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Initiating Noninvasive Management of Respiratory Insufficiency in Neuromuscular Disease

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SUPPLEMENT ARTICLE

Initiating Noninvasive Management of Respiratory Insufficiency in Neuromuscular Disease

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ABSTRACT

This is a summary of the presentation on initiating noninvasive management of respiratory insufficiency in neuromuscular disease, presented as part of the program on pulmonary management of pediatric patients with neuromuscular disorders at the 30th annual Carrell-Krusen Neuromuscular Symposium on February 20, 2008. *Pediatrics* 2009;123:S236–S238

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Abbreviations

DMD—Duchenne muscular dystrophy

NPPV—noninvasive positive-pressure ventilation

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RESPIRATORY INSUFFICIENCY IS one of the leading causes of morbidity and mortality in patients with neuromuscular disease. Recent advances in respiratory support technologies have led to devices that are able to support respiration and improve quality of life and survival in individuals with neuromuscular disease and, in particular, Duchenne muscular dystrophy (DMD).[1–3]

Respiratory failure in neuromuscular disease consists of several major functional problems (Fig 1 www.pediatrics.org/content/vol123/Supplement_4) that can lead to ventilatory failure, pneumonia, and death: (1) inspiratory failure (ventilatory failure) that occurs because of weakness of the diaphragm, external intercostals, and accessory muscles of breathing; (2) expiratory (cough) failure that occurs because of weakness of the abdominal and internal intercostal muscles; and (3) swallowing failure that can result from involvement with the bulbar muscles. In this article I concentrate primarily on ventilatory failure, the methods to support it, and factors around initiation of those support measures.

NOCTURNAL VENTILATORY FAILURE

Inspiratory failure is most commonly seen first at night because during rapid eye movement sleep, atony of muscles other than the diaphragm occurs and weakness of the diaphragm is exposed. In addition, upper airway muscles may also be weak, leading to obstructive sleep apnea. Individuals with neuromuscular disease are at risk for a wide variety of sleep-disordered breathing types including central and obstructive sleep apnea leading to hypercarbia and hypoxemia. Treatment of nocturnal sleep disturbance can result in improved quality of life, reduced daytime hypercarbia, and increased survival rates.[3–5]

INDICATIONS FOR INITIATING NOCTURNAL VENTILATORY SUPPORT

Nocturnal ventilatory support should be initiated when the presence of sleep-disordered breathing is detected. Symptoms of nocturnal hypoventilation should be sought for but can be subtle and include frequent nocturnal awakenings, morning headaches, fatigue, daytime hypersomnolence, and difficulty with concentration and schoolwork. The finding of hypercarbia during the daytime is a predictor of nocturnal sleep-disordered breathing and, if present, should prompt further investigation. In terms of evaluation of sleep-disordered breathing, the American Thoracic Society has published a position paper on the respiratory care of individuals with DMD.[2] They suggested the following:

1. Review sleep quality and symptoms of sleep-disordered breathing at every patient encounter.
2. Perform an annual evaluation for sleep-disordered breathing in patients with DMD, starting from the time they are wheelchair users and/or when clinically indicated.
3. When available, it is ideal to perform annual polysomnography with continuous CO_2 monitoring.
4. In areas where full polysomnography is not readily available, perform overnight pulse oximetry with continuous CO_2 monitoring, which provides useful information about nighttime gas exchange, although sleep-disordered breathing not associated with desaturation or CO_2 retention will not be detected.

A consensus conference of the American College of Physicians[6] has established generally accepted criteria for reimbursement for devices for nocturnal ventilatory support that most insurers follow that include the following

criteria: symptoms of sleep-disordered breathing such as morning headache, daytime fatigue or dyspnea, and 1 of the following: (1) a $Paco_2$ level of ≥45 mm Hg; (2) nocturnal oximetry demonstrating oxygen saturation at ≤88% for 5 consecutive minutes; (3) a maximal inspiratory pressure of <60 cm H_2O; or (4) a forced vital capacity of <50% predicted. It should be noted that these are criteria for reimbursement only and that the astute clinician must look for more subtle symptoms and signs of sleep-disordered breathing.

CONTRAINDICATIONS TO NONINVASIVE VENTILATORY SUPPORT

Although there are few contraindications to noninvasive positive-pressure ventilation (NPPV), inability to fit or tolerate the mask interfaces is an absolute contraindication. Relative contraindications include bulbar involvement and swallowing dysfunction, lack of patient motivation, and inability to clear secretions (despite assisted coughing maneuvers).[7,8]

EQUIPMENT FOR NOCTURNAL VENTILATORY SUPPORT

The equipment for nocturnal noninvasive ventilatory support consists of 2 major pieces of equipment: a pressure/volume generator that supplies the forces to provide ventilation (Fig 2 www.pediatrics.org/content/vol123/Supplement_4) and an interface that connects the ventilator to the patient (Fig 3 www.pediatrics.org/content/vol123/Supplement_4). In most cases, the pressure/volume generators are pressure-support bilevel-type devices that are adjusted for a set expiratory positive airway pressure that functions to prevent airway obstructions and a set inspiratory positive airway pressure that serves to ventilate the individual. In almost all cases, because of the presence of central sleep apnea in patients with neuromuscular disease, a back-up rate is supplied that functions to deliver a breath if the patient does not trigger an inspiratory effort. Volume-type ventilators can also be used for nocturnal ventilation.[9] They have the advantage of being able to supply higher pressures if needed but are not able to compensate for leaks as the pressure-support devices are.

