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# Respiratory Management of Patients With Neuromuscular Weakness Series: Review Recommendations and Best Practice

By Duane Reed, EdD, RRT, RCP

# Duane Reed, EdD, RRT, RCP

Dr. Duane Reed received his Doctoral degree in Education from Walden University. He completed his Master's degree in Adult Education from Central Michigan University and a Bachelor's degree in Advanced Respiratory Care from Weber State University. He is the Respiratory Care Program Director at Southern Crescent Technical College. His clinical experience entails over 20+ years as a Respiratory Care Practitioner at Grady Memorial Hospital in Atlanta, Georgia, working in critical care areas of surgical, cardiac, medical, and neurointensive care units. Over the last ten years, Dr. Reed has contributed to educational publishing companies as a clinical editor and senior reviewer. Additionally, Dr. Reed owns Pulmonary Education Consultants, LLC, a company providing respiratory care educational materials and tutoring services. He has authored study cards in areas of mechanical ventilation, which are nationally published, helping respiratory therapists around the country.



# Disclosures

- **Presenter Disclosure: Financial:** Dr. Reed is the owner of Pulmonary Education Consultants, LLC. He received an honorarium for presenting this course.  
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# Learning Outcomes

After this course, participants will be able to:

- Identify neuromuscular diseases that affect the respiratory system.
- Identify evidence-based recommendations for patients with neuromuscular diseases.
- Discuss best practice management for patients with neuromuscular diseases.

# Overview of Neuromuscular Diseases

- A neuromuscular disease (NMD) occurs when the nerves that control movement are adversely affected.
- Muscle weakness is a complication that is created from neuromuscular diseases including the respiratory system.
- Respiratory failure is a common complication associated with neuromuscular diseases.

# Overview of Neuromuscular Diseases

- Patients with NMD can have muscle weakness which can lead to lack of adequate ventilation, PCO<sub>2</sub> retention at night, and the lack of the patient's ability to mobilize secretions which can be a cause of death.
- There were according to the Cochrane Review no studies that compared invasive and noninvasive mechanical ventilation or positive versus negative ventilation for NMD. This lack of research has made it difficult to provide a single set of guidelines.

