SLEEP DISORDERED BREATHING IN NEUROMUSCULAR DISEASE

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University of Texas Southwestern Medical Center
Associate Professor

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Sleep Disordered Breathing in Neuromuscular Disease

Won Y. Lee, M.D.

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University of Texas Southwestern Medical Center
Dallas, Texas

I have no financial disclosures to declare.
Traditional Management of Patients with Neuromuscular Diseases

- Sleep medicine specialists
  - have expertise to IMPROVE quality of life
  - can also make mistakes to WORSEN quality of life

Neuromuscular Disorders (NMD)
- Overview of neuromuscular diseases
- Physiologic testing
  - Restrictive physiology and impaired forces

Noninvasive Ventilation (NIV)
- How to qualify for a respiratory assist device?
- The Polysomnogram – Friend or Foe?
  - The double edged sword

Longitudinal Management
- Practical pearls and lessons learned
  - “With great power, comes great responsibility”
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**Pathophysiology**

**Motor dysfunction**

**Neuropathies**
- Guillain-Barre syndrome
  - Acute inflammatory demyelinating polyneuropathy
  - Molecular mimicry/autoimmune
  - Injury/loss of myelin sheath
    - Myelin → coils around nerve

- Anterior horn cell
  - Poliomyelitis (infection)
  - Acute flacid myelitis (AFM)

- Myoneural junction
  - Myasthenia gravis (antibodies)

- Myopathies
  - Muscular dystrophy

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Muscular Dystrophy

- Duchenne muscular dystrophy
  - X-linked.
  - Absence of dystrophin protein
- Proximal muscle weakness
  - Age 5 → initial symptoms
  - Age 13 → most require wheelchair before their teenage years
  - Late teens/early 20’s → chronic respiratory failure
- Cardiomyopathies

Bushby K et al. Lancet Neurol 2009; published online Nov 30. DOI:10.1016/S1474-4422(09)70271-6.
http://www.cdc.gov/ncbddd/musculardystrophy/

Amyotrophic Lateral Sclerosis (ALS)
Lou Gehrig’s Disease

- NY Yankees
  - 17 seasons
    - 2130 consecutive games
    - 1938/1939 season
      - Decreased coordination, lack of power, batting average decline
- Diagnosed with ALS
  - Mayo Clinic in 1939
- Died in 1941, at age 37

Amyotrophic Lateral Sclerosis

- Relentless, progressive, and incurable
  - Median survival of 3 to 5 years
- All races, age 40-75, Men > women
  - 90% sporadic
  - 10% familial
- Idiopathic degeneration of cells/pathologic inclusions
  - motor cortex
  - anterior horn
  - corticospinal tracts
  - corticobulbar tracts
    - Dysarthria, dysphagia, sialorrhea

Why pulmonologists need to know...

- Severe restrictive physiology
  - Progressive dyspnea
- Need for ventilatory support
  - Acute Guillain Barre Syndrome
    - 20-30%
  - Myasthenia gravis
    - 15-28%
  - ALS
    - most will die from progressive respiratory failure

Mehta S. Respir Care. 2006;51:1016.
NMD → Respiratory Muscles

- Muscles
  - Diaphragm, external intercostels, scalene, sternocleidomastoid, trapezii
- Dyspnea, orthopnea, rapid shallow breathing
- Use of accessory muscles
- Hypercarbia, hypoxemia
- Nocturnal hypoventilation


ALS and Respiratory System

Physiologic Evaluation is Important

• Reason:
  1. Quantify respiratory muscle weakness
  2. Evaluate cough effectiveness
  3. Identify those who need ventilatory support

• Tools
  – FVC and MIP
  – MEP or Peak Cough Flow

ATS/ERS Statement on Respiratory Muscle Testing. 2002

Why restrictive physiology on PFTs?

• IRV is reduced  → due to weak inspiratory muscles
• ERV is reduced  → due to weak expiratory muscles

• FVC in the supine position is ~ 10% lower than upright
  – Can drop between 12 and 65% in NM disease patients.
MIP / MEP

- Mechanical pressure gauge connected to a mouthpiece
- Electronic devices available
- Should have a small hole (1mm diameter and 20-30 mm in length) which allows an air leak.
  - Prevents patient from generating pressure by using cheek muscles

