

Treating Respiratory Problems in ALS Patients Can Improve Quality of Life

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A 40-year-old artist diagnosed with amyotrophic lateral sclerosis had three small children and only wanted to live as long as he could paint to support his family. After starting noninvasive positive pressure ventilation, he painted 20 pieces of art over a three-year period. He referred to his NPPV as "his best friend" and the home respiratory therapist as "an angel."

Today, more patients with ALS consider it a chronic disability rather than a fatal disease and are interested in long-term survival and continued involvement in life with family and friends.¹ Dramatic improvements in the treatment of respiratory failure in ALS due to the use of NPPV, assisted cough and a better understanding of sleep-disordered breathing have made this possible.

ALS, a motor neuron disease that generally affects adults between the ages of 40 and 70, varies in its clinical pattern and rate of progression. ALS causes progressive muscle weakness and atrophy, eventually resulting in quadriplegia, loss of respiratory muscle strength, and death from respiratory failure, usually in three to five years.

However, treating respiratory muscle weakness with NPPV improves patients' quality of life and survival.²⁻⁴ NPPV, initiated early, can prolong survival for several additional years, and if patients with ALS elect to continue assisted ventilation via tracheostomy, they can survive 10 to 15 years or more. NPPV and tracheostomy ventilation are complementary; both are useful in the respiratory management of ALS.

THE TRIPLE THREAT

ALS is a triple threat for the respiratory system. It impairs the inspiratory muscles, particularly the diaphragm, resulting in hypoventilation; the expiratory muscles, resulting in poor cough and difficulty removing secretions; and the bulbar muscles responsible for pharyngeal and glottic upper airway function, resulting in dysphagia and dysarthria.⁵⁻⁷

Symptoms of respiratory muscle weakness may be of a general nature. They include excessive fatigue, low energy, anxiety and cognitive impairment. Specific respiratory symptoms include shortness of breath, orthopnea, weak cough, trouble clearing secretions, ankle swelling due to cor pulmonale, or aspiration due to bulbar impairment. Patients also may have sleep problems including poor sleep, frequent awakening, daytime sleepiness, restless legs syndrome and morning headache.

It's important to check for these symptoms regularly. Patients with ALS often don't report problems they have adapted to, even when they need to sleep sitting up. Orthopnea is very suggestive of a weak diaphragm.³

Often the first evidence of respiratory muscle weakness in ALS occurs at night, particularly during REM sleep. The intercostal and accessory muscles are flaccid during REM sleep, unable to assist a weak diaphragm at postural disadvantage in the supine position. The result is hypoventilation with hypoxemia and hypercapnia.

Initially, arousals occur resulting in sleep fragmentation. A cascade of events follows, including a blunted arousal response, reduced REM sleep, depressed chemoreceptors, progression of respiratory failure, cardiovascular and neurohormonal changes, and cognitive impairment.

EQUIPMENT TO USE

Supported by evidence-based recommendations, NPPV has become the preferred initial therapy for hypoventilation due to respiratory muscle weakness in ALS.^{2,5-8} The use of overnight oximetry can detect periods of hypoxemia. Most patients use nighttime bilevel NPPV to rest their respiratory muscles and prevent sleep-related progression of respiratory failure.

Oxygen shouldn't be used because hypoxemia in ALS is usually a manifestation of hypoventilation.⁹ It only should be used if there's an additional indication such as chronic obstructive pulmonary disease, heart failure, another lung disease causing a low PaO₂, pneumonia, or for palliative care.

Continuous positive airway pressure also shouldn't be used for respiratory muscle weakness and hypoventilation. CPAP doesn't provide sufficient inspiratory assistance, and it increases the expiratory work of breathing. CPAP is useful for ALS patients with sleep apnea, without respiratory muscle weakness and hypoventilation.

When assisted ventilation is needed, more than 50 percent of patients with ALS have some bulbar impairment. Most can use bilevel NPPV ventilation initially, even with bulbar impairment. Some may do better using a volume ventilator for NPPV.

BEGINNING NPPV

Patients should be able to try different home ventilators, and particularly a wide variety of interfaces, to select the most comfortable and effective system. Interfaces must be fitted skillfully by an experienced RT.¹⁰

When bilevel NPPV is started, comfortable and well-fitting interfaces are essential. Newer interfaces adapt better to facial contours and don't require tight straps. It's useful to rotate among different interfaces to avoid skin injury.

For daytime use and in cases of nasal congestion, NPPV with a mouthpiece is helpful for patients without significant bulbar impairment. A heated humidifier also may alleviate nasal congestion, as well as an inhaled nasal corticosteroid. Some people need to use a full-face mask if mouth leaks are a problem and a chinstrap isn't effective.