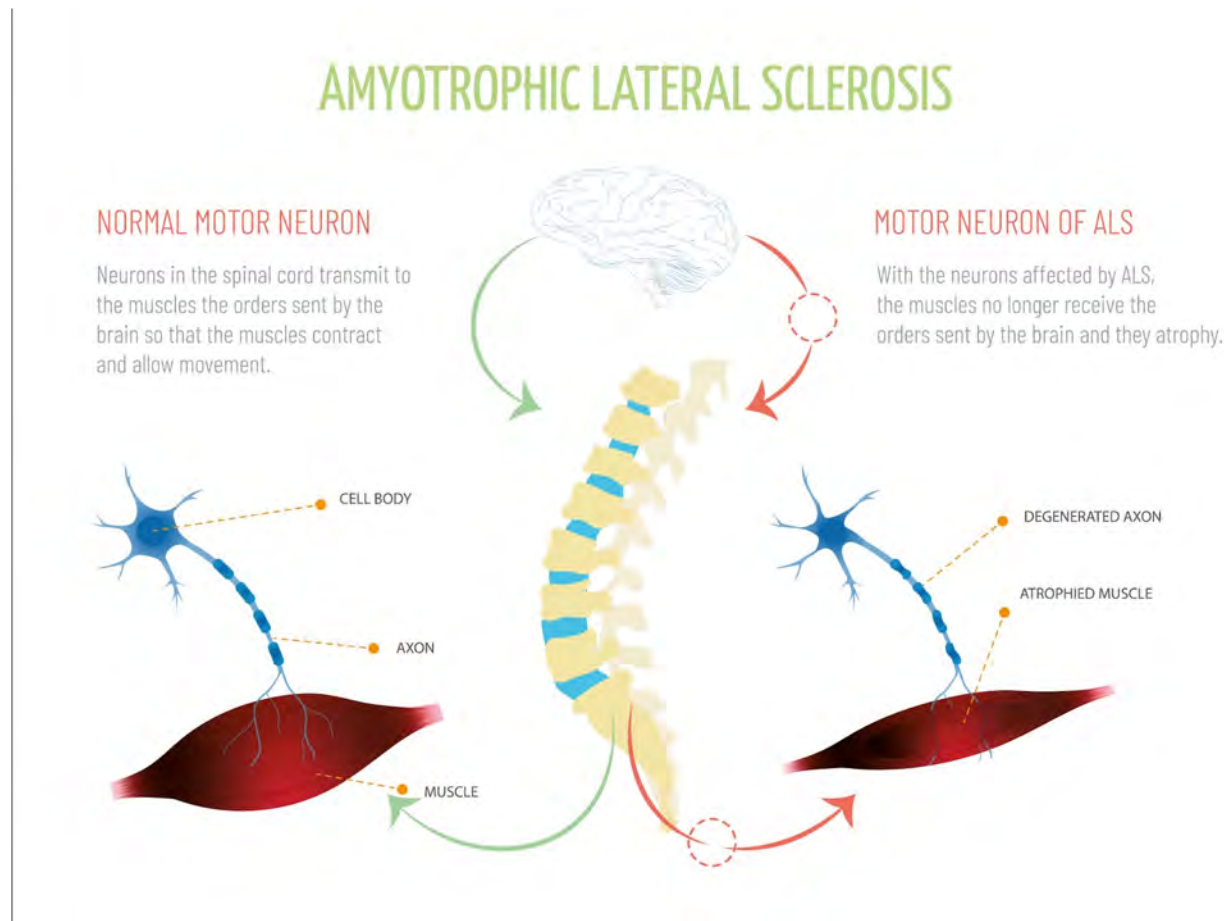
# Overview of Neuromuscular Diseases

Many forms of the neuromuscular disease exist, which are:

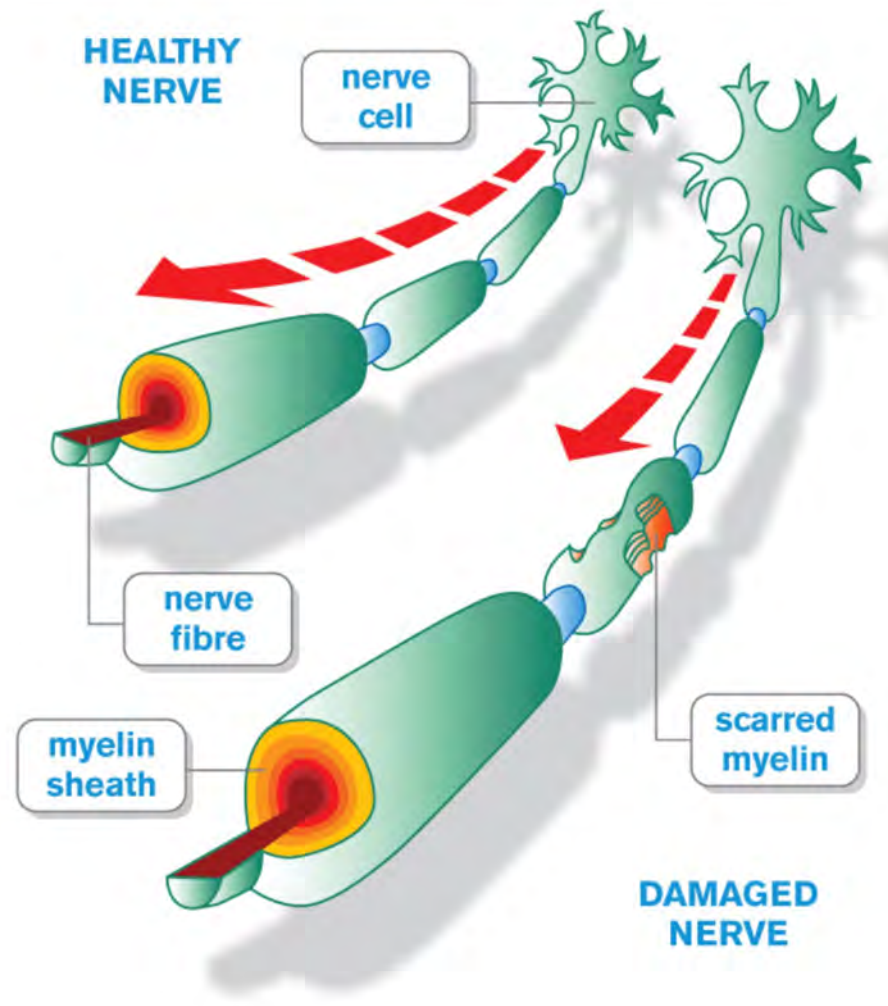
- Amyotrophic Lateral Sclerosis (ALS)
- Charcot Marie Tooth Disease (CMT)
- Multiple Sclerosis (MS)+
- Muscular Dystrophy



