

# Full-Time Noninvasive Ventilation: Possible and Desirable

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**Introduction: History of Noninvasive Ventilation**

**Problems With Tracheostomy Ventilation**

**Techniques for Full-Time NPPV**

**Ventilatory Support**

**Cough Support**

**Data Supporting NPPV Use in Neuromuscular Disease**

**Nocturnal Ventilation**

**Continuous NPPV**

**Summary**

**Noninvasive ventilation has been available for over 100 years. In the past 25 years, relatively lightweight, portable ventilators and comfortable interfaces have become available that have allowed for full-time noninvasive ventilation. In motivated individuals who have access to centers with expertise in nocturnal and diurnal ventilation, continuous noninvasive ventilation is quite feasible. There are several reasons continuous noninvasive ventilation may be preferable to invasive tracheostomy ventilation, including the lack of need for an expensive surgical procedure, and less risk of infectious and bleeding complications. This article reviews the techniques and rationale for full-time noninvasive ventilation. Key words: noninvasive positive-pressure ventilation, neuromuscular disease, respiratory failure, glossopharyngeal breathing, insufflation, exsufflation, tracheostomy, cough augmentation. [Respir Care 2006;51(9):1005–1012. © 2006 Daedalus Enterprises]**

## **Introduction: History of Noninvasive Ventilation**

The desire to support ventilation when breathing stops has been present for centuries. At the beginning of the 19th century, Dalziel<sup>1</sup> developed the first “body ventilator,” in the form of a negative-pressure device that functioned by

expanding the rib cage, creating negative pleural and alveolar pressure and drawing air into the lungs. The subject sat in an airtight box, with the head protruding through a circular leather seal. Negative pressure was created within the box by means of a bellows. Without electrical means to drive the bellows these negative-pressure ventilators were not efficient, and it was not until the early 1900s, with the development of the “iron lung” by Drinker and Shaw<sup>2</sup> and the polio epidemics, that these devices found widespread use. The fact that these ventilators were heavy and cumbersome led to the development of devices that were portable and could be used in the home. Lightweight negative-pressure ventilators were developed, including impermeable fabric shells such as the “poncho” and “raincoat” ventilators, the cuirass or “shell” ventilator, and even a lighter-weight version of the iron lung.<sup>3</sup>

Although negative-pressure ventilators are still occasionally used, their utility is limited by the fact that their use during sleep can be associated with obstructive sleep

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apnea due to asynchrony between diaphragm and upper-airway muscle contraction.<sup>4-6</sup>

In addition to negative-pressure devices, other types of body ventilators were also developed, including the rocking bed<sup>4</sup> and the pneumobelt.<sup>7</sup> The motorized rocking bed found wide use during the polio epidemics of the mid-20th century. It functions by moving the supine patient bed through an arc, in the rostral-caudal direction, with the point of rotation centered at the hips. With a rotational arc of 40°, gravity moves the abdominal contents and the diaphragm up and down, thus inflating and deflating the lungs, resulting in ventilation.

The pneumobelt also acts by displacing the abdominal contents and diaphragm. It has an inflatable rubber bladder inside of a canvas belt, which is periodically inflated by an attached positive-pressure generator. The belt is placed snugly around the anterior abdomen, and inflation of the bladder causes upward displacement of the abdominal contents and diaphragm, causing exhalation of gas from the lungs. Inhalation is accomplished passively, through the action of gravity, which causes the abdominal contents and diaphragm to drop, resulting in movement of air into the lungs. Neither the rocking bed nor pneumobelt is currently widely used, although they can be effective in performing ventilation.

Although noninvasive methods were used primarily during the majority of the polio epidemics of the 20th century, a particularly bad outbreak in Denmark in 1953 quickly outran the supply of negative-pressure ventilators. When the negative-pressure devices ran out, patients were then intubated and ventilated with anesthesia machines, run by hand, by medical staff.<sup>8</sup> The outcomes for these patients were better than those previously reported, and from that point on, invasive (endotracheal) intubation became the method of choice for acute polio. Fortunately, the advent of the polio vaccine shortly thereafter dramatically reduced the number of cases of acute poliomyelitis. Although some patients with chronic sequelae of polio were treated with body ventilators at home or in institutions in the 1960s and 1970s, it was not until the early 1980s that noninvasive positive-pressure ventilation (NPPV) experienced a renaissance, with the advent of positive-pressure ventilation via face mask.

NPPV had been used for centuries in the resuscitation of victims of drowning and fires. In 1530, Paracelsus described the use of a fireplace bellows to ventilate, via the mouth, patients who had drowned. This technique was used intermittently in Europe for centuries, and pneumatic mechanical resuscitators were used by fire departments in the United States around the turn of the 20th century.<sup>9</sup> Positive-pressure ventilation was used in conjunction with body ventilators during the polio epidemics. When a patient needed to be removed from an iron lung, the patient's head would be placed in a clear plastic dome, to which

