lung injury include airway obstruction, hyperinflation, and airway hyperreactivity
7-year-old female with **wheezing 1-2 days/week** and nighttime cough during all seasons

- Wheezing improves with albuterol and systemic steroid administration
- Additional symptoms: **chronic sniffing, itchy nose, mouth breathing, loud snoring**
- Denies purulent nasal discharge, headache or facial pain
- Daytime symptoms improved on a low-dose inhaled corticosteroid, but continues to have frequent nighttime cough

Family has a cat, but no smokers in the home
CASE 3: PHYSICAL EXAMINATION

- **Pertinent vital signs**: Weight and height at the 25th percentile
- **HEENT**: Infra-orbital edema, crease across his nasal bridge, nasal mucosal edema, clear nasal discharge
- **Respiratory**: Normal anterior-posterior diameter, no retractions or labored breathing, end-expiratory wheezes with a normal expiratory phase
- **Skin**: mild eczema in antecubital fossae bilaterally
CASE 3: ALLERGIC RHINITIS (AR)

- This child demonstrates atopy, has allergic rhinitis (AR) in addition to asthma.
- AR occurs in over 10% of children and is more common in children with asthma.
- Children with AR are 3 times more likely to develop asthma than are those without.
CASE 3: CHARACTERISTICS OF AR

- "Allergic shiners:” infraorbital crescents of edema and venous congestion
- **Dennie-Morgan lines:** creases below the lower eyelids indicative of allergic conjunctivitis
- Line across nasal bridge occurs with “**allergic salute**” from chronic upward wiping of the nose
- **Mouth breathing** may be due to chronic nasal obstruction by mucosal edema
- “**Cobblestoning**” of posterior pharynx caused by **lymphoid hyperplasia** is another common finding in allergic rhinitis
- **Nighttime cough** can result from post-nasal drainage caused by allergens such as dust mites, molds, pet dander or cockroaches
CASE 3: ENVIRONMENTAL ALLERGY TESTING

- **Skin or bloodwork testing to aeroallergens** is indicated in those with poorly-controlled AR and asthma.

- Therapy for AR can be initiated without additional diagnostic testing.

- Identifying specific allergens by performing immediate hypersensitivity testing (skin prick tests) or specific serum immunoglobulin E immunoassays can aid in avoiding potential triggers.

- **Environmental control measures** include removing allergen source and any potential reservoirs:
  - Success of allergen avoidance in reducing symptoms depends on specific allergens involved and aggressiveness of efforts to remove or avoid allergens.
  - Cat dander is difficult to remove from the home even after the cat has been relocated because the dander persists in carpet, upholstered furniture, and on clothing.
CASE 3: PATIENT UPDATE

- Environmental allergy testing results:
  - Very low/low level (0-1/1): Molds (4 types)
  - Moderate level (2): Dust mites, cockroach
  - High level (3): Dog/cat dander, grasses (5 types)
  - Very high level (4-6): Mouse/mouse epithelia, Trees (birch, beech, oak)
  - IgE was markedly elevated (3460.5 IU/mL)

- Patient is started on montelukast, anti-histamine and nasal corticosteroids in addition to already prescribed low-dose inhaled corticosteroid

- Follow-up appointment reveals marked improvement with chronic sniffling, itchy nose, and mouth breathing; but **loud snoring was unchanged**
CASE 3: WHAT TO DO WITH SNORING?

- Persistent, loud snoring in children **always warrants further evaluation**
  - Referral for diagnostic sleep study, sleep physician, pediatric pulmonologist and/or pediatric ENT

- The nose has the greatest resistance in the upper airway, but during sleep the pharynx has the greatest increase in airway resistance
  - This increased resistance is due to decreased pharyngeal size from decreased neuromuscular tone

- Normal children **snore infrequently**

- Reasons to consider further evaluation for **sleep-disordered breathing** other than snoring:
  - Daytime sleepiness or issues with attention/concentration
  - Restless sleep despite good sleep hygiene
  - Choking/gasping arousals
  - Observed sleep apnea during sleep
CASE 3: SLEEP-DISORDERED BREATHING

Sleep-disordered breathing (SDB) in children is associated with varied factors:
• Craniofacial anatomy including midface hypoplasia
• Adenotonsillar hypertrophy
• Obesity
• Upper airway inflammatory processes
• Environmental exposures
• Asthma
  • Prematurity
  • Genetic variation

Each of these factors contributes to SDB because of:
• Anatomy of upper airway
• Intrinsic compliance of upper airway
• Neuromuscular control
CASE 3: QUESTION

Children who **snore** warrant further evaluation with:

A. Diagnostic sleep study
B. Sleep physician or pediatric pulmonologist consultation
C. Pediatric ENT consultation
D. Any or all of the above
CASE 3: ANSWER

Children who *snore* warrant further evaluation with:

A. Diagnostic sleep study
B. Sleep physician or pediatric pulmonologist consultation
C. Pediatric ENT consultation
D. Any or all of the above
CASE 3: KEY POINTS