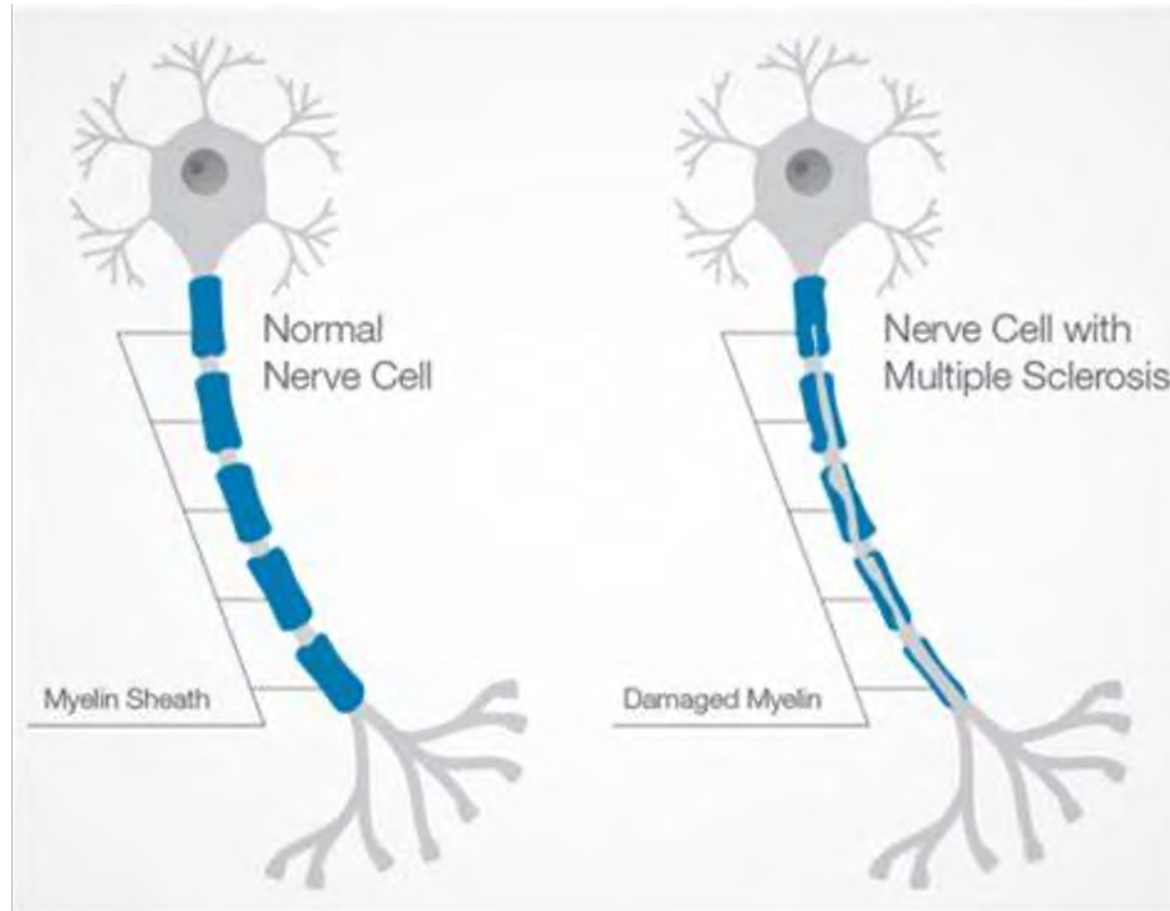
# Amyotrophic Lateral Sclerosis (ALS)



# Charcot Marie Tooth Disease



# Multiple Sclerosis



# Other Neuromuscular Diseases

- Myasthenia Gravis
- Myopathy
- Myositis
- Guillain Barre Syndrome (not a disease but when the immune system attacks the neuro system)

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

- Clinical experts (2023) comprised a multi-professional panel to conduct a systematic review to address patients with neuromuscular diseases.
- The panel focused on the respiratory management of NMD and applied the grading, recommendations, assessment development, and evaluations.
- Based on 128 studies, the panel generated 15 graded recommendations, one good practice statement and one consensus-based statement.

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 1: Use and Timing of PFT

For patients with NMD at risk for respiratory complications, the panel recommends Pulmonary Function Testing (PFT) to assist with the management

## Panel Suggestion: PFT is a low-cost intervention

Panel suggested that spirometry with FVC or SVC MIP/MEP, Sniff Nasal Inspiratory Pressure (SNIP) and Peak Cough Flow (PCF) be considered in patients with NMD

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 2: Use and Timing of PFT

For patients with NMD at risk for respiratory complications, the panel recommend PFT at a minimum of every 6 months as appropriate

## Panel Suggestion:

Panel suggested that one or more of the following be performed at least every 6 months, VC, SVC, MIP, MEP, SNIP, PCF

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## Panel Recommendation 3: Sleep-Related Disorder

For patients with NMD with normal PFT and overnight oximetry (ONO) panel suggest polysomnography to assess the need for Noninvasive Ventilation (NIV)

### Panel Suggestion:

Panel suggested that polysomnography will be a good tool to see if NIV is indicated for NMD patients



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## Panel Recommendation 4: NIV

For patients with NMD and chronic respiratory failure the panel recommended using NIV

## Panel Suggestion:

Panel mentioned that clinical indications for NIV although indicated, can vary depending on the NMD, age, and rate of disease progression

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## Panel Suggestion 4:

Panel also mentioned that any fall in FVC to  $< 80\%$  of predicted with symptoms or FVC  $< 50\%$  of predicted without symptoms or SNIP/MIP  $< -40$  CM H<sub>2</sub>O would indicate a need for NIV.