Table 1. Potential Disadvantages of Long-Term Tracheostomy

Expense of procedure
Higher risk of respiratory infection
Formation of granulation tissue
Airway stenosis/malacia
Tracheoinnominate-artery fistula
Tracheoesophageal fistula
Impairs speech and swallowing
Inability to stack breaths for cough augmentation
May require skilled assistance for suctioning
Social issues around stoma and tracheostomy tube

cyclic positive pressure was applied, resulting in ventilation.<sup>10</sup>

Ventilation through a mouthpiece rather than a face mask or chamber surrounding the head was first considered by Affeldt in 1953.<sup>10</sup> This was used with good success with patients with neuromuscular disease at some rehabilitation facilities, such as Ranchos los Amigos in California and the Goldwater Rehabilitation Center in New York. Patients used the mouthpiece during the day, but many of them also used it at night, even while asleep. This mouthpiece ventilation system has been used successfully to this day and is an important part of a program of full-time NPPV.<sup>11</sup>

Although for decades positive-pressure ventilation for resuscitation was delivered via mask, it was in the mid-1980s that nasal and face-mask ventilation became possible for longer-term management, with the development of commercial masks for continuous-positive-airway-pressure therapy. In addition, in 1990, bi-level pressure-support devices were developed, which allowed portable, easily managed home ventilation. These devices are pressure-regulated, which (very importantly) allows compensation for the leaks that almost invariably occur with nasal and oronasal masks. These devices have found widespread use in both acute and chronic situations for patients with neuromuscular disease and primary pulmonary diseases. In particular, there has been dramatic growth in the nocturnal use of these devices to treat the sleep-disordered breathing that is prevalent in patients with neuromuscular and chest-wall diseases.

### Problems With Tracheostomy Ventilation

What are the reasons that one might consider 24-hour NPPV for an individual who needs full-time ventilatory support? Although tracheostomy is critical for survival in patients with glottic dysfunction and respiratory failure (who are at high risk for aspiration), for those with ventilatory failure but without glottic dysfunction, tracheostomy ventilation has several drawbacks that can be avoided with NPPV (Table 1). These complications were detailed

Table 2. Indications for Tracheostomy in Neuromuscular Disease

Substantial glottic dysfunction
Increased risk of aspiration
Inability to clear secretions, despite cough augmentation
Inability to generate adequate cough flow, despite aggressive cough assistance
Recurrent pneumonia on full-time NPPV with adequate cough assist
Substantially elevated $P_{aCO_2}$ , despite optimal full-time NPPV
Patient preference
Lack of experienced health-care providers in continuous NPPV

NPPV = noninvasive positive-pressure ventilation

in an excellent review by Epstein and have been reported to occur in up to 65% of patients with long-term tracheostomy.<sup>12</sup>

The placement of an opening and indwelling tube in the trachea has a number of effects, many of which relate to the mechanical and physiological effects of a foreign body within the airway. Pressure on the wall of the trachea can ulcerate and scar the tracheal wall, which can lead to tracheal stenosis. This can lead to dyspnea and increased ventilator dependence, and it may be very difficult to treat. If the cartilage is damaged, a loss of structural rigidity and an increase in the collapsibility of the airway, known as tracheomalacia, can occur.<sup>13</sup>

One of the most common complications of tracheostomy is a higher rate of respiratory-tract infection and pneumonia. Ibrahim et al prospectively studied 3,000 patients in medical and surgical intensive care units in a nonteaching hospital.<sup>14</sup> There was a > 6-fold higher relative risk of ventilator-associated pneumonia in patients with tracheostomy, which appeared to be related to colonization of bacteria in the trachea and on the tracheostomy tube. The latter occurs because the plastic tracheostomy tube is an ideal site for biofilm formation and colonization with bacteria, and because the tube is associated with a decrease in function of the mucociliary escalator, which compromises the normal clearance of bacteria and other foreign material from the airway.<sup>15</sup> Also, with the cuff inflated, the tracheostomy tube has been reported to impair swallowing and increase the risk of aspiration.<sup>16</sup> Tracheostomy can also cause other mechanical problems, including erosion into the esophagus or the innominate artery, which can be disastrous.<sup>17</sup>

There are instances when tracheostomy ventilation should be considered (Table 2). These relate predominantly to patient characteristics, although lack of provider expertise with NPPV can be a factor as well. Impairment of glottic function with inability to protect the airway is a strong contraindication for NPPV and indication for consideration of tracheostomy ventilation. Tracheostomy does not prevent all aspiration events, but it probably prevents

Table 3. Forms of Noninvasive Ventilation

Mouthpiece ventilation
Nasal or full-face-mask ventilation
Negative-pressure ventilation
Cuirass
Tank ventilator
Suit ventilator
Pneumobelt ventilation
Rocking bed ventilation
Glossopharyngeal breathing

large-bolus aspiration events. Bach has suggested that tracheostomy ventilation is warranted only when assisted-cough maneuvers cannot achieve a peak cough flow of  $\geq 270$  L/min while the patient is well or  $\geq 160$  L/min during illness; below 160 L/min, secretion removal is inadequate.<sup>10</sup>

Lack of health-care providers experienced in or willing to institute a program of full-time NPPV may be one of the most common reasons full-time NPPV is not used. No data are available to address this question, but it is clear from surveys of Muscular Dystrophy Association clinics that these techniques are not routinely offered to patients with chronic neuromuscular disease.<sup>18,19</sup>

It should be noted that a consensus statement by the American College of Chest Physicians suggested that NPPV for  $\geq 20$  h/d is a relative indication for tracheostomy ventilation.<sup>20</sup> No reason for that suggestion was given in the consensus statement, although it has been suggested that mechanical ventilation might be more “securely” provided by tracheostomy ventilation (Edward A Oppenheimer MD, Pulmonary and Critical Care Medicine Division, Kaiser Permanente Medical Center, Los Angeles, California, personal communication, 2004).

