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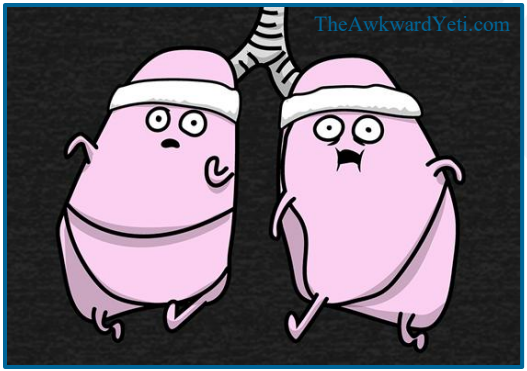
Considerations for Supporting Quality Sleep and Effective Airway Clearance in Children and Young Adults with Intellectual/Developmental Disabilities



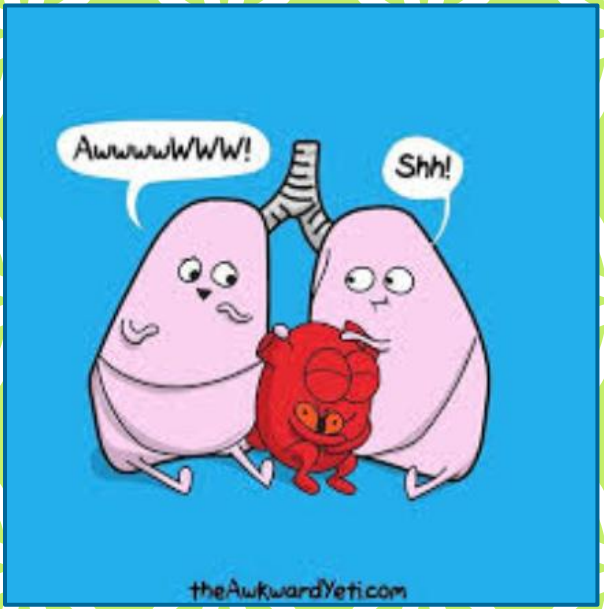
Kate Elizabeth Powers, D.O. {she/her}
Pediatric Pulmonologist, Bernard and Millie Duker Children's Hospital
Associate Professor of Pediatrics, Albany Medical College
Pediatric Director, Cystic Fibrosis Center

Overview

- **Review sleep-disordered breathing**
  - Central sleep apnea
  - Obstructive sleep apnea
  - Hypoventilation

- Discuss the impact of **limited mobility** and **decreased tone** upon breathing with sleep

- Examine ways to **improve sleep**:
  - Optimize airway clearance
  - Address sleep hygiene and more



Brief Introduction to Sleep

- Phases of Sleep
- Diagnostic Sleep Study
- Sleep Quality and Efficiency

Why Do We Care About Sleep?

- We spend about **1/3rd** of our **lives asleep**
  - Sleep supports growth and development in children
- Lack of sleep affects our **memory** and **ability to think clearly**
- Sleep deprivation:
  - **Neurological dysfunction** such as mood swings or hallucinations
  - Higher risk of developing **obesity**, **diabetes mellitus, CVD**

- Sleep difficulties → adverse effects on well-being, functioning, and quality of life
- Lack of or altered sleep disrupts family life, well-being, and the ability to care for children or oneself

Phases of Sleep

- Rapid Eye Movement (**REM; 25%**) and **non-REM (75%)**

- Non-REM is divided into 3 stages progressing into deeper sleep:
  - **Stage 1 (N1; 5%):** Drowsiness, light sleep ,transition into sleep
  - **Stage 2 (N2; 45%):** Light sleep, body movements/startles
  - **Stage 3 (N3, Slow wave sleep: 25%):** Deep sleep, difficult to wake, muscles relax and breathing slows

- **REM:** Rapid eye movements, increased brain activity, vivid dreams

Sleep Cycles

- Cycle through all stages ~4-6 times/night averaging 90 min/cycle
  - Sleep quality and time spent in each sleep stage can be altered by medications, aging, and underlying disorders
  - **SWS (N3)** is associated with loss of respiratory stimulation from the frontal cortex → relative **loss of respiratory drive**
  - **REM sleep** is associated with **loss of skeletal muscle tone** → loss of accessory muscles of respiration (diaphragmatic function is preserved)