- Know the history and physical findings that suggests that allergy is contributing to asthma
- Children with allergic rhinitis are 3 times more likely to develop asthma than are those without
- Environmental allergy testing is indicated for children with asthma and atopic features
- Normal children snore infrequently and warrant further evaluation with a diagnostic sleep study, sleep physician, pediatric pulmonologist and/or pediatric ENT consultation
CASE 4

- 15-year-old male with **exercise intolerance** over the past 2 years as he has been more active in track
  - Initially runs without difficulty
  - Develops shortness of breath and chest tightness, worse in cold weather
  - During episodes, he has frequent cough, feeling of suffocation, stridor and a hoarse voice

- Normal vital signs, height 60th percentile; weight 55th percentile

- Normal cardiac and pulmonary examination except for **mild pectus excavatum**

- Normal spirometry with FVC 103% predicted, FEV1 100% predicted, normal FEV1/FVC ratio
CASE 4: EXERCISE-INDUCED DYSPNEA

- This patient has normal spirometry and lung exam except pectus excavatum
  - Symptoms consistent with **vocal cord dysfunction** (frequent cough, feeling of suffocation, stridor and a hoarse voice)

- Vocal cord dysfunction (VCD) or inspiratory laryngeal obstruction (ILO) is often confused with asthma
  - Symptoms and triggers can be similar
  - Some individuals have both VCD and asthma

- **Cardiopulmonary exercise testing** would be an appropriate next step in evaluation of his exercise-induced dyspnea as well as concerns for vocal cord dysfunction
  - Exercise testing protocols vary and can be used to evaluate exercise-induced bronchospasm or cardiac and pulmonary limitation
CASE 4: PECTUS EXCAVATUM

- **Pectus excavatum**: congenital chest wall deformity characterized by concave depression of the sternum
  - Pectus excavatum contributing to current symptoms can be difficult to determine, but is rare
  - Clinical significance of mild-to-moderate pectus excavatum abnormalities is controversial

- **Restrictive ventilatory defect** has been seen associated with pectus excavatum
  - Restriction appears to improve initially following surgical repair, then re-develops, suspected to be related to chest wall growth restriction
  - Some surgeons suggest delaying surgery until post-pubertal growth to minimize this growth restriction
  - Often performed for aesthetic reasons rather than treatment of cardiopulmonary compromise

- Most common complaints associated with pectus excavatum: exercise intolerance and shortness of breath, which improve following surgical repair of the chest wall deformity

- Degree of restriction on spirometry does not correlate with severity of symptoms
CASE 4: VOCAL CORD DYSFUNCTION

- Common signs/symptoms:
  - Shortness of breath or difficulty getting air into the lungs
  - Throat or chest tightness
  - Frequent cough or throat clearing
  - Feeling of choking or suffocation
  - Noisy breathing: stridor, gasping, raspy sounds or wheezing
  - Hoarse voice

- VCD comes on suddenly, varies in severity and does not occur during sleep
- No hypoxemia
- Frequently misdiagnosed as anxiety or panic attacks
CASE 4: ELUCIDATING VCD

- **Triggers:** gastroesophageal reflux (GER), post-nasal drip, upper respiratory infections, exercise, strong odors/fumes, tobacco smoke, strong emotions and stress

- **Diagnosis:**
  - Suspected if symptoms are **not alleviated by bronchodilators**
  - **Flattened inspiratory loop** with pulmonary function testing or witnessed inspiratory stridor with CPET testing
  - Laryngoscope to evaluate for other etiologies of vocal cord damage: nerves, growths or other upper airway abnormalities

- **Management:**
  - Medicines are not indicated to control/prevent VCD
  - **Breathing techniques** that help control your vocal cords are the mainstay and are usually taught by a speech therapist or psychologist who is trained and experienced in VCD
  - **Treat the triggers:** sinus symptoms, GER, stress
  - Learn to **manage stress and strong emotions** if those are identified triggers with relaxation techniques, biofeedback and psychotherapy
CASE 4: QUESTION

Which of the following statements is most accurate.

A. Vocal cord dysfunction and asthma symptoms/triggers are not similar.
B. Vocal cord dysfunction is not associated with hypoxemia.
C. Vocal cord dysfunction occurs during sleep.
D. Vocal cord dysfunction is relieved with bronchodilator use.
CASE 4: ANSWER

Which of the following statements is most accurate.