### Techniques for Full-Time NPPV

Several technologies are available for full-time NPPV (Table 3). Many of these were developed during the polio epidemics of the mid-20th century and were used extensively during that period. Most of these technologies are still available, although body ventilators are infrequently used today.

### Ventilatory Support

Currently, the primary method for full-time NPPV is a combination of mouthpiece ventilation during the day and nasal-mask ventilation at night. However, it is possible to use either mask ventilation or mouthpiece ventilation continuously. Mouthpiece ventilation (Fig. 1) has been available for about 50 years.<sup>21</sup> It involves the use of a portable (usually volume-limited) ventilator set in the assist-control



Fig. 1. Mouthpiece ventilation. Left: Patient with mouthpiece and ventilator circuit in standard position. Right: Rear view of wheelchair with ventilator and mouthpiece circuitry in place.

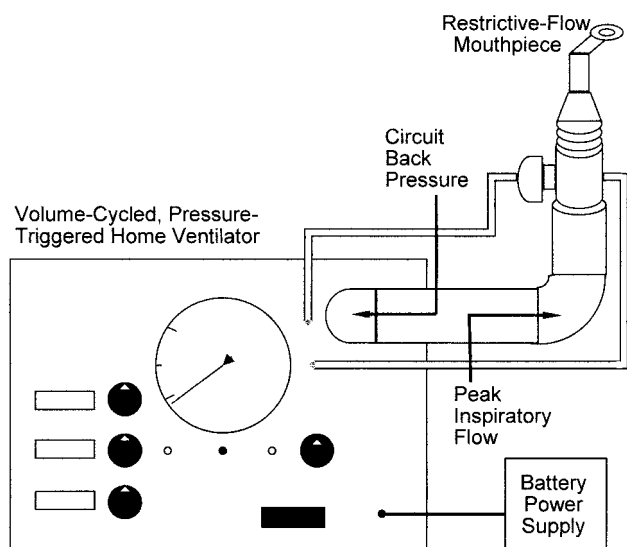


Fig. 2. Schematic of mouthpiece ventilation setup.

mode. The ventilator is attached to a standard ventilator circuit (Fig. 2), at the distal end of which is a mouthpiece that has either a narrow bore or an angled bend that allows back-pressure in the circuit to prevent low-pressure alarming. The circuit is open, but the pressure in the circuit prevents low-pressure alarm. The respiratory rate is generally set on the lowest possible setting, so the patient can take breaths as needed without the machine triggering 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 to leak around the lips, so the tidal volume is set higher than what would be set for tracheostomy ventilation. Tidal volume is set per the patient’s comfort; the range is generally 700–1,200 mL.

It is possible to use nasal-mask ventilation during the day with patients who cannot tolerate mouthpiece ventilation or chose not to use it. Patients with amyotrophic



Fig. 3. Setup for noninvasive positive-pressure ventilation with a nasal mask, for a patient with amyotrophic lateral sclerosis. Left: Front view. Right: Rear view, showing noise-dampening box that contains the bi-level ventilator and auxiliary battery.

lateral sclerosis, in particular, often have weakness of the lips, which prevents them from using mouthpiece ventilation. In my practice I have seen many patients with amyotrophic lateral sclerosis who have used nasal mask NPPV for the majority of the day and night and were able to set up bi-level devices on their wheelchairs (Fig. 3).

Nocturnal ventilation is most commonly accomplished with nasal mask, full face mask, or oronasal interface. These interfaces can be used with bi-level pressure ventilators or volume ventilators. Bi-level ventilators have the advantage that they can compensate for the leaks that almost invariably occur, at least intermittently, at night. Volume ventilators have the advantage that they can deliver higher pressure for those with poorly compliant respiratory systems. In general, volume ventilators also have trigger mechanisms that can be more easily adjusted. It is also possible to ventilate with a mouthpiece at night.<sup>22</sup> The mouthpiece can be held in place with a lip seal and head strap, though this may not be necessary, as some patients appear to hold the mouthpiece in without other support.

Glossopharyngeal breathing (also known as “frog breathing”) is an adjunctive therapy for full-time NPPV. It is a method whereby the patients can breathe for him or herself, even with minimal or no respiratory muscle function.<sup>23,24</sup> Some patients have been able to support themselves during the daytime entirely with glossopharyngeal breathing. Some have used it in conjunction with other ventilatory support methods or as an emergency, backup method, in case of failure of the ventilator or the power supply.