ATS/ERS Statement on Respiratory Muscle Testing. 2002

Normal MIPs and MEPs

- Men:
  MIP: – 100
  MEP: + 100

- Women:
  MIP: - 80
  MEP: + 80

<table>
<thead>
<tr>
<th>Age Group</th>
<th>MIP</th>
<th>MEP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children (ages 7 to 13)[2]</td>
<td>Male: 77 to 114</td>
<td>99 to 101</td>
</tr>
<tr>
<td></td>
<td>Female: 71 to 108</td>
<td>74 to 126</td>
</tr>
<tr>
<td>Adolescents (ages 13 to 35)[2]</td>
<td>Male: 114 to 121</td>
<td>131 to 161</td>
</tr>
<tr>
<td></td>
<td>Female: 85 to 93</td>
<td>92 to 95</td>
</tr>
<tr>
<td>Adults (ages 18 to 65)[3]</td>
<td>Male: 92 to 121</td>
<td>140*</td>
</tr>
<tr>
<td></td>
<td>Female: 68 to 79</td>
<td>95*</td>
</tr>
<tr>
<td>Older adults (ages 65 to 85)[4]</td>
<td>Male: 65 to 90</td>
<td>140 to 190</td>
</tr>
<tr>
<td></td>
<td>Female: 45 to 60</td>
<td>90 to 130</td>
</tr>
</tbody>
</table>

UpToDate
# Neuromuscular Disorders (NMD)
- Overview of neuromuscular diseases
- Physiologic testing
  - Restrictive physiology and impaired forces

# Noninvasive Ventilation (NIV)
- How to qualify for a respiratory assist device?
- The Polysomnogram – Friend or Foe?
  - The double edged sword

# Longitudinal Management
- Practical pearls and lessons learned
  - “With great power, comes great responsibility”

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**Respiratory muscle weakness**

- Noninvasive
  - Negative pressure ventilation
  - Positive pressure ventilation
- Invasive
  - Tracheostomy
- Palliative Measures
**Negative Pressure Ventilation (NPV)
The Iron Lung**

- Augments normal spontaneous breathing
  - Negative pressure
    - Rotary pumps causes thoracic expansion, pressure gradient

- Poliomyelitis epidemic
  - Copenhagen in 1952
  - 31 patients, 27 died
    - Within 3 days despite negative pressure ventilation.

West JB. J Appl Physiol 2005;99:424-432

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**Positive Pressure Ventilation**

- **Patient 32 (12 year girl)**
  - Dr. Bjorn Ibsen
    - tracheostomy
    - positive pressure ventilation
    - manual pressure from a rubber bag

- **Up to 1500 medical and dental students**
  - 6-8 hour shifts around the clock to deliver positive pressure ventilation

West JB. J Appl Physiol 2005;99:424-432
Best Clinical Practices

• Task Force → evidence and consensus-based standardized NIV titration guidelines

• Chronic Alveolar Hypoventilation syndromes secondary:
  – central respiratory control disturbances (CRCD)
  – restrictive thoracic cage disorders (RTCD) → scoliosis
  – neuromuscular diseases (NMD)
  – obesity hypoventilation syndrome (OHS)


Survival Benefit for use of NIV in ALS

<table>
<thead>
<tr>
<th>Author, year</th>
<th>Study design</th>
<th>NIV device</th>
<th>NIV started</th>
<th>Participants &amp; treatments</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pinto, 1985</td>
<td>NCT Bi-level PAP</td>
<td>Daytime hypercapnia or hyperpnea, Daytime orthopnea, hypoventilation or both</td>
<td>10</td>
<td>NIV 10 standard</td>
<td>3-year survival higher with NIV (87.5% vs 22.2%, P &lt; .004)</td>
</tr>
<tr>
<td>Aboussouan, 1997</td>
<td>Obs BiPAP ST mode or PLV-100</td>
<td>Respiratory symptoms, FVC &gt;50% predicted, or FEv &lt;15% in 3 months</td>
<td>21</td>
<td>NIV &gt;4th nocturnal intubated 15</td>
<td>Median survival 2 months in those NIV intolerant, 15 months NIV tolerant (P &lt; .001)</td>
</tr>
<tr>
<td>Keipea, 1990</td>
<td>Obs Bi-level PAP</td>
<td>Respiratory symptoms and FVC &lt;50% or MIP &lt;60 cm H2O</td>
<td>38</td>
<td>NIV&lt;4hrs/52 refused NIV</td>
<td>Mean survival 14.2 mo &gt;4hrs (P&lt;0.001), 7.0 mo &lt;4 hrs (P=0.038), 4.6 mo refused NIV</td>
</tr>
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<td>Grui, 2006</td>
<td>Obs Bi-level PAP, S mode</td>
<td>Orthopnea &amp; hypoxemia</td>
<td>18</td>
<td>NIV &gt;4 hrs nocturnal intubated 15</td>
<td>NIV tolerant deceased risk of death (HR 0.23) 95% CI (0.10, 0.54)</td>
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<tr>
<td>Bourke, 2006</td>
<td>RCT VPA® S/T, ST mode</td>
<td>Orthopnea &amp; MIP &lt;60% or hyperscapnia</td>
<td>22</td>
<td>NIV 19 standard</td>
<td>Median survival benefit 205 days with NIV (P = .006)</td>
</tr>
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</table>

