Mechanical Ventilation of the Patient with Neuromuscular Disease

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Neuromuscular Diseases Causing Respiratory Failure

- **Cerebral cortex**: stroke, tumor
- **Brainstem**: drugs, hemorrhage, anoxia, polio, multiple sclerosis, primary hypoventilation
- **Spinal cord**: trauma, tumor, tetanus
- **Motor nerves**: motor neuron disease (ALS, SMA), Guillain-Barré, critical illness neuropathy
- **Neuromuscular junction**: drugs, myasthenia gravis, toxins (botulism, snake bite)
- **Myopathies**: muscular dystrophy, myotonic dystrophy, Pompe disease
Guillain-Barre (reversible) and myasthenia gravis (treatable) are most common

Inspiratory/expiratory muscle weakness, upper-airway dysfunction

Bedside PFTs can predict need for mechanical ventilation:
- Vital capacity <15 mL/kg or < 1 L
- Maximal inspiratory pressure > -30 cm H₂O
- Maximal expiratory pressure < 40 cm H₂O

Insufficient evidence to recommend NIV in patients with Guillain-Barré syndrome or myasthenia gravis

Mehta, Respir Care 2006; 51:1016
ICU-Acquired Muscle Weakness

- Polyneuropathy and myopathy (coexist, myopathy most common); occurs in 25% of ventilated patients
- Risk factors: sepsis, corticosteroids, hyperglycemia, neuromuscular blockade, severity of illness
- Associated with adverse outcomes: mortality, longer time on ventilator, increased length of stay
Mobility in the ICU

Needham, JAMA 2008
Morris, Crit Care Med 2008; 36:2238
Burtin Crit Care Med 2009; 37:2499

Bailey, Crit Care Med 2007; 35:139
Schweikert, Lancet 2009; 373: 1874
Neuromuscular Respiratory Failure

- Inability to Ventilate
  - Inspiratory muscle weakness

- Aspiration Risk
  - Upper-airway muscle weakness

- Inability to Cough
  - Expiratory muscle weakness
  - Upper-airway muscle (glottic) weakness
  - Inspiratory muscle weakness

Benditt, Respir Care 2006; 51:829
Respiratory Muscle Training

- Training can increase respiratory muscle strength and endurance
  Leith and Bradley, J Appl Physiol 1976; 41:508;
  Lotters, Eur Respir J 2002;20:570

- Training occurs slowly – weeks?
  (unlikely the result of a few SBTs)
Cough Assist Techniques

- Evaluation of Cough
  - Cough flow < 160 L/min: initiate cough assist
  - Cough flow < 270 L/min: risk for secretion retention
  - Low cough flow associated with extubation failure
    (Salam et al, Intensive Care Med 2004)

- Cough Assist Techniques
  - Hyperinflation
  - Manually assisted cough
  - Mechanical In-Exsufflator (Cough Assist)
Noninvasive Ventilation
NIV and ALS

Patients with normal or moderately impaired bulbar function

Patients with severe bulbar impairment

Bourke, Lancet Neurol 2006;5:140
Indications for Chronic NIV

- Symptoms (fatigue, dyspnea, morning headache, orthopnea) and one of the following:
  - $\text{PaCO}_2 \geq 45 \text{ mm Hg}$
  - Nocturnal desaturation $\leq 88\%$ for 5 consecutive min
  - For progressive neuromuscular disease, maximal inspiratory pressures $> -60 \text{ cm H}_2\text{O}$ or FVC $< 50\%$ predicted

*Chest 1999; 116: 521–534*
Full-Time Noninvasive Ventilation: Possible and Desirable

Joshua O Benditt MD

Respir Care 2006; 51:1005
How to Choose Settings for NIV

- **Empiric**
  - Short-term symptoms: comfort, accessory muscle use
  - Long-term symptoms: less morning headache, fatigue, and daytime sleepiness
- **Physiologic:** tidal volume, gas exchange
- **Polysomnography**
  - Long wait time
  - Sleep labs less familiar with NMD than OSA
- **Overnight oximetry**
  - Does not assess sleep quality
NIV Settings for NMD

- Back-up rate (periodic breathing)
- Trigger, cycle, rise time per patient comfort
- EPAP: 3 – 4 cm H$_2$O (low as possible unless OSA)
- IPAP: 8 – 15 cm H$_2$O as tolerated; may need higher settings with acute illness
- Ramp off
- Unclear role for newer modes like AVAPS
NIV Settings for NMD

- FIO\textsubscript{2}: room air unless acute illness
- Humidity: routine
- Inhaled bronchodilators and steroids not necessary
- Nasal symptoms: humidity, OTC remedies, nasal steroids and anticholinergics
Approaches to Intolerance: Chronic Use

- Start with low settings
- Practice wearing mask without pressure
- Short periods with distraction (watching TV)
- Use during naps
- Short times at night, gradually increasing
- Personal motivation, family support
- Knowledge of the evidence (using NIV is life prolonging)
Indications for Tracheostomy

- Patient preference
- Inability to tolerate NIV or failing NIV
- Bulbar involvement: aspiration, pneumonia
- Inadequate cough despite cough assist
- Consider need for resources to manage trach and ventilator
Ventilator Settings for Trach Patient

- Volume control ventilation (avoid pressure support and pressure control)
- Rate and tidal volume per comfort, gas exchange, and safety
- PEEP 5 cm H$_2$O; room air
- Cuff deflation for leak speech; adjust PEEP, tidal volume, inspiratory time to improve speech
- Cough assist (MIE) for airway clearance
Ventilator Settings for Trach Patient

- Choose ventilator for size (fits on wheelchair) and battery life
- Transition to homecare ventilator in acute care setting
- Do not wean patient with progressive disease (e.g., ALS)
- Use lung protective ventilation strategies if patient develops acute lung injury (Crit Care Med 2007; 35:1815)
Summary

- A number of acute and chronic neuromuscular diseases have respiratory muscle involvement.
- Invasive and noninvasive ventilation, and airway clearance, can be life prolonging.