

Noninvasive Ventilation in Neuromuscular Disease: Equipment and Application

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Summary

Noninvasive support of ventilation is commonly needed in patients with neuromuscular disease. Body ventilators, which are used rarely, function by applying intermittent negative pressure to the thorax or abdomen. More commonly, noninvasive positive-pressure ventilation (NPPV) is used. This therapy can be applied with a variety of interfaces, ventilators, and ventilator settings. The patient interface has a major impact on comfort during NPPV. The most commonly used interfaces are nasal masks and oronasal masks. Other interfaces include nasal pillows, total face masks,

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helmets, and mouthpieces. Theoretically, any ventilator can be attached to a mask rather than an artificial airway. Portable pressure ventilators (bi-level positive airway pressure) are available specifically to provide NPPV and are commonly used to provide this therapy. Selection of NPPV settings in patients with neuromuscular disease is often done empirically and is symptom-based. Selection of settings can also be based on the results of physiologic studies or sleep studies. The use of NPPV in this patient population is likely to expand, particularly with increasing evidence that it is life-prolonging in patients with diseases such as amyotrophic lateral sclerosis. Appropriate selection of equipment and settings for NPPV is paramount to the success of this therapy. *Key words: bi-level ventilation, body ventilator, mask ventilation, mouthpiece ventilation, neuromuscular disease, noninvasive positive-pressure ventilation.* [Respir Care 2006;51(8):896–911. © 2006 Daedalus Enterprises]

Introduction

Support of ventilation is commonly needed in patients with neuromuscular disease. Body ventilators can be used, but they have been abandoned for the most part in favor of positive-pressure ventilators. Noninvasive positive-pressure ventilation (NPPV) is commonly used in patients with neuromuscular disease. This therapy can be applied with a variety of interfaces, ventilators, and ventilator settings. The purpose of this paper is to review the technical aspects of NPPV, with a focus on patients with neuromuscular disease and specific emphasis on the interface, the ventilator, and selection of ventilator settings.

Body Ventilators

Body ventilators function by applying intermittent negative pressure to the thorax or abdomen.^{1–3} These devices became popular during the polio epidemic of the 1950s. They fell out of favor with the increasing use of endotracheal intubation in the 1960s and the popularity of positive-pressure ventilation that followed. With modern meth-

ods of invasive and noninvasive ventilation, the use of body ventilators is today largely historical, although some patients continue to use these devices.

Negative-Pressure Ventilators

The tank ventilator, or iron lung, is the prototype negative-pressure ventilator (Fig. 1). It consists of a horizontal metal tank with side portholes. The patient lies supine on a foam mattress with the head protruding through a port-hole at the end. A neck collar is tightened and negative pressure (-10 cm H₂O to -35 cm H₂O at a rate of 15–25 breaths/min) is generated by a pump-driven leather bellows near the patient's feet. There is limited access to the patient while in the iron lung. Moreover, the size of the device limits portability for the patient.

The Portalong is a modified version of the iron lung. It is smaller and weighs less than the iron lung, and it fits on a standard bed. It is powered by a negative-pressure ventilator. The chest cuirass, or "tortoise shell," is a rigid shell that fits over the anterior portion of the chest, or over the chest and abdomen, and is connected to a negative-pres-

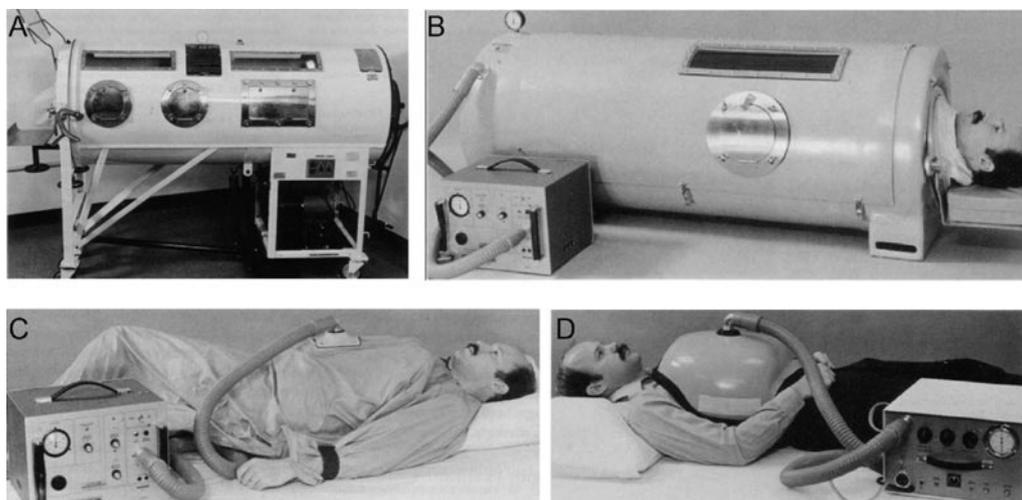


Fig. 1. Negative-pressure ventilators. A: Iron lung. B: Portalong. C: Pneumowrap. D: Cuirass. (From Reference 2, with permission.)

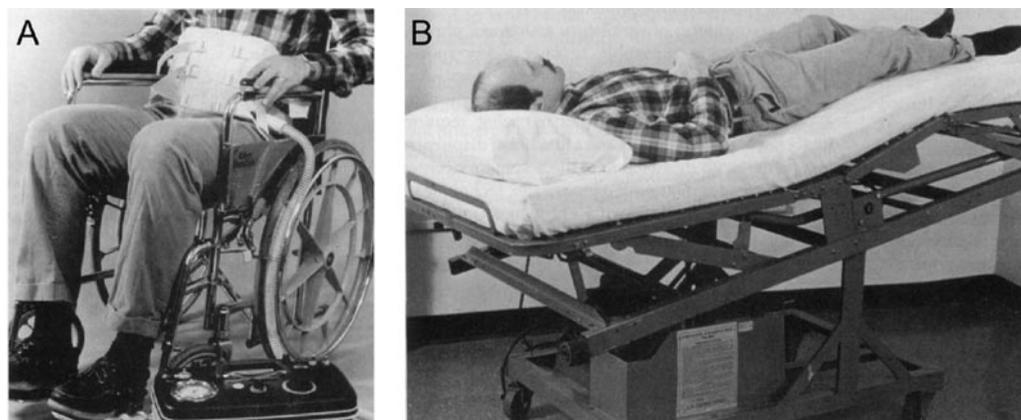


Fig. 2. Ventilators that displace the abdominal contents. A: Pneumobelt. B: Rocking bed. (From Reference 2, with permission.)

sure ventilator. It is less efficient than the iron lung or Portalung, but it has the advantages of portability and ease of application. Skin abrasions can occur if the device does not fit well.

The pneumowrap (also called the raincoat, poncho, or wrap) consists of a wind-proof, water-permeable nylon parka suspended over a rigid plastic or metal chest piece. It applies negative pressure over the anterior portion of the chest and abdomen. The patient must lie supine while using the device. It is easy to use. A common complaint with this device is coldness due to air circulation in the device.

An issue of concern with negative-pressure ventilators is the risk of upper-airway obstruction, especially during sleep. This, along with the practical considerations complicating their use, has made negative-pressure ventilators virtually obsolete. With the widespread use of NPPV, few clinicians have experience with the use of negative-pressure ventilators.