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 5: NMD and Sleep

For patients with NMD and sleep-related breathing disorders, the panel mentioned using NIV for treatment

### Panel Suggestion:

Panel suggested using the American Academy of Sleep Medicine (AASM) criteria for sleep disorder and hypoventilation for adults, and the European Respiratory Society (ERS) for pediatric patients

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 6: Parameters for NIV

For patients with NMB the panel suggested the use of diagnostic tests such as FVC, MIP/MEP, ONO, or sleep disorder breathing or hypoventilation on the polygraph to predict the timing of NIV initiation

## Panel Suggestion:

Panel suggested that Polysomnography is not necessary for adults to initiate NIV, and PFT criteria alone may be adequate

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 7: Treatment for NIV

For patients with NMB requiring NIV the panel mentioned that individualizing NIV treatment to achieve ventilation goals

### Panel Suggestion:

Panel suggested adjusting mode, inspiratory time, and inspiratory and expiratory pressures. No mode is suggested over the other

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Suggestion 7:

Panel also suggested that a backup respiratory rate may lead to better patient-ventilator synchrony and improved gas exchange.

It was noted that patients with **bulbar impairment** may not be able to tolerate NIV. Panel also suggested an ongoing assessment of sleep quality, digital downloads, leaks, oximetry (capnography), and secretion management

# Bulbar Impairment

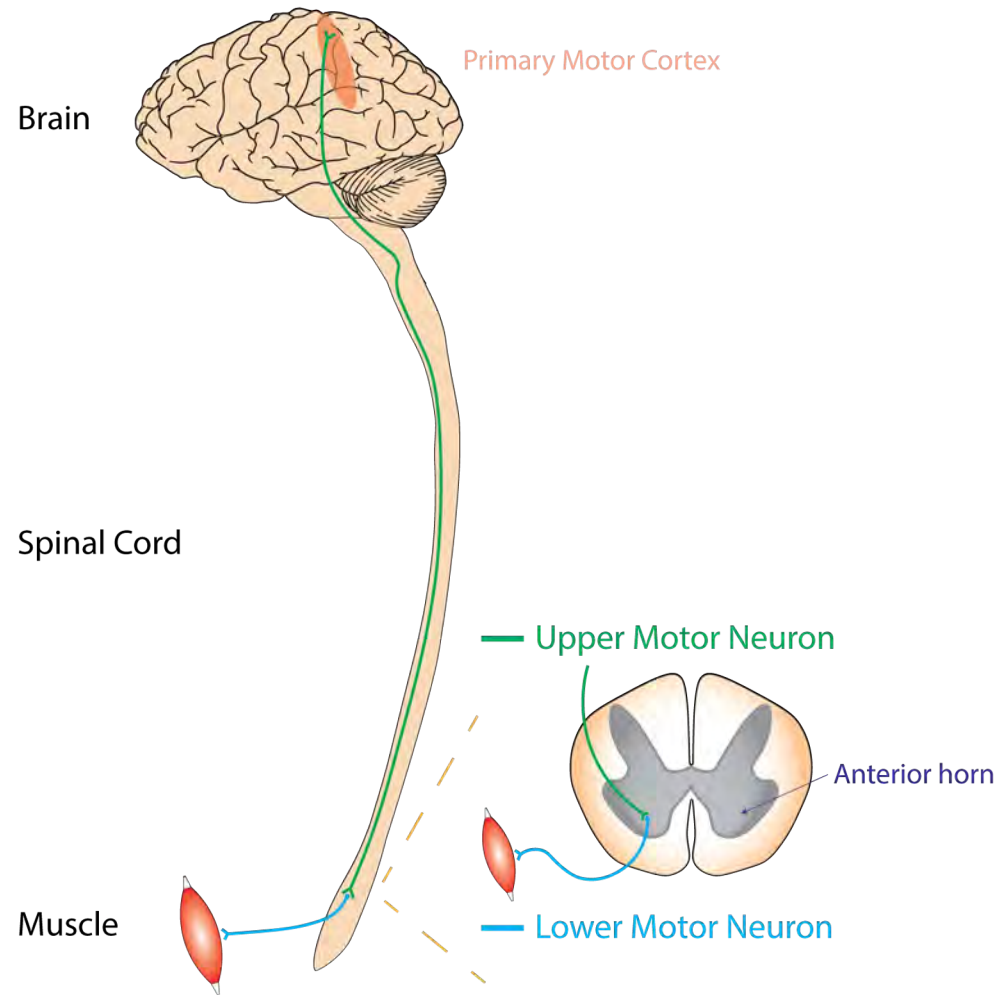


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## Panel Recommendation 8: NMD and Bulbar

For patients with NMB and preserved bulbar function using NIV the panel mentioned Mouthpiece Ventilation (MPV) for daytime as an adjunct to nocturnal mask NIV

## Panel Suggestion:

Panel suggested although MPV has been used for NMDs to delay mechanical ventilation although bulbar symptoms in some NMDs may limit its use



# Mouthpiece Ventilation



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## Panel Recommendation 9: Mechanical Ventilation

For patients with NMB that fail NIV, worsening bulbar function, aspiration, insufficient cough, and declining lung function, home MV via trach is a good alternative

## Panel Suggestion:

Panel suggested that MV be started as early as possible and include goals of care.

