Amyotrophic Lateral Sclerosis*

Prolongation of Life by Noninvasive Respiratory Aids

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Study objective: To describe prolongation of survival in patients with amyotrophic lateral sclerosis (ALS) by continuous noninvasive intermittent positive-pressure ventilation (NPPV) and mechanically assisted coughing (MAC) using oximetry as feedback.

Setting: A retrospective review of ALS patients visiting one center from 1990 to 2000.

Design: Patients were trained in mouthpiece and nasal NPPV when symptomatic for hypoventilation, and trained in MAC with oximetry feedback when assisted peak cough flow (PCF) levels decreased to < 270 L/min. Survival was considered to be prolonged when full-time NPPV was required with limited ventilator-free breathing tolerance.

Results: Of 101 patients who met the criteria for access to NPPV and MAC, 15 have not yet used them, and 11 patients with severe bulbar muscle dysfunction died without ever successfully using them. Three patients used NPPV full-time, and oximetry and MAC episodically, but did not yet require ongoing NPPV. Eighteen used NPPV part-time for a mean (± SD) duration of 3.8 ± 4.1 months. Nineteen others underwent tracheotomy after 4.7 ± 4.5 months of receiving part-time NPPV. Sixteen patients used part-time NPPV for 17.5 ± 13.0 months (maximum, 25 months), then full-time NPPV for 14.1 ± 12.6 months (maximum, 40 months) before undergoing tracheotomy. Nineteen patients used part-time and full-time NPPV for 25.2 ± 19.8 months (maximum, 114 months) and 17.5 ± 13.3 months (maximum, 87 months), respectively, without undergoing tracheotomy. Ten of these NPPV users died once bulbar dysfunction became severe.

Conclusion: We conclude that up to continuous use of NPPV, along with MAC when needed, can permit prolonged survival and delay the need for tracheotomy for a significant minority of ALS patients by > 1 year.

Key words: amyotrophic lateral sclerosis; exsufflation; noninvasive mechanical ventilation; survival; tracheostomy

Abbreviations: ALS = amyotrophic lateral sclerosis; MAC = mechanically assisted coughing; MIC = maximum insufflation capacity; NPPV = noninvasive intermittent positive pressure ventilation; PCF = peak cough flow; SpO₂ = pulse oximetric saturation; TPPV = tracheostomy intermittent positive pressure ventilation; VC = vital capacity

The mean length of survival from the onset of amyotrophic lateral sclerosis (ALS) has been reported as 2.4 to 4.1 years. Pulmonary complications and respiratory failure are responsible for at least 84% of deaths.1,2 Although survival can be prolonged for an average of 5 years by tracheostomy intermittent positive-pressure ventilation (TPPV),3 many clinicians have ethical reservations about recommending it for ALS patients and, at least in some states, < 10% of ALS patients are offered or consent to tracheotomy.4 Survival can also be statistically prolonged for up to 12 months by providing positive inspiratory pressure plus positive end-expiratory pressure, commonly known as bilevel positive airway pressure.5,6 However, this is typically used only overnight and at inspiratory-to-expiratory pressure spans of < 10 cm H₂O. Such low spans are inadequate for patients with advanced inspiratory muscle dysfunction or during intercurrent chest infections.7 Furthermore, patients using low-span bilevel positive airway pressure are not typically taught mechanically assisted coughing (MAC) to prevent respiratory failure from airway mucus accumulation; therefore, few go on to require continuous ventilatory support without tracheostomy.8

Patients with ALS progress through several stages of respiratory muscle dysfunction (Table 1). Once peak cough flow (PCF) levels decrease to < 270 L/min,
patients are at risk of respiratory failure from the inability to cough effectively, particularly from the aspiration of saliva or during intercurrent chest infections. When assisted PCF levels decrease to < 160 L/min, bulbar muscle dysfunction is severe; patients fail to clear airway secretions and typically fail extubation, and tracheotomy can be required for survival when the airway becomes sufficiently obstructed with debris to decrease the pulse oximetric saturation (SpO₂) baseline to < 95%. The ability to generate assisted PCF levels > 160 L/min and to hold an insufflation deeper than the vital capacity (VC) have been reported to be associated with the capacity to prolong survival by nontracheostomy methods.

