Respiratory Management of Patients with Neuromuscular Disease

Joshua O. Benditt, MD
University of Washington School of Medicine

My practice
- Adult pulmonary and critical care physician
- Adjunct appointment in Rehabilitation Medicine
- Outpatient clinic
  - 350 patient with various neurologic diseases
    - DMD
    - Myotonic dystrophy
    - SMA
    - Congenital myopathies
    - ALS
    - SCI
    - Post-polio syndrome

Topics
- Neuromuscular respiratory failure
  - Pathophysiology
  - Management paradigm
- Assessment and Monitoring
- Tracheostomy versus non-tracheostomy
- Transition of Care from pediatric to the adult clinic

Neuromuscular Respiratory Failure
- Ventilatory Insufficiency
- Cough Insufficiency
- Swallowing Insufficiency

Pertinent Anatomy of the Neurorespiratory system
- Ventilatory Insufficiency
  - Diaphragm
  - External intercostal muscles
  - Accessory muscles
- Cough Insufficiency
  - Abdominal muscles
  - Internal intercostals
  - Glottic muscles (compressive phase)
- Swallowing insufficiency
  - Glottic and pharyngeal muscles

Ventilatory Insufficiency Assessment
- History:
  - Symptoms of sleep disordered breathing
  - Symptoms of nocturnal or diurnal hypercarbia
- Clinical evaluation
  - Forcéd Vital Capacity
    - upright
  - supine (if diaphragm weakness suspected)
  - Maximal Inspiratory Pressure (MIP)
  - CO₂
    - Arterial
    - Transcutaneous
    - ETCO₂
  - Sleep evaluation
Inspiratory Failure and Sleep

- During sleep, particularly REM, central output to the respiratory muscles decreases
- Weak muscles + decreased drive results in significant hypoventilation with CO₂ retention
- Can result in daytime hypercarbia


**NEUROMUSCULAR WEAKNESS**

- Severe Nocturnal and Diurnal Hypoventilation
  - Depression of Respiratory Drive
  - HCO₃⁻ Retention
  - Alveolar Hypoventilation ↑
  - Periods of REM Sleep ↑
  - Sleep Deprivation

**Daytime Hyposomnolence and Fatigue**

Perrin C et al. Muscle Nerve 29:5; 2004

When is nocturnal ventilation needed?

- Symptoms of sleep disordered breathing and one of the following:
  - FVC < 50% predicted
  - MIP > -60 cm H₂O
  - Daytime PaCO₂ > 45 mm Hg
  - Nocturnal Hgb sat < 88% for 5 minutes or more continuously
  - Abnormal and consistent polysomnogram

Follow-up of nocturnal assisted breathing

- Symptom resolution
- Restoration of normal daytime PaCO₂
- Normalization of nocturnal oximetry
- Polysomnogram if adequacy of nocturnal support not clear

Nocturnal Noninvasive Ventilation

- Bilevel Pressure Support Ventilation
- Back-up rate critical
  - Central events during sleep are common
- Nasal mask or pillows most commonly used interface.
- If oral leak is significant
  - Full facemask
  - Chin strap
- Monitoring downloads is important

Efficacy of Nocturnal Nasal NPPV

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<th>Author</th>
<th>Reference</th>
<th>Type of Ventilation</th>
<th>Diagnosis</th>
<th>Symptoms</th>
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**Multivariate Outcomes**

Acute effects of NPPV Addition and Withdrawl

When is Daytime Ventilation Needed in Neuromuscular Disease?
- Persistent hypercarbia despite adequate nocturnal therapy (PaCO2 > 50 mm Hg)
- Diurnal dyspnea despite adequate nocturnal therapy
- Recurrent infections despite adequate nocturnal therapy
- Voice augmentation (larger vital capacity)

Potential Methods of Daytime Ventilation
- Invasive ventilation
  - Tracheostomy
  - Cuff inflated
  - Cuff deflated
  - Diaphragm Pacing
  - SCI
- Noninvasive Ventilation
  - NPPV
  - Mask
  - Mouthpiece
  - Body ventilators
    - Of historical interest mainly