NCT, nonrandomized controlled clinical trial; Obs, observational study; RCT, randomized controlled clinical trial; NIV, noninvasive positive-pressure ventilation; PAP, positive airway pressure; FVC, forced vital capacity; MIP, maximum inspiratory pressure; MV, maximum voluntary ventilation; MIP, MV, or negative inspiratory force are often used interchangeably; PLV-100, volume-controlled portable ventilator in assist-control mode (LifeCare Products, LaBretta, CO); BiPAP® Respironics, Inc.; Murname, Roff WPA® STI RespMed, UK Ltd, Abingdon, UK; SV, spontaneous breath mode; S, spontaneous mode; HR, heart rate; CI, confidence interval; cm, centimeters; h, hours; mo, months.

NIV improves ALS survival in those with preserved bulbar function

Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial

- ALS patients
  - Randomized
    - NIV (22 patients) vs. no ventilation (19)
  - Orthopnea, MIP < -60, or symptomatic hypercapnia
- NIV improved quality of life, sleep quality, and survival mostly in ALS patients without bulbar disease
  - Increased survival by 205 days
- Good bulbar function → 9.3 hours per day
- Poor bulbar function → 3.8 hours per day
- Settings
  - Average IPAP of 15 cm H2O
  - Average EPAP of 4 cm H2O


Why does optimal NIV settings/titration matter?

4.1.8 Attended NPPV titration with polysomnography allows definitive identification of an adequate level of ventilatory support for patients with NMD in whom NPPV treatment is planned. (Level A - Consensus)

- Preserve quality of life
  - Maintain ability to communicate
  - Improve sleep quality
- Reduce morbidity
  - Decrease carbon dioxide
  - Avoids morbidity involved with tracheostomy
- Reduce mortality
  - Extend duration of life

Do we need a DIAGNOSTIC sleep study to facilitate initiating a RAD for NMD?

- Common phone call
  - “I have a patient with a diagnosis of a NMD (ALS, muscular dystrophy, etc…) and hypercapnic respiratory failure.”
  - “The patient has done GREAT on bilevel PAP in the hospital.”
  - “I am told that the patient needs a diagnostic attended polysomnogram to get his bilevel PAP device.”
  - True or False?

Bilevel PAP Devices

**Coding**

- **E0470** – Respiratory Assist Device, Bi-Level Pressure, Without Backup Rate Feature
  - Delivers adjustable, variable levels of positive air (during single respiratory cycle) and supplements volume of air into the lungs
- **E0471** – Respiratory Assist Device, Bi-Level Pressure, With Backup Rate Feature
  - Has the same features as E0470, with the addition of timed backup feature to deliver air when insufficient inspiratory efforts fail

www.medicare.gov
Why is a sleep study NOT needed to initiate NIV?

Initial Coverage Criteria

- **Restrictive Thoracic Disorders**

  **A** Documentation of progressive neuromuscular disease, or severe thoracic cage abnormality

  1. Arterial blood gas (while awake) ≥ 45 mm Hg or
  2. O2 saturation ≤ 88% for at least 5 continuous minutes
  3. For progressive neuromuscular disease (only) maximal inspiratory pressure is < 60 cm H2O or Forced vital capacity is %50 predicted and

  **B**

  1. Chronic Obstructive pulmonary disease does not contribute significantly to beneficiary’s pulmonary limitations
  2. If criteria A-C are met, either E0470 or E0471 will be covered for the first three months.
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Sleep and breathing in neuromuscular disease

NORMAL SUBJECTS

- Decrease in minute ventilation
- Decrease in tidal volume
- Increase in respiratory rate

- NREM
- REM
- Tonic REM
- Phasic REM

Fig. 1: a) Minute ventilation, b) tidal volume and c) respiratory rate during wakefulness and sleep in 19 normal adult subjects. Data are presented as mean±SEM. * indicates lower during sleep compared to wake p<0.05; ** indicates lower during sleep compared to wake p<0.01. *: tidal volume lower during sleep compared to wake p<0.05; **: p<0.01; #: respiratory rate higher in rapid eye movement (REM) compared to non-REM eye movement sleep (NREM) or wake p<0.05. Data from O’Donnell et al. [65].
Is a sleep study needed?

