Clinical Guideline Update: NIV and Respiratory Care in Neuromuscular Disease

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Objectives

- Review recently published guidelines of respiratory management in patients with neuromuscular muscular disease (NMD)
- Understand the role of pulmonary functions tests in aiding with initiation of NIV
- Understand recommendations for sialorrhea management
- Understand the recommended modalities that aide in airway clearance

[Critical Care Guidelines and Consensus Statements]

CHEST

Respiratory Management of Patients With Neuromuscular Weakness

An American College of Chest Physicians Clinical Practice Guideline and Expert Panel Report

Akram Khan, MD; Lindsy Frazer-Green, PhD; Reshma Amin, MD; Lisa Wolfe, MD; Garner Faulkner, RRT; Kenneth Casey, MD; Girish Sharma, MD; Bernardo Selim, MD; David Zielinski, MD; Loutfi S. Aboussouan, MD; Douglas McKim, MD; and Peter Gay, MD

Endorsed by the American Academy of Sleep Medicine, the American Association for Respiratory Care, the American Thoracic Society, and the Canadian Thoracic Society

Chest 2023; 164(2): 394-413

Study Design and Methods

Expert Panel

Systematic Review

Applied Grading of Recommendations, Assessment, Development and Evaluations

Modified Delphi technique to reach consensus recommendations

- 128 studies
- 15 graded recommendations
- One good practice statement
- One consensus-based statement

Recommendations – 6 subsections

- Use and Timing of Pulmonary Function Testing
- Use of NIV
- Respiratory Parameters for Initiation of NIV
- Use of Mechanical Ventilation
- Sialorrhea Management
- Airway Clearance Techniques

Use and Timing of PFTs

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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).

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Question 1

- What measurements in PFTs are NOT typically measured in those with NMD?
  - FEV1 – Forced Expiratory Volume in 1 second
  - FVC – Forced Vital Capacity
  - MIP – Maximum Expiratory Pressure
  - SNIP – Sniff Nasal Inspiratory Pressure
  - PCF – Peak Cough Flow

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).

Use and Timing of PFTs

- Based on review of 1561 abstracts and 22 studies reviewed
- Commonly a low cost method to assess progression of respiratory failure and predictors of survival

Question 2

- How often do you order PFTs?
  - Every 3 months
  - Every 6 month
  - Once a year
  - Only when symptomatic

Use and Timing of PFTs

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).

Use and Timing of PFTs

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).

- Based on expert opinion
- No harm from performing test
- Rate of change differs depending on type of NMD

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).

Use and Timing of PFTs

- Abnormal Values
  - FVC < 80 % with symptoms or 50% without symptoms
  - MIP < - 60 cm H20
  - MEP < 40
  - PCF < 270 L/min

- SNIP and MIP can be substituted
- Unclear if to do in supine and upright positions.

Question 3

- When should polysomnogram be used to assess the need for NIV?
  - When symptomatic with normal PFT and ONO
  - When FEV1 is abnormal
  - When FVC is abnormal
  - When ONO is abnormal

Use and Timing of PFTs

3. 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 and Timing of PFTs

3. 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).

- Based on 2192 abstracts 5 studies
- No regular interval of timing recommended
- Consensus recommendation with low certainty of evidence to perform overnight oximetry, Home Sleep Test or Polysomnogram based on sleep history and symptoms
  - Transcutaneous or end tidal CO2 maybe helpful for detecting hypoventilation

Summary - Use and Timing of PFTs

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).

3. 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

Use of NIV

- 763 abstracts and 25 studies reviewed
- NIV vs no NIV – outcomes include survival, respiratory function, sleep cognitive function, QOL
- Overall: Desirable effects of NIV were moderate, with small undesirable effects, with no clear harm – thus a net benefit that favor NIV
- Most of the studies from ALS patient with some with DMD.

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).

5. For patients with NMD and sleep-related breathing disorders, we suggest using NIV for treatment (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).

- Indication for NIV can vary depending on disease and rate of progression
- Generally use:
- FVC < 80% with symptoms
- FVC < 50% without symptoms
- SNIP/MIP < -40 cm H20
- Or hypercapnia

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

- Panel suggest using AASM criteria for sleep disordered breathing and hypoventilation for adult patient and ERS criteria for pediatric patients

Respiratory Parameters for Initiation of NIV

Question 4

- True or False:
  - PFTs are the only way to qualify for NIV for NMD patients

Respiratory Parameters for Initiation of NIV

- 422 abstracts and 5 studies reviewed
- Parameters reviewed: AHI, Hypoventilation ($O_2 < 90\%$), $etCO_2$, $tcCO_2$, PFC, MIP, MEP, PCF, SNIP
- Outcomes reviewed: timing of NIV, patient preference, care use, symptom improvement

Respiratory Parameters for Initiation of NIV

Polysomnogram is not necessary for adult patient to start NIV if met by PFT criteria alone

6. For patients with NMD, we suggest the use of diagnostic tests such as FVC, MIP/MEP, ONO, or evidence of sleep-disordered breathing or hypoventilation on polysomnography to predict the timing of NIV initiation (Conditional Recommendation, Very Low Certainty of Evidence).

