Hypoventilation - Criteria and Case Studies in Pediatrics

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Conflict of Interest Disclosures for Speakers: Fauziya Hassan

1. I do not have any relationships with any entities producing, marketing, reselling, or distributing health care goods or services consumed by, or used on, patients, OR

2. I have the following relationships with entities producing, marketing, reselling, or distributing health care goods or services consumed by, or used on, patients.

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<td>Grant/Research Support</td>
<td>Jazz Pharmaceuticals, NIH</td>
</tr>
<tr>
<td>Consultant</td>
<td>Biogen</td>
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<td>Speakers’ Bureaus</td>
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3. The material presented in this lecture has no relationship with any of these potential conflicts, OR

4. This talk presents material that is related to one or more of these potential conflicts, and the following objective references are provided as support for this lecture:

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Outline

• Hypoventilation criteria in pediatrics and monitoring
• Trouble shooting equipment in the sleep lab
• Examine pediatric case with hypoventilation
• Approach to special cases in pediatrics with hypoventilation
• Treatment options with NIPPV
If electing to score hypoventilation during sleep if EITHER of the below occur:

- There is an increase in the arterial PCO\(_2\) (or surrogate) to a value >55 mmHg for ≥10 minutes
- There is ≥10 mmHg increase in arterial PCO\(_2\) (or surrogate) during sleep (in comparison to an awake supine value) to a value exceeding 50 mmHg for ≥10 minutes
- Pediatrics – CO\(_2\) values higher than 50 mm Hg for over 25% of the study time
End Tidal $\text{CO}_2$ monitoring

- In a normal lung $\text{CO}_2$ rapidly diffuses across the capillary alveolar membrane with ventilation and perfusion well matched.

- $\text{EtCO}_2$ reading is the $\text{PCO}_2$ equivalent to airway $\text{CO}_2$ concentration at the end of expiration which approximates arterial $\text{PCO}_2$ (slightly higher than by 4 mm Hg).
End Tidal CO$_2$ Errors

- Increase in dead space ventilation-underestimates arterial PCO$_2$
- Incomplete exhalation before next breath - small airways obstruction
- Smaller tidal volumes among children esp infants with tachypnea
- Leaks - mouth breathing or around the stoma of the tracheostomy
- Increase in physiologic dead space
  - low cardiac output states e.g. PE, blood loss
  - hypothermia
  - hyperventilation
- Technical
  - improper setup and calibration
  - moisture
High End Tidal CO$_2$

- Hyperthermia
- Injections of bicarb
- Contamination of sampling device
- Rebreathing exhaled gas
Transcutaneous \( CO_2 \) monitoring

- \( TCO_2 \) values has good correlation with \( PaCO_2 \), still it is recommended to compare with arterial blood gas at the time of sampling.
- Hypoperfusion with low cardiac output can cause poor skin perfusion and decreased values as can vasoactive medications.
- \( TCO_2 \) monitoring should be avoided in areas of increased thickness or edema of skin and / or subcutaneous tissue.
- Avoid using if there are skin blisters or burns and the sensor placement should not be placed peripherally as the perfusion can be inconsistent with cold extremities.

AARC Clinical Practice Guideline: Transcutaneous Monitoring of \( CO_2 \) and \( O_2 \)
Recommendations for Transcutaneous CO$_2$ Monitoring

- To check adequacy of ventilation esp when using NIPPV
- Long terms trends and typically not recommended for spot checks
- Physiologically
  - Use among infants
  - Use among patients with tracheostomy esp those with large stoma
  - Mouth breathing
  - Trying to use supplemental oxygen with NC in place
- Pathological conditions
  - Patients with severe obstructive lung disease
Errors in Transcutaneous CO₂ Monitoring

- There is a prolonged stabilization time required of the equipment which is required for mandatory heating of the electrode.
- Movement and lying on the sensor can lead to erroneous values.
- Improper calibration, trapped air bubbles, damaged membrane as well as too much contact gel and saline between sensor and skin leads to erroneous values—needs experienced RT.
- Thermal injury can occur at sensor placement site.
Case Study

- 6 week old male infant seen in clinic a week after discharge from hospital to establish care
- Parental concerns- loud and fast breathing using abdominal muscles
- Good sleeper and breathing better during sleep. No problem with feeding
- Low muscle tone
- Discharged from the hospital after week long stay with URI symptoms and was rhino-enterovirus positive
Lab Work

