Disclosure

- I have no relevant financial relationships with the manufacturers(s) of any commercial products(s) and/or provider of commercial services discussed in this CME activity.

- I do not intend to discuss an unapproved/investigative use of a commercial product/device in this presentation.
Question

Why do children with Neurological Impairment (NI) have frequent respiratory problems?

What are the pathophysiologic mechanisms?

Aspiration

- Oropharyngeal Motor Dysfunction (from above)
  - Aspiration of oral feeds
  - Aspiration of oral or upper respiratory secretions

Gastroesophageal Reflux (from below)
  - Food
    - Acidic/Non-acidic secretions
  - May cause apnea, laryngeal spasm, inflammation
Aspiration

Both can lead to Aspiration Pneumonia

Increased susceptibility to infection

Chronic lower airway inflammation

Worse airway secretion clearance

Bronchiectasis, lung parenchymal damage

Poor Cough and Airway Clearance

Difficulty coordinating expiratory abdominal and intercostal muscles with glottic muscles

Poor clearance of aspirated materials

Poor clearance of lower airway secretions, especially during respiratory infections

Atelectasis

Hypoxemia

Superinfection

Hypoxemia

Superinfection
Respiratory Muscle Weakness

Contributes to poor cough and airway clearance

Low tone in pharynx

Upper airway obstruction

Obstructive sleep apnea

Paradoxical breathing (chest in, belly out)

Lower respiratory muscle weakness

Hypoventilation

Kyphoscoliosis

Restrictive Lung Disease

V/Q mismatch

- atelectasis on one side
- overexpansion on the other

If occurs early in life, can limit lung growth
Brainstem Control of Breathing

Of particular concern in Chiari malformation and acute hydrocephalus

Central Sleep Apnea

Other factors

Asthma is common in all populations including kids with NI
  • Reflux and Aspiration worsens

Some causes of NI also cause respiratory problems
  • Prematurity → IVH & BPD

Oromotor dysfunction
  • Malnutrition → Respiratory weakness → Increased infections

Autonomic Dysfunction
  • Respiratory distress

Tonsil and Adenoid hypertrophy
  • Watch for Stertor
Case 1

5-year-old with cerebral palsy, seizure disorder, and G-tube presents with fever, tachypnea, hypoxia, and right sided crackles

- RVP is positive for rhino/entero
- CXR shows a RML infiltrate
- This is the 3rd such episode in the last year
- The patient is noted to have a weak cough

What else would you want to know?

How would you approach this patient’s treatment?

Symptoms of Aspiration

- Gagging, choking, apnea, stridor, cyanosis during feeding
- “Wet breathing” or “rattling” suggests laryngeal penetration
- Repeated aspiration can cause recurrent wheezing, hoarseness of voice
- Coughing may or may not be present
  - Cough reflex blunted in kids with NI, leading to silent aspiration
### Aspiration Pneumonia – Risk Factors

- Seizure
- Anesthesia, or other episode of reduced level of consciousness
- Neurologic disease
- Dysphagia
- Gastroesophageal reflux
- Alcohol or substance abuse
- Use of a nasogastric tube
- Foreign body aspiration

### Aspiration Pneumonia

May be caused by anaerobic oral flora

- Anaerobic *strepococci* (eg *Peptostreptococcus*)
- *Fusobacterium* spp
- *Bacteroides* spp
- *Prevotella melaninogenica*
## Diagnosis

### Swallow Study

### CXR
- Early PNA or silent aspiration may be negative
- Infiltrate in dependent lung segments, such as the superior or posterior basal segments of a lower lobe or the posterior segment of an upper lobe

### Nuclear Medicine
- Salivagram
- Gastroesophageal scintigraphy

### Scopes
- Laryngoscopy, Esophagoscopy, Bronchoscopy
- Bronchial-Alveolar lavage looking for Lipid-laden macrophages

### pH/impedence probes

## Aspiration Management

### Acute Pneumonia

#### Antimicrobials
- Amp/sulbactam or Clinda
- Moxifloxacin in older kids/young adults
- If known Gram neg colonized, consider meropenem, piperacillin-sulbactam

#### Oxygen

#### Pulmonary Toilet
- Airway Clearance
Treatments for Poor Airway Clearance

Chest physiotherapy
• Vibes in younger children

Bronchodilators

Secretion management: anticholinergics, suctioning

Vest Therapy

Cough Assist

May be used at baseline and increased during acute exacerbations

Chronic/Preventive Management

Reduction of GE reflux
• Positioning
• H2 blockers, PPI, low dose, pro-motility erythromycin
• Adjustment of enteral feeds
  • Smaller boluses/continuous feeds
  • Jejunal feeds vs. Nissen

Reduction of oral secretions
• Tastes of foods → increased swallowing
• Anticholinergics – glycopyrollate, scopolamine
• Botox
• Salivary Gland removal
• Laryngeal-Tracheal Separation
Case 2

3 year old with Trisomy 21 is admitted for gastroenteritis and dehydration. The patient is noted to have periods of apnea and desaturations during sleep on the monitor. Patient has some noisy breathing that is horse with sleep, but lungs otherwise sound clear. The patient had a normal echo during infancy.