The second major piece of equipment is the interface that connects the ventilator to the patient. Interfaces are available in a wide variety of forms including nasal mask, nasal pillows, full face mask, and oral interfaces (Fig 3 www.pediatrics.org/content/vol123/Supplement_4). The type of interface used will depend on the preference of the patient. We have found that many patients prefer starting with the nasal mask. If mouth leak is present, a chin strap can be used. Alternatively, a full mask device can be used. Some individuals who have trouble fitting the mask to their nose or who suffer from claustrophobia may prefer an oral interface. Proper fitting of the mask is of utmost importance in the overall success of the ventilator-support venture. It is crucial to have an experienced therapist with a wide array of masks available for the patient to try on. We often spend an hour or more in the clinic during the fitting procedure.

WHERE TO INITIATE NPPV

Nocturnal NPPV may be initiated in a number of locations. Often, it will be started in the sleep laboratory and can be accomplished by using part of the night for diagnosis and part for treatment application, a so-called split-night protocol. On some occasions, individuals will be admitted to the hospital for initiation of NPPV or will be transitioned off of invasive ventilation to nocturnal NPPV during a hospital stay. In our practice, once diagnostic criteria for sleep-disordered breathing are met, we will acclimatize the patient to the device and begin initial titration of the ventilator in the office. Once the patient has had a chance to become used to the device, final titration occurs in the sleep laboratory or in the home with oximetry and CO_2 monitoring.

DIURNAL NONINVASIVE VENTILATION

When to Initiate Diurnal Noninvasive Ventilatory Support

Many neuromuscular diseases such as DMD are progressive; therefore, even with optimal management of nocturnal hypoventilation daytime respiratory insufficiency is bound to develop. Three possibilities for management of this situation are to (1) do nothing and let nature take its course, (2) consider tracheostomy, or (3) use a noninvasive ventilator device during the daytime. In our clinic, we have chosen the latter and found it to be quite successful, as has been reported in the literature.[10–12] The criteria for timing the initiation of daytime support have not been well defined; however, we have used the following triggers to initiate diurnal support: (1) if diurnal hypercarbia develops despite maximal nocturnal ventilatory support; (2) development of daytime dyspnea; and (3) increasing infections despite adequate cough therapy.

Equipment for Diurnal Ventilatory Support

There are 2 major methods of performing diurnal positive pressure with either mask or mouthpiece ventilation on a continuous basis. Mouthpiece ventilation (Fig 4 www.pediatrics.org/content/vol123/Supplement_4) is a methodology that has been available for the past 50 years or more. It involves the use of a portable volume ventilator set in the assist-control mode. The ventilator is attached to a standard ventilator circuit (Figs 2 and 4 www.pediatrics.org/content/vol123/Supplement_4), at the distal end of which is attached a mouthpiece that has either a narrow bore or an angled bend that allows back pressure in the circuit to prevent the ventilator from activating the low-pressure alarm. The circuit is open, but the pressure in the circuit prevents alarming. The respiratory rate is generally set on the lowest possible setting so that the patient may take breaths as needed without the machine cycling when a breath is not needed. The patient activates the breath by placing his or her mouth on the mouthpiece and creating a small negative pressure in the circuit by "sipping" or inhaling. Some of the breath is lost in a leak through the lips; therefore, tidal volumes are set higher than those that would be set for tracheostomy ventilation. Tidal-volume

values are set per the patient's comfort but generally range between 700 and 1200 mL.

It is also possible to use nasal-mask ventilation during the day for those who cannot tolerate mouthpiece ventilation or chose not to use it. No direct comparison studies between diurnal mouthpiece and mask ventilation have been performed. The use of the device will depend in part on the experience of the health care team taking care of the patients.

COUGH SUPPORT

Although the focus of this article is on the initiation of noninvasive ventilation modalities, the importance of implementing cough support in parallel cannot be underestimated. In fact, some have considered adequate cough support to be the most important factor in maintaining lung health. In almost all cases we will initiate cough support in parallel with noninvasive ventilatory support.