Ventilators That Displace the Abdominal Contents

The pneumobelt (Fig. 2A) is a cloth corset that contains an inflatable rubber bladder. The bladder is fitted over the abdomen and inflated intermittently by a positive-pressure ventilator (15–45 cm H₂O). The pneumobelt functions effectively only when the patient is sitting at > 30 degrees. Inflation of the bladder pushes abdominal contents inward, displacing the diaphragm upward, and assisting exhalation.⁴ Deflation of the bladder allows passive downward motion of the diaphragm and an associated inhaled tidal volume (V_T).

The rocking bed (Fig. 2B) functions by rocking the patient in a vertical axis over an arc of 40–45 degrees. The force of gravity on the abdomen affects diaphragm motion and V_T. A greater rocking arc increases ventilation but is more uncomfortable for the patient. The rocking bed is

Table 1. Desirable Characteristics of an NPPV Interface

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
Quickly removable
Anti-asphyxia mechanism
Compatible with a wide range of ventilators

NPPV = noninvasive positive-pressure ventilation

limited by its bulkiness, lack of portability, and relative inefficiency.

The Interface for NPPV

The patient interface has a major impact on comfort during NPPV. A poorly-fitting interface decreases clinical effectiveness and patient adherence to therapy. Table 1 lists desirable characteristics of an NPPV interface. The interface is often the weak link in the application of NPPV. The most commonly used interfaces are nasal masks and oronasal masks (Fig. 3), and there are advantages and disadvantages of each (Table 2). Other interfaces (Figs. 4 and 5) include nasal pillows, total face masks, helmets, and mouthpieces. In the past, custom masks were occasionally molded to fit the anatomy of the patient's face. Today, a variety of sizes and designs are commercially available from a number of manufacturers, as both disposable and reusable designs. The variety of interfaces commonly available can be confusing for the clinician who is



Fig. 3. Left: Nasal mask. Right: Oronasal mask. (Courtesy of Respironics, Murrysville, Pennsylvania.)

only occasionally involved in the care of patients who need NPPV.

Nasal Interface

The nasal interface provides ventilation to the nose. Most common is the nasal mask, which 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 nasal masks are gel-filled and others use an open cushion with an inner lip, in which pressure inside the mask pushes the cushion against the face (Fig. 6).

Nasal pillows, or nasal plugs or cushions, are available from several manufacturers. This interface consists of soft

Table 2. Advantages and Disadvantages of Various Types of NPPV Interface

Interface	Advantages	Disadvantages
Nasal	<ul style="list-style-type: none"> Less risk of aspiration Easier secretion clearance Less claustrophobia Easier speech Patient may be able to eat Easy to fit and secure Less dead space 	<ul style="list-style-type: none"> Mouth leak Higher resistance through nasal passages Less effective with nasal obstruction Nasal irritation and rhinorrhea Mouth dryness
Oronasal	<ul style="list-style-type: none"> Better oral leak control More effective in mouth breathers 	<ul style="list-style-type: none"> Increased dead space Claustrophobia Increased aspiration risk Increased difficulty speaking and eating Asphyxiation with ventilator malfunction
Mouthpiece	<ul style="list-style-type: none"> Less interference with speech Very little dead space May not require headgear 	<ul style="list-style-type: none"> Less effective if patient cannot maintain mouth seal Usually requires nasal or oronasal interface at night Nasal leak
Total face mask	<ul style="list-style-type: none"> May be more comfortable for some patients Easier to fit (one size fits all) Less facial-skin breakdown 	<ul style="list-style-type: none"> Potential for orthodontic injury Potentially greater dead space Potential for drying of the eyes Cannot deliver aerosolized medications
Helmet	<ul style="list-style-type: none"> May be more comfortable for some patients Easier to fit No facial-skin breakdown 	<ul style="list-style-type: none"> Rebreathing Poorer patient-ventilator synchrony Less respiratory muscle unloading Risk of asphyxiation if ventilator malfunctions Cannot deliver aerosolized medications

NPPV = noninvasive positive-pressure ventilation



Fig. 4. Left: Nasal pillows. (Courtesy of Puritan-Bennett.) Center. Total face mask. (Courtesy of Respirationics, Murrysville, Pennsylvania.) Right: Helmet.

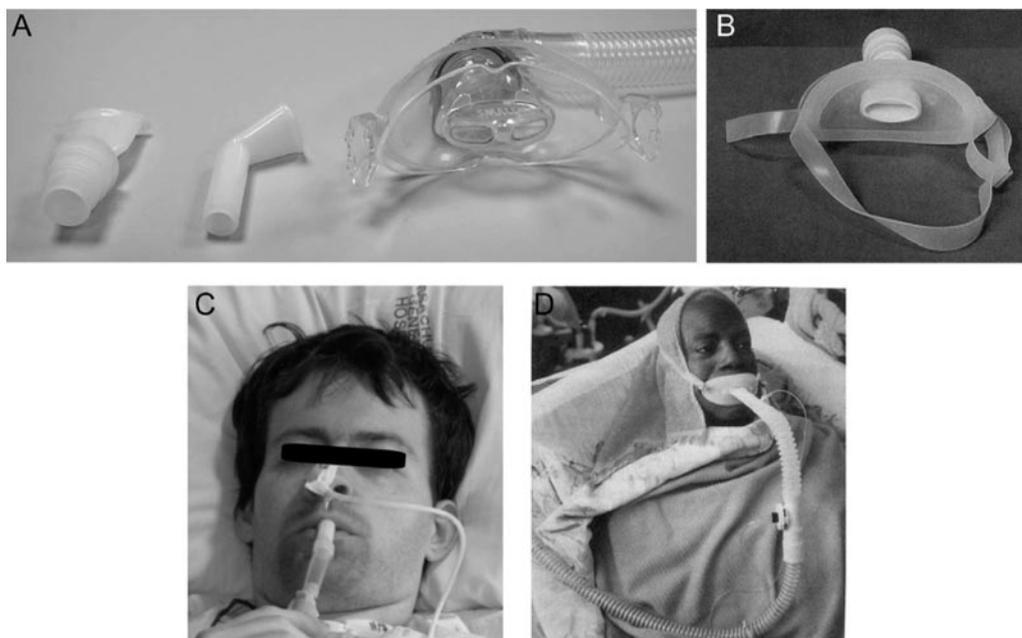


Fig. 5. A: Mouthpiece devices for noninvasive positive-pressure ventilation (NPPV). B: Mouthpiece with lip-seal. (From Reference 5, with permission.) C: Patient using an angled mouthpiece for NPPV. D: Patient using mouthpiece with lip-seal for NPPV. (From Reference 5, with permission.)

plastic plugs inserted into the nares, shaped in a way that the pressure applied during inspiration helps to seal the wall of the pillows against the inner surface of the nasal vestibule. The pillows are held in place with specifically manufactured headgear.