A. Vocal cord dysfunction and asthma symptoms/triggers are not similar.

B. Vocal cord dysfunction is not associated with hypoxemia.

C. Vocal cord dysfunction occurs during sleep.

D. Vocal cord dysfunction is relieved with bronchodilator use.
CASE 4: KEY POINTS

- Cardiopulmonary limitation due to pectus excavatum is rare
- Dyspnea with exertion warrants further assessment with exercise testing, especially if bronchodilators are not alleviating symptoms
- Vocal cord dysfunction is often confused with asthma or misdiagnosed as anxiety or panic attacks
- Unlike asthma, **VCD is not associated with hypoxemia and does not occur during sleep**
CASE 5

- 3 year-old male with a persistent cough that began in early fall
  - Coughs day and night, though cough is possibly worse at night
  - Cough increases with exercise and with upper respiratory tract viral infections
  - Diagnosed with pneumonia and treated with antibiotics 2-3 times in the past 4 months due to abnormal chest radiographs
  - No fever or chest pain, and does not produce sputum
  - No other history of recurrent infections

- Chest radiograph as follows:
CASE 5: CHEST RADIOGRAPH
CASE 5: CHEST RADIOGRAPH

Mild hyperinflation

Peri-bronchial cuffing/thickening

RML streaky and patchy sub-segmental opacities most consistent with atelectasis from mucus-plugging
CASE 5: ATELECTASIS

- These chest radiograph findings are common in asthma; **this child has asthma**
- Numerous conditions are associated with atelectasis in children, asthma is the most common
- **Atelectasis** is a reversible radiologically apparent loss of lung volume seen in a segment of lung, a lobe of lung, or occasionally in complete lung
  - Caused by increased accumulation of alveolar fluid
  - Usually secondary phenomenon
  - Occurs in young children frequently due to high compliance of their rib cage and small airways, which are more easily blocked with mucus and fluid
CASE 5: LOBAR ATELECTASIS

- Lobar atelectasis can occur in any lung area in children, **RML is most often affected in asthma**
  - RML bronchus has an acute angle take-off from bronchus intermedius and a narrow diameter, creating poor conditions for drainage
  - RML also has poor collateral ventilation which hinders re-inflation once atelectasis occurs
CASE 5: LOBAR ATELECTASIS

- Atelectasis is seen in frontal projection, but is frequently more distinctly seen on lateral projection on chest radiographs, as demonstrated in these radiographs.
CASE 5: CHEST RADIOGRAPH

- Mild hyperinflation
- Peri-bronchial cuffing/thickening
- RML streaky and patchy sub-segmental opacities most consistent with atelectasis from mucus-plugging
CASE 5: CHEST RADIOGRAPHY

- Chest radiograph is **not typically indicated in an uncomplicated asthma exacerbation** in a previously diagnosed patient.

- All children with suspected asthma should have a **chest radiograph at some point** in diagnostic process to exclude:
  - Parenchymal lung disease
  - Suggestion of foreign body in the airway
  - Congenital pulmonary abnormality

- Chest radiographs frequently obtained in asthma exacerbations:
  - Usually do not change diagnosis or management
  - Not always indicated
  - Findings are variable and not correlated with severity of acute exacerbation
CASE 5: LOCALIZED FINDINGS

- Chest radiograph can be helpful if asthma diagnosis is unclear or other diagnoses need to be considered:
  - Pneumonia
  - Foreign body aspiration
  - Pneumomediastinum
  - Pneumothorax

- If localized findings on chest examination, especially if there is no clinical improvement with acute asthma therapy, radiograph should be considered.
CASE 5: OTHER RADIOLOGIC FINDINGS

- **Pneumothorax (PTX)** is an uncommon complication of asthma in children
  - More often seen in the mechanically ventilated child
  - Can be life-threatening
  - Should be considered in a child with asthma and asymmetric breath sounds, chest or shoulder pain, or severe degree of tachypnea or hypoxemia
  - Chest radiograph is the initial diagnostic study to identify a PTX

- **Secondary pneumomediastinum** occurs due to excess pressure in alveolus leading to alveolar rupture and free air in mediastinum, can occur in asthma
  - Uncommon in children
  - Suspected if subcutaneous emphysema or crepitus is observed
True or False: Atelectasis is common with asthma exacerbations in young children and is most often seen in the right middle lobe.
True: Atelectasis is common with asthma exacerbations in young children and is most often seen in the right middle lobe.
CASE 5: KEY POINTS

- Chest radiographs are not usually indicated in acute exacerbations of asthma.
- Recognize indications for chest radiograph in a patient with acute asthma.
- In asthma exacerbations, chest radiograph is often normal, common findings:
  - Hyperinflated lung fields
  - Atelectasis from mucus plugging of the airways
  - Peri-bronchial thickening/cuffing
- Atelectasis is common with asthma exacerbations in young children.
- Atelectasis is most often seen in RML.
- Pneumothorax is rare in non-mechanically ventilated asthmatic.

Breathe easy!
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ADDITIONAL RESOURCES

- American Thoracic Society, Patient Information Series (www.thoracic.org)
- American Academy of Allergy, Asthma and Immunology (www.aaaai.org)
- State Department of Health has locally accepted version of Asthma Action Plan
- Your friendly local pediatric pulmonologists
THANK YOU FOR YOUR TIME AND ATTENTION. QUESTIONS?

A complete list of references can be supplied upon request.

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