### Cough Support

Support of ventilation is only one part of the process of full-time NPPV. Therapeutic interventions must also be



Table 4. Methods of Secretion Management

Cough augmentation
Manually assisted cough ("quad" cough)
Breath-stacking
Resuscitator bag
Glossopharyngeal breathing
With mouthpiece ventilator
Mechanical insufflation-exsufflation
Secretion mobilization
Chest-wall oscillation therapy
Intrapulmonary percussive ventilation

able to support cough and airway clearance. This is as important as, if not more important than, ventilatory support, because pneumonia is one of the leading causes of morbidity and mortality in patients with neuromuscular respiratory disease.

Cough depends on an intact neural and neuromuscular reflex arc. Stimulation of cough receptors in the respiratory system activates brainstem centers that lead to activation of a coordinated contraction of expiratory muscles of the abdomen and chest wall, with the glottis closed.<sup>25</sup> Gas is compressed and high pressure builds within the airways. The glottis then rapidly opens and an explosive release of gas occurs from the airways, past the vocal cords. High-velocity gas flow causes shearing of secretions away from the airway walls and propels secretions out of the lungs. This is essential for airway hygiene. Fortunately, when cough function is impaired, there are several ways to augment cough function that are completely adequate to maintain pulmonary hygiene without the use of a tracheostomy (Table 4).

Manually assisted cough is a method of applying a positive pressure to the abdomen, pleural space, and airway, and to provide an adequate cough flow. Several techniques are available by which an attendant can apply rapid abdominal thrusts, resulting in effective secretion clearance.<sup>26</sup> The patient can assist the attendant by taking a maximal inspiration and then breath-stacking prior to application of the abdominal thrust. This maximal achievable inspired volume has been labeled the "maximal insufflation capacity."<sup>27,28</sup> Increasing the volume of air in the respiratory system prior to the assisted breath increases the volume of gas in the lungs for exhalation and also increases the inward elastic recoil pressure of the lung and chest wall, which can increase the exhalatory pressure. The combination of the 2 methods increases peak cough flow.<sup>29</sup> Glossopharyngeal breathing can be used to augment the maximal inspiration in patients who cannot generate an adequate inspiratory effort.<sup>22</sup>

Mechanical insufflation-exsufflation is a very effective secretion-management technique that has been known for over 50 years but has only recently been used to any great extent. The device currently available to deliver mechan-



Fig. 4. CoughAssist In-Exsufflator, used with assistant's help.

ical in-exsufflation is the CoughAssist In-Exsufflator (JH Emerson, Cambridge, Massachusetts) (Fig. 4), the electric motor in which can generate positive and negative pressures of up to 50 cm H<sub>2</sub>O delivered to the airway of a patient who is unable to cough. The pressure is applied via face mask, connected to the device via flexible pressure tubing. Gas is insufflated into the respiratory system over a 1–3-second period, by exerting a positive pressure of 30–50 cm H<sub>2</sub>O. Then the device rapidly reverses the flow and generates a negative pressure of –30 to –50 cm H<sub>2</sub>O. Secretions are thus noninvasively suctioned from the airway.

Other noninvasive mechanical aids include devices that oscillate the chest wall or airway.<sup>30</sup> These have not been well studied in neuromuscular-disease in patients without tracheostomy, and their role in secretion management in these disorders is unclear.

The decision as to when to initiate cough assistance is determined by measurement of peak cough flow, which is easily accomplished with an asthma peak-flow meter and a mouthpiece or face mask (Fig. 5). A value of < 270 L/min while the patient is well is thought to be insufficient for secretion clearance.<sup>31</sup> Cough augmentation, using one or more of the above techniques, is recommended when cough peak flow falls below 270 L/min.

### Data Supporting NPPV Use in Neuromuscular Disease

#### Nocturnal Ventilation

The use of nocturnal ventilation in patients with neuromuscular disease has several benefits, including reduced

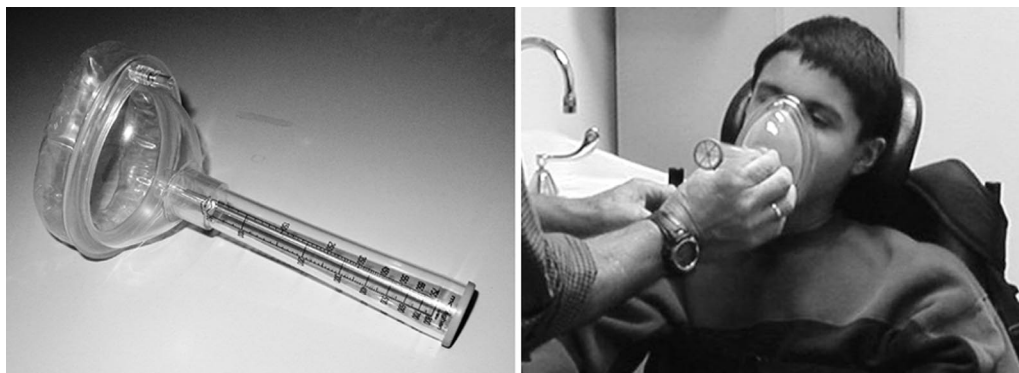


Fig. 5. Left: Cough-peak-flow meter, consisting of an asthma peak-flow meter and a face mask. B: Cough-peak-flow measurement being made with a patient.