Mouth Leak

Leak through the mouth is common with a nasal mask. This can affect comfort, cause dry mouth, result in less-

effective ventilation, affect patient-ventilator interaction (trigger and cycle), and disrupt sleep architecture. Navelesi et al⁶ assessed 26 stable hypercapnic patients who were naïve to NPPV and had restrictive thoracic disease or chronic obstructive pulmonary disease (COPD). In a cross-over design, each patient received 30-min trials with a nasal mask, oronasal mask, or nasal pillows. The nasal mask was better tolerated than the other interfaces, but P_{aCO_2} was lower with an oronasal mask than with a nasal mask, and minute ventilation was higher with the oronasal

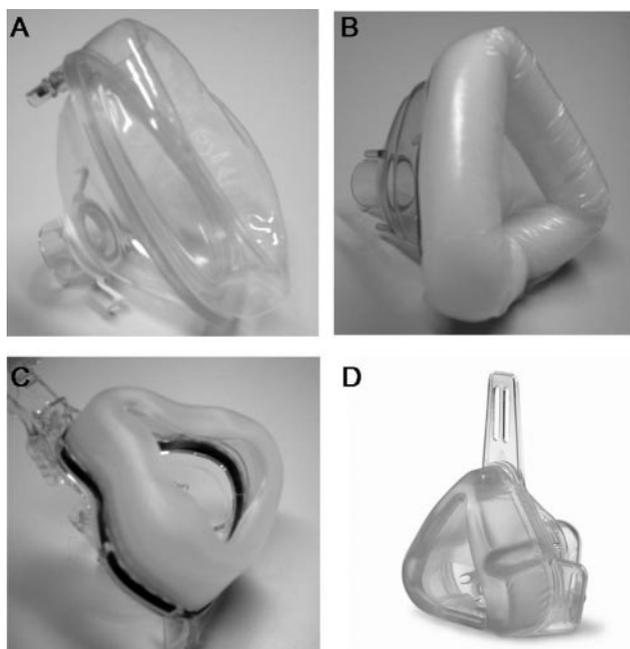


Fig. 6. A: Mask with inflatable cushion. B: Mask with foam-filled cushion. C: Mask with inner lip that fits to the face when pressure is applied to the mask. D: Mask with a gel-filled cushion.

mask than with the nasal mask because of greater V_T . A criticism of that study⁶ is that the researchers used a very comfortable mask (gel cushion) during nasal NPPV but a less comfortable mask (air inflatable cushion) during oronasal ventilation. In acutely ill patients, Kwok et al⁷ reported that nasal and oronasal masks performed similarly with regard to gas exchange and avoiding intubation, but the oronasal mask was better tolerated. Unsuccessful NPPV has been associated with mouth leak.⁸

Willson et al⁹ studied the effectiveness of a nasal mask versus an oronasal mask in patients with nocturnal hypoventilation. They reported that the type of interface did not affect gas exchange (oxygen saturation or transcutaneously measured P_{CO_2}), and arousal indices were comparable for both masks. Meyer et al¹⁰ assessed the effect of mouth leak on sleep quality in 6 patients with chest wall and neuromuscular disease using nocturnal nasal NPPV. All patients had air leak through the mouth for the majority of sleep. Air leaking through the mouth was associated with frequent arousals during lighter stages of sleep, which interfered with progression to deeper stages, thus compromising sleep quality. Teschler et al¹¹ studied the acute effect of sealing the mouth on sleep architecture and transcutaneously measured P_{CO_2} in 9 patients receiving long-term nasal ventilation with symptomatic mouth leak. On one night, the mouth was taped closed and on another night the mouth was untaped (allowing leak). P_{CO_2} was lower in 8 of 9 patients with the mouth taped. The arousal index was lower in every patient with the mouth taped

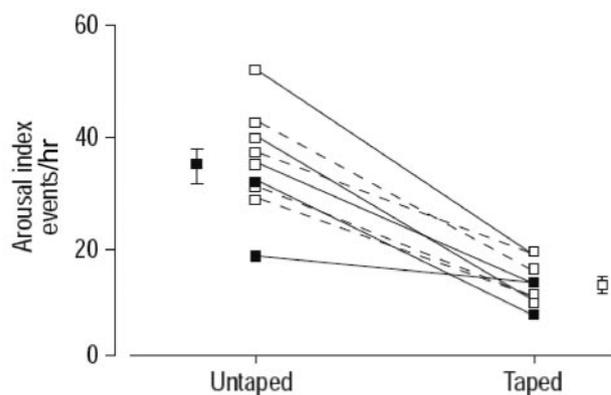


Fig. 7. Effect of taping the mouth on arousal index. The black squares represent data points from patients who had cardiac failure and Cheyne-Stokes breathing. The white squares represent data points from patients who had chronic obstructive pulmonary disease or kyphoscoliosis (of the latter there was only one). The solid lines represent patients who received supplemental oxygen. The dashed lines represent patients who did not receive supplemental oxygen. (From Reference 11, with permission.)

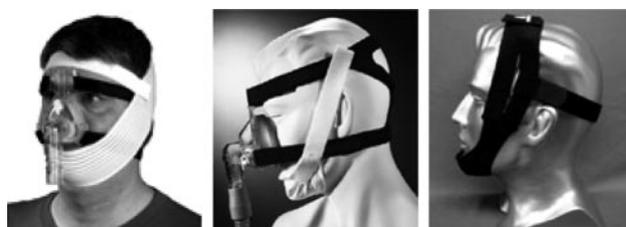


Fig. 8. Examples of commercially available chin straps. (Courtesy of Respirationics, Murrysville, Pennsylvania.)

(Fig. 7). The authors concluded that mouth leak reduced effective nasal ventilatory support, increased P_{CO_2} , and disrupted sleep architecture. These studies suggest that mouth leak during nasal ventilation may have an important effect on sleep quality during NPPV.

Several approaches can be used if excessive mouth leak occurs with a nasal interface. The patient can be coached to keep the mouth closed, but this is usually ineffective, particularly during sleep. A chin strap (Fig. 8) can be tried. Willson et al⁹ found the chin strap effective in 14 of 16 subjects. Gonzalez et al¹² reported that a chin strap can reduce air leak and P_{aCO_2} . However, the chin strap was only effective in about a third of patients. If persistent mouth leak occurs, an oronasal mask is often needed. In some patients, a nasal interface can be used during the daytime and an oronasal mask is used at night to minimize mouth leak and improve sleep quality.

Oronasal Interface

The oronasal mask should fit just above the junction of the nasal bone and cartilage to just below the lower lip.

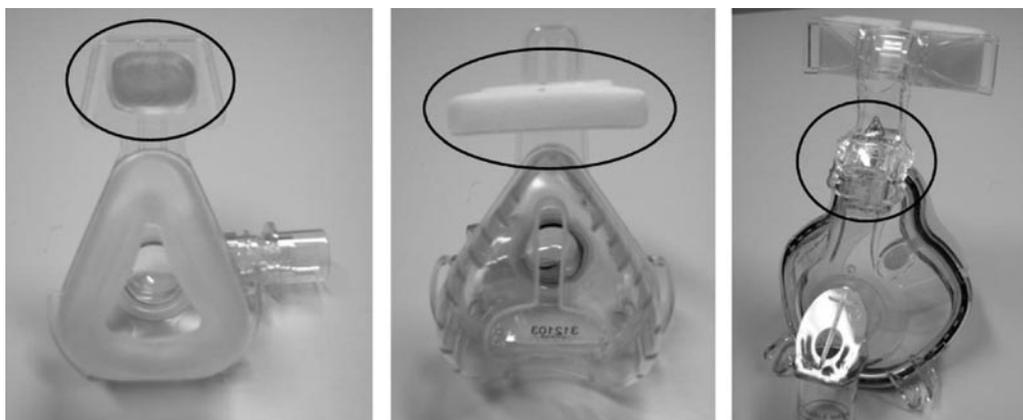


Fig. 9. Forehead spacer designs to decrease the risk of facial skin breakdown. Left: Gel spacer. Center: Foam spacer. Right: Adjustable forehead arm.