- "Daytime respiratory function has greater prognostic value than nocturnal measurements."

- In comparison to daytime lung function [and symptoms], nocturnal measurements are surprisingly WEAK predictors of survival.

- There is NO evidence that sleep studies improve the selection of subjects for NIV over and above symptoms and daytime respiratory function.

A diagnostic sleep study is NOT needed to initiate NIV therapy

- "...daytime respiratory function has greater prognostic value than nocturnal measurements."

- Confirmed NMD Diagnosis AND one of the following...
  - PaCO2 > 45 mm Hg
  - FVC < 50%
  - MIP < -60
  - SpO2 < 88% for 5 consecutive minutes (min 2 hour recording)

- These patients can EITHER:
  - DIRECTLY obtain a respiratory assist device
  - Go DIRECTLY to the sleep laboratory or hospital for optimal titration
Clinical Case #1

- 37 F with limb girdle muscular dystrophy.
  - Wheelchair limited. Marked dyspnea.
  - FVC 19% and MIP – 17

- Qualifies for NIV?

- Evaluated by a pulmonary/sleep specialist
  - Diagnosed with mild sleep apnea (AHI 5.1)
  - Titrated, then retitrated to CPAP 19 cm H2O
    - Choking, suffocating, dyspnea is markedly worse.

- We switched her from CPAP to NIV during nighttime and daytime mode ventilation
  - Marked improvement in quality of life
Clinical Case #2

• 35 F, diagnosed with bulbar ALS early this year and referred to discuss ventilation options.
  – She is getting more dyspneic and rapidly weakening.

• Spirometry and forces
  – FVC 45%
  – MIP -20

• ABG
  – pH 7.32, PaCO2 55mm Hg, and PaO2 of 62 mm Hg

• Qualifies for NIV?

• She had a diagnostic sleep study 8/1/17
  – "poor sleep efficiency, AHI of 1.5, no sleep related breathing disorder" so unfortunately it wasn't super helpful.

• Can you help?

Clinical Case #3

• 51 year old female
• Shrinking lung syndrome, SLE
• Gradual dyspnea
  – FVC 27%
  – MIPs -42
  – 2 diagnostic PSGs
    • Both showed an AHI < 5

• Which of the following is the next appropriate step?
  – A. Repeat a 3rd diagnostic sleep study
  – B. Order oxygen, she is not a candidate for noninvasive ventilation (NIV)
  – C. Order a bilevel PAP with back up rate at settings of 8/4 cm H2O with a rate of 10 and gradually increase as tolerated
  – D. Order a bilevel PAP in AVAPS mode
  – E. Perform a titration sleep study using bilevel PAP with back up rate to meet patients’ respiratory needs
NMD and Sleep Medicine

• **Strengths**
  - Expertise in noninvasive ventilation
    - Synchrony to optimize sleep quality, ventilation, and oxygenation
  - Expertise in mask interfaces
  - Compliance monitoring

• **Pitfalls of sleep medicine**
  - The current state of sleep medicine training, does not focus on NMD patient population
    - Excess focus on OSA
  - Complexities of respiratory physiology

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**Lessons Learned**
Protocols and equipment to accommodate for NM patients in the sleep lab

- Hospital bed
  - 2 of our beds
- Hoyer lift
- Suction
- Supplemental O2
- Call system
- Accommodations for a care giver
- Technical expertise
  - RRT and RPSGT
  - 1:1 if needed
**Goals of NIV**

1. Decrease work of breathing
2. Optimize ventilation and oxygenation
3. Tolerance to NIV
   - Minimize mask leakage
   - Good sleep quality

**Start Slow**

- Start 8/4 cm H2O with back up rate
- Increase IPAP to augment tidal volume
  - Goal tidal volume of 8-10 mL/kg
    - Ideal body weight
- Increase back up rate to match needs
- Example
  - 15/5 cm H2O
  - TV ~600 mL

**4.3 Recommendations for Initial and Maximum Pressures during NPPV Titration**

4.3.1. The recommended minimum starting IPAP and EPAP should be 8 cm H2O and 4 cm H2O, respectively. (Level A - Consensus)

