ALS SLA CANADA

FACT SHEET

VENTILATION: OPTIONS AND DECISION MAKING

As ALS progresses, the muscles involved in breathing and coughing become weaker, although the lungs remain unaffected. This muscle weakness leads to difficulties such as shortness of breath, fatigue, increased mucous production, sleep apnea, hypoventilation, and weakened coughs. Eventually, respiratory muscle failure may occur. Consequentially, it is crucial to monitor respiratory function regularly at ALS clinics. When breathing issues become significant, mechanical ventilation can provide necessary support by helping people living with ALS breathe better.

It is essential to discuss breathing support options with your healthcare team and caregivers to make informed decisions before breathing function has seriously declined. These decisions should be documented in an advanced directive, which helps ensure your preferences are followed during a respiratory crisis. It's important to revisit and update your ventilation preferences as your condition changes. Effective communication with your healthcare team and family is vital.

There are two types of mechanical ventilation: non-invasive ventilation and invasive ventilation.

NON-INVASIVE VENTILATION (NIV) FOR ALS

Non-invasive ventilation (NIV) is delivered through a removable mask over the nose and mouth, through nasal tubes, or through a mouthpiece. Types of NIV recommended for people living with ALS include:

- Bilevel positive airway pressure (e.g., BiPAP™): Delivers air at two pressures, one for inhalation and one for exhalation. It is mainly used with a suitable mask while sleeping and can help with inhaling and exhaling more fully. This is not the same as continuous positive airway pressure (CPAP), which is not effective for managing ALS respiratory symptoms.
- Volume-cycled ventilator: Delivers a set volume of air at higher pressures.

Advantages of NIV

- It does not involve any surgical procedures, making it less costly, easier to use, and often more comfortable than invasive ventilation.
- There are fewer complications and care is less complex than invasive ventilation.
- It reduces the risk of lung infections with proper lung hygiene practices.
- It can improve quality of life and extend survival when used consistently.

Challenges of NIV

- Finding the right mask can be difficult, especially with paralysis in the upper extremity.
- Severe bulbar impairment can make mask fitting challenging and NIV less effective.
- Some people living with ALS find it hard to adapt to the equipment.
- Gastric distention (bloating) may occur, causing discomfort.

Healthcare workers may be less familiar with NIV compared to invasive ventilation and might require more education.

INVASIVE VENTILATION FOR ALS

Invasive ventilation involves a tracheostomy, a surgically created hole in the windpipe through which air is delivered. This method is typically used when NIV is no longer sufficient, often at an advanced stage of ALS. It is important to note that ALS will continue to progress even with invasive ventilation.

Advantages of invasive ventilation

- It provides more secure ventilation for people who are ventilator-dependent.
- Survival time can be significantly extended.
- External headgear and straps are not used, which reduces skin irritation.

Challenges of invasive ventilation

- It is sometimes considered too invasive and will increase dependence.
- There is increased secretion production and infections from the tracheostomy therefore, tracheostomy tubes need regular care and maintenance to minimize the risk of infections.
- Frequent suction is needed, which can be uncomfortable and make coughing difficult.
- Care needs are more complex, more expensive, and require 24-hour support, which is not provided through homecare.
- While invasive ventilation can prolong life, it is possible to experience locked-in syndrome, meaning that someone living with ALS has progressed to total immobility while having intact cognition, potentially leading to a decreased quality of life.

KNOW THAT WE ARE HERE TO HELP

The ALS Society of Canada can assist in connecting people and families living with ALS in Ontario to support services, equipment, and ALS clinics. We also invest in the most promising Canadian ALS research, advocate federally and provincially for the needs of people affected by ALS, and provide information to empower Canadians affected by the disease. Learn more at als.ca where you can also find more resources in the "What is ALS?" section.

If you live outside of Ontario, please contact your provincial ALS Society for information on support available in your region.