- Blood Gas (VBG) - pH 7.39, PCO₂ 45, PO₂ 40
- CBC - WBC 8.5, Hg 14.2, Hct 39.3, Plts 325
- Basic Chemistry - Na 139, K 5.2, Cl 106, CO₂ 24, BUN 14, Creatinine < 0.1, Glucose 91, Ca 10.2
- Chromosomal microarray, acylcarnitine profile, Prader –Willi and Angelmann syndrome testing all negative
- Brain and spine MRI normal
Chest X-Ray
Confirmatory Lab Diagnosis

- 0 copies of SMN1 exons 7 and 8 detected
- 2 copies of SMN2 exons 7 and 8 detected
**Spinal Muscular Atrophy**

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<th>Age of Onset</th>
<th>Highest Function</th>
<th>Natural Age of Death</th>
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<td>0-6 mo</td>
<td>Never sits</td>
<td>&lt;2 y</td>
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<td>7-18 mo</td>
<td>Never stands</td>
<td>&gt;2 y</td>
</tr>
<tr>
<td>Type 3 (mild)</td>
<td>&gt;18 mo</td>
<td>Stands and walks</td>
<td>Adult</td>
</tr>
<tr>
<td>Type 4 (adult)</td>
<td>Second or third decade</td>
<td>Walks during adult years</td>
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Table 1. Clinical Classification of Spinal Muscular Atrophy

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Sleep Study Epoch
Sleep Study Epoch
Non REM Sleep
REM Sleep
Indications for CO$_2$ Monitoring with Neuromuscular Diseases

• Impaired respiratory muscle function
  ➢ FVC < 40%
  ➢ Peak inspiratory pressure < 15 cm H$_2$O
  ➢ Pharyngeal dysfunction-snoring, swallowing abnormalities

• Snoring, cor pulmonale, morning headaches, personality or behavioral changes, FTT, developmental delay

• Planning and implementation of nocturnal assisted ventilation

• Assess adequacy of home respiratory support

• Pre and post operative before upper airway, thoracic, abdominal or orthopedic surgery

Routine Respiratory Care for Duchenne Muscular Dystrophy (DMD)

- Once a year visit with pulmonologist starting between 4-6 years of age and before confinement to wheelchair with spirometry if possible (> 6 yrs) and evaluation of symptoms of sleep disordered breathing

- Visit with a pulmonologist twice a year after confinement to wheelchair, fall in FVC < 80% and / or age 12 years and every 3-6 months when on NIPPV

- Spirometry, MIP, MEP, pulse oximetry and end-tidal or transcutaneous CO₂ monitoring (esp when FVC < 50% or on NIPPV)

- Respiratory Care of the Patient with Duchenne Muscular Dystrophy ATS Consensus Statement
- The respiratory management of patients with Duchenne Muscular Dystrophy: A DMD care considerations working group specialty article
Muscular Dystrophy

- NIPPV can be used – BiPAP with back up rate or mechanical ventilation for sleep disordered breathing and night time hypoventilation
- Nocturnal NIPPV results in improved survival, improved quality of sleep, improved day time gas exchange and slower decline in pulmonary function
- CPAP has limited utility for OSA treatment with normal nocturnal ventilation
- Avoid oxygen use for treatment of hypoxemia
Intervention in SMA

- Normal breathing
- Inspiratory, expiratory, bulbar muscle weakness
- REM-related sleep-disordered breathing
  - REM-related sleep-disordered breathing
  - Ineffective cough, reduced peak cough flows
  - Swallow dysfunction
- NREM and REM sleep-disordered breathing
  - Chest infections
- Daytime ventilatory failure
  - Death

- Physical examination
  - Pulmonary function, cough peak flow, respiratory muscle strength
  - Chest x-ray, sleep study
- Intervention
  - Airway clearance with cough assistance
  - Nocturnal noninvasive ventilation
  - Nocturnal or continuous noninvasive ventilation
Sleep Study

- Sleep related hypoventilation is correlated with awake PaCO$_2$ of $\geq 45$ mm Hg
- In lab sleep studies have CO$_2$ monitoring, home sleep studies do not
- Recommendation
  - Review of sleep quality and symptoms of sleep disordered breathing at every patient encounter
  - Annual evaluation of sleep disordered breathing when in wheelchair - annual sleep study with CO$_2$ monitoring (?)
  - Pulse oximetry with CBG in the morning (?)
Central Apnea