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## Panel Suggestion 9:

Panel also suggested optimizing secretion management and airway clearance, using patient preference, treatment goals, quality of life considerations, and available resources to help make decisions

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 10: NMD and Sialorrhea

For patients with NMB and sialorrhea, panel recommends a therapeutic trial of an anticholinergic medication as first-line therapy

### Panel Suggestion:

The panel suggests an initial trial of an oral anticholinergic medication

# Sialorrhea

- Sialorrhea, which is also known as drooling occurs when there is excess saliva in the mouth beyond the lip margin.
- Sialorrhea can occur with several neurologic disorders such as ALS, Cerebral Palsy, Parkinson disease or side effects of medications
- Cerebral Palsy is the most common cause of sialorrhea in children

# Sialorrhea

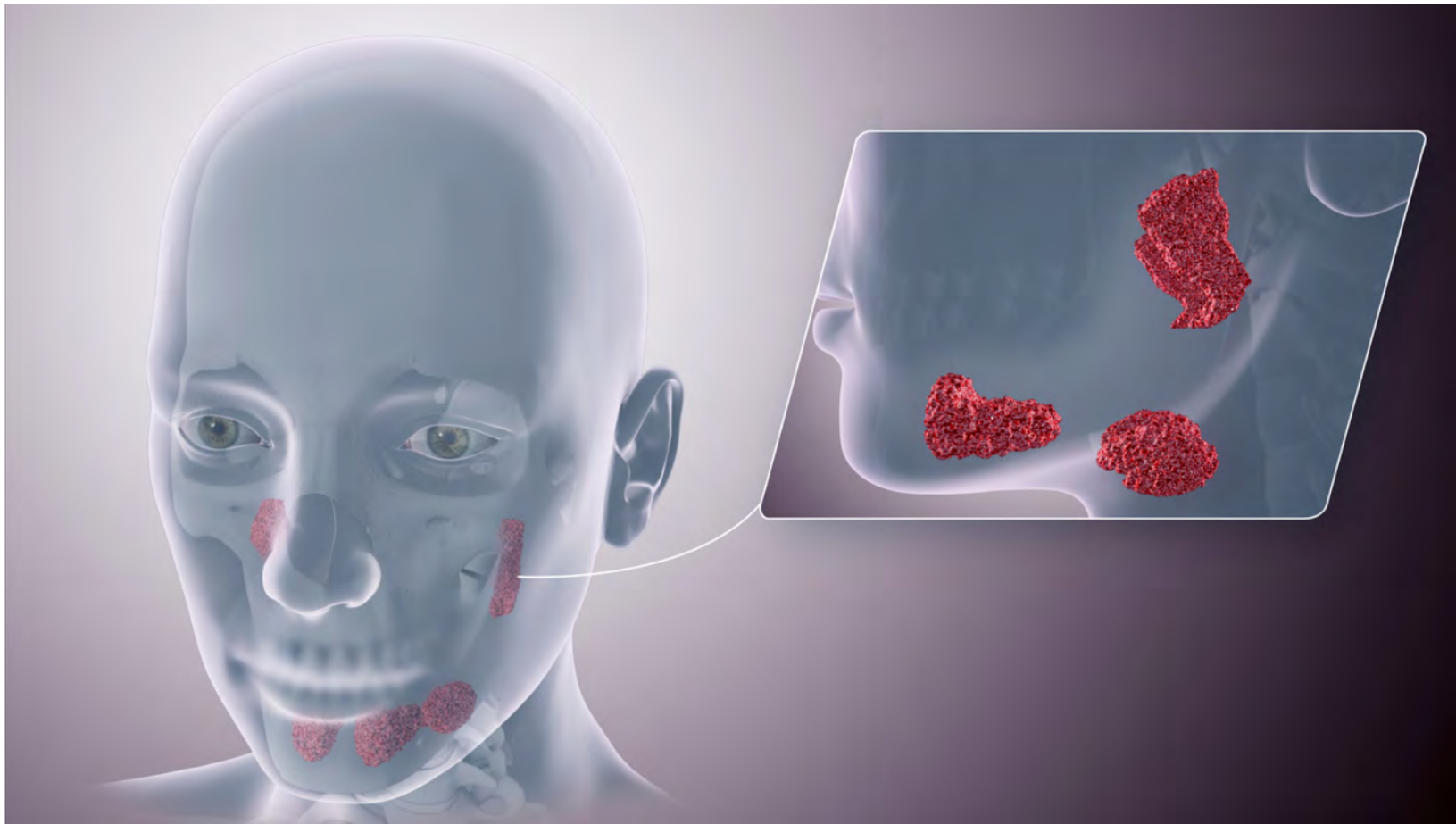


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# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 11: NMD and Sialorrhea