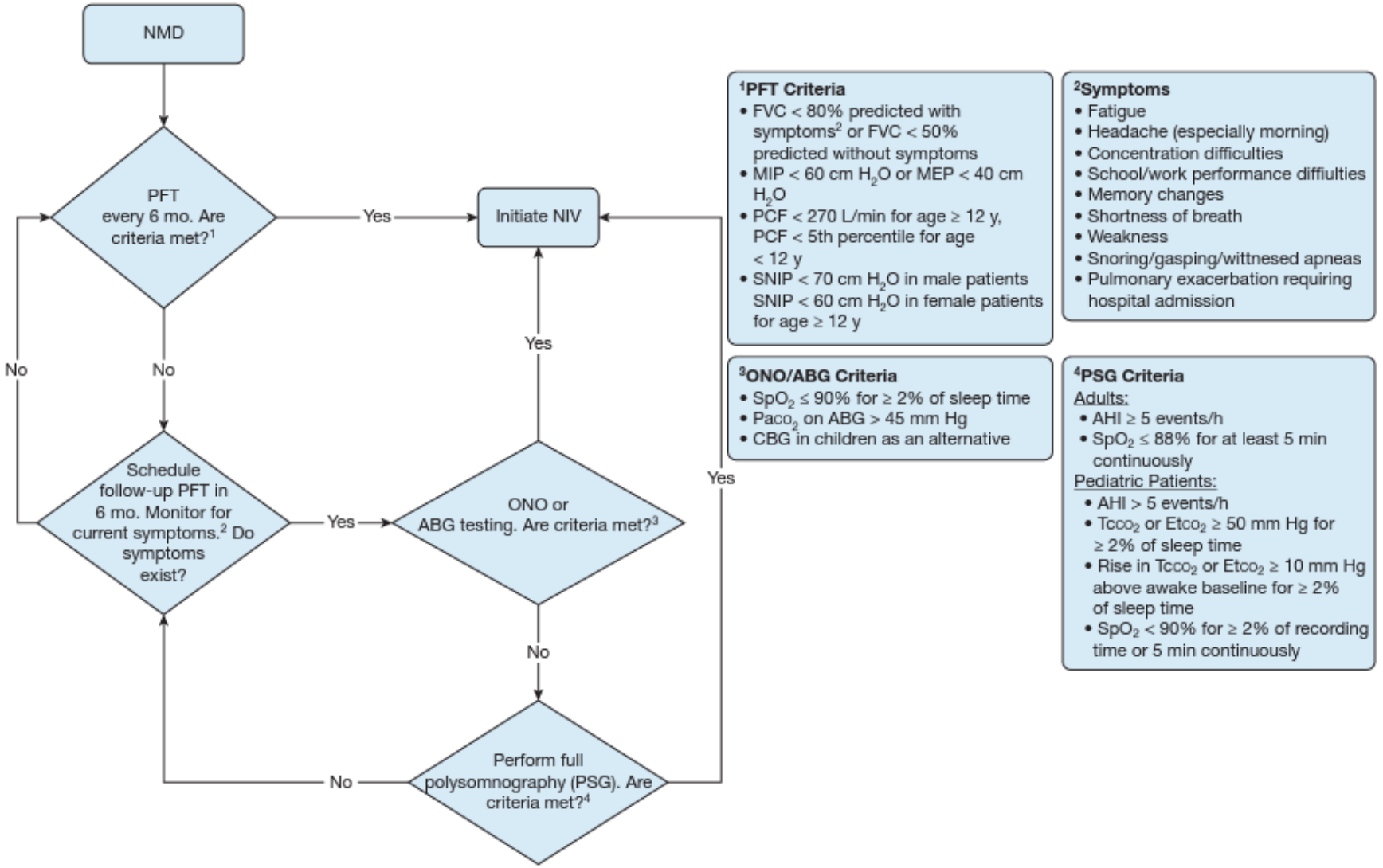

Figure 1 – *Flowchart for NIV initiation for patients with NMD showing respiratory failure symptoms. ABG = arterial blood gas; CBG = capillary blood gas analysis; $ETco_2$ = end-tidal co_2; MEP = maximum expiratory pressure; MIP = maximum inspiratory pressure; NIV = noninvasive ventilation; NMD = neuromuscular disease; ONO = overnight oximetry; PCF = peak cough flow; PFT = pulmonary function testing; SNIP = sniff nasal inspiratory pressure; SpO_2 = arterial oxygen saturation; $tcPco_2$ = transcutaneous Pco_2.*

Is there are preferred mode for ventilation?