$P_{aCO_2}$  and increased  $P_{aO_2}$  (on and off ventilator), decreased symptoms of respiratory failure, improved quality of life, and reduced morbidity and mortality. It has been clear for some time that intermittent ventilation may ameliorate symptoms of respiratory failure, reduce  $P_{aCO_2}$ , increase  $P_{aO_2}$  (even during periods off the ventilator), and prolong survival in patients with neuromuscular disease.

Nocturnal ventilation has become a widely accepted clinical practice in providing ventilatory assistance for patients while sleeping and allowing them to breathe on their own during the day. Curran reported on the initial use of nocturnal negative-pressure ventilation in patients with late-stage Duchenne muscular dystrophy.<sup>32</sup> In patients who had symptoms of ventilatory failure and  $P_{aCO_2} \geq 60$  mm Hg, nocturnal negative-pressure ventilation with a cuirass or tank ventilator significantly improved  $P_{aCO_2}$  (60.8 mm Hg before treatment vs 45.5 mm Hg after treatment) and  $P_{aO_2}$  (59.3 mm Hg before treatment vs 74.6 mm Hg after treatment). A number of other studies on nocturnal ventilation have supported those findings.<sup>33–40</sup>

The mechanism whereby intermittent nocturnal ventilation ameliorates respiratory failure has not been entirely elucidated but is probably multifactorial. During periods of mechanical ventilation there is a substantial reduction of diaphragm and accessory-muscle electromyographic activity,<sup>41,42</sup> which probably signifies a decrease in work performed and oxygen consumed by the respiratory muscles. Some authors have therefore postulated that nighttime ventilation rests fatigued respiratory muscles, allowing improved daytime functioning.<sup>43</sup> The rest provided by this reduction in work load may reverse the chronic respiratory-muscle fatigue that is thought to be present in these patients, allowing improved daytime functioning. In one study, daytime inspiratory-muscle *endurance* had increased from  $7.1 \pm 3.4$  min to  $14.8 \pm 7.6$  min 3 months after the initiation of nighttime ventilation.<sup>44</sup> The results of studies on improvement in muscle strength have been mixed, with some authors noting slight improvements and others finding no improvement.<sup>34,42</sup>

A second hypothesis for the improved daytime respiratory function associated with nocturnal ventilation is related to reversing the adverse effects of chronic neuromuscular disease on respiratory-system mechanics.<sup>45</sup> Improvements in lung compliance, increases in resting lung volume, and a decrease in the work of breathing have been reported<sup>42,45</sup> in patients with neuromuscular disease following positive-pressure ventilation. If these improvements are sustained throughout the day, they would constitute a reduced load on the respiratory muscles, which would ameliorate chronic fatigue. In addition, increases in end-expiratory lung volume to a more normal range would reduce atelectasis and improve oxygenation.

A third explanation involves the reversal of what has been referred to as “central fatigue,”<sup>46</sup> in which nighttime hypoventilation and hypoxemia are thought to lead to a blunting of central respiratory drive, resulting in “adaptive” daytime hypoventilation. It has been postulated that nighttime ventilatory intervention results in a “resetting” of central control mechanisms, with an increase in chemosensitivity and a reduction of the body bicarbonate pool.<sup>44</sup> Increase in  $P_{aCO_2}$  would be met with a more appropriate response in minute ventilation. Data to support this hypothesis are not currently available.

It is possible that all three of the above-described mechanisms are involved in the improvement of arterial blood gas values and daytime functioning in patients treated with nocturnal ventilation.

In addition to improvement in arterial blood gases, other measures of physiologic function improve with intermittent ventilation. Hoeppner and colleagues<sup>45</sup> found an increase in vital capacity, reduction in erythrocytosis, and improvement in right-sided heart failure following nighttime ventilation; the changes were maintained over a mean follow-up period of 3.4 years.

For ethical reasons, it is not possible to perform a randomized controlled trial of the effect of mechanical ventilation on survival in patients with neuromuscular disease. It is clear that, in most progressive neuromuscular dis-

eases, once an elevation in  $P_{aCO_2}$  and a decrease in  $P_{aO_2}$  are noted, cor pulmonale and death are inevitable within a short period. It is therefore accepted by most authors that mechanical ventilation in the home leads to improved survival in most patients.

### Continuous NPPV

As with nocturnal NPPV for neuromuscular disease, there have been no randomized controlled trials of continuous ventilation. In one cohort study, Bach et al compared 24 patients who used NPPV (14 of whom used the Bach et al protocol for continuous NPPV) to 22 patients who did not use the protocol because they failed to return for follow-up or were already tracheostomized and could not be converted to continuous NPPV.<sup>47</sup> When comparing the tracheostomized individuals to those who used continuous NPPV, there were significant differences in hospital days per year per patient ( $2.3 \pm 2.4$  d/patient vs  $0.3 \pm 2.4$  d/patient, respectively,  $p < 0.04$ ) and hospitalizations/year/patient ( $0.3 \pm 0.4$  hospitalizations/year/patient vs  $0.1 \pm 0.4$  hospitalizations/year/patient, respectively).