For patients with NMB and sialorrhea who have an inadequate response or are intolerant of the side effects of anticholinergic therapy

### Panel Suggestion:

The panel suggests a Botulinum toxin (BT) therapy to salivary glands

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 12: NMD and Sialorrhea

For patients with NMB and sialorrhea who have an inadequate response or are intolerant of the side effects of anticholinergic therapy

### Panel Suggestion:

The panel suggests salivary gland radiation



# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 13: NMD and Sialorrhea

For patients with NMB and hypoventilation the panel recommend **Glossopharyngeal Breathing** (GPB) for lung volume recruitment (LVR)

### Panel Suggestion:

The panel suggests LVR because of low cost and can be performed by patient independently with minimal assistance and training

# Glossopharyngeal Breathing (GPB)

- Glossopharyngeal breathing is a positive pressure breathing technique to assist with failing respiratory muscles.
- The glottis is used to trap the air in the lungs while the gulp of air is being processed.
- Glossopharyngeal breathing can be used to improve voice and cough efficacy.

# Glossopharyngeal Breathing (GPB)

## Glossopharyngeal breathing

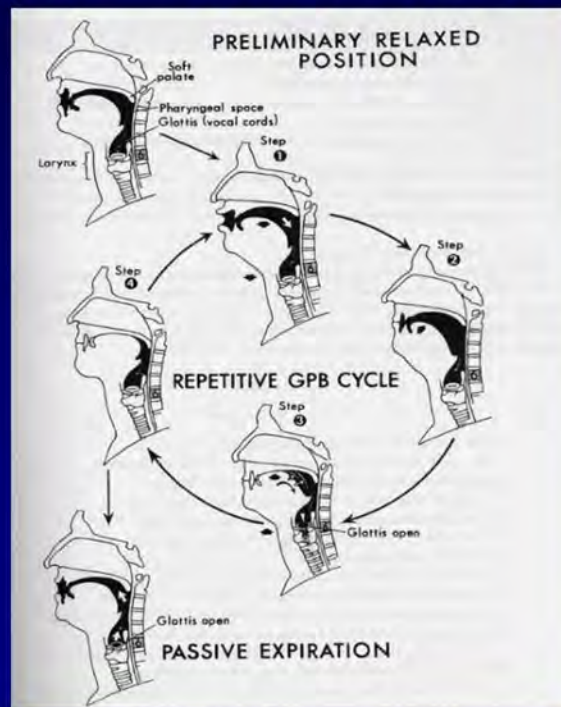


Image source: Warren, V.C. (2002)

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 14: NMD/reduced cough

For patients with NMB and reduced cough the panel recommend manually assisted cough techniques

## Panel Suggestion:

The panel suggests the techniques be done independently or added to other modalities such as LVR

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 15: NMD/reduced LVR

For patients with NMB and reduced LVR and cough, the panel recommends regular use of LVR using a handheld resuscitation bag or mouthpiece

## Panel Suggestion:

The panel suggests the technique is low-cost but requires assistance and training.

Manually assisted cough is more effective when added to volume recruitment or expiratory cough assist

# Advancing Research on Respiratory Management of Patients with Neuromuscular Weakness

## Panel Recommendation 16: NMD/reduced cough

For patients with NMB and reduced cough effectiveness which cannot be improved with alternative techniques

### Panel Suggestion:

The panel suggests the addition of regular mechanical insufflation-exsufflation (MI-E; cough assist device)