- Most children with progressive respiratory impairment develop episodes of **nocturnal hypoventilation** years before having daytime issues

Changes over Time and Gender Differences

- **Newborns:** Short sleep cycles with less consolidated sleep, significant time in both non-REM and REM
- **Infancy/Preschool:** Sleep cycles lengthen and deep sleep (N3) increases
- **School-Aged/Adolescents:** Sleep architecture matures, less deep sleep (N3) and more REM

- **Men:** Longer N1 and more nighttime awakenings
- **Women:** Longer N3 and more difficulties falling asleep

Diagnostic Sleep Study

- Utilizes an electroencephalogram (EEG), electro-oculogram (EOG), electromyogram (EMG), electrocardiogram (ECG), pulse oximetry, airflow, and respiratory effort
  - Sensors to measure brain activity, HR, nasal/oral air movement, SpO2, limb and eye movements, presence of snoring
  - Assesses sleep quality, sleep efficiency, and breathing
- Requires a minimum of 5-6 hours of sleep
- Measures **Apnea/Hypopnea Index (AHI)**
- **Gold standard test for diagnosing sleep-related breathing disorders**: obstructive sleep apnea, central sleep apnea, and sleep-related hypoventilation or hypoxemia
  - Can also evaluate nocturnal seizures, periodic limb movement disorder, narcolepsy, and REM-sleep behavior disorder

Sleep Quality versus Sleep Efficiency

- **Sleep quality:** subjective experience
  - How quickly you fall asleep
  - How long you stay asleep
  - How refreshed you feel upon waking

- **Sleep efficiency:** measurable percentage of time spent asleep while in bed
  - Total sleep time/Total time spent in bed x 100%
  - Normal $\geq$ 85%

Sleep-Disordered Breathing

- Central sleep apnea
- Obstructive sleep apnea
- Hypoventilation

Central Sleep Apnea (CSA)

- Pauses in breathing during sleep due to **improper signaling from brainstem → muscles** that control breathing
  - Less common than obstructive sleep apnea
  - More common in men and people >60 years of age

- **Neurological impairment**
- Certain conditions can contribute to CSA:
  - Medications (opioids), high altitude, kidney and heart disease, stroke, brain tumors, idiopathic

Obstructive Sleep Apnea (OSA)

- Pauses in breathing during sleep due to **partial or complete blockage in the upper airway**
  - Incidence peaks in childhood between 2-8 years of age
  - Males are increased risk post-puberty, similar between males and females pre-puberty

- Children often present differently than adults → snoring, mouth breathing, choking/gasping, restless, sleep, witnessed apneas, frequent nighttime awakenings, secondary nocturnal enuresis

- Disrupted sleep → hyperactivity, daytime sleepiness, irritability, napping, morning headaches, difficulty concentrating

Hypopnea and Hypoventilation

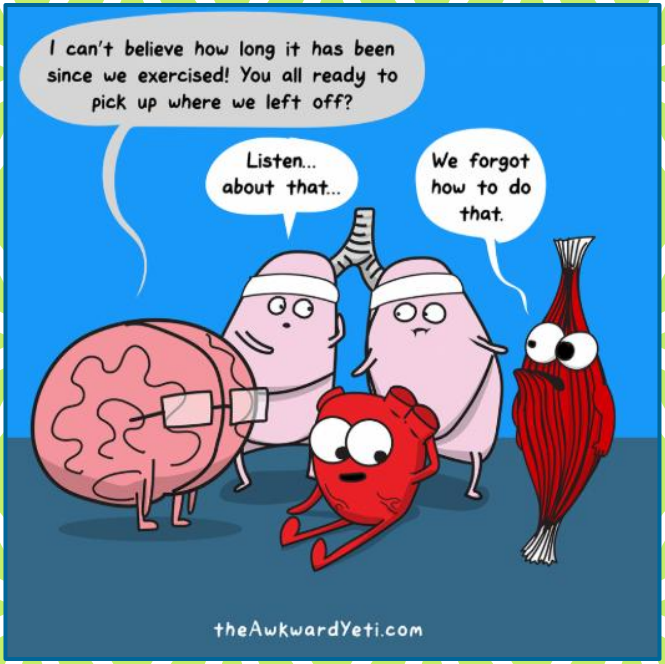
- **Shallow breathing or reduced airflow** during sleep with hypoxemia or arousal from sleep