Some commercially available oronasal masks have a soft air-filled or foam-filled cushion. Others have an inflatable cushion, in which air can be added or removed after it is fitted to the patient to improve mask fit.¹³ As with nasal masks, most oronasal masks designed specifically for NPPV have an open cushion with an inner lip. They are commercially available in a variety of sizes and are often equipped with anti-asphyxia valves and quick-release features.

Headgear

Appropriate headgear is needed to maintain correct position of the mask. Elastic straps with holes that attach to hooks are used with some oronasal masks. The hooks can be either on the outer edge of the mask or near the center of the mask. Attachment of the headgear to the outer edge of the mask may better distribute the pressure of the mask and facilitate a seal. Most modern masks designed specifically for NPPV use cloth straps and Velcro to secure the mask. The cloth straps fit through attachments at the sides and top of the mask. Velcro allows nearly infinite adjustments of the headgear. Some headgear uses a cap design to minimize movement of the straps. A common mistake is to fit the headgear too tightly. It should be possible to pass 1 or 2 fingers between the headgear and the face. Fitting the headgear too tightly may not improve the fit and always decreases patient comfort and compliance.

The design of some NPPV masks is such that the top of the mask is secured on the forehead rather than at the bridge of the nose. Forehead spacers are an important feature of this design (Fig. 9). These foam or gel cushions fill the gap between the forehead and the mask, thus reducing pressure on the bridge of the nose. Another design that decreases pressure on the nose is an adjustable forehead arm on the mask.



Fig. 10. Facial skin breakdown secondary to mask used for non-invasive positive-pressure ventilation. (From Reference 14, with permission.)

Facial Skin Breakdown

A potential problem with nasal and oronasal masks is facial skin breakdown, which most commonly occurs on the bridge of the nose (Fig. 10).^{14,15} Several approaches can be taken to address this issue. Perhaps most important is to avoid strapping the mask too tightly. Although large leaks around the mask are uncomfortable (particularly leaks toward the eyes), NPPV can be successful with small-to-moderate leaks, and most modern ventilators designed for NPPV adequately compensate for this leak. It is important that the mask size is appropriate for the patient. Too large or small a mask increases the likelihood of poor fit and facial soreness. Sometimes it helps to change to a mask made by a different manufacturer. A mask with a forehead spacer or an adjustable forehead arm can be used to reduce the pressure on the bridge of the nose. Wound-care tape (eg, Duoderm or Hydrogel) can be applied to the bridge of the nose,

but this is less effective after substantial skin breakdown has occurred. One can also consider the use of a different interface (eg, nasal pillows, mouthpiece, or total face mask).

Rebreathing

The interface can affect the degree of rebreathing during NPPV if the ventilator circuit has a leak port for exhalation. In a lung-model study, Schettino et al¹⁶ reported a lower volume of rebreathed CO₂ with the exhalation port in the mask, as compared to the exhalation port in the circuit. Saatci et al,¹⁷ also using a lung model, found that an oronasal mask with the exhalation port in the mask decreased the total dynamic dead space, compared to having the leak port in the circuit. Theoretically, with a nasal mask the patient can exhale through the mouth, which should decrease rebreathing.

Total Face Mask

The total face mask¹⁸ is an alternative for patients who are unable to obtain a good seal with a nasal mask, experience skin breakdown, or are claustrophobic. Because the total face mask covers the entire face, there are no pressure points around the nose to cause sores or skin breakdown. An effective seal is created around the outside of the face as air pressure from the system inflates the soft, flexible sealing layer. This minimizes leaks while providing optimal airflow through the nasal passages. Because the air pressure is able to circulate throughout the mask, it may be more comfortable for the patient.

Helmet

The helmet (a transparent, latex-free, polyvinyl chloride cylinder linked by a metallic ring to a soft collar that seals the helmet around the neck) has been proposed as an alternative to conventional face mask for NPPV in patients with acute respiratory failure.^{19–24} One concern with the helmet is the risk of rebreathing.²³ Also, the helmet is less effective in unloading inspiratory muscles than is a standard face mask.²⁴ This interface is probably not appropriate for patients with neuromuscular disease and chronic respiratory failure.

Mouthpiece

Mouthpieces are more often used in patients with chronic respiratory failure, but may be used occasionally during acute respiratory failure.^{5,25,26} Some patients

with neuromuscular disorders use volume-controlled ventilators and small angled mouthpieces or straw-type mouthpieces. Although a few individuals have learned to use such mouthpieces during sleep, most patients change to a nasal or oronasal mask at night. The mouthpiece can be mounted close to the head so that the patient can speak after each breath. Some mouthpieces are configured with a lip seal to minimize air leak. The patient may need nose plugs if nasal leak occurs. Orthodontic deformity⁵ and hypersalivation can occur with mouthpiece ventilation.

Considerable experience with mouthpiece NPPV has been reported by Bach et al.^{5,25} In a report of 257 patients with acute or chronic respiratory failure, mouthpiece NPPV was the predominant method of daytime ventilator support in most of the patients. Mouthpiece NPPV was also used at night by 163 patients, 61 of whom had little or no measurable vital capacity. A lip seal or custom orthodontic interface was used for nocturnal mouthpiece NPPV.

Mouthpiece ventilation can be provided with a bi-level ventilator or a portable volume ventilator. Currently available portable volume ventilators have low-pressure alarms to detect a disconnection. This makes open-circuit ventilation, such as that used with mouthpiece ventilation, difficult, because of low-pressure alarming. Open-circuit mouthpiece ventilation can be performed when sufficient peak inspiratory flow is used to create enough back-pressure against the mouthpiece to prevent a low-pressure alarm. When the set ventilator rate is sufficient to prevent an apnea alarm, the ventilator circuit can remain open for extended periods without either low-pressure or apnea alarming. The patient receives a ventilator-assisted breath as often as needed by making a “sip” effort through the mouthpiece to trigger the ventilator. This allows the patient to receive as much noninvasive ventilatory support as needed. Boitano and Benditt²⁶ determined which portable volume ventilators support mouthpiece ventilation and what peak inspiratory flows create adequate circuit pressure to prevent a low-pressure alarm. The following ventilators supported mouthpiece ventilation: Respironics Lifecare PLV-100 and PLV Continuum, Mallinckrodt Achieva PSO2, Pulmonetics LTV800, Newport HT50, and Uni-Vent Eagle 754.