In a retrospective study by Gomez-Merino and Bach, Duchenne muscular dystrophy patients who used continuous NPPV and a secretion-management protocol at home were compared to a group who did not have access to the protocol and were either tracheostomized or used NPPV nearly continuously.<sup>48</sup> The protocol consisted of using breath-stacking, mouthpiece positive-pressure ventilation, and mechanical in-exsufflation to maintain an oxyhemoglobin saturation of  $> 94\%$  (as measured with a home oximeter). Among the 34 full-time NPPV users with access to the protocol there were 3 deaths, all due to heart failure. Among the 31 patients without access to the protocol there were 27 deaths: 20 due to respiratory failure and 7 due to cardiac failure. Further studies of continuous NPPV are needed to assess whether this treatment is truly better than tracheostomy ventilation. A recent American Thoracic Society consensus statement on the respiratory care of Duchenne muscular dystrophy suggested that this method be considered when expertise is available for initiation of appropriate protocols.<sup>49</sup>

### Summary

Full-time NPPV is quite possible for selected individuals with neuromuscular respiratory failure, but the patient must be motivated and have relatively intact glottic function. The techniques are relatively easy to learn and have been available for decades. Full-time NPPV can be very rewarding for patients and providers alike.

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## Discussion

**Hess:** I would point out that you can do mouthpiece ventilation with a portable pressure BiPAP [bi-level positive airway pressure] ventilator as well as a volume ventilator. In fact, the patients that Bob Brown and I talked about yesterday are on BiPAP machines with mouthpiece, and use a nasal mask at night. One of our patients has gone to Puerto Rico several times with her BiPAP machine, using it on the airplane. Now the down side to that approach is that, although those devices are small and very portable, it's very difficult to find a suitable battery to power them. That's one of the things I've struggled with, and several patients have jury-rigged the battery supply, which ends up, at least in our experience, being larger than the BiPAP

machine on the back of the wheelchair.

**Benditt:** Yes, you can use a pressure-support machine for mouthpiece ventilation, but you can't do breath-stacking with pressure support.

**Hess:** Can't do it at all!

**Benditt:** Right, but I think one of the major benefits of the mouthpiece with a volume ventilator is that you can do breath-stacking, which makes the cough independent. So we've pretty much used volume ventilators. Now, this does bring up the issue of whether the insurer has to supply one ventilator for the wheelchair and one for the bed stand at night. In the United States, most people would be using a pressure ventilator at night and a volume ventilator during the daytime. We've written a lot of let-

ters of medical necessity to get 2 ventilators, but some insurers balk at that.

**Hill:** I think you are facing a situation for which there are many different solutions that are limited only by your imagination. The technology is out there to place a pressure-limited portable ventilator on the back of the wheelchair and connect it to a nasal mask, or to use a volume-limited ventilator with a mouthpiece.

In response to Dean's point about air travel, for the 1998 Lyon conference on Home Mechanical Ventilation, one of my patients flew over (I was in the jet with her). She had a BiPAP device powered by a marine battery. That was before 9/11 [September 11, 2001]. They asked the pilot if it was OK if she brought her battery along with her ventilator, and he said, "Yes. No problem." She



used the BiPAP for much of the flight. I guess after 9/11, it could be more of a problem.

**Hess:** We've had at least one patient I know of who within the last year was still traveling that way.

**Hill:** The size of the battery you can bring on board might be an issue after 9/11, though.

**Benditt:** I want to make a strong plug for the respiratory therapist in this matter. In terms of equipment, the therapist might be more creative than the physician. "Creativity" is the watchword. I am trying to disseminate this idea and to increase the popularity of these creative techniques through the therapists, rather than the physicians, because, as you know, you can lecture to people about neuromuscular disease and what it is, but you'll still see that they come to the emergency department and they get oxygen. Few people think about supporting ventilation until it's too late, and then they're intubated. But the therapists, who deal with these technologies all the time, are much more reasonable about it. I wish Louie Boitano were here, because he's pushed this a lot, and he comes up with all kinds of ideas about how to maneuver here.

I'm going to talk to Sam [Sam P. Giordano MBA RRT FAARC, Executive Director, American Association for Respiratory Care] about this, to try to get the Association to push for the care of the neuromuscular patient not so much from the physician but from the health-care team, with the respiratory therapist as a lead. It's such a natural. During the time of polio there was a lot more familiarity with everything we're talking about, and it's unfortunate that that familiarity was lost.

**Hill:** You're absolutely right. However, some therapists today don't have much knowledge in this area, so the home-equipment provider that you deal with has a lot to do with what

kind of technology you can put together. When I've ordered trays for ventilators on the backs of wheelchairs, it seems that I go through a lot of reinventing the wheel, and I have to get people in touch with therapists who know how to do it. The same applies to the CoughAssist In-Exsufflator; a lot of the therapists are not skilled with that device and don't know how to teach the technique very effectively. So you need to get therapists involved who do know.

**Panitch:** Families also add to this creative movement. We recently added mouthpiece ventilation for a young man who was requiring longer periods of support while he was awake. I wasn't familiar with the setup you showed, with the gooseneck holder for the mouthpiece. His family bought a microphone stand and bolted it to his wheelchair, and that created the holder for his mouthpiece.