While inspiratory muscle failure can be offset by noninvasive intermittent positive-pressure ventilation (NPPV), expiratory muscle dysfunction and cough impairment can be compensated for by MAC. MAC is the use of mechanical insufflation-exsufflation (In-exsufflator or Cough-Assist; J. H. Emerson Co; Cambridge, MA) with an abdominal thrust timed to exsufflation. This provides expiratory flow directly to the airways via oronasal interfaces or via invasive airway tubes when present. The insufflation-exsufflation device is used at about 40 to −40 cm H₂O insufflation-to-exsufflation pressures. The efficacy of MAC is seen by the expulsion of airway debris and by increases in VC and SpO₂ immediately after use. As early as 1970, the use of IPPV was delayed by 4 years and 3 months for one ALS patient by the 24-h use of mouthpiece NPPV. There have been no similar subsequent reports for ALS patients. We consider prolongation of life to be indisputable when ventilator use is necessary 20 to 24 h a day and breathing tolerance is very limited; i.e., when acute respiratory distress and blood gas derangements occur within 1 or 2 h and often within seconds of discontinuing ventilator use. The purpose of this study was to demonstrate the use of NPPV for prolonging survival and eliminating the need for tracheotomy for ALS patients with bulbar muscle function.

### Materials and Methods

ALS was diagnosed on the basis of characteristic clinical course, physical and electrodiagnostic findings, and absence of evidence of spondylotic myelopathy, paraproteinemia, hyperparathyroidism, Lyme disease, glycoprotein antibodies, and vitamin E toxicity. The conditions of ALS patients were managed at a Jerry Lewis Muscular Dystrophy Association Clinic. The patients were evaluated every 2 to 6 months, depending on the rate of disease progression, until they required 24-h ventilatory support, and then monthly by respiratory therapists in the home.

The patients initially underwent pulmonary function testing (Horizon Spirometer model 2450; SensorMedics Inc; Yorba Linda, CA). Initially and subsequently, VC was measured in both sitting and supine positions, as well as the maximum insufflation capacity (MIC), the difference between MIC and VC (Wright spirometer, Mark 14; Ferraris Development and Engineering Co, Ltd; London, UK), unassisted PCF levels and assisted PCF levels following a deep insufflation and an abdominal thrust (Peak Flow Meter, model 710; HealthScan Products Inc; Cedar Grove, NJ), end-tidal Pco₂ (Microspan 8090 capnograph; Biochem International; Waukesha, WI), and SpO₂ (oximeter model 3760; Dalek-Ohmeda; Louisville, CO). Nocturnal oximetry and end-tidal Pco₂ monitoring were performed in patients who had symptoms of hypventilation, daytime hypercapnia, oxymoglobin desaturation, limited ability to breathe when supine, or a ≥ 30% decrease in VC when going from a sitting to a supine position. Sought-for symptoms included dyspnea, frequent sleep arousal with dyspnea or tachycardia, nightmares, morning headaches, daytime somnolence, and fatigue. Exclusion criteria were lung disease on the basis of FEV₁/FVC < 70% or SpO₂ < 95% despite good bulbar muscle function (PCF levels > 200 L/min), normal CO₂, and absence of acute respiratory illness.

The following criteria were used for training and intervention. Once the VC was decreased from predicted normal levels, air stacking was prescribed for the patients (i.e., holding consecutively delivered volumes of air from a manual resuscitator, with a closed glottis, to maximum lung volumes [MICs] multiple times, three times a day). When PCF levels were found to be < 270 L/min, the patients were trained in and equipped to use MAC or were given rapid access to MAC. Oximetry was prescribed for spot checks, in the event of breathing difficulty or airway encumbrment, and for continuous monitoring during chest infections to guide the use of NPPV and MAC by the patients and care providers. Portable volume ventilators (PLV-100; Respirionics, Inc; Murrysville, PA) were used for mouthpiece and nasal NPPV, initially for 8 to 20 h per day (part-time) to treat symptomatic hypventilation, but often eventually for up to full-time noninvasive ventilatory support. Volume ventilators also permit independent air stacking. Assist-control mode was used with a backup rate of 10 to 12 breaths/min with delivered volumes of 800 to 1,500 mL. The large volumes were provided to more quickly compensate for insufflation leakage out of the nose or mouth during sleep, for more efficient air stacking, and to provide the

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**Table 1—Stages of Respiratory Muscle Dysfunction**