4.4.2 Recommendations for adjusting pressure support for low tidal volume or hypoventilation during sleep

4.4.2.1 The PS should be increased every 5 minutes if the tidal volume is below the acceptable goal. An acceptable tidal volume goal for most patients ranges from 6 to 8 mL/kg using ideal body weight (Figure 3). (Level A - Consensus)

4.3.5 The minimum and maximum incremental changes in PS during NPPV titration should be 1 and 2 cm H2O, respectively. (Level A - Consensus)

4.3.4 The recommended maximum IPAP should be 20 cm H2O for patients < 12 years and 30 cm H2O for patients ≥ 12 years. (Level A - Consensus)
BPAP ST Mode

- BPAP ST
  - NPPV in the spontaneous-timed (ST) mode provides a backup rate to ensure a minimum respiratory rate.
  - For example, if the back-up rate is 10 bpm, the time window following the previous breath is 6 seconds.
  - If a spontaneous breath does not occur, the device provides a machine triggered breath.


Pressure Control
Fixed Inspiratory Time

Selim B, Wolfe L, Coleman J, and Dewan N
Achieve Muscle Rest

- Bipolar electrodes
  - Surface diaphragm electrodes
    - 2 electrodes, 2 cm apart horizontally in 7-8th intercostal spaces in right anterior axillary line (reduce EKG artifact)
  - Sternocleidomastoid muscles
  - Right parasternal intercostal muscles
    - 2nd and 3rd intercostal spaces in mid-clavicular line

Initial settings

Bilevel PAP 8/4 cm H2O with rate 16
Better settings
Reduction in accessory muscle use

Suboptimal – Bilevel 8/4 with rate 10
Irregular breathing, low tidal volumes (250’s mL)...
**Optimal – Bilevel 16/5 rate 20**

Regular breathing and better tidal volumes (550 mL)...  

<table>
<thead>
<tr>
<th>Setting</th>
<th>Recommendation</th>
<th>Reason</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trigger</td>
<td>Sensitive</td>
<td>Weak inspiratory muscles</td>
</tr>
<tr>
<td>Inspiratory time</td>
<td>Make it longer</td>
<td>Recruit alveoli and avoid atelectasis</td>
</tr>
<tr>
<td>Tidal volumes</td>
<td>More is better</td>
<td>8-10 ml/kg</td>
</tr>
</tbody>
</table>
**Pearl**
Not all NMD can tolerate NIV

- **Bulbar disease**
  - May trigger vocal cord spasm
  - Sialorrhea (drooling)
    - medications (Robinul, Levsin, Scopolamine, atropine, BoTox)
  - Suboptimal mask fit, poor seal

- **Claustrophobia**
  - Myotonic dystrophy
  - Weakened upper extremity strength, inability to remove mask

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**Clinical Case**

- 32 year man
- Myotonic dystrophy
- FVC 82%
- MIP - 54

- Poorly tolerated bilevel PAP
  - Poor sleep efficiency
  - Severe desaturations
  - No REM
Pearl

NM patients may require several interfaces

- Sleep clinics/labs have access to a wide variety of interfaces
  - “creativity”

- Interface needs
  - Daytime
  - Nighttime
  - Chin straps may be necessary

Volume Assured Pressure Support (VAPS)

**BIPAP AVAPS**

**Technology**

AVAPS—advanced technology that simplifies patient care

It’s the only way of ensuring the delivery of targeted tidal volumes for continuous ventilation patients. Standardized for "Average Volume Assured Pressure Support." 8700 was the first to introduce it in respiratory ventilators. The exclusive AVAPS algorithm automatically regulates pressure support to meet changing patient needs while maintaining a target tidal volume.

Advanced leak-compensating technology with digital Auto-Trak

Our advanced univolved digital Auto-Trak algorithm enables clinicians to achieve optimal patient-ventilator synchrony.

- **Adjustment of Ti min** and **Ti max**
- Automatically adjusts its variable triggering and cycle deadtimes

**Improved humidification**

System line humidity control analyzes ambient temperature, relative humidity and patient flow to deliver optimal humidity—real ultimate comfort—to the patient while also dramatically reducing mucus.

