

Noninvasive Ventilation for Neuromuscular Disease



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KEYWORDS

• Face mask • Mouthpiece ventilation • Neuromuscular disease • Noninvasive ventilation

KEY POINTS

- Noninvasive ventilation (NIV) is standard practice for patients with neuromuscular respiratory failure.
- The interface distinguishes NIV from invasive ventilation.
- Selection of an appropriate interface and ventilator settings requires a close working relationship between clinicians, patients, and families.

INTRODUCTION

Noninvasive ventilation (NIV) is standard practice for patients with neuromuscular respiratory failure. Observational studies in patients with Duchenne muscular dystrophy¹ and a randomized controlled trial in patients with amyotrophic lateral sclerosis (ALS)² support that NIV in patients with neuromuscular disease is life prolonging. NIV with airway clearance therapy, such as cough assist, are considered standard practice for patients with neuromuscular respiratory failure.³⁻⁵ This review addresses issues related to NIV, particularly the technical aspects of NIV.

INDICATIONS

Vital capacity (VC) is commonly used to assess respiratory muscle weakness in patients with neuromuscular disease. VC less than 50% predicted is commonly used as an indication for NIV, and reimbursement is linked to this value (Box 1).⁶ Maximum inspiratory pressure (P_{imax}) greater than -60 cm H₂O is also used as an indication to initiate NIV. Mendoza and colleagues⁷ reported that there were no cases among 161 patients whereby VC less than 50% occurred before P_{imax} greater than -60 cm H₂O. Patients reached the P_{imax} criterion 4.0 to 6.5 months

earlier than the VC criterion. In patients with diaphragm weakness, supine VC (orthopnea) may be a better indicator of the time to initiate NIV than an erect VC. A sniff nasal pressure can also be used, which is measured through a plug occluding one nostril during sniffs through the contralateral nostril.⁸

Overnight oximetry may be useful, as desaturation suggests nocturnal hypoventilation. Polysomnography to identify the need for NIV and titration of setting is controversial, although it was recommended in one consensus statement.⁹ Recommendations from a consensus committee suggest that NIV should be initiated in patients with neuromuscular disease with symptoms (such as fatigue, dyspnea, morning headache) and one of the following physiologic criteria: P_aCO₂ 45 mm Hg or greater, nocturnal oximetry demonstrating oxygen saturation at 88% or less for 5 consecutive minutes; for progressive neuromuscular disease, maximal inspiratory pressures greater than -60 cm H₂O or forced vital capacity less than 50% predicted.¹⁰

NIV can be used to provide respiratory support during gastrostomy tube placement in patients with neuromuscular disease.¹¹ Some patients with neuromuscular disease can be successfully extubated to NIV,¹² and some patients with neuromuscular disease who have received

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Box 1**Medicare coverage criteria, noninvasive ventilation for restrictive thoracic disorders**

Documentation in patients' medical record of a neuromuscular disease (eg, ALS) or a severe thoracic cage abnormality (eg, post-thoracoplasty for tuberculosis) and

1. An arterial blood gas P_{aCO_2} , done while awake and breathing the patient's fraction of inspired oxygen (F_{IO_2}), is 45 mm Hg or greater; or sleep oximetry demonstrates oxygen saturation of 88% or less for 5 minutes or longer (minimum 2-hour recording time), done while breathing patient's prescribed F_{IO_2} ; or for a neuromuscular disease (only), maximal inspiratory pressure is greater than -60 cm H_2O or forced vital capacity is less than 50% predicted and
2. Chronic obstructive pulmonary disease does not contribute significantly to patients' pulmonary limitation.

If all of the aforementioned criteria are met, either an E0470 or E0471 (based on the judgment of the treating physician) will be covered for patients within this group of conditions for the first 3 months of therapy.

From Hess DR. The growing role of noninvasive ventilation in patients requiring prolonged mechanical ventilation. Respir Care 2012;57(6):907. [discussion: 918–20]; with permission.

tracheostomy can be successfully decannulated to NIV.¹³ These patients might also be successfully transitioned to NIV with a capped tracheostomy tube and ultimately decannulated (Fig. 1).¹⁴

INTERFACE

The interface distinguishes NIV from invasive ventilation. Desirable characteristics of an interface are



Fig. 1. NIV via nasal pillows, with a capped tracheostomy tube. (From O'Connor HH, White AC. Tracheostomy decannulation. *Respir Care* 2010;55(8):1080; with permission.)

listed in **Box 2**.¹⁵ Interfaces are commercially available in a variety of styles and sizes (Fig. 2).^{16,17} They can provide nasal ventilation (nasal mask or nasal pillows) and oronasal ventilation; they can fit over the entire face (total face mask); they can provide ventilation directly to the mouth (mouth-piece or oral mask); and they can be of hybrid designs (nasal pillows with oral mask). There are advantages and disadvantages of each (Table 1).

Types

The nasal mask fits just above the junction of the nasal bone and cartilage, directly at the sides of both nares, and just below the nose above the upper lip. Some are gel filled; others use an open cushion with an inner lip, in which pressure inside the mask pushes the cushion against the face. Nasal pillows, sometimes called nasal cushions, are available from several manufactures. This interface consists of soft plastic plugs inserted into the nares, shaped in a way that the pressure applied during inspiration helps to seal the walls of the pillows against the inner surface of the nasal vestibule.

Several interfaces fit over the nose and mouth. The oronasal mask fits just above the junction of the nasal bone and cartilage and extends below the lower lip above the jaw. The total face mask, as the name implies, fits over the entire face of patients, extending from the forehead to the jaw. A

Box 2**Desirable characteristics of an interface for noninvasive ventilation**

- Low dead space
- Transparent
- Lightweight
- Easy to secure
- Adequate seal with low facial pressure
- Disposable or easy to clean
- Nonirritating (nonallergenic)
- Inexpensive
- Variety of sizes: adult and pediatric
- Adaptable to variations in facial anatomy
- Ability to be removed quickly
- Antiasphyxia mechanism
- Compatible with wide range of ventilators

From Hess DR. Noninvasive ventilation in neuromuscular disease: equipment and application. Respir Care 2006;51(8):898; with permission.

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