Ventilation

FACT SHEET | WWW.ALS.CA

Very similar to other muscles in the body, people with ALS may also experience weakness of the breathing muscles. This can lead to a weak cough making it more difficult to clear secretions in the throat and chest while also increasing the risk of developing pneumonias. Weaker breathing muscles can also lead to hypoventilation which means the lungs are not able to take in enough air to get oxygen but more importantly cannot breathe out enough air to get out the carbon dioxide. There are assistive devices to address both these weaknesses that can reduce symptoms related to weakened breathing muscles, improve quality of life and in some instances prolong survival.

Decisions about what forms of breathing support you would like to be treated with, or not, are some of the most important that will have to be made during your ALS journey. An advance care plan can ensure your wishes are respected, and that your loved ones are aware of your preferences. It can help to begin thinking about these decisions relatively soon after diagnosis, so they are in place if you begin to experience breathing symptoms. Almost everyone who dies of ALS does so as a result of respiratory difficulties, so these decisions are very important.

ASSISTIVE DEVICES AVAILABLE FOR AIRWAY CLEARANCE

When breathing muscles that affect the strength of a cough weaken, people with ALS often experience chest congestion, mucus in the back of the throat and at times recurrent pneumonias. In order to help alleviate these symptoms, there are two options to help generate a stronger cough:

Lung volume recruitment with manual assisted cough

This is a non-mechanical method of increasing the volume of air that can be taken in when the breathing muscles get weaker. It involves a bag and mask and the assistance of a caregiver to pump air into the lung then subsequently help the abdominal muscles push air out of the lungs to generate a cough. It is recommended this be done 2-3 x per day.

Mechanical insufflator-exsufflator (Cough Assist device)

As the disease progresses and the cough gets weaker, a mechanical option for generating a cough is required. This device is pre-set and involves a small machine connected to a hose and a mask and is used minimum twice daily. It slowly pushes air into the lungs then quickly sucks it out generating a strong cough. Regular use of this device has been shown to reduce the risk of pneumonias and reduce hospitalizations if a person with ALS were to get a pneumonia.

ASSISTIVE DEVICES AVAILABLE FOR VENTILATION SUPPORT

Assistive devices can help with breathing when the muscles are too weak to do so effectively. There are two main types of ventilation devices available to people with ALS: non-invasive ventilation (e.g. BiPAP) and invasive ventilation (tracheostomy - a curved plastic tube placed surgically in the windpipe below the voice box).

Non-Invasive Ventilation

Non-invasive ventilation (NIV) involves placing a mask over the person's nose and/or mouth to deliver air to the lungs. Initially, this device is worn at night however as the disease progresses, NIV can be used during the day to help alleviate shortness of breath. The goal of non-invasive ventilation is to rest the breathing muscles at night, so they work more efficiently during the day.

The mask is fully removable, and the treatment can easily be started or stopped as the person wishes. NIV can also be delivered during the day though a mouthpiece resembling a straw for those needing NIV during the day. Studies have shown that NIV can prolong survival compared to no treatment, and improves quality of life in most people with ALS.[1]

There are both risks and benefits to NIV. The benefits include easier verbal communication, the ability to remove the device at one's will, and prolonged survival compared to no treatment at all. The risks are that NIV may not be suitable for all patients, especially those at a later stage of the disease, and in particular, those with impaired speech and swallowing (bulbar ALS). The person must also be able to maintain a lung hygiene routine in order to keep the lungs clear. People with ALS who have increased oral secretions often have more difficulty tolerating NIV through a mask. The decision to proceed with NIV should be made in conjunction with your healthcare team and caregivers.

The most common form of NIV is pressure ventilation. The device delivers air which inflates the lungs to a certain set pressure (IPAP-inspiratory positive airway pressure), providing or assisting an adequate breath volume for ventilation. There is usually a little background pressure as well (EPAP-expiratory positive airway pressure) which splints the upper airway open much like CPAP for sleep apnea. The difference between IPAP and EPAP is the pressure support which provides the breath. A breath in may be as a result of spontaneous effort or the device may provide a timed (machine-delivered) breath e.g. 12 breaths per minute, if you do not initiate a breath on your own. In other words, the device can breathe for you, providing respiratory muscle rest as needed. Generally, such a back up rate is strongly recommended by international guidelines.

Non-invasive ventilation is more suitable for the earlier stages of respiratory dysfunction in ALS. When a person requires round the clock breathing support, they may consider invasive ventilation.

Mouthpiece Ventilation

Mouthpiece ventilation (MPV) is another complementary form of NIV that can be used during the day in a carefully selected population of ALS patients that are requiring NIV at night and during the day. You must be able to retain a plastic mouthpiece in your mouth, secured by an arm-like holder and manage enough breaths to maintain normal breathing. People with weak facial muscles or lips, and difficulty speaking and swallowing (the majority) are not capable of using mouthpiece ventilation. Contact your respiratory team for more information.

