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Oximetry and Indications for Tracheotomy for Amyotrophic Lateral Sclerosis*

John Robert Bach, MD, FCCP; Carlo Bianchi, MD; and Elaine Aufiero, MD

Study objective: To explore the use of oximetry as a guide for using respiratory aids and tracheotomy in the treatment of patients with amyotrophic lateral sclerosis (ALS).

Setting: A retrospective review of all ALS patients presenting to a neuromuscular disease clinic since 1996.

Methods: Patients who were symptomatic for nocturnal hypoventilation were prescribed noninvasive ventilation (NIV). Patients with assisted cough peak flows of < 300 L/min were prescribed oximeters and access to mechanically assisted coughing (MAC) to prevent or reverse decreases in baseline pulse oximetric saturation (SpO2) levels of < 95%. The number of decreases in baseline SpO2 that could be normalized by any combination of NIV and MAC and the duration of normalization were recorded. When the baseline was not or could not be normalized, the time to acute respiratory failure and tracheotomy or death were recorded.

Results: Twenty-five patients became dependent on NIV, including 13 patients who received NIV continuously for a mean (± SD) period of 19.7 ± 16.9 months, without desaturation (group 1). For another 76 patients, the daytime baseline SpO2 level decreased to < 95% 78 times. For 41 patients, the baseline level was corrected by NIV/MAC (group 2) for a mean duration of 11.1 ± 8.7 months before desaturation reoccurred for 27 patients. Of the latter patients, 11 underwent tracheotomy, 14 died in < 2 months, and 2 had their condition again corrected by the addition of MAC therapy. For 35 patients, the desaturation was not or could not be normalized (group 3). Thirty-three of these 35 patients required tracheotomy or died within 2 months. The only significant difference between groups 1 and 2 and group 3 was significantly poorer glottic function in the patients in group 3.

Conclusion: Tracheotomy or death is highly likely within 2 months of a decrease in baseline SpO2 that cannot be corrected by NIV or MAC. The long-term use of NIV and MAC, and the avoidance of tracheotomy is dependent on glottic function rather than on inspiratory or expiratory muscle failure. (CHEST 2004; 126:1502–1507)

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

Abbreviations: ALS = amyotrophic lateral sclerosis; ANOVA = analysis of variance; BPV = bilevel pressure ventilation; CPF = cough peak flow; EtCO2 = end-tidal carbon dioxide; MAC = mechanically assisted coughing; MIC = maximum insufflation capacity; MI-E = mechanical insufflation-exsufflation; NIV = noninvasive mechanical ventilation; SpO2 = oxyhemoglobin saturation; VC = vital capacity

For patients with amyotrophic lateral sclerosis (ALS), ventilatory failure and death can occur in as little as 2 months from the onset of symptoms.1 The mean duration of survival from the time of diagnosis was 15 to 20 months in studies of 708 ALS patients2 and 194 ALS patients,3 respectively. Pulmonary complications and respiratory failure are responsible for the vast majority of deaths.4,5 Survival can be prolonged for an average of 5 years by tracheotomy for ventilatory support and airway suctioning.6 However, many clinicians have ethical reservations about the use of tracheotomy for ALS patients, and, at least in some states, < 10% of ALS patients are offered this therapy.7