Respiratory Parameters for Initiation of NIV

7. For patients with NMD requiring NIV, we suggest individualizing NIV treatment to achieve ventilation goals (Conditional Recommendation, Very Low Certainty of Evidence).

Based on review of 1383 abstracts and 5 studies

NIV should be tailored for the patient using mode of ventilation, Ti, IPAP, EPAP.

No evidence for preferred mode

Having a back up rate leads to better gas exchange and patient ventilator synchrony

Bulbar impairment may not tolerate NIV

Need ongoing assessment of sleep quality, digital downloads, leaks, oximetry, capnography, optimal synchrony settings with secretion management

What about mouth piece ventilation?

Respiratory Parameters for Initiation of NIV

- 44 abstracts and 4 studies reviewed
- Mouth piece ventilation can potentially delay tracheostomy, improve speech, improve cough effectiveness, improve swallowing and coordination of breathing
- No identified harmful consequences
- Bulbar symptoms may limit use

8. 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).

Summary - Respiratory Parameters for Initiation of NIV

6. For patients with NMD, we suggest the use of diagnostic tests such as FVC, MIP/MEP, ONO, or evidence of sleep-disordered breathing or hypoventilation on polysomnography to predict the timing of NIV initiation (Conditional Recommendation, Very Low Certainty of Evidence).

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When do you transition from NIV to mechanical ventilation?

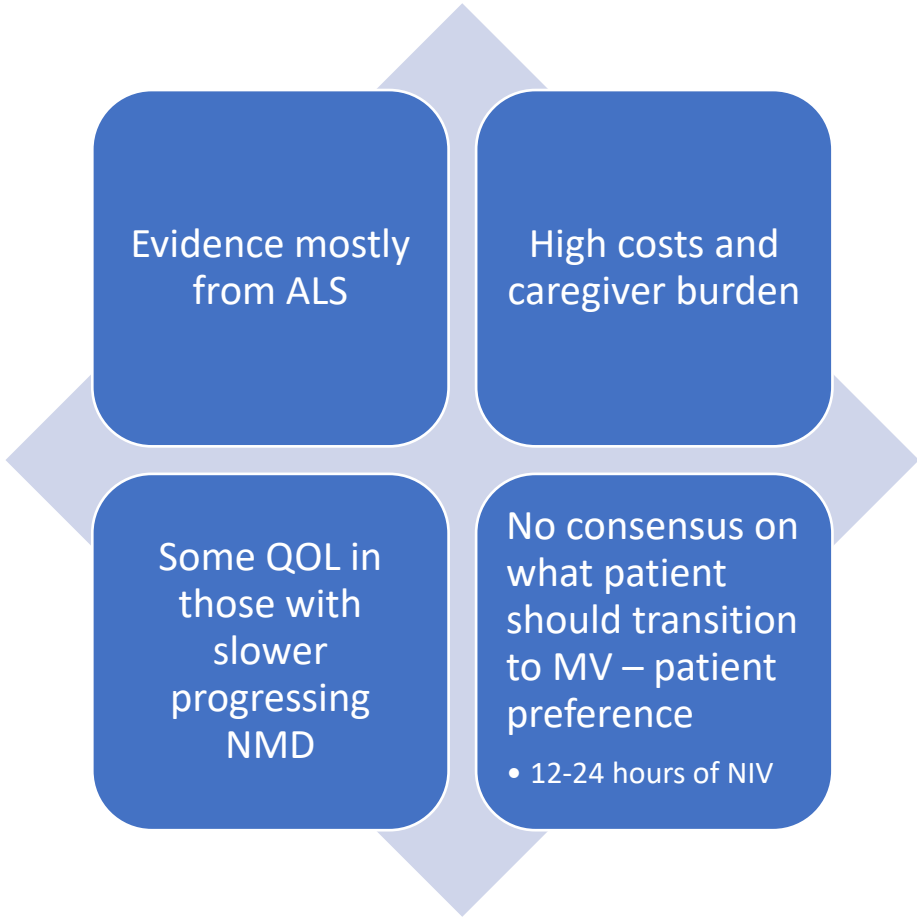
Use of Mechanical Ventilation

Use of Mechanical Ventilation

- Based on review of 390 abstracts and 10 studies
- Large improvement in survival and quality of sleep over no support, minimal benefit of NIV
- Undesirable effect – increase hospitalization, lower QOL, care giver burden
- Evidence favors home MV via tracheostomy as treatment option with progressive respiratory failure, unable to clear secretions or cognitive decline
- Continuous NIV can be done, with augmentation of mouthpiece ventilation

Use of Mechanical Ventilation

9. 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).



