Pulmonary Care in Neuromuscular Disease: Considerations & Resources

A Resource for Clinicians

Background

- Most patients with NMDs develop respiratory complications and/or failure due to muscle weakness, and tends to worsen with progressing disease.
- Early recognition of patient risk for pulmonary complications and integration of a pulmonologist into a coordinated multidisciplinary care can optimize outcomes
- This document highlights key points from an MDA webinar with a pulmonologist with expertise caring for patients with NMD. View the companion webinar here

Respiratory Involvement in NMD

Respiratory failure[1]
- Hypoventilation leading to poor gas exchange
- Increasing blood carbon dioxide tension ($PaCO_2$)
  - Initially during sleep and progressing to night and day

Acute[2]
Sudden loss of lung function leading to inadequate gas exchange, arterial oxygen tension <60 mm Hg, normal or low arterial CO_2 tension, or increased arterial CO_2 tension >50 mm Hg

Chronic[3]
Failure of the inspiratory respiratory muscles to continue normal ventilation and maintain normal arterial $PaCO_2$

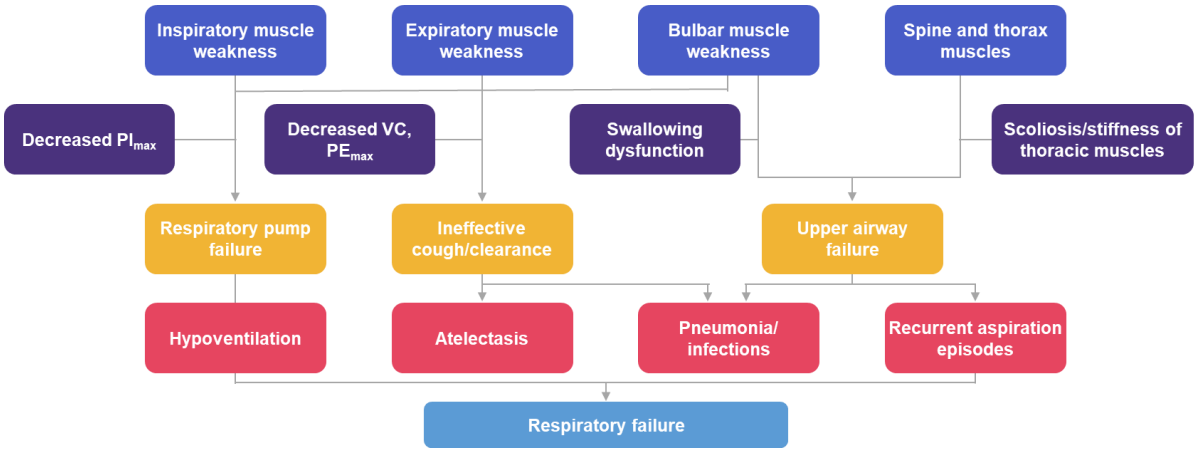
NMDs with **unavoidable** occurrence of respiratory failure[4]
- DMD
- ALS
- Some muscular dystrophies
- Some myofibrillar myopathies

NMDs with **frequent** occurrence of respiratory failure[4]
- SMA2
- DM1
- GBS
- MG
- Some congenital muscular dystrophies
- Some limb-girdle muscular dystrophies
- Some congenital myopathies
- Congenital myasthenic syndromes

ALS, amyotrophic lateral sclerosis; DM1, myotonic dystrophy type 1; DMD, Duchenne muscular dystrophy; GBS, Guillain-Barré syndrome; MG, myasthenia gravis; NMD, neuromuscular disease; PaCO2, partial pressure of carbon dioxide; SMA2, spinal muscular atrophy type 2.
1. Toussaint M, et al. Respir Med. 2021;80:106373. 2. Elhidsi M, et al. Ro J Neurol. 2022;21(3):219-224. 3. Carmona H, et al. Annu Rev Med. 2023;74:443-455. 4. Racca F, et al. Neurol Sci. 2020;41(3):497-508.

Pathophysiology of Respiratory Failure in NMD

Most patients with NMDs develop respiratory complications and failure due to muscle weakness[1,2c]



NMD, neuromuscular disease; PEmax, maximum expiratory pressure; PImax, maximum inspiratory pressure; VC, vital capacity.
1. Voulgaris A, et al. Pulm Med. 2019;2019:2734054. 2. Elhidsi M, et al. Ro J Neurol. 2022;21(3):219-224.

Developed with the expertise of Jeanette Brown, MD, PhD (University of Utah School of Medicine)
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Signs and Symptoms: Impaired Respiration

Recognizing early signs and symptoms of impaired respiration can aid in timely testing and intervention[1-3]

Inspiratory and expiratory muscle weakness

- Tachypnea
- Ineffective cough
- Paradoxical breathing

Bulbar muscle weakness

- Difficulty while swallowing and chewing
- Impaired mouth opening can hamper pulmonary function testing

Spine and thorax involvement

- Thoracic deformities, such as bell-shaped chest, common in patients with SMA
- Stiffness of the chest muscles

SMA, spinal muscular atrophy.
1. Boentert M. Curr Opin Neurol. 2017;30(5):529-537. 2. Ambrosino N. Eur Respir J. 2009;34(2):444-451. 3. Davidescu L. IntechOpen. 2019. doi: 10.5772/intechopen.77173.