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Inspiratory, expiratory, and bulbar muscle dysfunction can result in pulse oximetric desaturation (i.e., pulse oximetric saturation \([\text{SpO}_2]\) of <95%) due to hypercapnia, cough dysfunction, and overwhelming aspiration of saliva, respectively. Noninvasive ventilation (NIV), usually either intermittent positive-pressure ventilation or high-span bilevel pressure ventilation (BPV) delivered via nasal or oral interfaces, can be used to normalize \(\text{Pco}_2\) and \(\text{SpO}_2\). The generation of effective cough flows is critical for the prevention of pneumonia. Cough flows increased by manual and mechanical assistance can reverse desaturation due to airway mucus, provided that glottic function is adequate. Manually assisted coughing involves patients receiving and holding consecutively delivered volumes of air (i.e., air stacking) with a closed glottis to inflate the lungs to a maximum insufflation capacity (MIC). An abdominal thrust is then applied as the patient coughs. These “assisted” cough peak flows (CPFs) are measured by peak flowmeter. A manually applied abdominal thrust during the exsufflation cycle of mechanical insufflation-exsufflation (MI-E) [CoughAssist; J. H. Emerson Co; Cambridge, MA] is called mechanically assisted coughing (MAC). The MI-E is applied via oronasal interfaces or via invasive airway tubes when present, generally at pressures of +40 to −40 cm H\(_2\)O. Glottic closure is necessary for both unassisted and manually assisted cough. Upper airway patency is important for both coughing and MI-E. Both glottic control and airway patency are dependent on bulbar muscle function. Bulbar muscle dysfunction is severe when the glottis cannot be closed to permit air stacking or coughing (i.e., MIC = vital capacity [VC]) or sufficiently opened to permit adequate CPF and effective MI-E. Thus, bulbar dysfunction correlates with decreasing MIC, MIC-VC difference, CPFs, and assisted CPFs. Indeed, CPFs of at least 160 L/min are required to clear airway debris for successful extubation. Advanced bulbar dysfunction can ultimately result in overwhelming saliva aspiration that causes persistent desaturation that cannot be reversed by MAC. Thus, desaturation can be reversed by using NIV and MAC, bulbar function permitting. Otherwise, the result is pneumonia, respiratory failure, and tracheotomy or death. The purpose of this study was to explore oximetry as a guide in the use of NIV and MAC to avoid tracheotomy for managing patients with ALS. Factors that predict a successful intervention with NIV and MAC were explored.

**Patients and Methods**

All of the ALS patients presenting to a Jerry Lewis Muscular Dystrophy Association Clinic since 1996 were studied. ALS was diagnosed on the basis of characteristic clinical course, electrodiagnostic findings, and the absence of evidence of spondylotic myelopathy, cancer, paraproteinemias, hyperparathyroidism, Lyme disease, glycoprotein antibodies, and vitamin E toxicity. The 25 group 1 patients and 73 of the 76 patients who experienced desaturations had a mean (±SD) time to the onset of symptoms 8.9 ± 8.1 months and 10.9 ± 8.7 months, respectively, before the diagnosis was established. Three outlier patients apparently became symptomatic > 4 years before experiencing rapid progression.

**Patient Evaluation**

All patients were questioned about symptoms of respiratory muscle weakness including dyspnea, frequent arousals from sleep, nightmares, morning headaches, hypersonolence, and fatigue. They underwent routine initial pulmonary function testing including the measurement of forced expiratory flows. The following items then were monitored in the clinic every 2 to 6 months until the patient required continuous ventilatory support: symptoms; manual muscle testing; VC in sitting and supine positions; MIC; MIC-VC difference (Wright spirometer, Mark 14; Ferraris Development and Engineering Co, Ltd; London, UK); unassisted and assisted CPFs (Peak Flow Meter, model 710; Health Scan Products Inc; Cedar Grove, NJ); end-tidal \(\text{CO}_2\) (Et\(\text{CO}_2\)) level (Microscan 8090 capnograph; Biochem International; Waukesha, WI); and \(\text{SpO}_2\) (model 3760 oximeter; Ohmeda; Louisville, CO). Once patients began requiring continuous NIV, their clinic visits were less frequent, but respiratory therapists performed Et\(\text{CO}_2\), \(\text{SpO}_2\), spirometry, and CPF measurements, and monitored ventilator use monthly in the home (data not included here). Exclusion criteria were the presence of lung disease, based on an FEV1/FVC ratio of <70% or an \(\text{SpO}_2\) of <95% on initial referral despite adequate bulbar muscle function (ie, MIC more than VC or assisted CPFs of >300 L/min), normal or low \(\text{Pco}_2\), and the absence of acute respiratory illness. No patients presented in heart failure.