Goals of care should be discussed early in disease course, including desire for institutionalization and burden on caregivers

Use of Mechanical Ventilation

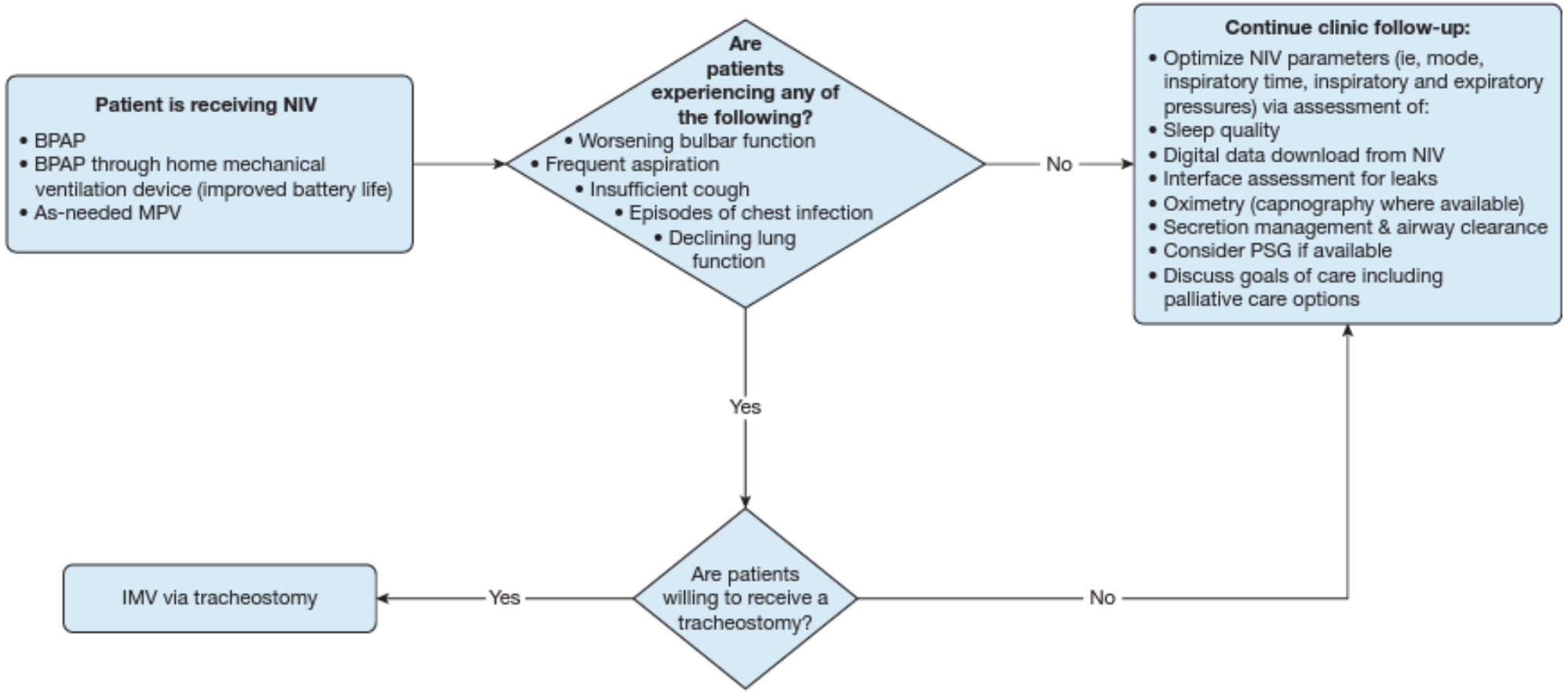


Figure 2 – *Flowchart for NIV intolerance for patients with NMD showing respiratory failure symptoms. BPAP = bilevel positive airway pressure; IMV = invasive mechanical ventilation; MPV = mouthpiece ventilation; NIV = noninvasive ventilation; NMD = neuromuscular disease; PSG = polysomnography.*

Sialorrhea Management

Sialorrhea (i.e. Hypersalivation)

- Commonly seen in NMD
- Cause of distress, reduced QOL
- Increase risk of aspiration
- Increase risk of pneumonia