- Children have higher airway integrity have less risk of complete collapse like adults → Children have **more hypopnea than apnea, which is still associated with diagnosis of OSA**

- **Hypercarbia** can be present in the pediatric population (often absent in adults) → state of **chronic hypoventilation**
- **Hypoventilation:** decrease in rate and/or depth of breathing → hypoxemia and/or hypercarbia

Apnea/Hypopnea Etiologies

- **Enlarged tonsils and/or adenoids/Macroglossia**
- **Craniofacial anatomic anomalies** that narrow airways including achondroplasia, Pierre-Robin sequence, Apert syndrome
- **Prematurity**
- **Neuromuscular disorders:** Trisomy 21, cerebral palsy and other neurological conditions that have decreased oropharyngeal muscle tone
- **Obesity** leads to increased tissue volume around airways → more susceptible to narrowing/collapse
- **Medications** → sedatives/opioids can relax airway muscles
- **Nasal congestion** due to allergies, polyps can restrict airflow



Limited Mobility and Decreased Tone

Discuss the impact upon breathing with sleep

Limited Mobility and Decreased Tone

- **Physical movement** helps mobilize pulmonary secretions

- Central muscle tone impacts the trunk, the neck, and sometimes extremity movement
- **REM sleep** is associated with **loss of skeletal muscle tone**

- **Baseline central hypotonia** can impact sleep, more often REM
  - Restless sleep and frequent awakenings
  - Decreased restful/deep sleep → daytime symptoms

- **Central hypotonia can also contribute to poor clearance of respiratory secretions**

Airway Clearance Mechanisms

- Airway clearance of the lungs involves 2 mechanisms:
  - **Mucociliary clearance escalator** removes debris from peripheral airways
  - An **effective cough** clears the more proximal airways

- Effective **mucociliary clearance** requires normal respiratory dynamic movement:
  - Narrowing of the intrathoracic airways on exhalation
  - Increasing expiratory airflow and movement of mucous towards the mouth

Effective Cough and Retained Secretions

- An **effective cough** includes 3 muscular phases: the use of inspiratory, upper airway/bulbar, and expiratory muscles
  - Impairment of any phase can impact the clearance of secretions from proximal airways

- **Impaired tone** → inability to generate normal tidal volumes leading to smaller breaths and negatively impacts the mucociliary clearance escalator

- **Poor mucociliary clearance** → Retained pulmonary secretions → chronic atelectasis and mucus trapping from decreased expiratory flow

Chronic Aspiration Makes It Worse…

- Bulbar dysfunction results in **impaired recognition of secretions**, **swallowing dysfunction**, and increased subsequent aspiration
- **Chronic aspiration** further contributes to recurrent respiratory infections, respiratory epithelial and ciliary damage
- **Ciliary damage** further decreases mucociliary clearance → more respiratory infections and further ciliary damage
- In people with low neuromuscular tone can have **hypoventilation** and mucus plugging → **hypoxemia** and **hypercapnia** especially during sleep

Ineffective Airway Clearance

- Retained secretions can be corrosive to lung tissue → recurrent tracheitis/bronchitis, pneumonia, and chronic airway inflammation
  - In some cases, retained secretions and recurrent infection → permanent lung damage (bronchiectasis)
  - Effective clearance of airway mucus would decrease the risk of recurrent respiratory infections and delay/prevent permanent lung tissue damage
  - Additionally, frequent antibiotic use to treat infections → resistance and limitations in future treatment options

- **Ineffective airway clearance may also impact sleep**

Improve Sleep

Optimize airway clearance

Address sleep hygiene and more

What About Airway Clearance?

