AMYOTROPHIC LATERAL SCLEROSIS SOCIETY OF CANADA
SOCIÉTÉ CANADIENNE DE LA SCLÉROSE LATÉRALE AMYOTROPHIQUE
3000 Steeles Avenue East, Suite 200, Markham, Ontario L3R 4T9
Tel. 905-248-2052 • Fax: 905-248-2019 • Toll Free:1 800 267-4257 • www.als.ca

VENTILATION: OPTIONS AND DECISION MAKING

| Fact Sheet

Over time, ALS impairs the muscles involved in breathing and coughing but does not impair the lungs. Impaired breathing function makes a person more prone to respiratory infections and failure and can cause shortness of breath, fatigue, increased mucous secretions, obstructive sleep apnea, hypoventilation and weakened coughs. Failure of the respiratory muscles may eventually occur. Because respiratory function is affected by ALS, it is usually assessed at each visit to an ALS clinic/centre. When breathing function declines and symptoms become problematic, medical equipment to help one breathe better (mechanical ventilation support) can be used.

If you have ALS, it is advised that you discuss breathing support options with members of your ALS health-care team, as well as caregivers at home to decide what might be right for you. This decision should be made well before breathing function has seriously declined and communicated in a personal health-care directive (advance directive) so that the care you desire is received if a respiratory crisis occurs. Ensure you give a copy of the directive to your doctor and inform your relatives. Without informing relatives and writing an advance directive, you could be placed on invasive ventilation if you have a respiratory crisis even if you do not want this kind of intervention.

The following information will increase your understanding of types of ventilation support available and what to consider in your decision-making process. Your decision may change over time, so it is important to revisit your options and if you change your mind, you need to state that in your advance directive and inform your doctor and relatives of your new decision.

o There are two types of mechanical ventilation, non-invasive and invasive

1. Non-invasive ventilation is delivered through a removable face mask placed around the nose and mouth, or through nasal tubes

2. Invasive ventilation is delivered through a tracheostomy, a surgically-created hole in the trachea (windpipe) through which air is forced (intubation)

o There are four forms of non-invasive ventilation recommended for people with ALS

1. Pressure-cycled ventilation delivers air at a set pressure level with a variable volume of air on a timed cycle

2. Bilevel positive airway pressure (e.g., BiPAP™) delivers air at two pressures, one for inhalation and one for exhalation

3. Volume-cycled ventilators deliver a set amount of air at greater pressures and volumes than pressure-cycled ventilators

4. Negative pressure ventilators consist of a corset-like device that wraps around the chest and creates negative pressure to allow the lungs to inflate and deflate on a timed cycle

o Non-invasive ventilation is often only initiated at night

o Non-invasive lung hygiene techniques such as lung volume recruitment and assisted cough are usually used during the day

o As ALS progresses, increased breathing support may be needed during the day, which can be provided by a mouth piece with volume ventilator

o Full-time use of non-invasive ventilation is unsustainable for extended periods of time - if a person requires 16 or more hours of ventilation daily, they will need to make a decision about invasive ventilation

o Severe bulbar impairment may make non-invasive breathing support more challenging to use

o Invasive ventilation involves the following:

o The ventilator delivers air on a timed cycle to ensure you take the minimum number of breaths per minute

o Many ventilators can be can be adjusted to either respond to a person's own efforts to breathe or override these efforts

Over

o If you can still speak, a tracheostomy speaking valve, the Passy-Muir™, will allow you to continue speaking

o Because it is often impossible for people with ALS to recover breathing function on their own after being intubated, the decision to discontinue invasive ventilation will result in respiratory failure

o Invasive ventilation is regarded as the most reliable means of delivering air to the lungs at an advanced stage of ALS

o A problem with invasive ventilation is that it interferes with the body's mechanisms for clearing mucus. Assisted cough devices and suctions can alleviate this problem

o Infections can occur at the tracheostomy site (stoma)

o Invasive ventilation requires 24-hour support from trained caregivers

Through clinical research innovations for alternatives to traditional non-invasive and invasive mechanical ventilators are on the horizon. The Diaphragmatic Pacing System (DPS), which involves implanting muscle stimulating electrodes on the diaphragm and has been shown to be effective in those with spinal cord injuries, is being tested to see if it is an effective method for breathing support and better survival in people with ALS. One of its advantages is that it does not require a face mask, nasal tubes, or tracheostomy. In January 2007, DPS trials began at Vancouver General Hospital, the second site in the world to become a trial centre. For more information on DPS, read VGH's backgrounder at http://www.vch.ca/news/docs/2007_dps_backgrounder.pdf.

Mechanical Ventilation: Things to Consider

Type of Ventilation	Non-Invasive Ventilation	Invasive Ventilation (With Tracheostomy)
Advantages	o Nose and mouth offer convenient routes for the delivery of breathing support and therefore do not involve any kind of surgical procedure o Easier to use than invasive ventilation o Usually more comfortable o Associated care is less complex and with fewer complications o If using a mouth piece with the volume ventilator your cough will be more effective o Longevity may be increased o Less costly overall o Little or no problem with aspiration o With appropriate lung hygiene regimen, lung infection occurs less frequently o Easiest to stop therapy	o More secure system if you are ventilator dependent o Has been found to provide much longer survival o No interface required therefore the face free of headgear, straps, and skin pressure problems o Doctors, nurses, and respiratory therapists tend to be more familiar with tracheostomy care and invasive ventilation
Disadvantages	o Initially, finding the optimal interface selection may be challenging, specially with upper extremity paralysis o Custom-fitted silicone-molded nasal masks which may provide a better seal at higher pressures, require additional time and expertise o Some people with ALS find it difficult to adapt o If bulbar impairment is severe, non invasive breathing support may be more of a challenge to use o Gastric distention may occur o Most centres are familiar with traditional invasive ventilation however, few have experience with day time mouth piece ventilation	o Some people feel it is too invasive, and increases their disability and dependence o The tracheostomy tube is a foreign object in the body thereby increases secretion production and infection occurrences o Secretions require suctioning with a catheter through the tracheostomy, during the day and at night which is uncomfortable o Coughing to clear the airways is difficult o The tracheostomy site (stoma) can become infected, bleed, or develop granulations that need to be removed o Associated care is more complex than non-invasive ventilation, therefore requiring more skills o Some people have difficulty with speech and swallowing o More costly overall