Question

- Which of the following treatments are NOT recommend for sialorrhea?
  - Anticholinergic
  - Beta Agonist
  - Botulinum Toxin injection
  - Radiation Therapy

Sialorrhea - Anticholinergic

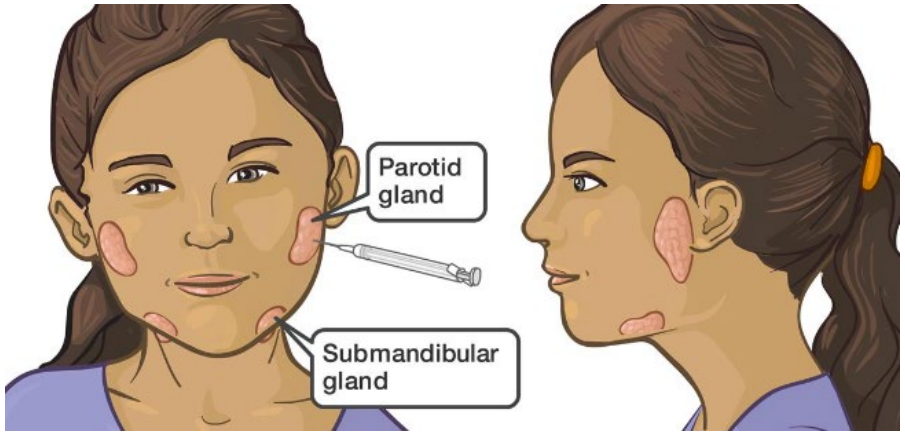
10. For patients with NMD and sialorrhea, we suggest a therapeutic trial of an anticholinergic medication as first-line therapy with continued use only if benefits are perceived to outweigh potential side effects (Conditional Recommendation, Very Low Certainty of Evidence).

- Recommend trial of oral anti - cholinergic
- Continue to use if find benefit and without side effects
- Anticholinergic available in patch form

Sialorrhea – Botulinum toxin

11. 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).

- Injected into salivary gland
- Inexpensive, but lasting effects
- Associated with viscous saliva and pain
- Limited evidence



Courtesy University of Iowa
Carver College of Medicine

Sialorrhea – Radiation Therapy

- Variable regimens
- Improvement at 1 month, 3 month and 6 months
- Long lasting relief – however, side effect of irreversible dryness

12. 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).

Sialorrhea

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Airway Clearance Therapies

Airway Clearance Therapies

- Glossopharyngeal Breathing
- Manually Assisted Cough
- Lung Volume Recruitment
- Mechanical Insufflation-Exsufflation
- High frequency chest wall oscillation