- Focuses on medications and interventions to help clear mucus and secretions from lungs contributing to:
  - Improved baseline breathing
  - Reduced risk of infection
  - Optimized pulmonary reserve → potentially improved sleep
- **Medications:** open airways and thin secretions
- **Interventions:** mobilize secretions through vibration and/or augment a cough

Respiratory-Related Medications

- **Bronchodilators** maximally open airways to enhance airway clearance
- **Hypertonic saline** is a hyperosmolar agent that can alter the physical properties of mucus and the airway fluid surface level
- Inhaled **anticholinergic drugs** (ipratropium) can block increased mucus secretion that can occur with increased inflammation
  - Especially important for children with thicker secretions → may benefit from switching bronchodilators to inhaled anticholinergics

Chest Physiotherapy (CPT)

- CPT provides assistance in loosening retained mucus in the lungs → moving secretions from small to larger airways
- Chest PT modalities can include:
  - Manual percussion
  - Intrapulmonary percussive ventilation (IPV)
  - High-frequency chest wall oscillation via vest therapy (HFCWO)
  - Oscillatory positive expiratory pressure therapy (OPEP)

Intrapulmonary percussive ventilation (IPV)

- Combines a high frequency ventilator with a jet nebulizer to deliver intermittent positive-pressure mini-bursts of gas with high frequency to promote clearance of distal bronchial tree
- IPV expands the lungs, vibrates, and enlarges airways facilitating greater gas delivery therefore reducing persistent atelectasis
  - To increase the percussive effect of IPV, a higher frequency and pressure with shorter inspiratory times is encouraged
- IPV can also improve the efficacy of nebulized therapy

High-frequency chest wall oscillations (HFWCO/Vest)

- Vest therapy is supplied by rapid inflation/deflation producing chest compressions at variable frequencies and pressures
  - Frequencies >3 Hz cause cough-like forces on mucus to move secretions towards the mouth
  - Frequencies around 10 Hz reduces mucus viscosity
- HFCWO is a mucus-mobilization technique that needs to be in tandem with cough or other mucus extraction techniques to clear the airways
- HFCWO may be better tolerated if IPV combined with Cough Assist causes hypoxemia from presumed alveolar de-recruitment

Oscillation positive expiratory pressure (OPEP)

- OPEP devices help clear excess mucus in airways and improve breathing by creating positive airway pressure when the patient exhales → vibrations in the airways to thin and loosen mucus allowing mucus to more easily move up the mucociliary clearance escalator and be coughed out

- OPEP improves airway clearance especially in patients with bronchiectasis and can reduce acute exacerbations when utilized preventatively

Cough Assist Device

- Mechanical insufflation-exsufflation also known as In-Exsufflator, Coughalator, or CoughAssist device clears airway secretions from the lungs by applying cycles of positive and negative pressure to the airway generating a high expiratory flow

- Simulates a cough → brings secretions closer to the mouth where they can be safely removed by suctioning or swallowing

- Safer, non-invasive, more effective alternative to deep invasive suctioning, which can lead to airway bleeding and obstructive granulomas

CPT versus Cough Assist

- CPT is an effective method of loosening and moving secretions to larger airways
- Cough Assist moves secretions further from the lower airway into the upper airway to be coughed, suctioned, or swallowed
- In those with low central tone, CPT should be followed immediately with Cough Assist
  - CPT and Cough Assist are NOT substitutes for each other and NOT exchangeable
  - For patients with weak cough, the two devices should be used in concert to achieve optimal airway clearance

Sleep Hygiene

- **Regular sleep schedule:** go to bed and wake up around the same time daily, even on weekends
- **Relaxing bedtime routine:** engage in calming activities – read a book, take a bath, avoid screens
- **Sleep environment:** ensure bedroom is dark, quiet, and cool

- **Manage stress/anxiety:** employ relaxation techniques like deep breathing or meditation
- **Limit daytime naps and stimulants** (caffeine/alcohol)

Medications to Consider for Snoring

- **Leukotriene inhibitors (montelukast)** may be appropriate for pediatric patients who are diagnosed with mild-moderate OSA
  - Montelukast has been shown to decrease adenotonsillar size significantly after 3 months of treatment → AHI decrease