The newer bilevel ventilators for NPPV have many features including rise time and inspiratory/expiratory sensitivity. These can facilitate improved comfort and adjustment to assisted ventilation, when properly monitored by an experienced RT.

It usually takes time for patients to acclimate to NPPV. Start at an IPAP of about 8 to 10 cm H₂O with a minimal EPAP of 2 to 4 cm H₂O. Over two to eight weeks, IPAP can be adjusted gradually up to an effective level, usually 12 to 18 cm H₂O. IPAP-EPAP span (DP) should be 10 cm H₂O.

The goal is patient comfort and effective ventilation that achieves an oximetry O₂ saturation 95 percent without added oxygen. Thus, follow-up oximetry spot tests are important.

Patients should use a bilevel ventilator with a back-up rate set for their usual nighttime breathing rate, so they don't have to regularly trigger the ventilator. A physician's order of "titrate to patient comfort" with a parameter range and the targets, just discussed, will assist this process.

The RT should communicate the treatment progression with the prescribing physician every step of the way to build a collaborative team process.

Most patients with ALS begin using bilevel NPPV at night while asleep. This improves survival as well as sleep quality, quality of life, daytime function and arterial blood gases.

As ALS progresses, the patient's ventilatory support will need further adjustment, and the number of hours of assisted ventilation will increase. Some patients use NPPV 16 to 24 hours/day. This requires special attention to safety.

EFFECTIVE COUGH

Effective cough is critical. NPPV often fails if airway secretions can't be cleared. Cough becomes impaired as expiratory and bulbar muscle weakness progress.

Cough strength can be measured by instructing the patient to forcibly cough through a face mask attached to a peak flow meter. When peak cough flow falls below 270 Lpm, caregivers need training in manually assisted cough techniques to clear secretions.

This involves increasing the lung volume before the cough, with a resuscitator bag or volume ventilator, and then administering an upper abdominal thrust ("quad cough") or lateral chest wall compression timed with the cough effort.

When manual methods aren't working, mechanical insufflation-exsufflation devices are effective.¹¹ They're usually set to deliver initial lung inflation with +40 cm H₂O and then a negative exsufflation cycle at -40 cm H₂O (adjusted for each person).¹²

Severe bulbar dysfunction can result in glottic obstruction that greatly limits the effectiveness of either a spontaneous or assisted cough maneuver.

WORKING WITH THE PATIENT

Regular follow-up including simple noninvasive pulmonary function tests by the RT is advisable after NPPV is initiated. A careful approach would be RT visits or phone follow-up daily for the first week, then twice a week for the next three weeks, and then every one to three months.

Patients can use simple devices at home to periodically monitor some aspects of respiratory muscle function, particularly during a respiratory tract infection. These include an incentive spirometer and a peak flow meter. They should keep a written log of the results for the RT and physician to check.

Current guidelines on when to start mechanical ventilation in ALS are based on the American College of Chest Physicians' consensus conference report (1999), the practice parameters from the American Academy of Neurology (1999), and Medicare.^{2,8,13}

Intervention with NPPV earlier than the current standard of care may result in improved quality of life.¹⁴

For equipment reimbursement, Medicare requires a diagnosis of ALS and symptoms related to respiratory muscle weakness medically documented by abnormality of at least one of four tests: FVC < 50 percent, MIF < -60 cm H₂O, PaCO₂ 45 mm Hg, or sleep oximetry SpO₂ 88 percent for at least five minutes.¹³

A large number of patients are interested in using NPPV when they have care that's experienced, hopeful and encouraging, plus the needed resources. In preparation for decision making about respiratory care options, patients with ALS can benefit from an education and training program that explores all the choices.

It's critical that this occurs before NPPV is needed. This process, along with good communication among patients, the RT and the physician, avoids crisis hospitalization and improves outcomes.

WHEN NPPV IS NO LONGER EFFECTIVE

Some patients are unable to tolerate NPPV, usually due to bulbar impairment or equipment problems. As bulbar impairment progresses, there will come a point when NPPV is no longer effective. Before this occurs, decision making regarding long-term tracheostomy ventilation is needed.

A growing number of patients elect tracheostomy ventilation. With good care, they report that they're satisfied with their quality of life and would make the same decision again. If NPPV is becoming ineffective and tracheostomy isn't desired, then hospice referral and palliative care are important.

We encourage respiratory professionals to be informed and enthusiastic about the treatment of patients with ALS and their families. The rewards far outweigh the professional and emotional challenges.

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RESOURCE

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