<table>
<thead>
<tr>
<th>Stage</th>
<th>VC</th>
<th>Symptoms</th>
<th>Pco₂, mm Hg</th>
<th>Spo₂</th>
<th>PCF, L/s</th>
<th>MIC, mL</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>&gt; 50%</td>
<td>None</td>
<td>&lt; 45</td>
<td>&gt; 95%</td>
<td>&lt; 6</td>
<td>&gt; 1,500</td>
</tr>
<tr>
<td>2</td>
<td>10-50%</td>
<td>Dyspnea</td>
<td>&gt; 45</td>
<td>&lt; 95%</td>
<td>&gt; 3</td>
<td>&gt; 500</td>
</tr>
<tr>
<td>3</td>
<td>&lt; 10%</td>
<td>VFBT &lt; 5 min</td>
<td></td>
<td></td>
<td>&gt; 3</td>
<td>&gt; 500</td>
</tr>
<tr>
<td>4</td>
<td>&lt; 10%</td>
<td>VFBT &lt; 5 min</td>
<td></td>
<td></td>
<td>&lt; 3</td>
<td>&lt; 500</td>
</tr>
</tbody>
</table>

*Pco₂ = maximum nocturnal end-tidal pco₂; Spo₂ = mean nocturnal Spo₂; VFBT = ventilator-free breathing time.
ability to independently vary tidal volumes. A variety of nasal interfaces and lip seals were offered to the patients, and many patients alternated these interfaces nightly. Simple flexed mouthpieces (Respironics, Inc) or, when buccal musculature was inadequate, nasal interfaces were used for daytime ventilatory support. Four patients with inadequate buccal musculature for mouthpiece NPPV also used intermittent abdominal pressure ventilators for daytime aid. Supplemental oxygen was not used except when patients required intubation or when, in two cases, patients with no measurable assisted PCF levels chose to die at home rather than undergo tracheotomy.

Besides attending routine reevaluations, the patients were instructed to present for evaluation when unable to maintain $S_pO_2$ at $>94\%$ especially when they had chest infections. Thus, they were taught to use NPPV and MAC to maintain normal $S_pO_2$, or immediately return $S_pO_2$ levels to normal (ie, $>94\%$) by eliminating airway secretions and marked hypoventilation. Besides being used for feedback, oximetry was used to screen for atelectasis and other pulmonary complications. Oximetry was used only overnight or episodically. As weakness progressed and ventilator-free breathing tolerance was lost, patients spontaneously increased NPPV use up to 24 h per day. The $t$ test was used for all statistical comparisons.

### Results

#### Patient Demographics

One hundred sixty-six ALS patients presented from 1990 to 2000. Fifty-seven visited our clinic one or more times and were informed about the utility of noninvasive aids, but they have not returned or do not yet meet the criteria for using respiratory aids. Four patients were given a prescription for NPPV but did not obtain it. Four patients presented using TPPV. The remaining 101 patients (62 men and 39 women) met or would eventually meet the criteria for use of NPPV and MAC. The data are presented as the mean $\pm$ SD. These patients had onset at 51.4 $\pm$ 14.4 years of age (range, 22 to 85.1 years), received diagnoses at 52.7 $\pm$ 14.8 years, and became dependent on a wheelchair at 54.4 $\pm$ 13.8 years. Sex differences were not significant for the demographic data. No ALS patients met the criteria for exclusion from the study.

#### Pulmonary Function

The 101 patients underwent a total of 461 pulmonary function evaluations in the clinic, the results of which included VC levels of 1,749 $\pm$ 327 mL when sitting and 1,475 $\pm$ 298 mL when supine. In four cases, the sitting VC levels were as much as twice those measured in the supine position, and patients who had no difficulty breathing when erect or sitting had no breathing tolerance when supine and required nocturnal NPPV. The data for monthly spirometry evaluations performed at home were not included.