The Ventilator for NPPV

Type

Any ventilator can be attached to a face mask or other NPPV interface, rather than an artificial airway. Critical-care ventilators can be used to provide NPPV in the hospital setting, with the advantages of precise control of fraction of inspired oxygen (F_IO₂), various modes and in-

Table 3. Considerations in the Selection of a Ventilator for NPPV

 Leak compensation

Trigger and cycle coupled to patient's breathing pattern

Rebreathing

Oxygen delivery (acute care)

Monitoring

Alarms (safety vs nuisance)

Portability (size, weight, battery)

Tamper-proof

Cost

 NPPV = noninvasive positive-pressure ventilation

spiratory flow patterns, and separation of inspiratory and expiratory gases to limit rebreathing. Critical-care ventilators have extensive monitors and alarms, which may be desirable during invasive ventilation but can be distracting and annoying for patients and clinicians during NPPV. The greatest disadvantage of critical-care ventilators is that they have difficulty dealing with the leaks that occur during noninvasive ventilation. Some modern ventilators can be set to provide either invasive or noninvasive ventilation, providing the benefits of NPPV within a critical-care ventilator. Table 3 lists considerations in the selection of a ventilator for NPPV.

Conventional home-care ventilators have been used to provide NPPV. These ventilators function well when little patient-ventilator interaction occurs, such as in patients with neuromuscular disease. In the past, the triggering on these ventilators was poor and inspiratory flow was fixed, which limited their use for NPPV. These ventilators are generally intolerant of leaks, although, to some extent, leak compensation can be made by increasing the set V_T . These ventilators have a limited number of alarms, and they operate from battery power, both of which are benefits for home use. These ventilators have a true exhalation valve, so rebreathing is not a problem. Newer home-care ventilators are much improved and offer a variety of modes and features desirable for NPPV. Some are small and light-weight and thus portable.

Portable pressure ventilators (bi-level positive airway pressure) are available specifically for NPPV (Fig. 11). Their major advantage is the ability to function correctly with leaks. In fact, they require a leak to function correctly. They are blower devices that vary inspiratory and expiratory pressure in response to patient demand. These ventilators typically provide pressure-support ventilation (PSV), but most can also provide pressure-controlled ventilation (PCV). They do not provide volume-controlled ventilation (VCV). PSV (or PCV) is achieved by setting the inspiratory positive airway pressure and expiratory positive airway pressure (EPAP). The mathematical difference between inspiratory positive airway pressure and

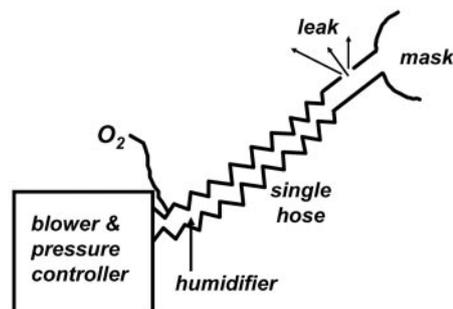


Fig. 11. Components of a bi-level ventilator used for noninvasive positive-pressure ventilation.

EPAP is the level of pressure support (or pressure control). They typically provide modest inspiratory pressure (≤ 30 cm H₂O) and expiratory pressure (≤ 15 cm H₂O). Evaluations of these have reported that they perform as well as, and sometimes better than, critical-care ventilators.²⁷⁻³⁴

A concern with the portable pressure ventilators is the potential for CO₂ rebreathing.³⁵⁻³⁷ These ventilators use a single hose that does not have a true exhalation valve. Expired gas passes through a fixed leak established in the device. Particularly with low flow from the ventilator, as may occur with a low level of positive end-expiratory pressure (PEEP), there is inadequate flushing of CO₂, and consequent rebreathing. This problem can be resolved by increasing the PEEP (≥ 4 cm H₂O) or using a valve that prevents rebreathing. Increasing the leak flow also flushes CO₂ from the system. A fixed leak in the mask may produce less rebreathing than a fixed leak in the hose.¹⁷ A nonbreathing exhalation valve can also be used, although Hill et al³⁸ reported no benefit from the use of such a valve in patients receiving long-term nasal ventilation, probably because of leak through the mouth.

Supplemental oxygen is usually not necessary in patients with neuromuscular disease who require NPPV, unless they develop an acute process such as pneumonia. Precise oxygen administration is difficult with some ventilators used for NPPV. Oxygen is typically titrated into the circuit at the ventilator outlet or into the mask.^{39,40} The F_{IO_2} is determined by the oxygen flow, ventilatory pattern, and leak. Because of the high flow from the ventilator, it is generally difficult to achieve an F_{IO_2} greater than 0.60. Newer noninvasive ventilators allow a precisely set F_{IO_2} .

Mode

There are advantages and disadvantages to VCV, PSV, and PCV for NPPV (Table 4). PSV is commonly used for NPPV. A theoretical advantage of PSV is that it varies the inspiratory flow to meet patient demand, which should improve patient comfort during NPPV. Girault et al⁴¹ com-

Table 4. Comparison of Volume Ventilator and Pressure Ventilator for NPPV in Patients With Neuromuscular Disease

Volume Ventilator
More complicated to use
Wide range of alarms
Constant tidal volume
Breath-stacking possible
No leak compensation
Can be used without PEEP
Rebreathing minimized
Pressure Ventilator
Simple to use
Limited alarms
Variable tidal volume
Breath-stacking not possible
Leak compensation
PEEP (EPAP) always present
Rebreathing possible

NPPV = noninvasive positive-pressure ventilation

PEEP = positive end-expiratory pressure

EPAP = expiratory positive airway pressure

pared VCV and PSV in 15 patients with hypercapnic respiratory failure. VCV and PSV similarly improved breathing pattern and gas exchange. The inspiratory work load was less with VCV, but patient comfort was greater with PSV. In patients with stable cystic fibrosis, Fauroux et al⁴² found that both PSV and VSV decreased respiratory-muscle unloading. The available evidence does not show any mode to be clearly superior to another for providing NPPV in patients with neuromuscular disease (Table 5).^{5,43-48}

A new mode, proportional-assist ventilation (PAV), has been used effectively and may improve patient tolerance of NPPV during acute respiratory failure.⁴⁹⁻⁵¹ In patients with chronic respiratory failure due to neuromuscular disease and chest-wall deformity, Hart et al⁵² found that PSV and PAV produced similar improvements in physiologic variables. However, greater diaphragm unloading was observed with PSV than with PAV, which was associated with greater symptomatic benefit. Porta et al⁵³ compared the short-term physiologic effects of PSV and PAV in 11 patients with clinically stable COPD and 7 patients with restrictive chest-wall diseases. They reported that noninvasive PAV, set at the patient's comfort, was not superior to PSV in unloading the inspiratory muscles. Winck et al⁵⁴ compared the tolerance and physiologic effects of a 5-night treatment with either nasal PAV or PSV in patients with chronic ventilatory failure (4 with COPD and 10 with restrictive thoracic diseases). PAV and PSV had similar patient tolerance and were equally effective in reducing daytime hypercapnia and improving nocturnal oxygen saturation and symptoms. PAV was associated with less nasal and oral dryness but with more alarm noise.

Trigger

Modern ventilator triggers are very sensitive to patient effort, so auto-triggering can be problematic. Auto-triggering can occur because of leaks, which is less of a problem with a bi-level ventilator.⁵⁵ Failure to trigger is usually due to muscle weakness, intrinsic PEEP, or a high level of support. Fanfulla et al⁵⁶ reported the presence of ineffective trigger efforts with a high level of pressure support in patients with neuromuscular disease. Central apnea was found to be more prevalent with PSV in normal subjects using a nasal mask,⁵⁷ in intubated patients,⁵⁸ and in patients being evaluated in an out-patient sleep laboratory.⁵⁹ For these reasons, a back-up rate is recommended during NPPV, particularly with nocturnal applications.