# Mechanical insufflation-exsufflation (MI-E; cough assist device)



Image source: Example of a MI-E device

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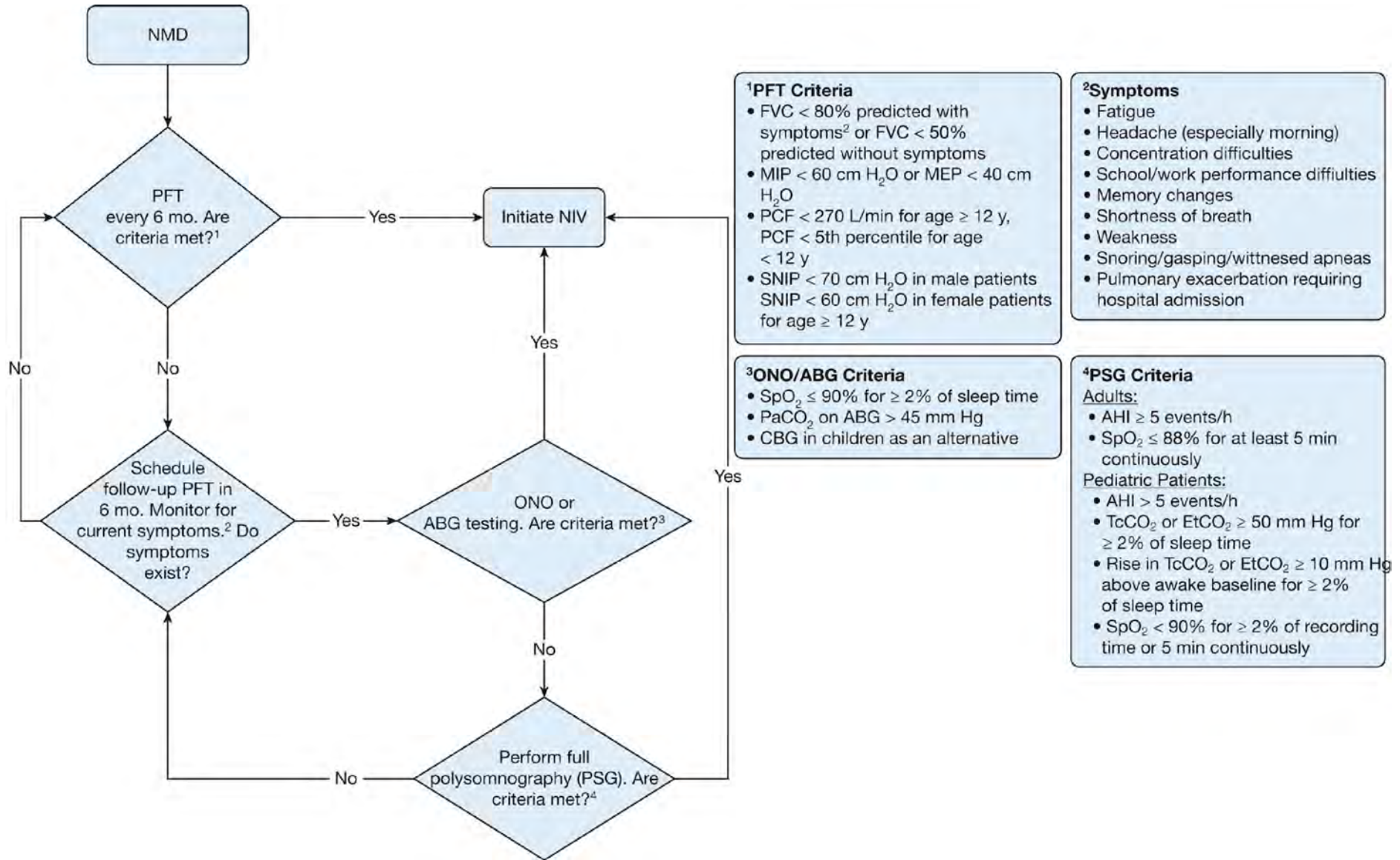
## Panel Recommendation 17: NMD/secretions

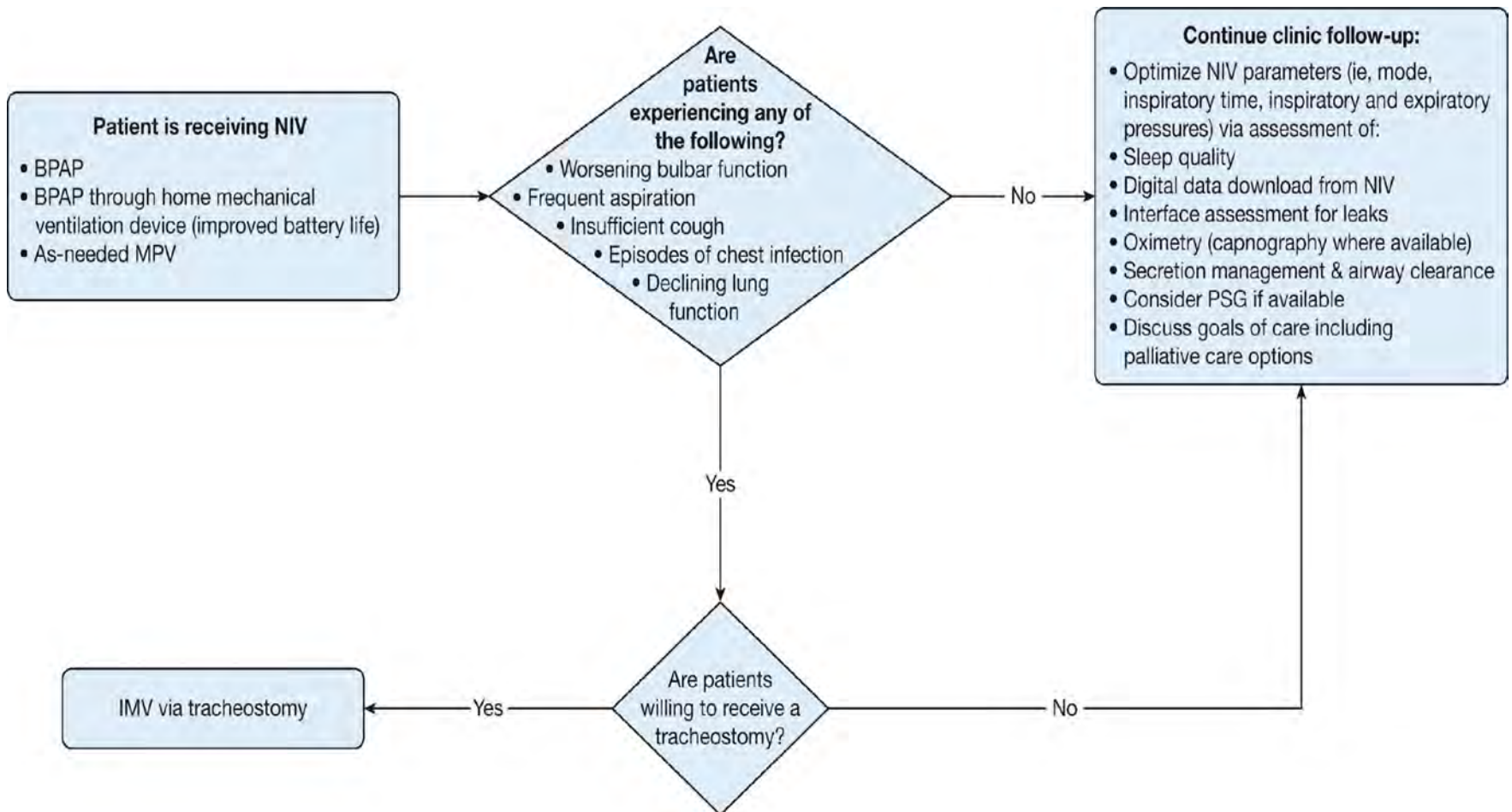
For patients with NMB and difficulties with secretion clearance the panel recommend High Frequency Chest Wall Oscillation (HFCWO)