Mouthpiece Ventilation - MPV
- "Sip Ventilation"
- portable ventilator
  - volume-cycled, assist-control configuration
  - Mouthpiece with "elbow" or narrow caliber to increase circuit resistance to prevent pressure alarm
  - ventilator
    - Pressure alarm set at minimum sensitivity
    - Rate set at 4-6 breaths/min
    - Respironics/Phillips ventilator now FDA approved for the indication

Mouthpiece-Ventilator Alternatives
**24 Hour Per Day NPPV Combination Therapies**

- Nocturnal ventilation
  - nasal or oronasal ventilation with pressure or volume
  - mouthpiece ventilation (Europe)
- Daytime support
  - If needed
    - dyspnea during day
    - PaCO$_2$ remains elevated despite nocturnal therapy
  - mouthpiece ventilation
  - occasionally nasal ventilation (ALS)

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**Diurnal ventilation and Survival in DMD. Toussaint et al Eur Resp J 2006 28:549**

- Followed 45 DMD patients on MPV
- 50% survival 31 years
- Main cause of death was cardiac or sudden death

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**Expiratory (Cough) Failure**

- Abdominal and internal intercostal weakness
- Inability to close glottis
- Reduced peak cough airway flow
- Inability to clear secretions
- Atelectasis and infection

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**Evaluating Cough Strength**

**Peak Expiratory Cough Flow:**

- Measures global cough strength
- Simple testing format
- Correlates with pneumotach cough flows >270 L/min.

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**Expiratory/Cough Failure Assessment**

- Peak Cough Flow
  - Normal > 500 lpm
  - < 160 lpm ineffective clearance
  - < 270 lpm when healthy
    - drops to < 160 seen during infection
- Maximal expiratory pressure
  - ? Minimal value predictive of problems
**Expiratory or Cough Assistance**

- Cough augmentation
  - Assisted cough
    - Abdominal or lateral ribcage thrust
  - Postural maneuvers
  - Resuscitation bag
    - breath stacking
    - elastic recoil augments cough function
  - Cough-Assist (Inexsufflator)
    - requires relatively intact bulbar function
  - Mouthpiece ventilator
    - breath stacking also possible

**Manual Cough Augmentation**

- Quad Cough Maneuver
  - Improves expiratory cough flows
    - Kirby NA, Arch Phys Med Rehabil 1966;47:705-10
  - Abdominal splint prevents paradoxical motion
  - Requires skilled caregiver and coordinated effort
  - Inspiratory volume dependent

**Manual & Mechanical Cough Augmentation**

- Manual hyperinflation
  - Increased expiratory flow volume and velocity
  - Intrathoracic pressure
  - Portable therapy
  - Oral strength limitation
  - Skilled caregiver

**Mechanical Cough Augmentation**

- Breath stacking and Mouthpiece Ventilation
  - Taking multiple breaths prior to exhalation
  - Stores elastic energy in chest wall and lung
  - Allows higher cough velocities
  - Allows independence in cough function- no assistance needed
Indications For Cough Augmentation

- Peak Cough Flow (PCF) < 160 Liters/minute is the level at which secretion clearance is inadequate
  - However, during respiratory infection expiratory muscle strength drops and peak cough flow < 270 Liters/minute when well often drops to 160 LPM or less when sick
- Difficulty clearing secretions
- Hospitalizations for pulmonary congestion associated with respiratory infection

Treatment of Swallowing Problems

- Head down
- Thick liquids
- Avoidance of dry, bulky foods
- Secretion management
- Tracheostomy
- Gastrostomy tube

Treatment of Swallowing Problems Gastrostomy Tube

- When glottic function is impaired
  - prevents excess muscle loss
  - decreases risk of aspirating particulates
  - reduces family and patient anxiety
  - outpatient procedure
  - can be performed with assistance of noninvasive ventilation

Disadvantages of Invasive Ventilation

- Surgical Procedure
- Infection Risk
- Bleeding Risk
- Interference with speech/swallowing
- Increased cost due to attendants for suctioning