**Giordano:\*** I first want to comment on traveling with oxygen and CPAP [continuous positive airway pressure] devices. We, the AARC [American Association for Respiratory Care], along with ATS [American Thoracic Society], ACCP [American College of Chest Physicians], and several patient groups, have been working with the Department of Transportation to get these devices designated as "personal medical equipment devices," which differ from nonessential electronic devices such as laptop computers in that designated medical devices do not have to be turned off during takeoff or landing. There's been quite a bit of "to and fro" about the power sources for these devices. That's been the big issue, especially from the patient groups, and I think it's a "chicken-and-egg" situation, because if we're successful in getting a rule change and

not abandoning the traditional oxygen-supply issues, then there'll be more investment in the technology, just as there was investment in the laptops, and with that investment will come better batteries.

So I think we're at step number 2, and we probably need to get to step number 150 on this project. Naturally, it's not going to be a walk in the park, because, with the Air Transport Association of America, which has constituents in the major airlines, after 9-11—even before 9-11—it's been difficult to get change. However, it's not actually been as bad as we thought it would be, mostly because of new electronic aspects, such as the new portable oxygen concentrators. Traditional delivery devices, however, are a different story and will require more than a rule change—perhaps legislation. We are looking for scientific evidence that proves the safety of portable liquid-oxygen systems. Fortunately, our friends in the Air Force have a lot of experience using liquid-oxygen systems on the C130 planes that they use to transport our wounded. We've had some discussion with them about this issue.

With regard to plugging the therapist into the process, it's not unlike a discharge-planning situation, where if you as physicians could order that these patients be assessed by respiratory therapists, in anticipation of their limitations at home, then at least the therapists might be able to get the DME [durable medical equipment] people in the hospital and have that discussion before you discharge the patient. I'm assuming this will be a post-acute-care situation. Perhaps the therapists might save you the time and grief of cobbling together all those people, by leveraging the respiratory therapist's ability to innovate.

**Rajiv Dhand:** Great presentation, Josh. I just wanted to clarify one comment that you made. You really pointed out the problem with the tracheostomy and the end-of-life issues

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that come up, because, like I said yesterday, the choice then is whether to do a tracheostomy or to do nothing. My submission to you yesterday was that we are presenting the tracheostomy in a very negative light, based on data that are actually quite old. There's a study in which they compared the complication rate of tracheostomies before and after 1986. I think their end-year was 1997.<sup>1</sup> They found that in the latter phase of the study, the complication rates from tracheostomy were much lower than what had been seen before 1986.

So what I'm trying to say is that there is a huge physician bias; you know, that first of all, the physicians present the tracheostomy in a very negative light; the patients don't really know how to interpret the information that is available, and so there is a strong negative bias toward getting a tracheostomy, which I feel is probably not well founded. Because the information is really not of good quality, and if we really need to do good service to our patients, then either (1) we should get better data or (2) we should not present it in a negative light. You know, we should say, "Well, it's your preference. These are the complications that we know are associated with this procedure, but it's up to you to decide whether you want to go ahead." And very often, what I've seen is physicians actually tell the patient, "You're not going to have good quality of life with this; there's going to be this and that complication." Therefore the family and the patients shy away from getting the procedure.

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**Benditt:** I completely agree with you that there is something about tracheostomy that is different and brings up a lot of issues, and I think part of it is that we're presenting an overly nega-

tive impression about tracheostomy complications. But there is something more in the patient's mind, and it has to do with artificial life support and that this is a technologically-driven intervention to prolong life.

It's funny; this just never comes up with noninvasive ventilation, other than that sometimes in the intensive care unit there's a debate about palliative care and noninvasive ventilation. But this is quite different, and it may be because with noninvasive ventilation you can decide you don't want to use it and just stop using it. I don't know. But with regard to mouthpiece ventilation, you never hear patients say, "Well, I'm not going to do that; that's going to prolong my life." Nobody talks about mouthpiece ventilation that way, but about tracheostomy I hear things like, "Oh, my God! This is a major decision point, where it's artificial life support versus not." But I would say they're very similar. Why is there that bias? It can't all be because tracheostomy is a procedure and creates a wound in the neck. It's gotta be something more than that. I don't think we know what it is.

Following up on Sam's comment, I know legislation has been proposed to get respiratory care reimbursed in the home setting. There's a lot of trouble about this now, because the DME providers, who have been providing some respiratory care, are under pressure now, but I would like to see respiratory therapists get reimbursed for work in the home, because a lot of this occurs in the home, not in the hospital. And our goal is to avoid hospitalizations.

**Hill:** And now it's even worse, because with this capped rental legislation that's just been passed, the DME companies are under even greater pressure to cover their respiratory-therapy cost. It could spell financial disaster for them and health-care disaster for patients. Because to manage things like this you need skilled respiratory therapists working for DME companies,

and if they're not compensated, they won't be there. I don't know how I would manage a lot of these patients in-home without their input.

**Jubran:** Josh, do any data indicate that noninvasive ventilation improves respiratory-muscle function in patients with neuromuscular disease?