Guidelines to Manage Lung Function in NMD

The American College of Chest Physicians® released a clinical guideline on respiratory management of patients with neuromuscular weakness in 2023, covering the following topics.

Mouthpiece ventilation[1]

- With NMD progression, there is a need to extend nocturnal NIV into daytime NIV using MPV

Transition to home MV[2]

- Indicated for chronic respiratory failure with alveolar hypoventilation
- Can be NIV or invasive

Sialorrhea management[3]

- Excessive salivation from either:
  - Increased production of saliva
  - Difficulty in saliva clearance
- Patients with NMD develop sialorrhea from bulbar dysfunction
  - Poor coordination of the tongue and palate
- Leads to poor performance with NIV

Airway clearance therapies[4]

- Mucociliary clearance and cough mechanisms may be compromised
- Respiratory physiotherapy is essential in managing airway clearance

Please see guidelines for full recommendations.

MPV, mouthpiece ventilation; MV, mechanical ventilation; NIV, noninvasive ventilation; NMD, neuromuscular disease.
1. Toussaint M. Respir Med. 2021;180:106373. 2. Park S. Acute Crit Care. 2020;35(3):131-141. 3. Sahni AS. Respir Care. 2018;63(5):601-608. 4. Chatwin M. Respir Med. 2018;136:98-110.

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Screening & Testing of Pulmonary Function*

PFT is recommended for patients with NMD to assist with management decisions*

- The following PFT parameters are recommended for spirometry as predictors of clinical outcomes:
  - Forced or slow vital capacity
  - MIP/MEP or SNIP and PCF
- For patients with NMD at risk for respiratory failure, PFT is recommended at a minimum of every 6 months†
- Clinicians should adjust the testing frequency based on the progression rate of the individual NMD

PFT is a low-cost intervention that can be performed in an office or home-based setting

- The following criteria values are considered abnormal parameters:
  - FVC <80% predicted
  - MIP <60 cm H_2O
  - MEP <40 cm H_2O
  - PCF <270 L/min in individuals ≥12 years of age
- Limited data to support PFT in supine vs. erect postures

For symptomatic patients with NMD, having normal PFT and ONO:

- Clinicians should consider PSG to assess whether NIV is clinically indicated*
  - Clinical indications may vary based on the patient's age and disease progression
  - PSG in adults may not be needed if PFT or ONO supports use of NIV
  - Full PSG should be considered in pediatric and adult patients with daytime tiredness, fatigue, or apneic episodes

- The use of PSG in patients with NMD requires an appropriate testing facility with ADA access, NMD protocols to address hypoventilation, equipment for NMD, and space for bedside caregivers
  - ONO or PFT may be used as alternatives to PSG to help with NIV decision-making
- No data available for frequency or timing of PSG, ONO, or home sleep testing

Not exact and have been summarized; review the publication for full recommendations.
Khan A, et al. Chest. 2023;164(2):394-413.

Other Clinical Considerations for Pulmonary Care

- Procedures that require sedation and anesthesia put patients at increased risk for:

Respiratory failure	Cardiac dysrhythmias	Congestive HF
Atelectasis	Hypoventilation	Upper airway obstructions

- Consult with anesthesia team prior to procedures for consideration of which medications to avoid and the need for NIV support during sedation, as well as post-extubation NIV following general anesthesia

- Patients should be up to date on vaccinations for respiratory viruses
- If they are admitted to the hospital, they should bring their NIV device and cough assist device with them, because the facility may not have their specific device and their device is already adjusted for them
- Patients who are hypoxic should be evaluated for hypercapnia prior to having supplemental oxygen placed because this can mask hypoventilation, leading to respiratory failure

HF, heart failure; NIV, noninvasive ventilation.
1. Birnkrant DJ, et al. Chest. 2007;132(6):1977-1986. 2. Myotonic Dystrophy Foundation. Practical Suggestions for the Anesthetic Management of a Myotonic Dystrophy Patient. https://www.myotonic.org/sites/default/files/pages/files/MDF_PracticalSuggestionsDM1_Anesthesia2_17_21.pdf.

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Resources

The American College of Chest Physicians®: Respiratory Management of patients with neuromuscular weakness. Khan A, et al. Chest. 2023;164(2):394-413.

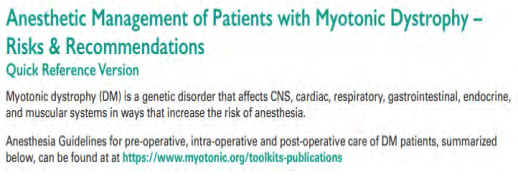
Published in the journal ***CHEST***®, the guideline contains:

- 15 evidence-based recommendations
- A good practice statement
- An ungraded consensus-based statement

Endorsed by:

- American Association for Respiratory Care
- American Thoracic Society
- American Academy of Sleep Medicine
- Canadian Thoracic Society

Anesthetic Management: Risks & Recommendations



Alert Card

Myotonic Dystrophy Foundation

Network of Ventilator User Resources

IVUN
INTERNATIONAL VENTILATOR USERS NETWORK
an affiliate of Post-Polio Health International

Emergency Alert Cards

MDA® Muscular Dystrophy Association

DMD Extubation Protocol

DMD Anesthesia Protocol

Parent Project Muscular Dystrophy

Access companion MDA webinar here

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