Rise Time

Rise time (pressurization rate) is the amount of time required to reach the pressure target at the onset of inhalation with PSV and PCV. With a slow rise time it takes longer to reach the pressure target, and with a fast rise time the pressure target is reached sooner. The rise time during NPPV can be adjusted on some ventilators. A faster rise time has been shown to better unload the respiratory muscles of patients with COPD (Fig. 12), but this may be accompanied by substantial air leaks and poor tolerance.⁶⁰ In patients with neuromuscular disease, a slower rise time is often better tolerated. Rise time should be set to maximize patient comfort.

Cycle

The term "cycle" refers to the change-over from the inspiratory phase to the expiratory phase. During PSV, the inspiratory phase terminates when flow falls to a predetermined fraction of peak inspiratory flow. Hotchkiss et al⁶¹ used a mathematical model to evaluate the effect of mask leak during PSV with NPPV. They found that PSV, applied in the presence of an inspiratory leak, can be accompanied by variations in the duration of the inspiratory phase and the development of intrinsic PEEP. Several approaches can be taken to address this issue. First, mask leak should be minimized. Second, some ventilators allow setting the maximum inspiratory time, which is useful in the presence of leaks.^{62,63} Third, on some ventilators the flow cycle criteria can be adjusted to mitigate issues with leaks.⁶⁴

Ramp

Ramp is a feature available on many bi-level ventilators. Ramp causes the pressure to increase gradually, from a low level to the prescribed level. This may be useful in patients with obstructive sleep apnea (OSA) who require a

NONINVASIVE VENTILATION IN NEUROMUSCULAR DISEASE

Table 5. NPPV Studies That Compared Ventilator Modes in Patients With Chronic Respiratory Failure

First Author	Study Design	Results
Navalesi ⁶	Crossover study. 26 stable hypercapnic patients with restrictive or obstructive pulmonary disease	No differences in V_T , blood gas values, or breathing pattern using PSV or VCV
Restrick ⁴³	Crossover study. 12 patients with nocturnal hypoventilation	PSV was as effective as VCV
Meecham Jones ⁴⁴	Crossover study. Patients with stable chronic respiratory failure	PSV was as effective as VCV
Schonhofer ⁴⁵	Crossover study. 30 patients with chronic respiratory failure (variety of diagnoses). Compared VCV and BiPAP in timed mode	Timed mode inadequate in some patients
Chadda ⁴⁶	Crossover study. 13 patients with neuromuscular disease	PSV, PCV, and VCV had similar effects on minute ventilation and respiratory muscle unloading. 5 patients preferred PSV, 4 preferred VCV, and 4 preferred PCV.
Windisch ⁴⁷	Crossover study. 5 patients with COPD and 5 patients without COPD	Nocturnal PCV and VCV had similar effects on gas exchange and sleep quality. More gastrointestinal adverse effects with VCV.
Munoz ⁴⁸	Retrospective study. 110 patients with chronic respiratory failure (neuromuscular disease, kyphoscoliosis, or post-tuberculosis)	Assist/control or control mode with VCV comparable

V_T = tidal volume
 NPPV = noninvasive positive-pressure ventilation
 PSV = pressure-support ventilation
 VCV = volume-controlled ventilation
 PCV = pressure-controlled ventilation
 BiPAP = bi-level positive airway pressure

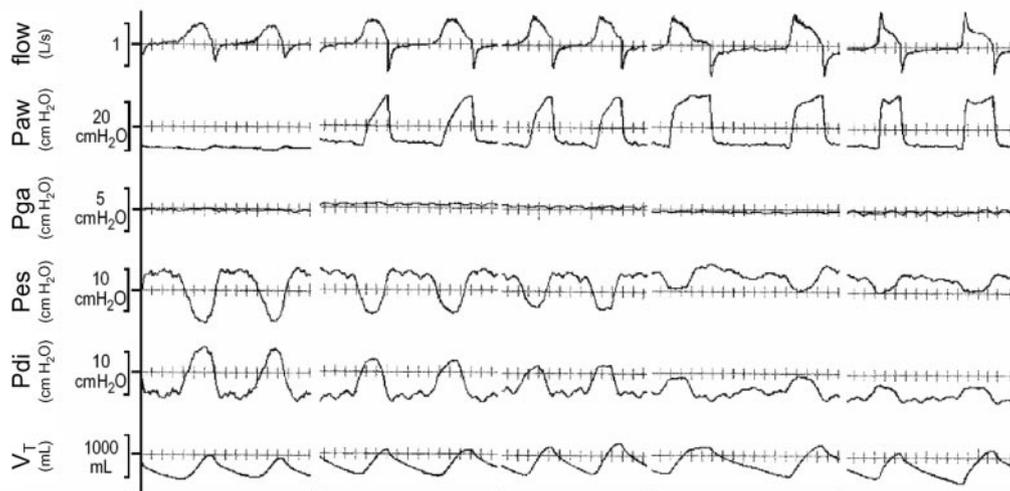


Fig. 12. Patient graphics illustrating greater respiratory-muscle unloading with a faster rise time. P_{aw} = pressure at the airway opening. P_{ga} = gastric pressure. P_{es} = esophageal pressure. P_{di} = transdiaphragmatic pressure. V_T = tidal volume. (From Reference 60, with permission.)

high level of continuous positive airway pressure, to improve tolerance of the pressure and mask when first applied. In patients with neuromuscular disease who require ventilatory assistance, the use of ramp is undesirable because it can delay the onset of effective therapy. In patients who are having difficulty acclimating to the mask and pressure, however, ramp may help the patient adjust to the therapy. Once the patient is comfortable with the therapy, ramp should be discontinued.

Humidification

Upper-airway symptoms of dryness and nasal stuffiness commonly occur during NPPV. This is particularly problematic with use of a nasal mask and mouth leak.^{65–68} Mouth leaks can increase nasal resistance.⁶⁵ Issues related to mouth leak and humidification have been described with a nasal mask and continuous positive airway pressure for treatment of OSA, and this may have implications in

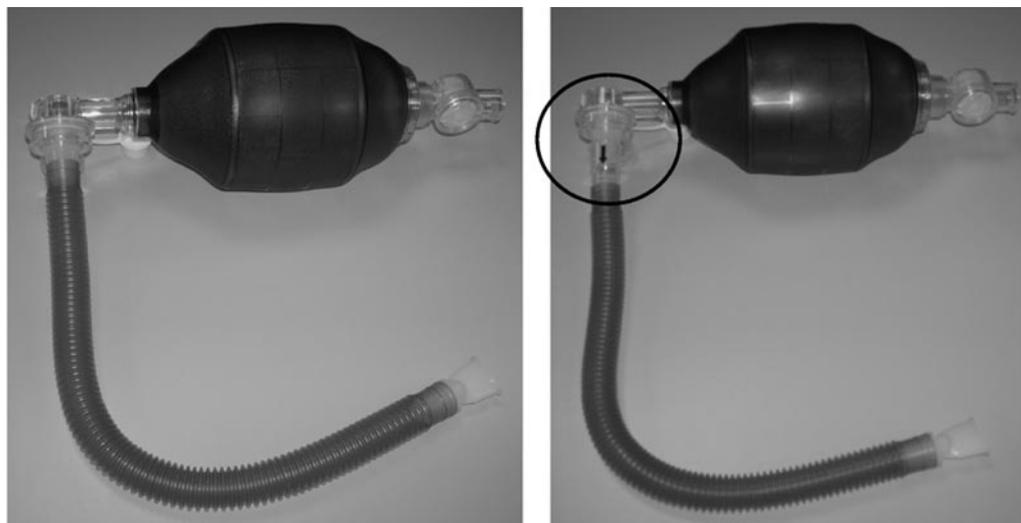


Fig. 13. Left: Bag-valve resuscitator configured for mouthpiece ventilation. Right: Bag-valve resuscitator with one-way valve to allow breath-stacking.