**Therapeutic Protocol**

Once the VC decreased from predicted normal levels, the patients with glottic function performed air stacking three times a day in MIC using a manual resuscitator. A trial of nocturnal NIV was undertaken for patients with reduced supine VC and symptoms suggestive of nocturnal hypventilation. Nocturnal high-span (i.e., inspiration, 16 to 22 cm H\(_2\)O; expiration, 2 cm H\(_2\)O) BPV (BiPAP-ST; Respironics Inc; Murrysville, PA) was used for the treatment of patients with insufficient glottic function for air stacking (i.e., MIC equal to VC). Portable volume ventilators (PLV-100, Respironics Inc; or LTV900; Pulmonetics Systems Inc; Colton, CA) were used on assist-control mode (respiratory rate, 10 to 12 breaths/min), with delivered volumes of 800 to 1,500 mL for all patients who could air stack (i.e., MIC more than VC). The high delivered volumes were provided to more quickly compensate for nose or mouth leakage during sleep, for more efficient air stacking, coughing, and raising of voice volume, for more effective respiratory muscle rest, and for more physiologic varying of tidal volumes. A variety of nasal and oral interfaces (Lipsseals; Respironics) was offered, and many patients alternated interface use. Simple 15-mm flexed mouth pieces (Respironics Inc., Murrysville, PA) or, when buccal muscularity was inadequate, nasal interfaces or intermittent abdominal pressure ventilators (Exsufflation Belts; Respironics Inc) were used when daytime ventilatory support became necessary. Four patients used the latter. Ventilator use of 8 to 20 h per day was considered to be part-time use. Ventilator use of >20 h per
day was considered to be full-time use. Survival was only considered to be prolonged by full-time ventilatory support. Supplemental oxygen, sedatives, and narcotics were not used except when patients required intubation, and in two cases when patients with diminished SpO2 chose to die at home rather than undergo tracheotomy.

When CPFs were found to be <270 to 300 L/min (depending on the rate of disease progression), the patients were trained in and provided access to MAC15,16 and home oximetry. The latter was used whenever the patients experienced respiratory difficulty. Thus, the SpO2 guided the use of NIV and MAC. The goal was to return SpO2 to normal (ie, >94%) without oxygen therapy.17–20 Thus, oximetry screened for severe hypventilation and airway congestion, and when these could not be corrected by treatment, the resulting gross atelectasis and pneumonia.1,13,17,18,21 In general, SpO2 was normalized by NIV when EtCO2 was high. SpO2 was normalized by MAC when the desaturation was caused by airway congestion. For these latter cases, the EtCO2 was usually normal or low. The use of MAC was recommended up to every 5 min around-the-clock (when awake) as needed during intercurrent respiratory tract infections. The patients were told to notify us for sustained SpO2 levels of <95%.

**Data Collection and Analysis**

The following groups were compared: the 25 patients in group 1 who developed symptomatic hypventilation and used NIV up to full-time without developing diurnal decrease in SpO2; the 41 patients in group 2 whose 43 episodes of persistent desaturation were corrected; and the 35 patients in group 3 whose desaturation episodes could not be corrected with treatment. For patients whose SpO2 levels were normalized with treatment, the duration of normalization until the next sustained desaturation was noted. For those patients whose SpO2 could not be normalized, the length of time until acute respiratory failure, tracheotomy, or death was noted.

A series of 13 univariate one-way analyses of variance (ANOVAs) was conducted to evaluate the relationships among the three groups with the dependent variables being age, months from diagnosis, SpO2 minimum, SpO2 maximum, EtCO2, VC in sitting position, VC in supine position, MIC, MIC-VG difference, CPF, assisted CPF, duration of part-time ventilator use, and duration of full-time ventilator use. In order to control for type I errors from the 13 univariate ANOVAs, the Games-Howell method for pairwise comparisons was used.