**Benditt:** I don't think there's been any evidence that it improves respiratory muscle function in patients with Duchenne muscular dystrophy. Some people have suggested that respiratory-system compliance may be improved by repeated hyperinflations, but that has not been established. Some say that with ALS patients, instituting noninvasive ventilation earlier may prevent decline, or change the slope of the curve, but I think that's also debated by many people. So the answer is no; there are no substantial data that it changes muscle function.

**Jubran:** So the improvement in  $P_{CO_2}$  is not a function of respiratory muscles, but a controller function?

**Benditt:** Absolutely. I'll let Nick answer.

**Hill:** I've spent a lot of time scouring the literature and thinking about this issue, and the only evidence that I think has any validity is from the study by Goldstein et al,<sup>1</sup> who found that the maximum sustainable minute volume went up a little bit following initiation of noninvasive ventilation in a mixed group of patients with neuromuscular disease and chest-wall deformities. And there are a few studies that have initiated noninvasive ventilation and shown some improvements in maximal inspiratory pressure, but those data are very inconsistent, and one problem in interpreting these studies is that the  $P_{CO_2}$  values drop, and some groups have found that severe hypercarbia impairs respiratory-muscle function.

So are the muscles getting better because of the muscle-resting effect of the noninvasive ventilation, or is it just because of the improved gas exchange? Also, many of these patients have acute illnesses that they're recovering from when they start noninvasive ventilation, so their infections clear, their nutritional status improves, and other aspects get better, so respiratory-muscle function might improve for reasons besides the noninvasive ventilation.

One of the studies I did during the 1980s looked at indices of respiratory-muscle performance, mainly vital capacity and the like.<sup>2</sup> We showed that over time, even though  $P_{CO_2}$  improved and then remained stable, the forced vital capacity progressively declined. I think that's a pretty clear indication that respiratory-muscle strength isn't as important as resetting the  $CO_2$  stat in maintenance of the improved gas exchange in these patients.

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**Hess:** Back to the tracheostomy issue. An area where I think we fumble a bit from time to time is in transitioning patients from 24-hour noninvasive ventilation to tracheostomy. That requires a lot of planning by a lot of people, to get the patient transitioned from mask ventilation to the tracheostomy—all the teaching that's needed about the tracheostomy and ventilator and in-exsufflation is something that we haven't always done very well.

**Mehta:** Have you had any experience with ongoing noninvasive venti-

lation with a mini-tracheostomy for secretion clearance?

**Benditt:** I have no experience with the mini-tracheostomy and noninvasive ventilation.

**Hill:** There is interest in that approach. I'm aware of one industry group that is trying to develop a mini-tracheostomy technology that would hook up to a small portable ventilator and could be used in COPD patients.

I'd like to get back to the issue of when you do a tracheostomy, and tracheostomy versus noninvasive ventilation. I'll counter a little bit of what you were saying, Rajiv. I don't believe that mouthpiece ventilation and tracheostomy ventilation are the same thing. On average, if you can manage someone noninvasively, you're better off. It's easier to manage these patients; they have fewer infections; they stay out of the hospital; and there's less stress on caregivers. Once you go to a tracheostomy, a lot more care is necessary, and often patients who previously were able to stay at home with noninvasive ventilation end up in the hospital. They can't go home because you can't provide them with enough support. The level of technological complexity is just greater. I have a lot of experience on both sides. Now, there are people who are simply not good candidates for long-term noninvasive ventilation, often because of glottic dysfunction or personal preference, so I certainly manage patients using tracheostomies, and I don't try to talk people out of having them, but I think the realistic view is that they're harder to manage and you run into more complications, mainly of an infectious nature.

Now, on the issue of our Consensus Conference,<sup>1</sup> you pointed out that the need for more than 20 hours was considered an indication for tracheostomy placement. I was there, and I think it was a suggestion rather than a recommendation. When someone needs

round-the-clock noninvasive ventilation, there are situations where there may be advantages to having a tracheostomy, so it was a suggestion to think about it. You also had a slide that said that when  $P_{aCO_2}$  is greater than 50 mm Hg on noninvasive ventilation, consider a tracheostomy. But I have patients whose  $P_{aCO_2}$  is in the 60s—up to 70 mm Hg in one case—and managed successfully with long-term noninvasive ventilation. So where do you think the  $P_{aCO_2}$  should be?

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**Benditt:** I took 50 mm Hg from the consensus statement. Some people use 45 mm Hg, some use 50 mm Hg. I agree that some people do just great with a higher  $P_{CO_2}$ . But I always worry about your other field of investigation, which is the pulmonary artery, and how it responds to  $CO_2$  continuously.

**Hill:** Not a problem.

**Benditt:** In the range of 50 mm Hg to 60 mm Hg, my personal practice, which is not really evidence-based, is to try to keep them at least below 55 mm Hg, and preferably below 50 mm Hg, but I have patients who are at 58 mm Hg or 60 mm Hg, and they feel fine. Their sleep studies and overnight oximetry look good without desaturation. They're using the mouthpiece during the day. I've thought, "Wow! Should I put these patients on a respiratory stimulant?" And then I think, "Hmmm. They're going to sense more dyspnea; probably not." So I don't do that. And they do great. So I think you're correct that maybe we're kind of treating our own anxiety rather than the patient. But there you have it.