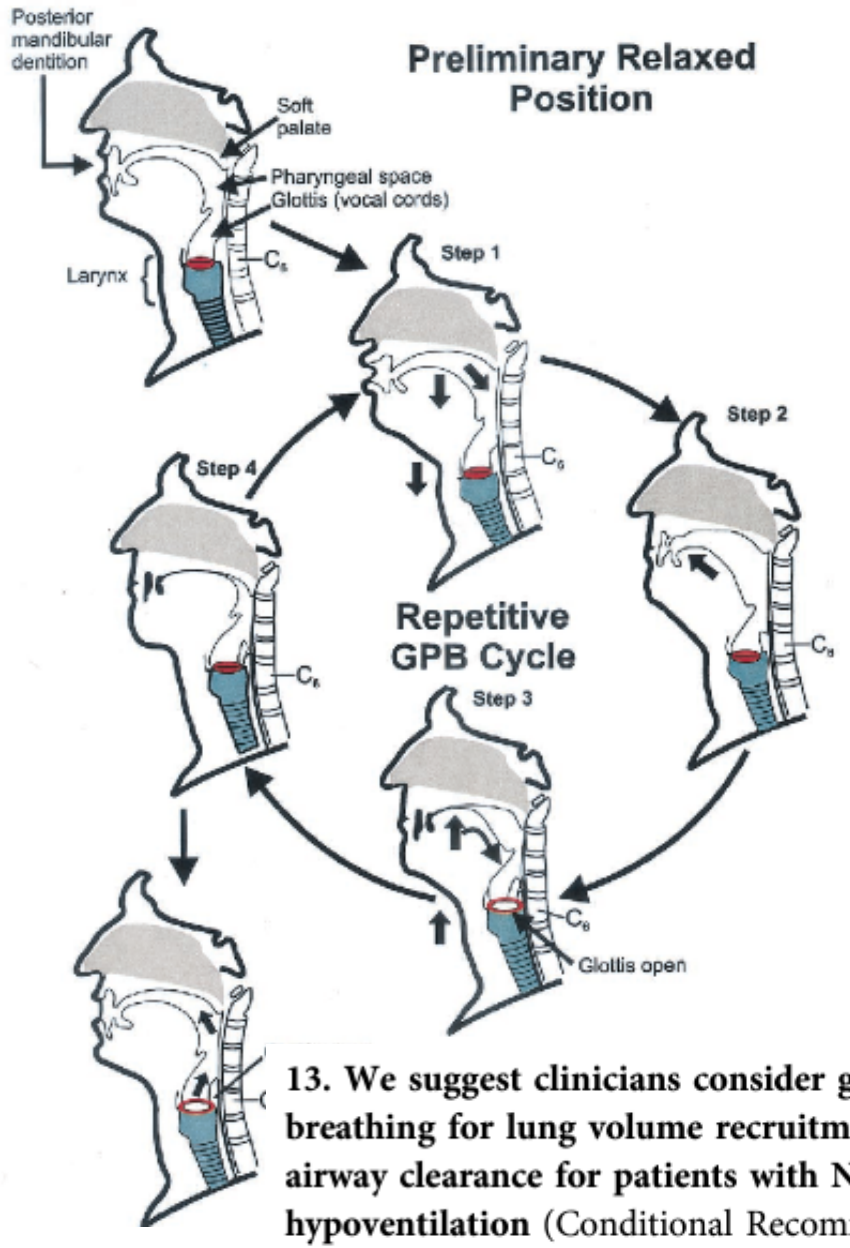
Question

- Which of the following airway clearance therapies are recommended according to guidelines?
  - A) Glossopharyngeal Breathing
  - B) Manually Assisted Cough
  - C) Lung Volume Recruitment
  - D) MIE
  - E) High Frequency Chest Wall Oscillation
  - F) All of the above
  - G) None of the above

Glossopharyngeal Breathing

"Glossopharyngeal breathing (GPB), also called "frog breathing", is a positive pressure breathing technique that uses muscles of the mouth and pharynx to propel small volumes of air ("gulps") into the lungs."

A LoMaruo et al, ERS European Respiratory Journal 2019 54: 1801938;



13. We suggest clinicians consider glossopharyngeal breathing for lung volume recruitment (LVR) and airway clearance for patients with NMD and hypoventilation (Conditional Recommendation, Very Low Certainty of Evidence).

Published in Physical Therapy 2002

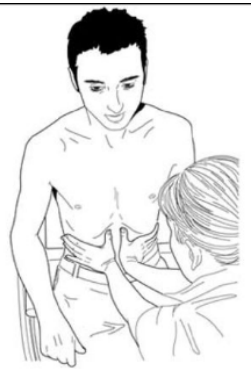
Glossopharyngeal and neck accessory muscle breathing in a young adult with C2 complete tetraplegia resulting in ventilator dependency.

V. Warren

Manually Assisted Cough

14. For patients with NMD and reduced cough effectiveness, we suggest manually assisted cough techniques independently or added to other modalities such as LVR (Conditional Recommendation, Very Low Certainty of Evidence).

A



- Park the wheelchair against a wall and make sure the brakes are engaged.
- With your child sitting upright, place your hands around the bottom of their ribs.
- Push inwards when your child coughs.

B

- With your child sitting upright, place one hand under your child's ribs and the other hand on their back to stabilize.
- Push up and inwards when your child coughs.

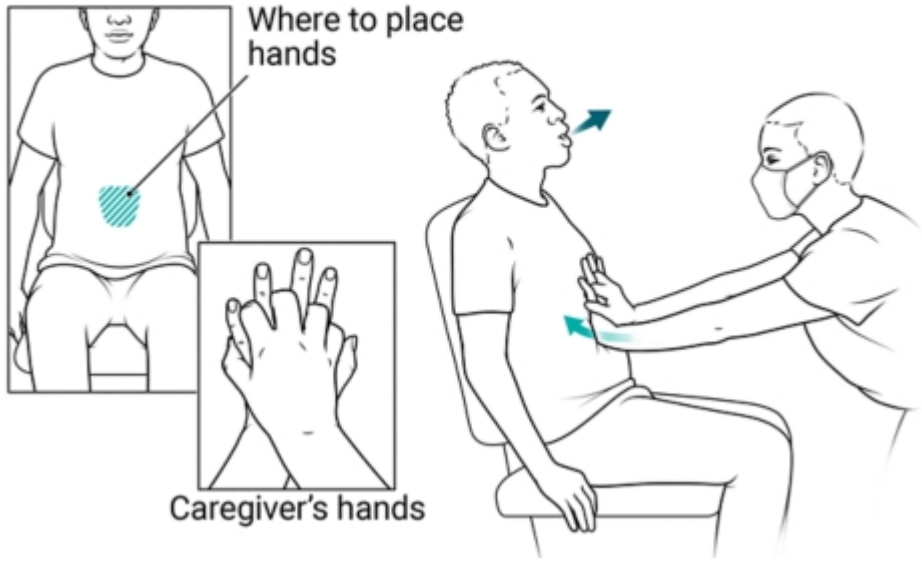
C

- With your child lying down or semi reclined in chair.
- Place the palm of your hand under the ribs and the other arm over the ribs to stabilize the chest.
- Push up and inwards when your child coughs.