**RESULTS**

**Patient Demographics**

A total of 165 ALS patients presented from 1996 to May 2003. There were 32 patients who had normal SpO2 levels and did not require NIV or MAC at their last evaluations, 19 patients with normal SpO2 values who were prescribed nocturnal NIV but were unavailable for follow-up, and 9 patients who either died or underwent tracheotomy without obtaining oximeters. Ten other patients were referred with tracheostomy tubes. Six of the 10 patients who underwent decannulation and switched to NIV/MAC were considered in this report. Three of those patients had undergone decannulation despite continuous ventilator dependence. All six patients maintained normal SpO2 levels immediately following decannulation, and their data were considered from this point forward. Three patients with exercise-induced bronchospasm or COPD with oxygen requirement and diminished forced expiratory flows were excluded.

Thus, 101 patients (59 males and 42 females) were studied over a mean (± SD) follow-up period of 3.9 ± 3.1 years. They underwent a total of 578 clinic evaluations. Descriptive data for the three patient groupings are presented in Table 1. The data were recorded at the last visit for the patients in group 1, and when persistent desaturation occurred for patients in groups 2 and 3. For the three groups, the mean VC sitting was 1,238 ± 836 mL (range, 40 to 3,830 mL), and the mean VC recorded with the patient in the supine position was 928 ± 738 mL.

### Table 1—Descriptive Data*

<table>
<thead>
<tr>
<th>Variables</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, yr</td>
<td>53.8 ± 11.8 (38–83)</td>
<td>53.5 ± 11.0 (34–74)</td>
<td>55.6 ± 12.7 (29–86)</td>
</tr>
<tr>
<td>Time from diagnosis, mo</td>
<td>16.6 ± 19.4 (0–70)</td>
<td>19.2 ± 15.3 (0–60)</td>
<td>18.5 ± 23.9 (0–131)</td>
</tr>
<tr>
<td>SpO2, %</td>
<td>Minimum</td>
<td>96.0 ± 0.7 (95–97)</td>
<td>92.3 ± 2.1 (84–94)</td>
</tr>
<tr>
<td></td>
<td>Maximum</td>
<td>97.1 ± 0.8 (96–99)</td>
<td>94.1 ± 1.9 (58–96)</td>
</tr>
<tr>
<td></td>
<td>CO2, mm Hg</td>
<td>41.0 ± 7.5 (32–66)</td>
<td>41.8 ± 9.8 (18–64)</td>
</tr>
<tr>
<td></td>
<td>VC, mL</td>
<td>Sitting</td>
<td>1,235 ± 877 (50–3,020)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Supine</td>
<td>940 ± 813 (20–2,920)</td>
</tr>
<tr>
<td></td>
<td>MIC, mL</td>
<td>2.02 ± 1.007 (120–4,340)</td>
<td>2.008 ± 1.008 (500–4,200)</td>
</tr>
<tr>
<td></td>
<td>MIC-VG difference, mL</td>
<td>831 ± 1,009 (0–4,290)</td>
<td>555 ± 681 (0–2,700)</td>
</tr>
<tr>
<td></td>
<td>CPF, L/min</td>
<td>2.31 ± 2.23 (0–6.7)</td>
<td>2.64 ± 2.17 (0–9.3)</td>
</tr>
<tr>
<td></td>
<td>Assisted CPF, L/min</td>
<td>3.55 ± 2.14 (0–7.6)</td>
<td>3.62 ± 2.34 (0–9.3)</td>
</tr>
</tbody>
</table>

*Values given as mean ± SD (range). Age = age at time of first contact; Time from diagnosis = time from establishment of the diagnosis until the last evaluation for group 1 or until desaturation occurred for groups 2 and 3; SpO2 minimum = minimum saturation over 5 min of observation during quiet breathing; SpO2 maximum = maximum saturation over 5 min of observation during quiet breathing.