### Outcomes With NPPV, MAC, and Oximetry

Three of the four patients who already required continuous TPPV on presentation had residual bulbar function and underwent decannulation. Two of the three patients who underwent decannulation came from out of state specifically because they were advised that they might be offered the option for decannulation even though they had no breathing tolerance. They were switched to continuous NPPV. Of the three patients who had undergone decannulation, one had required 24-h bilevel positive airway pressure for 2 years before undergoing a tracheotomy. One month post-tracheotomy, with relatively stable bulbar function, he referred himself to us and underwent decannulation. He had assisted PCF levels of 380 L/min. He has now used NPPV continuously for 78 months. A second patient underwent tracheotomy during an acute episode of respiratory failure and then used it for nocturnal ventilatory assistance for 4 months before undergoing decannulation. He had an assisted PCF level of 210 L/min. He began to require full-time NPPV 6 months later. His assisted PCF levels became unmeasurable, and when his $S_pO_2$ baseline level decreased, the patient was informed that he required retracheotomy but he preferred to die at home. The third patient in whom decannulation was performed had undergone tracheotomy during an episode of respiratory failure 2 years earlier but did not require ventilator use and had intact bulbar muscle function when we recommended decannulation. Two years after undergoing decannulation, the patient’s assisted PCF levels became unmeasurable. He refused repeat tracheotomy during another episode of respiratory failure and died.

Of the remaining 101 trained and equipped patients, 15 have not yet required use of the equipment and 11 patients with severe bulbar muscle dysfunction died without successfully using the equipment. Two of the patients who died had become demented. Three patients used full-time NPPV and MAC during chest infections and to clear airway secretions in general but do not require ongoing use. Eighteen patients have used part-time NPPV for 3.8 $\pm$ 4.1 months. Nineteen other patients underwent tracheotomy after 4.7 $\pm$ 4.5 months of part-time NPPV. In all but one case, assisted PCF levels decreased to $<160$ L/s, and the use of NPPV and MAC could no longer maintain normal $S_pO_2$ levels. The patient with greater PCF levels was demented and uncooperative while using noninvasive methods during a chest infection.

Sixteen other patients lost all autonomous ability to ventilate their lungs, but retained sufficient bulbar muscle function (and assisted PCF levels) to use...
MAC and eventually continuous NPPV. They used part-time NPPV for 17.5 ± 13.0 months (maximum, 25 months) and full-time NPPV for 14.1 ± 12.6 months (maximum, 40 months) before PCF levels became ineffective and they underwent tracheotomy. Thus, 34 of 35 patients underwent tracheotomies only after their assisted PCF levels had declined to < 160 L/min, and 28 had MIC levels that could not exceed VC. Thus, tracheotomy was delayed despite the fact that patients had < 5 min of ventilator-free breathing tolerance. Tracheotomy was considered only after assisted PCF levels fell to < 160 L/min, the airways were encumbered, and the SpO₂ baseline was at < 95%. Of the 35 patients who underwent tracheotomies, 23 died after surviving for a mean of 4.7 ± 3.8 years, and 12 are still alive and have had a tracheostomy for 2.2 ± 3.4 years.

Nineteen patients who have not undergone tracheotomy have benefited from part-time and full-time NPPV for 25.2 ± 19.8 months (maximum, 114 months) and 17.5 ± 13.3 months (maximum, 87 months), respectively. This included one patient who used part-time NPPV for 39 months, then full-time NPPV for 87 months, with a VC of 10 mL for > 6 years. His assisted PCF levels continue to exceed 160 L/min. Ten of these 19 patients died. In all cases, they died after their assisted PCF levels had decreased to < 160 L/s but they had refused tracheotomy. Thus, these 19 patients, the 16 patients described above and 1 other patient who underwent decannulation (27 men and 9 women) used NPPV without breathing tolerance. Their survival was prolonged without tracheotomy for a mean duration of 17.5, 14.1, and 87 months, respectively. Data comparing the 36 patients in these latter groups who were successful in using full-time NPPV with limited ventilator-free breathing ability and the 19 patients who used part-time NPPV but were unsuccessful in using full-time NPPV are presented in Table 2. The only significant differences (p < 0.05) in the data are as follows: (1) those who did not use full-time NPPV also could not use part-time NPPV as long; (2) they underwent tracheotomy with higher VCs; and (3) when requiring tracheotomy, they had significantly lower assisted PCF levels and MIC-VC difference (p < 0.005) than the successful full-time NPPV users (Table 1).