Early Complications of Tracheostomy

Durbin Respiratory Care 2005;50:511

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<th>Complication</th>
<th>Patients Who Experienced the Complication (%)</th>
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<td>Pneumothorax</td>
<td>0-4 0-4</td>
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<tr>
<td>Accidental decannulation</td>
<td>0-15 0-4</td>
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<tr>
<td>Subcutaneous emphysema</td>
<td>0-4 0</td>
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<tr>
<td>Stoma infection</td>
<td>0-43 0-10</td>
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<tr>
<td>Difficult intubation</td>
<td>0 0-27</td>
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<tr>
<td>False placement</td>
<td>0 0-4</td>
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<tr>
<td>Hypoxia</td>
<td>0-8 0-25</td>
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<tr>
<td>Loss of airflow/death</td>
<td>0 0-8</td>
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</tbody>
</table>
Late Complications of Tracheostomy

* Epstein Resp Care 2005;50:452 *

- Tracheal stenosis
- Granulation tissue
- Tracheoinnominate artery fistula
- Tracheoesophageal fistula
- Pneumonia
- Aspiration
- Reported to occur in up to 65% of patients

When Must Tracheostomy Be Considered?

- Significant glottic dysfunction with increased risk of aspiration
- Recurrent pneumonia on full-time NPPV
- Elevated PaCO2 despite optimal full-time noninvasive therapy
- Patient preference
- Lack of experienced healthcare providers
- (> 20 hours/day requirement for ventilation)

Transition from Pediatric to Adult Care

- This is a big problem
  - All children grow up and will transition to adult-centered health care
  - For children with special needs this can be an enormous and difficult transition
  - It is estimated that there are 10 million youth age 0-17 in the US with some form of special needs
    - Examples: Cystic fibrosis, neuromuscular diseases, autism spectrum, renal failure, rheumatologic conditions, survivors of neonatal intensive care (BPD, etc.)
  - There are a number of barriers to transitioning effectively and without breakdowns in medical care:
    - Training issues for physicians and other healthcare providers
    - Financial and equipment coverage issues

Medical Education and Transition of Care

- Include adult centers in advance planning.
- Involve family and patient in advance planning.
- 14 to look at readiness for transition
- Will be transitioning and to follow after transition
- Transfer of medical records to adult center

What to do in terms of medical education?

- Development of of transition curricula at all levels
- Undergraduate: teaching on long-term survival of diseases
- Graduate: Learn to care for long-term survivors and about transition processes
- Maintenance of Certification: learn how to incorporate and implement transition into practice
- Include other healthcare providers and patient and families
- Focus on NHCTC 6: National Healthcare Transition Center 6 core elements

NHCTC 6

http://www.gottransition.org

- Develop transition policy
- Establish tracking system to identify youth who will be transitioning and to follow after transition
- Conduct regular assessments at beginning age 14 to look at readiness for transition
- Develop and regularly update plans for transition. Involve family and patient in advance planning. Include adult centers in advance planning.
- Transfer of care: Set date for transfer and review transfer of medical records to adult center
- Transfer completion: Pediatric center call patient 3-6 months after last pediatric appointment to confirm follow up arrangements and success.
Suggestions for Financial Issues

- Ensure that essential health benefits are weighted to include transition services.*
- Expand Medicaid under the ACA to all 50 states.
- Require that ACO include specific care coordination activities to support transition.
- Compare ACO prospectively.
- Establish transition quality measures.

Our Current Practice

- Transition patients are identified by pediatric institution
- Annual review of patients with 2 year advance planning
- Chart records sent over 3-6 months in advance
- Pediatric and adult institution pulmonary providers are in agreement on noninvasive management pathway

Issues our patients have faced

- Financial
  - Insurance for transitioning patients changes
  - Pediatric hospital has internal durable medical equipment provider (DME)
    - All transitioning patients need a new “adult” DME provider
    - For college, patients may need new, non-family care providers
- Psychosocial
  - Patients are sometimes overwhelmed by adult hospital and different clinics
  - Parents may have difficulty with patients seeing patients on their own

What does the future hold

- Organization for transition at the institution level
- ACO will be key
- Training of interested adult physicians
- Med-Peds specialists ?
- Role of advanced respiratory therapists as case managers increased.