†Indicates significant difference at p < 0.01 by comparison with group 3.
(range, 0 to 3,780 mL). The mean VC was significantly greater when recorded with the patient in the sitting position than when recorded with the patient in the supine position for all three groups (p < 0.01). Of the 76 patients in groups 2 and 3, the SpO₂ had been documented to have been normal on one or more clinic visits before 63 of their desaturations.

**Group 1**

Only 5 of the 25 patients in group 1 began using NIV with diurnal hypercapnia (EtCO₂, 45 to 66 mm Hg). However, most patients could not breathe unaided when reclining. Only 4 of the 25 patients had gastrostomy tubes because of severe dysphagia. These 25 patients used NIV part-time for a mean duration of 12.8 ± 14.1 months (range, 1 to 58 months), and 13 patients went on to require NIV continuously for another mean period of 19.7 ± 16.9 months (range, 1 to 70 months) and continued to do so at the last evaluation. The mean MIC for the subgroup of 18 patients who could air stack was 2,294 ± 1,090 mL, compared to a mean VC of 1,161 ± 881 mL. The mean subgroup unassisted CPFs were 2.59 ± 2.21 L/min (range, 0 to 6.7 L/min), and the mean subgroup assisted CPFs were 4.37 ± 2.22 L/min (range, 2.3 to 7.1 L/min), indicating good glottic function.

**Group 2**

Forty-three of 78 episodes of desaturation were corrected by therapy with NIV/MAC. Patients in 30 episodes had their conditions corrected for a mean duration of 11.1 ± 8.7 months (range, 1 to 37 months) before desaturation reoccurred, and conditions could not be corrected in 28 patients. Thirteen of these 28 patients underwent tracheotomy, and 15 died, with all but 1 patient dying or undergoing tracheotomy in < 61 days. Eleven other successfully treated patients have not experienced desaturation again for a mean duration of 4.2 ± 7 months (range, 1 to 13 months).

Considering all 43 episodes of corrected baseline SpO₂ levels, patients in 12 episodes had their conditions corrected by NIV therapy alone, including 5 patients whose EtCO₂ exceeded 50 mm Hg, 9 by MAC alone, and 22 by the combination of NIV and MAC. Thirty-four of the 41 patients used NIV daily. Thirty-two patients used part-time NIV for a mean period of 11.1 ± 18.1 months (range, 0.5 to 32 months), and 23 used full-time NIV for a mean period of 8.3 ± 12.7 months (range, 1 to 29 months).

Of the 13 patients in group 2 who were not provided with MAC devices (CoughAssist; J. H. Emerson Co) because of airway collapse and stridor, despite having chronic airway congestion, 10 had MIC equal to VC, and 12 had assisted CPF equal to CPF, indicating poor glottic function. Nine of these 41 patients had gastrostomy tubes in place when they experienced desaturation and were successfully treated. Twenty-four of the 28 patients who later experienced desaturation and whose desaturations could not be corrected had gastrostomy tubes in place at that time.

**Group 3**

Of the 35 patients in group 3, 8 either refused or were not equipped soon enough to possibly avert respiratory failure, and 2 patients were lost to follow-up until they developed respiratory failure. The other 25 patients were prescribed and used ventilators, were prescribed MAC, or both. Twenty-one patients used part-time NIV for a mean period of 7.9 ± 12.3 months (range, 0.3 to 35 months), and 9 patients used full-time NIV for a mean period of 12.2 months (range, 0.4 to 84 months).

With desaturation, 14 patients developed acute respiratory failure and underwent tracheotomy in a mean time period of 22 ± 25 days (range, 1 to 80 days), and 20 patients died in a mean time period of 48 ± 61 days (range, 1 day to 9 months). One continuously NIV-dependent outlier patient aspirated and arrested suddenly 21 months after her baseline SpO₂ decreased to < 95% (Table 2). Twenty-one of the 35 patients had gastrostomy tubes when their baseline SpO₂ decreased, which is consistent with severe bulbar muscle dysfunction.