Considering the entire population of ALS patients, those evaluated when their assisted PCF levels were > 160 L/min (n = 153) had a mean (± SD) VC of 1,197 ± 1,302 mL; MIC, 1,945 ± 1,921 mL; and MIC minus VC of 748 ± 811 mL. Of the 534 evaluations done on patients with assisted PCF levels  < 160 L/min, there were 462 evaluations in which MIC and VC both equaled 942 ± 857 mL; in 72 evaluations, the MIC was 861 ± 923 mL and the VC was 788 ± 894 mL, for a difference of 73 ± 121 mL. Thus, patients with effective assisted PCF levels also had significantly greater MIC than VC (p < 0.001), whereas patients with ineffective assisted PCF levels usually had no difference between MIC and VC and, therefore, severely dysfunctional bulbar musculature.

Table 2—Comparison of ALS Patient Groups Successful or Unsuccessful at Using Full-Time Noninvasive Ventilation*

<table>
<thead>
<tr>
<th>Patient Characteristics</th>
<th>Successful Users (n = 36)</th>
<th>Part-time Users (n = 19)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset, yr</td>
<td>49.5 ± 13.2 (22–66)</td>
<td>52.4 ± 15.0 (26–85.1)</td>
</tr>
<tr>
<td>Age at diagnosis, yr</td>
<td>51.3 ± 13.0 (22.7–69.5)</td>
<td>53.6 ± 15.1 (28.1–85.2)</td>
</tr>
<tr>
<td>Time from onset, yr</td>
<td>1.8 ± 4.3 (0.1–19.5)</td>
<td>1.2 ± 1.3 (0–6.1)</td>
</tr>
<tr>
<td>Age at loss of ambulation, yr</td>
<td>52.1 ± 13.0 (25.6–70.1)</td>
<td>55.3 ± 14.1 (28–76.9)</td>
</tr>
<tr>
<td>Time from diagnosis, yr</td>
<td>1.9 ± 2.4 (0–10)</td>
<td>1.6 ± 2.0 (0–8.4)</td>
</tr>
<tr>
<td>Time from onset, yr</td>
<td>2.6 ± 3.1 (0.1–14.6)</td>
<td>2.8 ± 2.7 (0.2–14.6)</td>
</tr>
<tr>
<td>Age at onset of vent aid, yr</td>
<td>54.3 ± 11.1 (25.4–70.3)</td>
<td>56.8 ± 13.7 (31.7–55.2)</td>
</tr>
<tr>
<td>Time from onset, yr</td>
<td>4.9 ± 5.7 (0.5–24.0)</td>
<td>4.5 ± 4.8 (0.1–25.3)</td>
</tr>
<tr>
<td>Time from diagnosis, yr</td>
<td>3.0 ± 3.0 (0.3–13)</td>
<td>3.4 ± 4.6 (0–25)</td>
</tr>
<tr>
<td>Noninvasive aid use, mo</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nocturnal only</td>
<td>22.5 ± 17.6</td>
<td>4.7 ± 4.5†</td>
</tr>
<tr>
<td>Full-time NPPV</td>
<td>17.2 ± 35.4</td>
<td>—</td>
</tr>
<tr>
<td>VC at trach, mL (n = 16)</td>
<td>274 ± 301 (10–800)</td>
<td>1,164 ± 523 (640–1,930)</td>
</tr>
<tr>
<td>MIC at trach, mL (n = 16)</td>
<td>274 ± 301 (10–800)</td>
<td>1,193 ± 590 (640–2,080)</td>
</tr>
<tr>
<td>MIC – VC difference at trach (n = 16)</td>
<td>0</td>
<td>29 ± 58</td>
</tr>
<tr>
<td>Recent VC, mL (n = 19)</td>
<td>292 ± 380 (10–615)</td>
<td>145 ± 258 (0–520)</td>
</tr>
<tr>
<td>Recent MIC, mL (n = 19)</td>
<td>840 ± 436 (480–2,330)</td>
<td>—</td>
</tr>
<tr>
<td>Recent MIC – VC difference (n = 19)</td>
<td>548 ± 701</td>
<td>—</td>
</tr>
<tr>
<td>Assisted PCF at trach, L/min (n = 16)</td>
<td>30 ± 24</td>
<td>26 ± 25</td>
</tr>
<tr>
<td>Recent assisted PCF, L/min (n = 9)</td>
<td>192 ± 92</td>
<td>—</td>
</tr>
</tbody>
</table>

*Data presented as mean ± SD (range). Vent = ventilatory; trach = tracheostomy.  
†p < 0.05.
In one patient, VC sitting, VC supine, and MIC were 520 mL, 350 mL, and 5,600 mL, respectively. He had no measurable unassisted PCF levels, but assisted PCF levels were 390 L/min. Over the next year, he lost all breathing tolerance except by glossopharyngeal breathing.\textsuperscript{21} As his VC decreased to 400 mL sitting and 250 mL supine, he maintained 5,240 mL of MIC and 370 L/min of assisted PCF levels. One year later, his VC dropped to 50 mL and the MIC to 4,340 mL, but his assisted PCF level was then < 160 L/min. One month later, he developed a respiratory infection and died after refusing a tracheotomy.