Invasive Ventilation

Invasive ventilation can be considered if increasing use of non-invasive ventilation is required due to progressive muscle weakness or acutely in the intensive care unit if there is a condition such as pneumonia. Chronic invasive ventilation uses a tube inserted surgically into a hole through the neck (a tracheostomy) to assist a person with breathing and is usually worn throughout the day. While some people can preserve verbal communication for a short period of time, most require assistive devices to communicate. Your speech language pathologist may be able to identify options that will work for you. Those receiving chronic invasive ventilation receive nutrition through a tube inserted directly in the stomach [PEG (percutaneous endoscopic gastrostomy)].

Making the decision to use invasive ventilation is a big choice for you and your family. It should be discussed early in the disease progression and should align with the person's advance care plan. Your healthcare team is an excellent resource to discuss options as the involvement of ALS and the breathing muscles continue. The decision for acute ventilation when there are sudden changes in health such as pneumonia should also be considered part of advance care planning.

Those choosing chronic invasive ventilation require 24-hour support from trained caregivers. This may take place in one's own home, a nursing home, or a hospital with the support of family or paid caregivers. The treatment interferes with body's mechanisms for clearing mucus. Techniques such as assisted cough (the Cough Assist device) and suction can help. Studies have shown that invasive ventilation can prolong survival for people with ALS compared to non-invasive ventilation for approximately 17 months, to a total of 47 months. With no treatment, the survival time is approximately 30 months.[2]

Invasive ventilation can prolong survival for people with ALS for approximately 17 months, to a total of 47 months. With no treatment, the survival time is approximately 30 months.[2]

People with ALS may opt to have their ventilator removed at any point during the illness. If this is a life-limiting decision symptom management (palliative care) will be provided to prevent significant breathlessness or anxiety. It is important to note that even with the use of invasive ventilation unfortunately ALS continues to progress and may lead to a point where communication is no longer possible.

WHAT DO PEOPLE WITH ALS FIND IMPORTANT IN THE VENTILATION DECISION MAKING PROCESS?

Breathing support is one of the most important and challenging decisions a person with ALS and their loved ones will have to make. People with ALS have to decide whether they would like to pursue breathing support, and whether invasive versus non-invasive ventilation is best for them.

When asked by researchers about the considerations when making decisions about ventilation, people living with ALS identified the following:

- **Autonomy.** They wanted to feel like they could make this decision themselves, in advance, rather than having it made for them in an emergency.
- **Relationships.** They took their loved ones' considerations into account, and did not want to burden them physically, emotionally, or financially.
- **Communication.** They wanted to maintain their ability to communicate verbally.
- **Survival.** They considered the survival implications of their decision.
- **Quality of Life.** They wanted their choice to improve their quality of life.[3]

People with ALS say it is important for them to understand the progression of respiratory failure in ALS, as well as all their options for ventilation. This information is best discussed early in the disease course to allow people with ALS to make an informed and independent decision. Your ALS clinical care team is always available to provide guidance.

YOUR ALS SOCIETY IS HERE TO HELP.

Making decisions about breathing support can be challenging. The ALS Society of Canada or your provincial ALS Society can connect you to support groups where you can meet with others who are going through the same process.

SUMMARY

- People with ALS and their families face important decisions surrounding breathing support.
- Breathing troubles caused by ALS can be distressing, but assisted ventilation can often relieve these fears.
- There are two major forms of breathing support: non-invasive ventilation, which is worn externally, and invasive ventilation, inserted through a tracheostomy.
- People with ALS can often maintain communication while receiving breathing support, with the use of assistive devices
- People living with ALS take many considerations into account when making breathing support decisions, including their autonomy, relationships, communication, survival, and quality of life.

ADDITIONAL RESOURCES

[1]Early use of non-invasive ventilation prolongs survival in subjects with ALS
[2]Tracheostomy mechanical ventilation in patients with ALS
[3]ALS and assisted ventilation: How patients decide
Quality of life & psychosocial issues in ventilated patients with ALS & their caregivers
Mechanical ventilation for ALS
Effects of non-invasive ventilation on survival & quality of life in patiend with ALS
Home mechanical ventilation for ALS
Associative increases in ALS survival duration with non-invasive ventilation initiation and usage protocols
Respiratory challenges in ALS - coughing
Home mechanical ventilation for patients with ALS: A Canadian thoracic society clinical practice guideline
ALS guide: A resources for people living with ALS

KNOW THAT WE ARE HERE TO HELP I For people and families living with ALS in Ontario, ALS Canada can assist in connecting you to support services, equipment, and ALS clinics. Whether you are a person living with ALS, a family member or a caregiver, we will strive to support you along this journey. If you live outside of Ontario, please contact your provincial ALS Society for information on support available in your region. Learn more at www.als.ca.

Special thanks to everyone who helped write and review this fact sheet.

* Last updated 11/2020