D

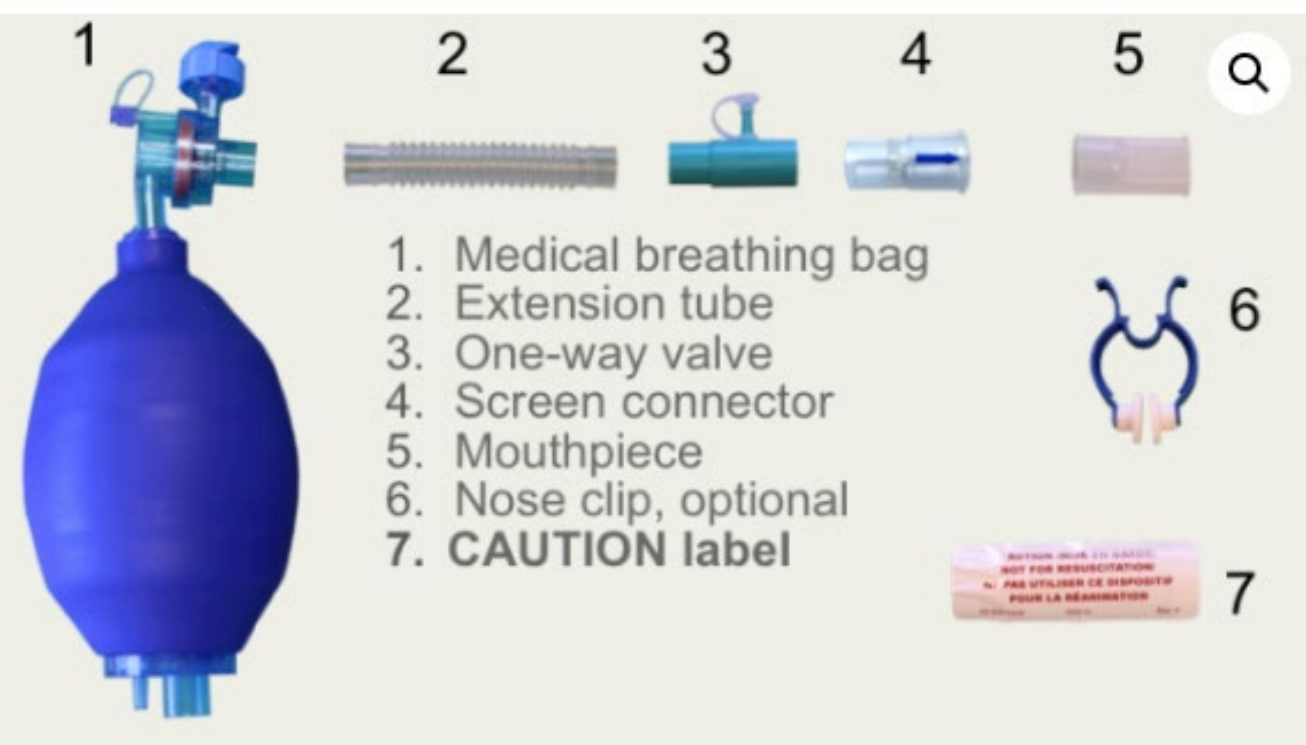
- With your child sitting up or lying down, place hands on your child's ribs at either side of their chest.
- Push in on the ribs as your child coughs.

Assisted cough: Abdominal thrust assist



Respiratory Care December 2018, 63 (12) 1520-1527;