No patients who were symptomatic for hypventilation were noncompliant with NPPV; however, patients who were dyspneic due to airway secretion encumberment never used NPPV. No patient intentionally withdrew treatment with NPPV. Because patients were always offered the use of at least three or four nasal interfaces and were encouraged to try lip-seal use for nocturnal NPPV, perinasal skin pressure sores were never a persistent problem. Several patients reported nasal congestion. This was successfully treated by using heated humidification and, at times, vasoconstrictors. Portable volume-cycled ventilators were used when patients could perform air stacking (MIC > VC). Otherwise, a bilevel positive-pressure device (BiPAP-ST; Respironics Inc) often was used for nocturnal-only NPPV. The mean follow-up period for the 101 patients was 3.4 ± 3.1 years.

**Discussion**

NPPV can be used as an alternative to TPPV provided that adequate ventilation volumes are maintained, assisted PCF levels exceed 160 L/min, supplemental oxygen is avoided, the use of sedatives is avoided or minimized, and the aspiration of airway secretions is not so severe as to cause a persistent decrease in SpO\textsubscript{2} levels to < 95%.\textsuperscript{10} Because the assisted PCF levels and difference between MIC and VC correlate with bulbar muscle function,\textsuperscript{15} the ability to use full-time NPPV over the long term is a function of residual bulbar muscle function and is independent of VC or the extent of the need for ventilatory support.\textsuperscript{10} While tracheotomy needs to be considered only when assisted PCF levels decrease to < 160 L/min, likewise, decannulation can be safely performed when assisted PCF levels exceed this level.\textsuperscript{10} Besides being able to use mouthpiece and nasal NPPV continuously to prevent ventilatory failure, patients with adequate bulbar muscle function can use MAC to prevent respiratory failure from airway encumberment.\textsuperscript{7,9}

In the literature on ALS and neuromuscular disease, investigators have sought to demonstrate statistically prolonged survival by using NPPV\textsuperscript{5,6} or TPPV\textsuperscript{3} only at night and have sought to compare the two approaches.\textsuperscript{22} However, limiting NPPV to nocturnal-only bilevel positive airway pressure\textsuperscript{5,6,22} renders the development of ventilatory failure inevitable. Because severe bulbar dysfunction is essentially inevitable in ALS patients, respiratory failure must eventually develop from saliva obstructing the airways or during intercurrent chest infections.\textsuperscript{9,17} In addition, NPPV and TPPV are not mutually exclusive methods. In the acute setting, it has been demonstrated that NPPV can be used to avoid intubation, but that when NPPV is insufficient, intubation can be performed without untoward effects from the attempt at NPPV.\textsuperscript{23} Likewise, for ALS patients, once assisted PCF levels decrease to < 160 L/min and the SpO\textsubscript{2} baseline levels decrease to < 95%, respiratory failure becomes imminent and patients must consent to tracheotomy to further prolong survival. Our data suggest that about 36 of 166 patients, or > 20%, can use MAC as needed and NPPV continuously for prolonged survival.\textsuperscript{7,9} Most other patients develop severe bulbar dysfunction before becoming dependent on continuous ventilatory support. Several patients were referred to our center specifically for noninvasive management, and may not have been referred had they had severe bulbar ALS. This may have skewed the number of successful full-time NPPV users to some degree.

Patients with ALS have been routinely treated with aerosols, bronchodilators, methylxanthines, continuous positive airway pressure, chest physical therapy, and tracheal suctioning\textsuperscript{24,25} and with supplemental oxygen\textsuperscript{26} without evidence of prolonged survival. Indeed, besides decreasing ventilatory drive, exacerbating hypercapnia, and increasing the risk of pneumonias and hospitalizations for respiratory failure,\textsuperscript{17} oxygen therapy can hinder the utility of oximetry as feedback for clearing airway secretions by MAC, maintaining alveolar ventilation by NPPV,\textsuperscript{7,9} and for monitoring alveolar ventilation during periods of autonomous breathing.

