



Optimal Use of Nasal Ventilation in People with Amyotrophic Lateral Sclerosis (ALS)

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Amyotrophic lateral sclerosis (ALS) is a degenerative disease of the motor nerves, occurring mostly in adults. Ultimately, this results in progressive respiratory muscle weakness, causing progressive inability to move air in and out of the lungs. This eventually leads to breathing failure, unless respiratory weakness is effectively treated with mechanical ventilation to maintain breathing.

Until the 1990s, people with ALS in the United States used tracheostomy positive pressure ventilation (TPPV), usually as a result of emergency hospitalization, without advance decision-making. A tracheostomy is a surgical opening made in the neck; ventilation is provided through a tube inserted in the windpipe, the trachea. Tracheostomy ventilation is "invasive" life support, and through this alternative, people with ALS may choose to prevent respiratory failure. This may allow life to continue as long as five to ten or more years in some cases.¹

In the past decade, however, nasal positive pressure ventilation (NPPV) has become the treatment of choice for ALS and often an alternative to tracheostomy. Nasal ventilation can be used

successfully to treat respiratory symptoms, as well as to prevent and treat respiratory failure in ALS, thus increasing survival in selected individuals. Use of nasal ventilation also provides experience and time for people with ALS to

decide whether to have a tracheostomy, if nasal ventilation fails.

Nasal ventilation is a noninvasive treatment in which a portable ventilator delivers positive pressure air through the nose, into the lungs and inflates the lungs. The ventilator uses air from the room and delivers the air to the lungs, through a long plastic tubing attached to a nasal interface. A nasal interface is either a small silicone mask fitted around the nose, or "nasal pillows" that are cushioned inserts that fit comfortably inside the nostrils. The nasal interface is anchored to the nose by wearing special straps or headgear. Some people can use a mouthpiece (straight, angled, or one with a lip seal) in the daytime if their lip muscles can hold on to the mouthpiece without air escaping from the mouth, or by using straps. Rotating from one interface to another can help promote comfort and to avoid pressure to the same areas of skin.

People who have trouble keeping their mouth closed may need to use a chinstrap to close the mouth. Full-face masks are also available to alleviate problems due to air leaks from

the mouth or congested nasal passages. These are not used continuously because they impair the ability to talk, as well as increase the risk of aspirating oral secretions.

The best candidates for nasal ventilation are people who are "nonbulbar" (those who can talk and swallow and do not have excessive oral secretions).^{1,2,3} In fact, people who are nonbulbar are sometimes able to successfully use nasal ventilation for up to 24 hours a day, unless bulbar symptoms become advanced.^{1,2} Although people who are bulbar can use nasal ventilation,^{1,2,3} overall, they are less tolerant than the nonbulbar^{1,2,3} and are a higher risk for aspiration.³ Studies have shown that some nonbulbar ALS patients who have used nasal ventilation have prolonged their survival for more than five years, until bulbar impairment became severe.^{1,2} Thus, nonbulbar users of NPPV tend to live longer than those who are bulbar, unless tracheostomy is used.

Factors for successful NPPV use include: the absence of excessive oral secretions, effective airway clearance, properly fitting nasal or oral interfaces, willingness to wear interfaces, the desire to live, and good caregiver support.¹

Initiating nasal ventilation at the appropriate time is essential in order to treat early symptoms,

prevent respiratory failure, avoid emergency decision-making, and prevent unwanted tracheostomy. Nasal ventilation should be initiated when respiratory decline is detected. Symptoms of respiratory impairment vary widely and may include: shortness of breath, the inability to take a deep breath, a weak cough force, discomfort in breathing in any position except sitting or standing upright and a low voice volume.¹ Some people experience headaches, particularly in the morning, daytime drowsiness or restlessness.¹ Because signs of respiratory muscle weakness may be overlooked, careful and accurate monitoring of the respiratory status is necessary.

People with ALS should have periodic pulmonary function tests (PFTs) and an evaluation by a pulmonary physician. This will help assess the degree of respiratory muscle involvement, the progression of symptoms and when to start nasal ventilation. One of the simple tests that indicates significant respiratory impairment is a vital capacity (VC) of 50% or less.¹ People with ALS should know the results of their pulmonary tests and what they mean. This will allow ALS individuals and their families to participate collaboratively in decision making with their doctors.

Noninvasive nasal or mouthpiece ventilation can be provided with either a volume cycled or a bi-level ventilator. The volume ventilator delivers a preset volume of air for each breath cycle. The bi-level ventilator delivers two preset levels of pressure: one level of air pressure is for inhalation, called the "IPAP" (inspiratory positive airway pressure) and the other for exhalation, called the "EPAP" (expiratory positive airway pressure). To achieve and maintain adequate ventilation and relief of respiratory symptoms,

appropriate ventilator settings are essential.⁴ The IPAP setting should be set high enough to adequately inflate the lungs, while the EPAP should be low, since the higher the EPAP, the more difficult it may be to exhale.⁴ To maintain ventilation, people with ALS who use a bi-level ventilator should use one that includes a Spontaneous / Timed (S/T) mode. The Spontaneous mode assists one's breaths, while the Timed mode provides breaths if one is unable to do so.

People with ALS should not use CPAP (continuous positive airway pressure) machines because this increases the work of breathing out. CPAP is used to treat obstructive sleep apnea, rather than for neuromuscular disorders where "assisted ventilation" is needed.⁴ Also, oxygen should not usually be used as a substitute for positive pressure ventilation.⁴ Use of oxygen alone can result in respiratory failure when assisted ventilation is needed to treat respiratory muscle weakness.⁴ Sometimes oxygen will be needed in addition to assisted ventilation, if a person has pneumonia or a lung disease.

Nasal ventilation is usually initially used when sleeping at night and during naps during the day. The amount of hours necessary to relieve symptoms and to maintain optimal ventilation depends on each person's changing respiratory status. Thus, NPPV is not needed "only" when sleeping. The progression of respiratory muscle weakness will gradually increase the need to use NPPV if maintaining ventilation is desired. Although the physician must prescribe the type of ventilator and the settings, the respiratory care professional should work in conjunction with the physician and patient to help evaluate and identify optimal settings. The ventilator settings should

be adjusted to achieve optimal comfort and relief of symptoms. Only the patient can determine what is most comfortable.

The bi-level ventilator was first introduced in 1990 by Respironics, Inc. (Pittsburgh, PA, USA) and their original model with the spontaneous / timed mode was called , the Respironics BiPAP® S/T, meaning "Bi" for the two levels of pressure, and "PAP" for positive airway pressure. As the pioneer of bi-level ventilation, the Respironics "BiPAP" became widely used. Other bi-level machines became available later.

Bi-level ventilators are simple to use, lighter weight and less expensive. They also compensate for air leaks. Although they have no built-in battery, most are able to be adapted to run on an external battery that is kept charged. The volume-cycled ventilators may be more complex to use and heavier in weight. However, they have an internal battery that automatically works for a few hours, in the event of a power failure, and they attach easily to an external battery. Whatever ventilator is used, one should have the right to indicate their ventilator preference, know their prescribed ventilator settings and what they mean.

People with ALS and their families should have the right to accurate, necessary and understandable information for making the best life choices and timely interventions. It is wiser to plan well ahead than to be one minute too late.

References:

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