Respiratory Management of Patients With Neuromuscular Weakness

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- Respiratory failure is a common complication in patients with NMD.
- Respiratory muscle weakness in patients with NMD can lead to inadequate ventilation, hypoventilation at night, and the inability to mobilize secretions that frequently are the cause of death in this population.
- Sleep-related breathing disorders are common initial symptoms of NMD.[4]

- Data on the treatment of respiratory failure in NMD are limited.
- NMDs present at different ages and progress at variable rates, making it difficult to provide a single set of guidelines.

- Patients with NMD often are cared for by interprofessional teams consisting of pulmonologists, neurologists, respiratory therapists, physical and occupational therapists, pediatricians, internists, and family physicians.

Criteria and timing for initiation of respiratory assistance in NMD

- In adults, NMDs are due either to adult-onset diseases such as amyotrophic lateral sclerosis (ALS) and spinal cord injury or childhood-onset diseases such as Duchenne muscular dystrophy (DMD) and cerebral palsy where survival into adulthood is increasingly common .
- The most common NMDs in childhood are DMD, spinal muscular atrophy, and congenital muscular disorders including a large group of muscular dystrophies. In adults, ALS, myotonic dystrophy, and limb-girdle muscular dystrophy are the most common NMDs.
- When considering criteria and timing for initiation of respiratory support devices, it is useful to differentiate between slowly progressive and rapidly progressive disease

TABLE 1 Rates of progression of select NMDs.

Slow (>15 years)	Intermediate (5–15 years)	Rapid (0–3 years)
SMA type 2	DMD	SMA type 1
SMA type 3	Myotonic dystrophy	ALS
Acid maltase deficiency	LGMD	Traumatic SCI

SMA, spinal muscular atrophy; DMD, Duchenne muscular dystrophy; LGMD, limb-girdle muscular dystrophy; ALS, amyotrophic lateral sclerosis; SCI, spinal cord injury. (Adapted from Pinto et al (8).

- Goals of NIV include the improvement of symptoms of hypoventilation, sleep quality, and overall quality of life. In some instances the use of NIV in patients with NMD may prolong survival.

- The initiation of NIV is based on a combination of patient symptoms and objective measures using pulmonary function testing, arterial blood gas measurements, and polysomnography or desaturation studies.

- In the United States, a respiratory assist device for NIV may be limited by the Centers for Medicare and Medicaid Services (CMS) reimbursement criteria:
- (1) Documentation of a restrictive thoracic disorder.
- (2) One of the following:
- a. $PaCO_2$ on awake arterial blood gas ≥ 45 mmHg while breathing the prescribed fraction of oxygen
- b. Sleep oximetry with SpO_2 ≤ 88% for five minutes or more while breathing the prescribed fraction of oxygen. c. Either a maximal inspiratory pressure (MIP) < 60 cmH_2O or forced vital capacity (FVC) < 50% predicted.

- initiation criteria for NIV and evidence supporting NIV use for three of the common etiologies of chronic respiratory failure in NMD:

- ALS
- DMD
- spinal cord injury

Amyotrophic lateral sclerosi

- The European Federation of Neurological Societies guidelines recommend initiation of NIV if any one of the following is present :
- (1) One respiratory clinical symptom related to muscle weakness
- (2) FVC < 80%
- (3) MIP < −60 cmH2O
- (4) Sniff nasal pressure (SNP) < 40 cmH2O
- (5) Significant nocturnal desaturation
- (6) PaCO2 > 45 mmHg on a morning arterial blood ga

- An early retrospective analysis suggested that 4 h a day of NIV, usually initiated during sleep, was beneficial
- while a more recent retrospective analysis shows a potential benefit of NIV use for at least 8 h a day

Duchenne muscular dystrophy

- Treatment with NIV appears to improve quality of life and reduce morbidity and mortality in patients with DMD