The failure to use noninvasive respiratory aids effectively often results in intubation. Intubated patients who fail to wean typically undergo tracheotomy. However, tracheostomy impairs physiologic airway secretion clearance mechanisms, leads to chronic mucus accumulation and atelectasis, is associated with a significantly higher incidence of respiratory complications and hospitalizations for ALS and other neuromuscular disease than is dependence on continuous NPPV, and often results in continuous...
ventilator dependence.\textsuperscript{17,27} Tracheotomy can also necessitate the presence of licensed health-care professionals.\textsuperscript{28}

The use of noninvasive respiratory aids, on the other hand, is preferred by most patients and care providers over invasive approaches.\textsuperscript{28} Noninvasive aids can decrease the patient’s and family’s feelings of helplessness when shortness of breath or airway congestion occurs in the home. The use of NPPV and MAC test the family’s resolve and commitment before tracheotomy needs to be considered. Patients who are initially managed by noninvasive aids and later undergo tracheotomy may be better prepared to do so, and NPPV users can withdraw from ventilatory support without necessarily requiring personal assistance.

Although the medical literature has not demonstrated any clear advantages to the use of pressure-limited or volume-limited ventilators for managing ventilatory insufficiency, air stacking cannot be performed with pressure-limited machines. This was the primary reason that we used volume-limited machines for ALS patients capable of air stacking. Volume-limited machines were also more practical for daytime support because they have internal batteries and function readily with external batteries. ALS patients referred to us using pressure-limited machines for nocturnal-only breathing assistance and who were unable to perform air stacking, generally continued using pressure-limited machines for assisted ventilation and used manual resuscitators or positive pressure (Cough-Assist) at pressures of $\geq 40$ cm $H_2O$ for regular lung expansion.

Unlike patients with Duchenne muscular dystrophy,\textsuperscript{9} spinal muscular atrophy,\textsuperscript{7} and most other neuromuscular diseases, bulbar dysfunction in patients with ALS eventually necessitates tracheotomy to prolong survival. Thus, we do not offer decannulation unless bulbar muscle function is well-preserved for an ALS patient who is nonbulbar initially or, at least, has assisted PCF levels $> 160$ L/min and relatively stable bulbar function. ALS patients also tend to be more depressed than patients with other neuromuscular diseases,\textsuperscript{29} tend to have less familial support than pediatric patients, and, because of the lack of definitive diagnostic tests, tend to go “diagnosis shopping” rather than seek interventions that will prevent future complications. All of these factors may explain why one third of the patients may not return for eventual use of NPPV and MAC. On the other hand, it should be pointed out that much of patients’ despair is due to the very negative counseling and lack of hope usually offered them when their conditions initially are diagnosed, the extraordinary burdens placed on family caregivers, and the emphasis on institutionalizing or “warehousing”\textsuperscript{30} self-directed severely disabled people for lack of a national policy for personal assistance services. A more positive approach, as recommended by the American Academy of Neurology,\textsuperscript{31} a better explanation of respiratory management, and more emphasis on social services and personal care assistance would result in better effect.

In summary, NPPV and TPPV are not mutually exclusive approaches. Most ALS patients eventually require continuous ventilatory support to survive. This study demonstrates that, in addition to the use of nocturnal-only bivelve positive airway pressure, about 20% of ALS patients can use NPPV up to a continuous level to prolong survival for an additional 14 to 17 months, and in some cases for $> 7$ years. Noninvasive aids should not be considered extraordinary or heroic measures. Most ALS patients can benefit from their use before requiring tracheotomy. Because of an often short window of opportunity to institute these techniques and avoid premature intubation, the techniques should become accessible to ALS patients as PCF levels decrease to $< 270$ L/min. The assisted PCF levels may be the best measure of bulbar muscle function in patients with neuromuscular weakness, and they can be related to pneumonia risk and the need for considering tracheotomy or decannulation. Clinicians who are unfamiliar with how to institute NPPV or MAC, the limitations of the various techniques, or how to prepare and fit interfaces are referred to the literature.\textsuperscript{11,32–34}

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