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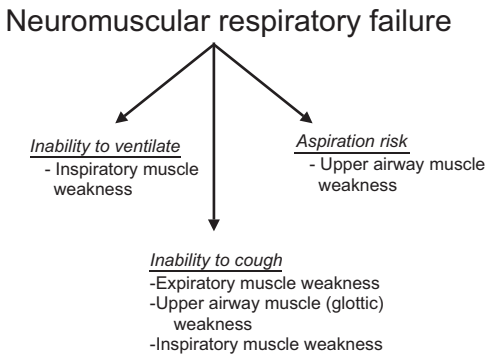

FIGURE 1
Components of neuromuscular respiratory failure.

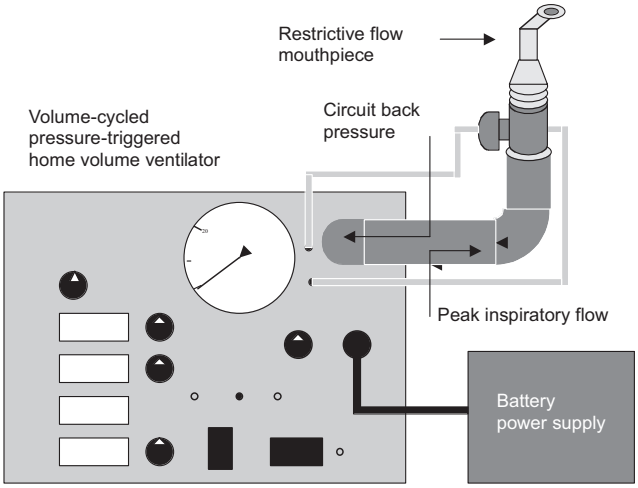

FIGURE 2
Schematic of ventilator setup for mouthpiece ventilation.

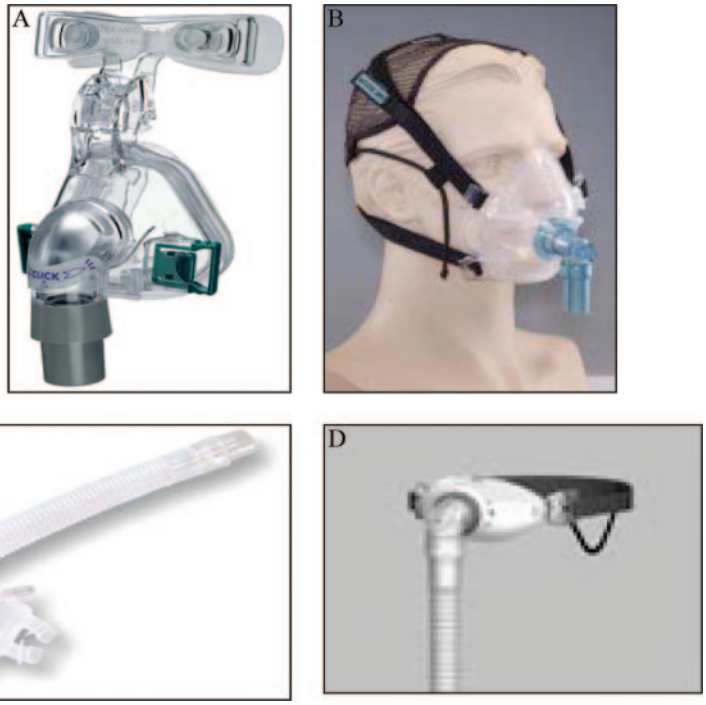

FIGURE 3
Interfaces for noninvasive mask ventilation. A, nasal mask; B, full face mask; C, nasal pillows; D, oral interface with lip seal.

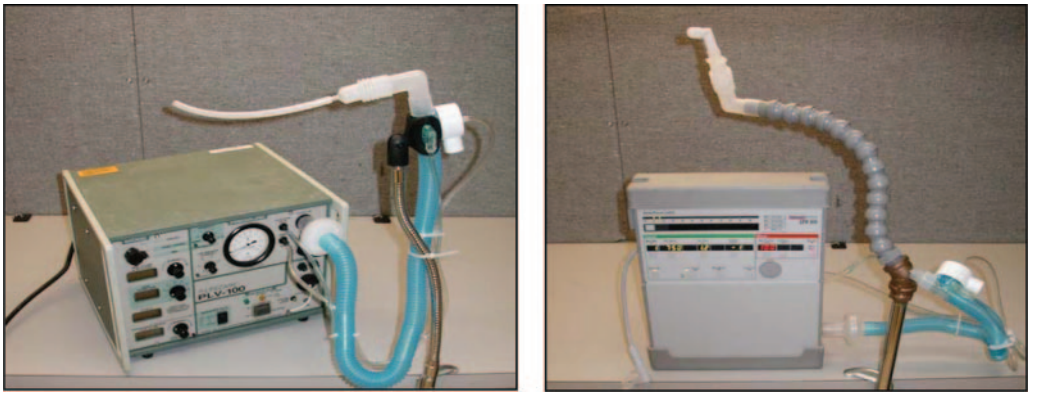

FIGURE 4
Example of ventilators with mouthpiece setup in place.

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