**All Groups**

In summary, 79 of the 101 patients used part-time NIV for a mean period of 10.7 ± 13.1 months (range, 0.5 to 35 months), and 45 patients required full-time NIV for a mean period of 12.4 ± 23.2 months (range, 1 to 84 months). No patient voluntarily withdrew from using NIV or MAC. Since patients were always offered the use of at least three or four nasal interfaces and were encouraged to try an oral interface for nocturnal NIV, perinasal skin pressure sores were never an insurmountable problem. Several patients reported nasal congestion. This was successfully treated by heated humidification, by switching from BPV to volume-cycled ventilators (to eliminate excessive airway drying by continuous airflow and the circuitry of the BPV device), and, at times, by vasoconstrictor medications.

In all, once desaturation appeared, if it could not be quickly corrected by therapy with NIV/MAC, 61 of 63 patients died or required tracheotomies within 61 days. Forty-five of the 63 patients whose desaturations could not be corrected had gastrostomy tubes in place, which is consistent with severe glottic dysfunction. Likewise,
cough muscle weakness was severe for patients in all three groups, because none of the full-time NIV users or the patients who experienced desaturation had sufficient abdominal muscle strength to perform sit-ups with or without arms extended.

**Group Differences**

The ANOVA performed on the variables in Table 1 demonstrated that the MIC, MIC-VC difference, CPFs, and assisted CPFs were significantly different (p < 0.02) between groups. Age, months from diagnosis, VC recorded with the patient in the sitting and supine positions, and EtCO₂ levels were not significantly different, as determined by Games-Howell multiple-group comparisons. Indeed, there was no significant difference between groups 1 and 2 for any parameter. Patients in groups 1 (0.043) and 2 (0.045) used NIV full-time significantly longer than did patients in group 3. Significant intergroup differences between groups 1 and 3 and groups 2 and 3 are noted in Table 1.

**Discussion**

The survival of 45 patients was prolonged for up to 84 months by continuous therapy with NIV. Since VC and EtCO₂ values were not significantly different between groups, and most patients had little function of abdominal and intercostal muscles, the extent of inspiratory and expiratory muscle impairment did not predict who would respond effectively to NIV and MAC. On the other hand, the MIC, MIC-VC difference, CPFs, and assisted CPFs were significantly greater in the groups that used NIV and MAC successfully than in the patients in group 3. Clearly, these variables correlate directly with glottic function. Not surprisingly, the group 1 patients with the best glottic control used both part-time and full-time NIV for the longest time. Group 3 patients with severely dysfunctional glottic musculature would not be expected to be able to protect their airways from the essentially continuous aspiration of saliva that eventually resulted in persistent desaturation and respiratory failure despite access to NIV and MAC therapy.

Irrespective of the extent of hypercapnia or airway congestion, ALS patients are typically treated with supplemental oxygen and are not trained in assisted coughing. Besides decreasing ventilatory drive, exacerbating hypercapnia, increasing the risk of pneumonias and hospitalizations for respiratory failure, and possibly rendering nocturnal NIV less effective, oxygen therapy can hinder the utility of oximetry as feedback for titrating NIV use and clearing airway secretions by MAC, and it does not prolong survival. On the other hand, we have previously demonstrated that when oximetry is used as a guide in the use of NIV and MAC, hospitalizations, pneumonias, respiratory failure, and need for tracheotomy can be avoided, despite continuous ventilator dependence, for 91 of 91 patients with Duchenne muscular dystrophy, for 29 of 33 patients with spinal muscle atrophy type 1, and for > 640 other patients. In these conditions, bulbar function is rarely lost to the extent that speech is lost, and baseline Spo₂ decreases persistently to < 95% despite the optimal use of NIV and MAC. While we showed that many ALS patients also can benefit from continuous NIV therapy as an alternative to tracheotomy, those with severe bulbar involvement developed respiratory failure despite NIV and MAC therapy. While the failure to attain adequate assisted CPFs had been reported to be a reason to consider treatment with tracheotomy, this study demonstrated that the conditions of some ALS patients with unmeasurable unassisted and assisted CPFs and no ability to air stack can be maintained without tracheostomy tubes, whereas those of other ALS patients cannot. Thus, only persistent desaturation despite NIV and MAC therapy clearly indicates the need to consider tracheotomy to