patients with neuromuscular disease using NPPV. Upper-airway symptoms can be addressed by use of an oronasal mask or heated humidification. Mador et al⁶⁸ reported that addition of heated humidification was associated with fewer symptoms attributable to the upper airway. The use of humidification during NPPV in patients with neuromuscular disease should be based on patient comfort. If a heated humidifier is used, it is important to avoid excessive condensate in the circuit, which could accidentally dump into the mask. A heat-and-moisture exchanger should not be used with NPPV, because it would increase dead space and resistance, which might interfere with triggering and cycling.^{69,70} Moreover, if a nasal interface is used, much of the exhaled gas may escape from the mouth, thus making the heat-and-moisture exchanger virtually ineffective. If upper-airway symptoms persist despite adequate humidification, they can be addressed with saline spray and nasal steroids.

Safety

The role of alarms and monitoring for noninvasive ventilators is controversial. Many patients can sustain adequate spontaneous breathing for short periods without ventilatory support. Nonetheless, disconnect and power-loss alarms are recommended. Airway pressure and volume monitors are desirable, but not mandatory, for acutely ill patients using NPPV. Newer ventilators used for NPPV provide sophisticated monitoring with graphics and alarms.

Battery backup is desirable, particularly with patients who require full support by NPPV. For patients who use only nocturnal NPPV, battery backup is less important and may not be necessary. Battery power may also be neces-

sary to provide more independence for patients who require full ventilatory support. Ideally, the battery should be lightweight, allow extended use on a single charge, and recharge quickly. Use of a battery with a portable pressure (bi-level) ventilator is not straightforward and often requires some “jury-rigging” by the patient or the home-care provider. Use of a battery with a portable volume ventilator is more straightforward, and these ventilators typically come with internal batteries.

For patients who require full ventilatory support via NPPV, the family and other care providers should be trained in the use of a manual bag-valve resuscitator. For patients who use mouthpiece ventilation, the bag-valve resuscitator can be set up with a mouthpiece (Fig. 13). Family members and other care providers should be trained in the proper use of a bag-valve resuscitator with a mask. The bag-valve resuscitator can also be modified to provide breath stacking, by placing a one-way valve at the outlet of the device.⁷¹ If this is done, it is important to remind the patient and care providers that the one-way valve must be removed to provide bag-valve ventilation via mouthpiece or mask.

Glossopharyngeal Breathing

Glossopharyngeal breathing (“frog breathing”) involves the use of the tongue and pharyngeal muscles to produce a V_T by projecting boluses of air past the glottis.⁷² The glottis closes with each gulp of air. Each V_T consists of 6–9 gulps of 60–100 mL each. Most individuals need considerable instruction and encouragement to learn this technique, as well as hours of practice to master it. This can provide an individual who has weak inspiratory mus-

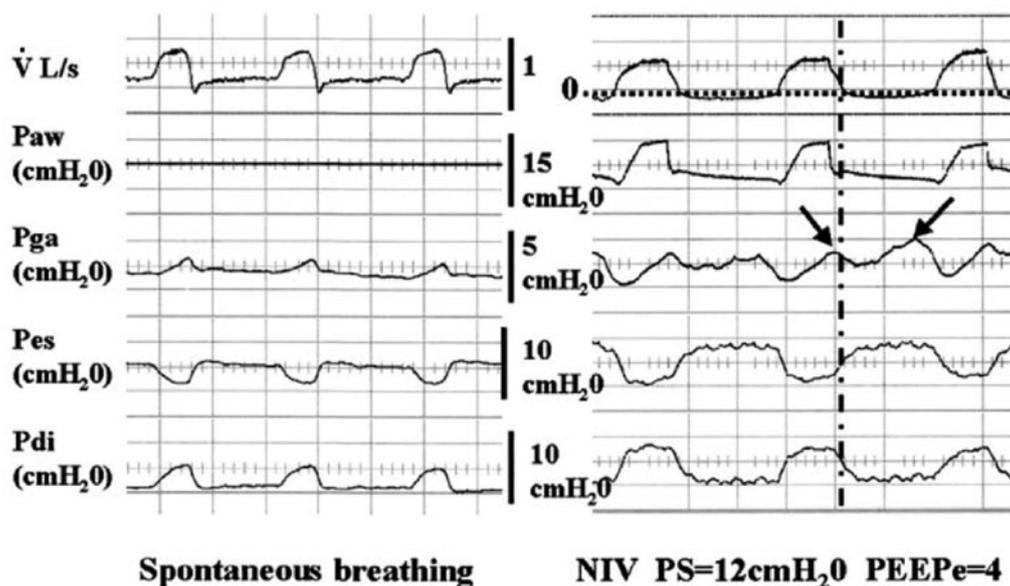


Fig. 14. Respiratory mechanics during spontaneous breathing and noninvasive positive-pressure ventilation (NPPV), illustrating expiratory-muscle activation. The dotted line represents the baseline zero flow. The vertical dotted-and-dashed line represents the beginning of expiration. Expiratory-muscle recruitment is shown as the rise in gastric pressure (P_{ga}) during expiration, from its end-expiratory level (left arrow) to the maximum at end-expiratory (right arrow). P_{aw} = airway pressure. P_{di} = transdiaphragmatic pressure. P_{es} = esophageal pressure. \dot{V} = airflow. (From Reference 56, with permission.)

cles ventilator-free time or a backup form of ventilation in the event of ventilator failure. It can also be used to improve cough effectiveness. Glossopharyngeal breathing is limited by oropharyngeal muscle weakness; the muscles of the tongue, soft palate, pharynx, and larynx must be functional. This breathing technique is rarely taught, since few health-care professionals are familiar with it.

Selection of Settings for NPPV

Selection of settings for NPPV in patients with neuromuscular disease is often done empirically, and is symptom-based. Initially, the NPPV settings are selected based on short-term symptoms such as chest expansion, accessory muscle use, and comfort.⁵⁷ Often, the prescribed settings are a compromise between those likely to be therapeutic and those tolerated by the patient. In some cases, the initial settings may be subtherapeutic, but are then increased as the patient becomes tolerant of the mask and the pressure in the mask (a process called desensitization). As the patient becomes increasingly tolerant of the therapy, settings are further adjusted, with the goal of improving symptoms of morning headache, fatigue, and daytime sleepiness.

The NPPV settings can be selected based on physiologic measures,⁵⁷ including V_T , minute ventilation, and arterial blood gas values, with the aim of improving daytime P_{aCO_2} . Sophisticated measurements of respiratory-muscle unloading, such as transdiaphragmatic pressure,

can also be used, but these are generally limited to research protocols.