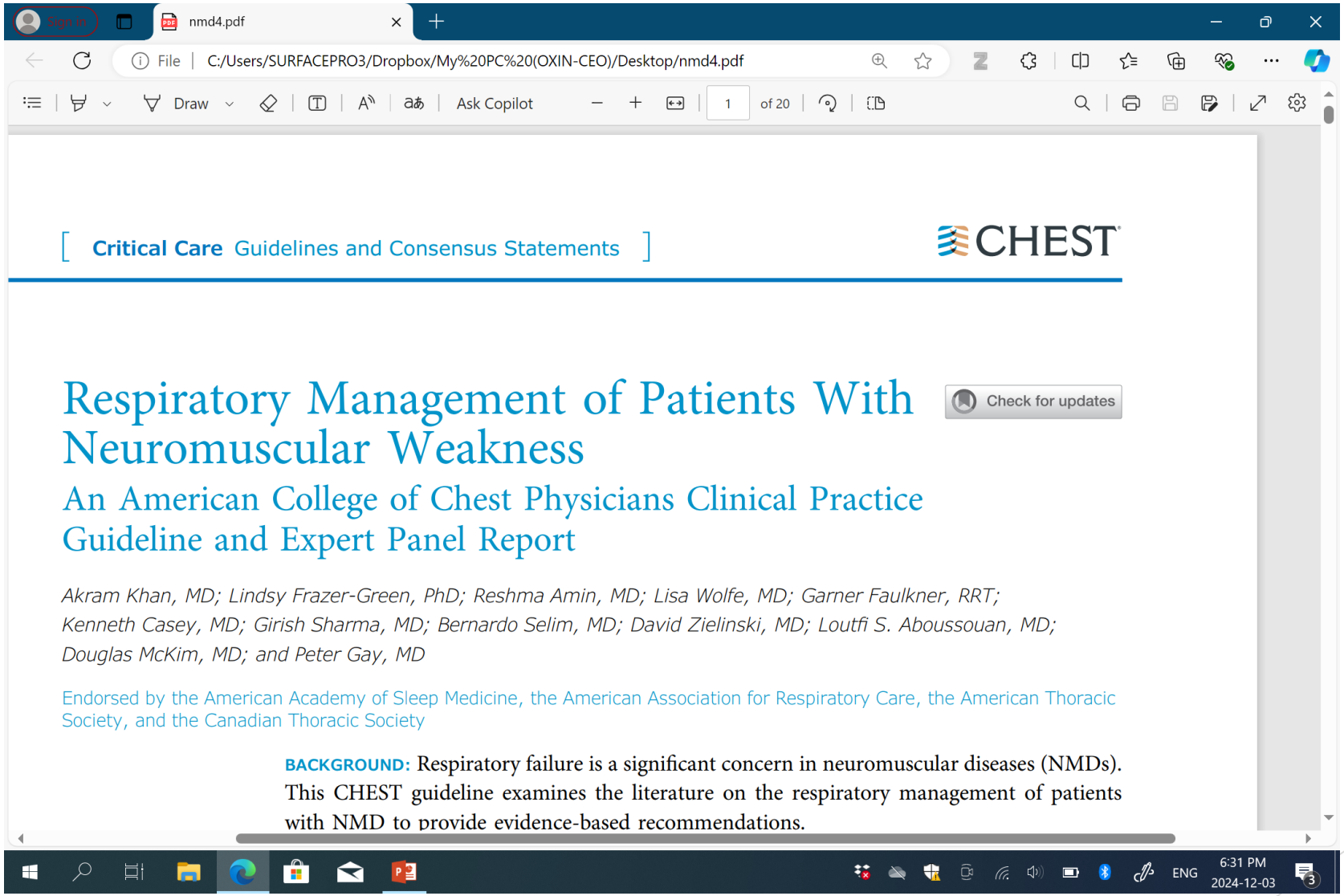
- A DMD care considerations working group recommends nocturnal ventilation with any of the following :
- 1) Signs or symptoms of hypoventilation
- (2) Baseline SpO_2 < 95% and/or end-tidal $PaCO_2$ > 45 mmHg while awake
- (3) Apnea-hypopnia index > 10/hour on polysomnography OR four or more episodes of SpO_2 < 92% OR drops in SpO_2 of at least 4% per hour of sleep.

Spinal cord injury

- The diaphragm is the principal muscle of inspiration and is innervated by the cervical nerve roots C3-C5.
- The principal muscles of expiration (and therefore cough) are the internal intercostals, innervated by the corresponding thoracic nerves, and the abdominal muscles, innervated by T5-L1

- In a traumatic spinal cord injury, ventilatory impairment depends on the involved spinal cord level and the completeness of the injury .
- Mechanical ventilation is usually required following injury to C3-C5