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**Table 2—Outlier With Long-term Desaturation**

<table>
<thead>
<tr>
<th>Patient Visit Date</th>
<th>Spo₂, %</th>
<th>CO₂, mm Hg</th>
<th>VC, mL</th>
<th>Sitting</th>
<th>Supine</th>
<th>CPF</th>
<th>aCPF*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/3/02</td>
<td>96–97</td>
<td>39</td>
<td></td>
<td>1,400</td>
<td>1,050</td>
<td>4.2</td>
<td>4.4</td>
</tr>
<tr>
<td>2/21/02</td>
<td>94–96</td>
<td>39</td>
<td></td>
<td>1,270</td>
<td>840</td>
<td>3.3</td>
<td>3.3</td>
</tr>
<tr>
<td>4/25/02</td>
<td>94–94</td>
<td>37</td>
<td></td>
<td>1,000</td>
<td>740</td>
<td>2.7</td>
<td>2.7</td>
</tr>
<tr>
<td>7/3/02</td>
<td>94–95</td>
<td>39</td>
<td></td>
<td>940</td>
<td>700</td>
<td>3.0</td>
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<tr>
<td>9/3/02</td>
<td>94–95</td>
<td>37</td>
<td></td>
<td>680</td>
<td>500</td>
<td>2.1</td>
<td>2.6</td>
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<td>93–95</td>
<td>44</td>
<td></td>
<td>420</td>
<td>340</td>
<td>1.0</td>
<td>2.1</td>
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<tr>
<td>12/18/02</td>
<td>92–93</td>
<td>39</td>
<td></td>
<td>440</td>
<td>340</td>
<td>1.0</td>
<td>2.1</td>
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<tr>
<td>2/5/03</td>
<td>93–94</td>
<td>38</td>
<td></td>
<td>580</td>
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<td>8/14/03</td>
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<td>400</td>
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<td>0.1</td>
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<td>12/8/03</td>
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<td></td>
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</table>

*aCPF = assisted CPFs.*
prolong survival. The $SpO_2$ usually decreases to $< 95\%$ for $> 24$ h before patients are hospitalized for acute respiratory failure. Thus, the prescription of oximeters for home use for all patients with ALS who satisfy the criteria warranting access to NIV and MAC therapy is justified.

Of 101 patients, 13 in group 1, 23 in group 2, and 9 in group 3 required continuous ventilatory support without the use of tracheostomy tubes. Thus, tracheotomy can be delayed or eliminated for ventilatory support for about 45\% of ALS patients by the use of NIV and MAC therapy. While it has been reported that survival can be statistically prolonged for up to 12 months by providing nocturnal-only low-span BPV,\textsuperscript{27,28} inspiratory-to-expiratory pressure spans of $< 10$ cm H$_2$O are inadequate for patients with advanced disease, especially during intercurrent chest infections.\textsuperscript{18} At such times, either high BPV spans or volume-cycled ventilators at adequate delivered volumes for air stacking need to be used along with MAC around the clock.

In conclusion, NIV therapy should be offered to patients who are symptomatic for hypoventilation, and oximetry feedback with access to MAC should be offered to those patients who have assisted CPFs of $< 270$ to $300$ L/min. These interventions become critical once the baseline $SpO_2$ decreases to $< 95\%$. Tracheotomy needs to be considered when the baseline $SpO_2$ is $< 95\%$ and cannot be normalized by some combination of NIV and MAC. The introduction of noninvasive intermittent positive-pressure ventilation and MAC, the limitations of the various techniques, and how to prepare and fit interfaces have been described.\textsuperscript{8,9,11,29}

ACKNOWLEDGMENT: The authors thank Dr. Scott Millis for his assistance in the statistical analyses.

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