

Respiratory problems

Evidence of reduced respiratory muscle strength can occur in patients with ALS. It is important that the diagnosis of respiratory muscle weakness is made early. This allows adequate treatment options to be discussed with the patient to prevent emergency presentation in life-threatening acute respiratory failure.

Patients with significant diaphragm and intercostal muscle weakness will often complain of difficulty breathing when they are lie down because this position displaces the abdominal contents into the thorax. For this reason they need more than 1 pillow to sleep. As weakness progress and become severe, the volume of speech declines and they complaint of breathlessness, first on exercising and later at rest. Episodes of hypoventilation can occur during sleep and cause repeated arousal and sleep fragmentation. This manifests excessive daytime sleepiness, lack of concentration, mood disturbance and loss of appetite. Eventually hypoventilation leads to the elevation of arterial carbon dioxide and decreased arterial oxygen and produces morning headaches. It is important to communicate your symptoms to your doctor for adequate testing and treatment to be started.

There are a number of pulmonary measurements to detect early respiratory muscle, including maximal inspiratory pressure, vital capacity and nocturnal oximetry. According to the American Academy of Neurology Practice Parameter on ALS if the vital capacity (VC) decreases to 50% of predicted, non-invasive ventilation (BiPap) is the treatment of choice to relieve symptoms of sleep disturbances. Several studies indicate that noninvasive ventilation improves the symptoms of hypoventilation, thereby improving quality of life and increasing survival of patients with ALS. The BiPap machine costs approximately \$3,500.00, and with a medical prescription most of the DME companies can provide this to you.



Inability to cough

The production of a normal cough requires the patient to inspire an adequate volume of air and to use the expiratory muscles to forcefully exhale the air. Weakness of the expiratory muscles and of the inspiratory muscles can produce an ineffective cough. The expiratory muscles can be augmented by using manually assisted coughing techniques (manual abdominal thrust applied by an assistant as the patient coughs) or using mechanical insufflation-exsufflation (**In-Exsufflator cough machine**). This applies a deep insufflation using positive pressure, followed by an immediate exsufflation using negative pressure, extracting excess mucus from the airway.

(PICTURE OF IN-EXSUFFLATOR)

Invasive ventilation:

Tracheostomy and positive pressure ventilation may be used when non-invasive ventilation has become ineffective, or the patient cannot tolerate it but has severe respiratory insufficiency. Tracheostomy on its own, without a ventilator, may be used when there are significant problems related to bulbar involvement: bronchial clearance of secretions, dysphagia, aspiration, and ineffective cough. Small portable ventilators are available and you should receive advice from your doctor about which one to use. Cost of home mechanical ventilation depends on resources used: equipment, supplies, attendant care or nursing care and time of care. It may be between \$10,000.00 and \$20,000.00 per month, but is more cost effective than acute hospital cost. As an alternative some specific nursing homes accept patients with ALS using ventilatory support.



Tracheostomy



Ventilator