- Systemic glucocorticoids are NOT effective in treatment of pediatric OSA
- **Intranasal corticosteroid (fluticasone)** for 6 weeks improves AHI; treatment option for patients with mild-moderate OSA

- When intranasal steroids are combined with leukotriene inhibitors, most patients →clinically-significant decrease in AHI

What if We Do Nothing for Sleep Apnea…

- Complications of **sustained hypoxemia** → increase pulmonary vasoconstriction → **pulmonary hypertension** and **right heart failure** at an early age
- **Cognitive dysfunction**, **impaired learning**, and **poor school performance** are associated with undiagnosed and untreated pediatric OSA
- Labored breathing → **failure to thrive** seen in children

- If there is **adenotonsillar hypertrophy** in children, the most effective treatment is **adenotonsillectomy**

Final Thoughts from this Pediatric Pulmonologist

- Sleep is essential → address sleep hygiene first
- Do you suspect OSA? Consider Soft tissue neck imaging, empiric medication trial
- Consider respiratory and non-respiratory etiologies
  - Do they have asthma or another underlying chronic lung disease? Are they well-controlled?
  - Do they have limited mobility/hypotonia: Is ineffective airway clearance the issue?

- Is it time to get a Diagnostic sleep study and/or refer to ENT or Pulmonology?

Questions for you…

You got this!

Question 1:

What 2 medications are considered the "medical management for snoring" in children and adolescents?

A. Antileukotriene: Montelukast
B. Bronchodilator: Albuterol
C. Nebulized mucolytic: 3% Hypertonic saline
D. Nasal corticosteroids: Fluticasone

Question 1 Answer:

What 2 medications are considered the “medical management for snoring” in children and adolescents?

A. **Antileukotriene: Montelukast**
B. Bronchodilator: Albuterol
C. Nebulized mucolytic: 3% Hypertonic saline
D. **Nasal corticosteroids: Fluticasone**

Question 2:

What are possible chest physiotherapy (CPT) options to use at home for people with ineffective airway clearance? (Select all that apply):

A. Manual percussion
B. Oscillatory positive expiratory pressure (OPEP)
C. Cough Assist Device
D. Intrapulmonary percussive ventilation (IPV)
E. High frequency chest wall oscillations (HFCWO)/Vest therapy

Question 2 Answer:

What are possible chest physiotherapy (CPT) options to use at home for people with ineffective airway clearance? (Select all that apply):

A. **Manual percussion**
B. **Oscillatory positive expiratory pressure (OPEP)**
C. Cough Assist Device*
D. **Intrapulmonary percussive ventilation (IPV)**
E. **High frequency chest wall oscillations (HFCWO)/Vest therapy**

*Unless using oscillatory setting component with cough assist device as that IS considered CPT. The oscillatory setting is CPT.

Question 3:

What is the most common form of sleep-disordered breathing in children?

A. Hypoventilation
B. Obstructive sleep apnea
C. Central sleep apnea
D. Sleep fragmentation

Question 3 Answer:

What is the most common form of sleep-disordered breathing in children?

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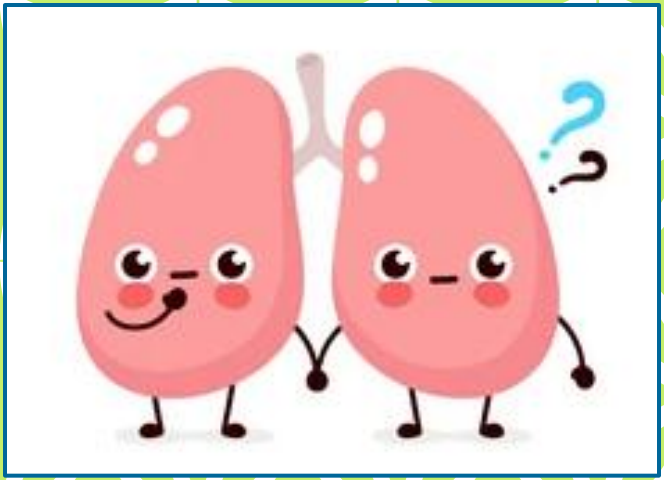

Thank you for your time and attention.
If you have questions, email me at:
powersk4@amc.edu

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