

# Respiratory Failure: Noninvasive Ventilatory Support

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Respiratory failure is one of the most serious consequences of ALS, and is the most common cause of death in individuals with this disease. It results from progressive weakness of the muscles of respiration as a result of the loss of motor neurons supplying those muscles. Although we have no means of stopping or reversing motor neuron loss, there are mechanisms for helping ALS patients with respiratory muscle weakness.

Breathing is controlled by the diaphragm and by muscles known as the accessory muscles of respiration (chest muscles and some neck muscles). Respiratory muscle weakness may develop at any point in the course of ALS. Most commonly it is a late consequence of the disease and develops gradually, with the affected individual initially unaware of it. Although most patients with ALS notice respiratory symptoms when the forced vital capacity (see explanation below) falls to 50% or less of normal, some patients with ALS may have values as low as 38% of normal, yet be unaware. In rare instances, however, shortness of breath may be an early consequence of ALS, or even the first symptom.

Most ALS clinics monitor strength of breathing regularly during the course of ALS, usually by measuring the forced vital capacity (FVC), a measure of how much air the individual can blow out. The American Academy of Neurology, in its guidelines for the care of patients with ALS, recommends measuring FVC at diagnosis and once every 3 months thereafter. FVC is measured as the percentage of normal for someone of the same height, weight, and sex, and usually is between 80% and 120% if the respiratory muscles are unaffected. Sometimes patients with ALS have no shortness of breath when standing or sitting, but become short of breath when lying down. Other signs of early respiratory failure are frequent nighttime awakenings, excessive daytime sleepiness, and morning headaches. For such individuals, measurements of FVC when lying down, or a measurement of the ability to pull air into the lungs (maximal inspiratory pressure, or MIP) may be a more useful method of assessing respiratory capacity. A MIP of at least 60 cm of water is considered normal. Sleep studies may be used as well, recording levels of carbon dioxide or oxygen saturation levels in the blood, but these are used uncommonly in determining the need for respiratory support in individuals with ALS.

When should ventilatory support be initiated? Most neurologists recommend a trial of noninvasive positive pressure ventilation (NIPPV, also called bilevel positive airway pressure or BiPAP) when FVC falls below 50% of the predicted value, regardless of the presence of absence of symptoms such as

shortness of breath. NIPPV is also recommended when individuals have shortness of breath or the more subtle signs of respiratory failure discussed above. Most commonly NIPPV is initially used at night, when breathing disturbances are likely to be most pronounced. Studies to determine more precisely when to initiate NIPPV, and in particular whether earlier institution of NIPPV may be beneficial, are underway. Patients with ALS who tolerate NIPPV for more than 4 hours per 24 hour period show prolonged survival compared to those using NIPPV for less than 4 hours or those who do not use it at all. NIPPV also appears to improve respiratory symptoms (shortness of breath, sleeping problems, etc), quality of life, and cognitive (thinking) function.

It is important to keep in mind that NIPPV is not at all the same as mechanical ventilation through a tracheostomy. NIPPV can be used only as long as the individual can breathe on his or her own. When the patient inhales, this triggers the machine to pull air in to increase the size of the breath. Similarly, when the individual exhales, the machine pulls out more air than the individual is capable of exhaling on his or her own. So, an NIPPV device acts to increase the size of breaths and the amount of air exchange. The machine is connected to a face mask which forms a tight seal over the nose and mouth, or to nasal "pillows" which form a tight seal around the nostrils. Although NIPPV can prolong life, it cannot do so indefinitely. In contrast, a mechanical ventilator requires no respiratory effort on the part of the patient. It "breathes" for the patient, usually via a connection to the trachea (windpipe) which is made surgically, and can maintain patients alive for years after they have lost their breathing ability. This will be discussed in more detail in a future column.