## Panel Suggestion:

The panel suggests HFCWO to be combined with airway clearance therapist such as cough assistance or LVR







# Clinical Scenarios: Question 1

When caring for a patient with an NMD, the panel suggested that a PFT be done at least how often?

- A. Once a year
- B. Every 3 months
- C. Every 6 months
- D. PFTs are not recommended for NMD patients

# Clinical Scenarios: Answer 1

C. Every 6 months

## Rationale:

For patients with NMD at risk of respiratory failure the panel suggest PFT at a minimum of every 6 months as appropriate to the course of the specific NMD

## Clinical Scenarios: Question 2

For patients with systemic NMD who have a normal PFT and overnight oximetry (ONO), what is suggested by the panel to assess whether noninvasive ventilation (NIV) is needed?

- A. Polysomnography
- B. Chest X-Ray
- C. ECG
- D. EEG

# Clinical Scenarios: Answer 2

## A. Polysomnography

### Rationale:

Polysomnography can be used to assess whether NIV is indicated in symptomatic patients with a normal PFT and ONO.

# Clinical Scenarios: Question 3

For the treatment of patients with NMD and sleep-related breathing disorders, the panel suggests which of the following?

- A. Tracheostomy
- B. Bronchodilator therapy
- C. PFT
- D. Noninvasive Ventilation (NIV)

# Clinical Scenarios: Answer 3

## D. Noninvasive Ventilation

### Rationale:

Noninvasive ventilation is suggested using AASM criteria for sleep-disordered breathing and hypoventilation for adults and ERS criteria for pediatric patients



# Clinical Scenarios: Question 4

For patients with NMD and sialorrhea, the panel suggests the use of which treatment?

- A. Sympathomimetic bronchodilator medication
- B. Anticholinergic medications
- C. Steroidal medications
- D. Nonsteroidal anti-inflammatory drug

# Clinical Scenarios Answer 4

## B. Anticholinergic medications

### Rationale:

- The panel suggested an initial trial of an inexpensive oral anticholinergic medication

# Clinical Scenarios: Question 5

After evaluating patients with NMD and difficulties with secretion clearance, the panel recommends using which for secretion mobilization?

- A. Suctioning with large catheter
- B. High-Frequency Chest Wall Oscillation (HFCWO)
- C. Incentive spirometer
- D. Tracheostomy

# Clinical Scenarios: Answer 5

## B. High Frequency Chest Wall Oscillation (HFCWO)

### Rationale:

The panel suggested using HFCWO for secretion mobilization. In addition, the panel suggested that HFCWO be combined with other airway clearance therapies.

# Summary

- Overview of respiratory management of patients with neuromuscular diseases.
- Evidence-based suggestions for patients with neuromuscular diseases.
- Evidence-based recommendations for patients with neuromuscular diseases.

# Discussion

# Thank You

**Duane Reed, EdD, RRT, RCP**

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