Myhealth.alberta.ca



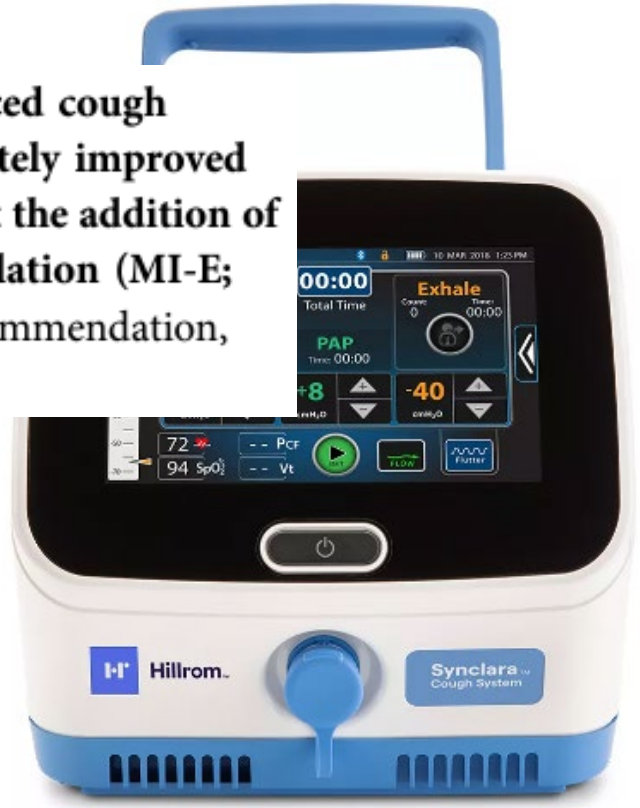
Lung Volume Recruitment

- Helps with expectorating
- Helps with blowing nose
- If strong enough, patients can squeeze it themselves

15. For patients with NMD and reduced lung function or cough effectiveness, we suggest regular use of LVR (breath stacking) using a handheld resuscitation bag or mouthpiece (Conditional Recommendation, Very Low Certainty of Evidence).

Mechanical Insufflation Exsufflation

16. For patients with NMD and reduced cough effectiveness, which cannot be adequately improved with alternative techniques, we suggest the addition of regular mechanical insufflation-exsufflation (MI-E; cough assist device) (Conditional Recommendation, Very Low Certainty of Evidence).



High Frequency Chest Wall Oscillation

17. For patients with NMD and difficulties with secretion clearance, we suggest using high-frequency chest wall oscillation (HFCWO) for secretion mobilization. In addition, we suggest that HFCWO be combined with airway clearance therapies such as cough assistance or LVR (Conditional Recommendation, Very Low Certainty of Evidence).

Airway Clearance Therapies

TABLE 5] Recommended Airway Clearance Therapies

Technique	Indications	Description	Remarks
GPB ("frog breathing")	Hypoventilation	Positive pressure breathing method using muscles of the mouth, tongue pharynx, and larynx	• Low cost • Performed by the patient independently
MAC	Reduced cough effectiveness	Abdominal thrust or lateral costal compression to generate expiratory flow.	• Low cost • Requires caregiver assistance
LVR ("breath stacking")	Reduced lung function or cough effectiveness	Handheld resuscitation bag or mouthpiece to inflate lungs to maximum inspiratory capacity without intervening expiration	• Low cost • Requires caregiver assistance
MI-E (cough assist device)	Reduced cough effectiveness not improved with alternative techniques	Alternating positive and negative pressure using a facemask or artificial airway. Effective for both upper and lower airway secretions.	• Expensive MI-E device • Requires caregiver assistance • Reduces morbidity and hospitalization, can have procedure intolerance
HFCWO combined with cough assistance or LVR	Difficulties with secretion clearance	Fit-tested vest that produces vibrations to mobilize peripheral airway secretions that then are cleared with cough or LVR to improve expiratory airflow	• Expensive HFCWO device • Requires caregiver assistance • Can have procedure intolerance

GPB = glossopharyngeal breathing; HFCWO = high-frequency chest wall oscillation; LVR = lung volume recruitment; MAC = manually assisted cough; MI-E = mechanical insufflation-exsufflation.

Airway Clearance Therapies

13. We suggest clinicians consider glossopharyngeal breathing for lung volume recruitment (LVR) and airway clearance for patients with NMD and hypoventilation (Conditional Recommendation, Very Low Certainty of Evidence).

15. For patients with NMD and reduced lung function or cough effectiveness, we suggest regular use of LVR (breath stacking) using a handheld resuscitation bag or mouthpiece (Conditional Recommendation, Very Low Certainty of Evidence).

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Based on review of 2714 abstracts and 36 studies

17. For patients with NMD and difficulties with secretion clearance, we suggest using high-frequency chest wall oscillation (HFCWO) for secretion mobilization. In addition, we suggest that HFCWO be combined with airway clearance therapies such as cough assistance or LVR (Conditional Recommendation, Very Low Certainty of Evidence).

Summary

- Update guidelines give directions on monitoring patients respiratory function
- Give clear indications for NIV
- Give clear indication for IMV and Mechanical Ventilation
- There are options for control of sialorrhea
- Airway clearance techniques are recommended

Questions?

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