What else would you want to know?

How would you approach this patient’s treatment?

Obstructive Sleep Apnea

Risk factors

• Adenoid/tonsillar hypertrophy
• Obesity
• Medical, neurological, or dental conditions that:
  • Reduce upper airway size
  • Affect the neural control of the upper airway
  • Impact the collapsibility of the upper airway

Consequences

• Behavioral problems
• Growth delay
• Pulmonary hypertension, right heart failure
## OSA - Diagnosis

**Diagnostic criteria from the American Academy of Sleep Medicine (AASM)**

**Both A and B criteria should be present**

<table>
<thead>
<tr>
<th>A criteria</th>
<th>B criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>The presence of one or more of the following clinical symptoms:</td>
<td>The polysomnogram (PSG) demonstrates one or both of the following:</td>
</tr>
<tr>
<td>Snoring</td>
<td>One or more obstructive apneas, mixed apneas, or hypopneas, per hour of sleep. These respiratory events are defined according to the AASM Manual for the Scoring of Sleep and Associated Events.</td>
</tr>
<tr>
<td>Labored, paradoxical, or obstructed breathing during the child’s sleep</td>
<td>A pattern of obstructive hypoventilation, defined as at least 25 percent of total sleep time with hypercapnia (PaCO₂ &gt;50 mmHg) in association with one or more of the following: snoring, flattening of the nasal pressure waveform, or paradoxical thoracoabdominal motion</td>
</tr>
<tr>
<td>Sleepiness, hyperactivity, behavioral problems, or learning problems</td>
<td></td>
</tr>
</tbody>
</table>

## Management Options

- **Surgical reduction of airway obstruction**
  - Tonsillectomy,
  - Adenoidectomy
  - Other airway surgeries
- **Supplemental O2**
- **Noninvasive Ventilation**
  - CPAP
  - BiPAP
- **Tracheostomy +/- ventilation**

*Children with NI may have intrinsic lung diseases and/or central sleep apnea, too, influencing management*
Case 3

A 2-year-old ex-24-week infant with mild CP, BPD, and tracheostomy presents with increased secretions from the trach.

- Patient is afebrile, and desaturating during sleep to the low 80s
- Chest X-ray has baseline chronic changes, but no acute changes

What else would you want to know?

How would you approach this patient’s treatment?

Artificial Airway-Associated Tracheobronchitis Pathogens

- *Staphylococcus aureus*
- *Streptococcus pneumoniae*
- *Haemophilus influenzae*
- *Moraxella catarrhalis*
- *Acinetobacter* species
- *Pseudomonas aeruginosa*
- *Klebsiella pneumoniae*
- *Escherichia coli*
- *Serratia marcescens*
- *Enterobacter* species
- *Stenotrophomonas maltophilia*
- Other gram-negative enteric organisms
Diagnosis

Centers for Disease Control and Prevention (CDC)/National Healthcare Safety Network (NHSN) surveillance definition

- Absence of clinical or radiographic evidence of pneumonia AND
- A positive culture obtained by deep tracheal aspirate or bronchoscopy AND
- ≥2 of the following signs or symptoms with no other recognized cause:
  - Fever >38°C (taken rectally in infants ≤1 year of age)
  - Cough
  - New or increased sputum production
  - Rhonchi and/or wheezing
  - In infants ≤1 year of age: respiratory distress, apnea, and/or bradycardia

Management

To choose antibiotic, consider

- The severity of illness
- Gram stain of the tracheal aspirate
- Susceptibilities of pathogens previously identified in the patient (most useful if recent)
- Likelihood of having multidrug-resistant flora (children with recent hospitalizations or residing in long-term care facilities)
Other Key Points

Be careful about using anticholinergics during acute infections, which can thicken secretions and increase plugging.

Change trach. Plugs can be hiding in them.

Pulmonary Toilet regimen may need to be increased.

Home vs. hospital depends on both disease severity and home care capabilities.

Some Take-Home Messages

NI increases risk for respiratory disease from a variety of mechanisms.

Aspiration in children with NI can come from both above and below, cause both acute and insidious disease.

Don’t forget about airway clearance regimen both acutely and chronically.

Children with NI are at higher risk for OSA and needing more invasive treatments for OSA.

Trach cultures should not be treated in a vacuum.
Resources


UptoDate articles on Aspiration, Obstructive Sleep Apnea, and Tracheitis are well-done