- Event lasting 2 missed breaths with associated arousal, awakening or ≥ 3% oxygen desaturation
- Or events lasts ≥ 20 seconds without physiologic consequence
- Absent inspiratory effort
- In infants under one year there is decrease in heart rate to < 50 beats per minute for 5 secs or < 60 for 15 secs

Physiologic
- Post arousal centrals
- During phasic REM sleep
**Modes of Non-Invasive Ventilation**

- Non-invasive positive pressure ventilation
  - BiPAP (bi-level positive airway pressure)
  - BiPAP with ST mode (spontaneous-timed)
  - BiPAP with T mode (timed)
  - BiPAP with PC mode (pressure control)
  - Average Volume Assured Pressure Support (AVAPS)® with S mode
  - AVAPS ® with ST mode
  - AVAPS ® with PC mode
Modes of Non-Invasive Ventilation

- Non-invasive positive pressure ventilation
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  - BiPAP with ST mode (spontaneous-timed)
  - BiPAP with T mode (timed)
  - BiPAP with PC mode (pressure control)
  - Average Volume Assured Pressure Support (AVAPS) with ST mode
  - AVAPS with PC mode
From the Old to the New
Options for Mechanical-Assisted Ventilatory Support at Home

- BiPAP or AVAPS
  - If only required ventilatory support during sleep
  - Spontaneous timed or timed modes should only be used as there is no spontaneous respiratory drive
BiPAP with Inability to Trigger IPAP
Trigger to IPAP
BiPAP with ST (Lower Tidal Volume with Spontaneous Breaths)
BiPAP with ST mode
BiPAP in PC vs. ST mode

Pressure Control Machine and Assist Breaths

Pressure Control Machine and Assist Breaths, and Pressure Support Patient Breaths
AVAPS

• AVAPS (Average Volume Assured Pressure Support) is available in
   S (spontaneous)
   S/T (spontaneous/timed)
   PC (pressure control)
   T (timed) modes

• Helps maintain the tidal volume by automatically controlling the pressure by varying the IPAP between the IPAP minimum and IPAP maximum settings
AVAPS automatically adjusts the pressure support according to the patient’s needs to maintain an average tidal volume.
AVAPS at Target Tidal Volume
## Trilogy vs AVAPS Settings

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<thead>
<tr>
<th>Feature</th>
<th>Trilogy in AVAPS mode</th>
<th>AVAPS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tidal volume (ml)</td>
<td>50 to 2000</td>
<td>200 to 1500</td>
</tr>
<tr>
<td>IPAP minimum (cm of water)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>IPAP maximum (cm of water)</td>
<td>50</td>
<td>25</td>
</tr>
<tr>
<td>Rise (ms)</td>
<td>1 to 6</td>
<td>1 to 6</td>
</tr>
<tr>
<td>EPAP (cm of water)</td>
<td>4 to 25</td>
<td>4 to 20</td>
</tr>
<tr>
<td>I time (seconds)</td>
<td>0.3 to 5.0</td>
<td>0.5 to 3</td>
</tr>
<tr>
<td>Rate (maximum)</td>
<td>60</td>
<td>30</td>
</tr>
<tr>
<td>AVAPS rate</td>
<td>1-6</td>
<td>none</td>
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<tr>
<td>Other features</td>
<td>Dual prescription</td>
<td>none</td>
</tr>
<tr>
<td></td>
<td>Back up battery</td>
<td></td>
</tr>
<tr>
<td></td>
<td>NIPPV and IPPV</td>
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Need for Daytime Ventilation

- Daytime ventilation should be considered when awake $\text{PaCO}_2 > 50$ mm Hg
- And/or hemoglobin saturation $<$92% while awake
- Tracheostomy should be considered when contraindications or patient aversion to NIPPV are present or when NIPPV is not feasible due to severe bulbar weakness
Options for Mechanical-Assisted Ventilatory Support at Home

- Positive pressure ventilation (PPV) via tracheostomy
- Modes of ventilation
  - Non-invasive - Sip and puff
  - AVAPS in PC mode via tracheostomy
  - Volume ventilation or pressure ventilation via tracheostomy
Michigan Difference

Pediatric Cardiology
Pediatric Neurology
Pediatric Pulmonology
Physical Medicine and Rehabilitation
Palliative Care

Nutrition
Nursing
Respiratory Care
Social Work
Physical & Occupational Therapy
Wheelchair Seating and Orthotics