The role of polysomnography for selection of NPPV settings in patients with neuromuscular disease is unclear and controversial. Selection of settings for daytime use, as occurs with empirical approaches, may result in settings that are not appropriate for nighttime use. Some have recommended selection of settings from sleep studies for patients with neuromuscular disease,^{73,74} whereas others have suggested that this is not necessary.^{75,76} Several practical issues also deserve consideration. There are long wait times for many sleep laboratories, which delay the initiation of NPPV. Also, many sleep laboratories lack experience in issues related to neuromuscular disease, because their attention is primarily on OSA rather than respiratory-muscle unloading. Given the evidence that daytime NPPV settings may not be ideal for nocturnal use in some patients, it may be prudent to consider polysomnography in patients with neuromuscular disease who remain symptomatic after acclimation to NPPV. Overnight oximetry may be useful to assess nocturnal gas exchange on NPPV, but sleep-disordered breathing may be present and sleep quality may be poor despite adequate gas exchange.

In patients with neuromuscular disease, higher NPPV settings are not necessarily better. These patients usually do not require PEEP unless they also have OSA or COPD. In patients with neuromuscular disease, higher PEEP (EPAP) can result in expiratory-muscle activation (Fig. 14).⁵⁷ Moreover, higher PEEP results in a higher

inspiratory pressure, which may decrease patient tolerance. It is noteworthy that bi-level ventilators commonly used for NPPV have a threshold PEEP (EPAP) setting of 4 cm H₂O to minimize rebreathing. This PEEP level may be unnecessary and uncomfortable for some patients with neuromuscular disease. There can also be problems from too high an inspiratory pressure, including greater leak, less comfort, ineffective inspiratory efforts, central apnea, and glottic closure.^{77–81} For most patients with neuromuscular disease and otherwise normal lung function, a PEEP (EPAP) of 4 cm H₂O, or lower if possible, and an inspiratory positive airway pressure of 12–14 cm H₂O is often sufficient. This results in a pressure support of about 10 cm H₂O. Because of the potential for ineffective triggers and central apnea, a backup rate should be set at about 12–16 breaths/min.

Summary

NPPV is commonly used in patients with respiratory failure associated with neuromuscular disease. The use of NPPV in this patient population is likely to expand, particularly with increasing evidence that this therapy is life-prolonging in patients with diseases such as amyotrophic lateral sclerosis.⁸² Appropriate selection of equipment and settings for NPPV is paramount to the success of this therapy. The selection of equipment for NPPV is based on the physiologic needs of the patient, the clinician's familiarity with NPPV, the desires of the patient, and the availability of equipment.

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Discussion

Deem: Dean, how did pressure ventilators evolve the way they did, with the single-circuit? Was it a matter of cost? Were they trying to keep things simple? Because it seems that the single-hose setup is prone to causing re-breathing.

Hess: I think that it evolved out of CPAP [continuous positive airway pressure] for sleep. I think these devices were originally designed not as ventilators the way that we use them today, but to treat OSA [obstructive sleep apnea]. Nick, can you comment on that?

Hill: Sure. The original BiPAP device was created by Respironics, and “BiPAP” is a proprietary term that belongs to Respironics. Their idea was to have a CPAP device that would be better tolerated by patients, because

you can lower the expiratory pressure below the pressure needed to stent the upper airway during inspiration. And it occurred to them along the way that they were also creating a potentially useful pressure-limited ventilator.

The first study I did on noninvasive positive-pressure ventilation in COPD patients used the prototypical BiPAP device.¹ It didn’t have a patient trigger. We also did a study² that found that it was a pressure-limited ventilator. And we also reported its use in some neuromuscular patients who had been using volume-limited ventilators and—for one reason or another—wanted something different: some because of the nuisance alarms, some because they wanted to travel, and some because carrying around a 30-pound volume-limited ventilator was a nuisance. So that’s where BiPAP came from. They were never originally designed as ventilators, and they preserved the single-limb circuit that

the CPAP devices had originated. But as long as you do relatively simple things to minimize the re-breathing, the single circuit is not a problem, and, as Dean pointed out, in that study we did looking at nasal masks,³ most of the CO₂ that otherwise would be rebreathed goes out the mouth. That’s why you don’t have re-breathing problems with nasal masks during nocturnal ventilation.

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Hess: I think the problem with re-breathing was a bigger problem when we weren't aware of it. I think now that most of us are aware of it, we make sure that it won't be a problem, by taking steps to prevent it.

King:* Dean, I think another important consideration, particularly for patients who are starting to use support more than 16–20 hours a day, is what will the device do if the power goes out? Does it have a power-loss alarm? And does it have some form of automatic internal battery that will continue to run the device?

Hess: Good point. Bob, do you want to tell our story now? This is how we approached the problem of power outage.

Brown: Dean and I look after a patient who has a neuromuscular disorder that's rendered her almost totally paralyzed. We weren't looking after her at the time that it was recommended to her that she have full-time assisted ventilation. She decided that she didn't want to have a tracheostomy tube, so her fiancé went to the Home Depot and bought some hoses and plumbing and plastic

tubing and so on, and rigged up a connection to her ventilator that's much like some of the mouthpieces that Dean showed you on some of his slides. She keeps a mouthpiece in her mouth during the day at all times. She has a family member nearby in case it falls out, which it rarely does. And she uses a face mask for BiPAP ventilation at night. She's an *amazing* woman. Last June, she and her mouthpiece and ventilator flew on a flight from Boston to Puerto Rico to visit family. Anyway, one day there was a big storm in Boston, and the power went out in her house. She had a battery, but she and her sister high-tailed it off to the local fire station, worried that the battery would run out (although they also have a bag-valve-mask) and the people in the fire station had no idea what to do with the ventilator, so they went to the Massachusetts General Hospital emergency room, where (I'm embarrassed to say this) they also didn't know what the heck to do with this portable ventilator and battery power. But they managed with a bag-valve-mask, once the battery ran out.

Hess: If I could interject: we had taught her sister how to use the bag with the mouthpiece and with a mask.

Brown: Actually, Dean taught her sister how to do it. He deserves all the credit, and so they bag-ventilated for

a long, long time beyond the fire station and to the emergency room. So bag-valve-mask is a very useful device.

Years ago I had a patient who had very severe COPD and was ventilator-dependent and lived at one of the Veterans Affairs hospitals in Boston. One day he told me how embarrassing it was for him to have to go to the bathroom sitting on the commode at the bedside, and asked if I couldn't do something to make it possible for him to go to the bathroom in the usual way. He had to use the commode because his ventilator was too big to fit through the door of the bathroom, and there wasn't a place to plug it in, and he really wanted to be able to use the regular toilet. So I taught him to disconnect himself from his ventilator and to bag-ventilate himself, using his right hand as his ventilator. He got really good at this, so that he could disconnect himself from the ventilator, hook himself up to the bag, walk to the bathroom, do his thing in the bathroom while ventilating himself, go back to bed, and re-hook himself up to the ventilator. And it goes beyond that, because he had a crush on one of the nurses in the unit, and he used to disconnect himself and go up to the nursing station to schmooze with this nurse from time to time. He was delighted with how effective the bag was for these purposes.

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