- though most patients with a lesion at C4 or below can be weaned
- Injuries to C6-C8 compromise the ability to generate forced exhalation and cough. For thoracic spine injuries, cough effectiveness improves as the level of injury decreases.
- Sleep disordered breathing is more prevalent in individuals with spinal cord injury; formal sleep study is recommended with complete tetraplegia and should be considered in patients with incomplete tetraplegia and paraplegia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CHEST
Respiratory Management of Patients With Neuromuscular Weakness
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An American College of Chest Physicians Clinical Practice Guideline and Expert Panel Report
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Endorsed by the American Academy of Sleep Medicine, the American Association for Respiratory Care, the American Thoracic Society, and the Canadian Thoracic Society
BACKGROUND: Respiratory failure is a significant concern in neuromuscular diseases (NMDs). This CHEST guideline examines the literature on the respiratory management of patients with NMD to provide evidence-based recommendations.
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Summary of Recommendations and Suggestions

- **Use and Timing of Pulmonary Function Testing**
- 1. For patients with neuromuscular disease (NMD) at risk of respiratory complications, we recommend pulmonary function testing (PFT) to assist with management decisions (Good Practice Statement).

- 2. For patients with NMD at risk of respiratory failure, we suggest PFT at a minimum of every 6 months as appropriate to the course of the specific NMD (Conditional Recommendation, Ungraded Consensus-Based Statement)

- For symptomatic patients with NMD who have normal PFT and overnight oximetry (ONO) findings, we suggest that clinicians consider polysomnography to assess whether noninvasive ventilation (NIV) is clinically indicated (Conditional Recommendation, Very Low Certainty of Evidence)

Use of NIV

- 4. For patients with NMD and chronic respiratory failure, we recommend using NIV for treatment (Strong Recommendation, Very Low Certainty of Evidence).

- The clinical indications for NIV can vary depending on NMD, patient age, and rate of disease progression.

Any fall in FVC to < 80% of predicted with symptoms or FVC to < 50% of predicted without symptoms or SNIP /MIP to < –40 cm H2O or hypercapnia would support the initiation of NIV or further testing as clinically indicated for individual NMD

Additional Comments and Implementation Recommendations

- The evidence was predominantly from older children ($ 12 years) and adults with NMD, with most studies in adults with ALS, although patients with Duchenne muscular dystrophy also are included

- 5. For patients with NMD and sleep-related breathing disorders, we suggest using NIV for treatment (Conditional Recommendation, Very Low Certainty of Evidence).

- The panel suggests using the American Academy of Sleep Medicine criteria for sleep-disordered breathing and hypoventilation for adult patients

- and the European Respiratory Society criteria for pediatric patients

- 6. For patients with NMD and preserved bulbar function using NIV, we suggest mouthpiece ventilation (MPV) for daytime ventilatory support as an adjunct to nocturnal mask NIV (Conditional Recommendation, Very Low Certainty of Evidence).

Use of Mechanical Ventilation

- 7. For patients with NMD in whom NIV fails or who are intolerant of NIV (including extended daytime NIV use), who have worsening bulbar function, frequent aspiration, insufficient cough, episodes of chest infection despite adequate secretion management, or declining lung function, we suggest invasive home MV via tracheostomy as an alternative to NIV (Conditional Recommendation, Very Low Certainty of Evidence).

Sialorrhea Management

- 8.For patients with NMD and sialorrhea, we suggest a therapeutic trial of an anticholinergic medication as first-line therapy with continued use only if benefits are perceived to outweigh potential side effects (Conditional Recommendation, Very Low Certainty of Evidence)

- 9. For patients with NMD and sialorrhea who have an inadequate response or are intolerant of the side effects of anticholinergic therapy, we suggest botulinum toxin (BT) therapy to salivary glands (Conditional Recommendation, Very Low Certainty of Evidence)

- See individual studies for doses of BT. It is unclear whether clinicians should consider BT or radiation therapy (RT) first and can base their decision on local expertise.

- 10. For patients with NMD and sialorrhea who have an inadequate response or are intolerant of the side effects of anticholinergic therapy, we suggest salivary gland RT (Conditional Recommendation, Very Low Certainty of Evidence).

Summary

- Respiratory failure is common and often is the final cause of death in patients with NMD
- These guidelines have several limitations:
- NMDs are a heterogeneous group of disorders, and it is difficult to provide a unique set of recommendations for each specific NMD
- In addition, randomized controlled trials on specific NMDs are limited.

- Furthermore, a significant portion of the evidence is based on ALS, because data on other slowly progressive diseases are limited and elements of the guidelines may need to be individualized according to the rate of progression of an individual patient's illness.

- Finally, the guideline focus on diagnosis and initiation of therapies and do not go into the details of NIV or MV, which would require separate guidelines.

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