**Easy access to patient data**

- Respiratory parameters, flow, tidal volume
- **Easy access to patient data**

**Volume Assured Pressure Support (VAPS)**

**S9 VPAP ST-A clinical settings — IVAPS mode**

**Primary Settings**

<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>DEFAULTS</th>
<th>DESCRIPTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Target Tl</td>
<td>5.2 L</td>
<td>Target tidal ventilation (L) is the main parameter that helps to determine the amount of pressure support required.</td>
</tr>
<tr>
<td>BIP</td>
<td>4 x 0.75</td>
<td>BIP is the pressure delivered when the device is cycled into expiration.</td>
</tr>
<tr>
<td>Height</td>
<td>37.5 cm</td>
<td>The patient’s height or arm span is needed to determine dead space.</td>
</tr>
<tr>
<td>Target Fio2</td>
<td>0.95</td>
<td>Target patient rate to the reference point that VPAP uses to determine the range for the backup rate. This should be set at or near the patient’s actual respiratory rate (RPM).</td>
</tr>
</tbody>
</table>

**Synchronization Settings**

<table>
<thead>
<tr>
<th>PARAMETER</th>
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<th>DESCRIPTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>T Min</td>
<td>0.0 seconds</td>
<td>Sets the maximum limit of the time the device operates in BIP.</td>
</tr>
<tr>
<td>T Max</td>
<td>15.0 seconds</td>
<td>Sets the minimum limit of the time the device operates in BIP.</td>
</tr>
<tr>
<td>Min P&amp;</td>
<td>0 x 0.75</td>
<td>Minimum pressure support in IVAPS mode.</td>
</tr>
<tr>
<td>Max P&amp;</td>
<td>20 x 0.75</td>
<td>Maximum pressure support in IVAPS mode.</td>
</tr>
</tbody>
</table>

- **Adjustment of Ti min** and **Ti max**


Volume Assured Pressure Support (VAPS)

Volume-assured pressure support modes (VAPS)
AVAPS (assured volume assured pressure support; ResMed) targets expiratory tidal volume
IVAPS (intelligent volume assured pressure support ResMed) targets alveolar ventilation (minute ventilation minus death space ventilation)

Selim B, Wolfe L, Coleman J, and Dewan N

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Longitudinal Management
- Practical pearls and lessons learned
  - “With great power, comes great responsibility”
Sleep clinics have expertise obtaining download data

1. Minute ventilation
   - Exhaled tidal volumes
   - Respiratory rate

1. Percent patient trigger

1. Hours of usage

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### Lessons Learned

Assessment of compliance data is important

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#### Table

<table>
<thead>
<tr>
<th>Variable</th>
<th>Median</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minute ventilation</td>
<td>10.5</td>
<td>8.5</td>
<td>12.5</td>
</tr>
<tr>
<td>Exhaled tidal volumes</td>
<td>10.5</td>
<td>8.5</td>
<td>12.5</td>
</tr>
<tr>
<td>Respiratory rate</td>
<td>10.5</td>
<td>8.5</td>
<td>12.5</td>
</tr>
</tbody>
</table>

#### Assessments

- **Assessment of compliance data:**
  - Compliance Summary
  - Hours of usage
  - Pressure

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Is an FVC $< 50\%$ too low?

How good are we in providing NIV for NM patients?

Missed opportunities?

The #1 Way to treat ARDS: Low tidal volume ventilation

<table>
<thead>
<tr>
<th>Low Tidal Volume</th>
<th>Traditional Tidal Volume</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>3.1 l/min</td>
<td>3.1 l/min</td>
<td>.002</td>
</tr>
<tr>
<td>Ventilator peak inspiratory pressure (cm H$_2$O)</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>Oxygen saturation (%)</td>
<td>93</td>
<td>92</td>
</tr>
</tbody>
</table>

Ventilator strategy in LUNG SAFE:
- 1/3 of patients never recognized to have ARDS
- $P_{aO_2}$ measured in 40%
- Less than 2/3 received TV $\leq$ 8 mg/kg

Angela Rogers, MD – CTS 2019
Summary Slide

1. Neuromuscular disorders benefit from NIV
   - Quality of life
   - Morbidity
   - Mortality

1. Pulmonary physiology determines obtaining a NIV
   - PaCO2 > 45
   - FVC < 50%
   - MIP < -60
   - PaO2 < 88% for 5 minutes

3. An FVC of < 50% may be “too late,” but...
   - Make sure to check supine FVC
   - Make sure to get an MIP

4. A diagnostic sleep study is unnecessary to obtain a NIV
   - However a TITRATION sleep study can be very helpful

5. Sleep trained clinicians can make an important and beneficial impact on NM patients

Traditional Management of Patients with Neuromuscular Diseases

- Sleep medicine specialists
  - have expertise to IMPROVE quality of life.
Sleep Disordered Breathing in Neuromuscular Disease

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