Breathing and ALS

The majority of ALS patients will eventually experience difficulty breathing. Unlike typical patients with respiratory problems, ALS patients rarely have trouble pulling oxygen out of the air they breathe and therefore the use of supplemental oxygen is typically not appropriate. The problem ALS patients face is simply the inability to move a sufficient amount of air in and out of the lungs due to respiratory muscle weakness. While this can result in low blood oxygen levels, it also results in an equally dangerous buildup of carbon dioxide in the lungs and blood, which the use of supplemental oxygen does not address. The solution to respiratory problems in ALS patients is either non-invasive mechanical ventilation called NIPPV or BIPAP, or invasive ventilation called a ventilator.

BIPAP

BIPAP stands for bi-level positive air pressure. It is a machine with two levels of air pressure—higher when you breathe in and lower when you breathe out. The machine senses when you are breathing in and pumps additional air into your lungs via a mask or nasal pillows. In essence, the BIPAP helps the breathing muscles to do their job. Most ALS patients initially use BIPAP only while sleeping.

The BIPAP is connected to a nasal mask, a full-face mask, or nasal pillows. The nasal mask covers the entire nose and is held in place by straps connected to headgear. There are a large variety of manufacturers, models and sizes available and comfort depends on the individual. Some patients use several masks, rotating between them to avoid repeated pressure on the same area of the skin. Some patients who have trouble keeping their mouth closed may need to use a chinstrap to hold the mouth closed. Full facemasks are also available to alleviate problems due to air leaks from the mouth or congested nasal passages. These masks cover both the nose and mouth. These are not used frequently because they impair the ability to talk. Nasal pillows are cushioned inserts that fit comfortably inside the nostrils. Nasal pillows, while not as secure as a nasal mask, have the advantage of being more comfortable for many patients. They also have the advantage of permitting the patient to wear eyeglasses. Some patients use nasal pillows during the day and a nasal mask while sleeping.

The BIPAP is usually recommended when the ALS patient’s breathing capacity falls below 50% of normal. BIPAPs are usually covered under most health insurance policies.

Ventilator

Invasive ventilation does all the work of the respiratory muscles. The ventilator involves a machine that provides complete ventilation to a patient via a tube down the person’s throat (usually this is temporary) or a tracheostomy. A tracheostomy is a surgical opening into the trachea or windpipe into which a plastic or metal tube is placed.
A tracheostomy can affect your ability to speak. For some patients utilizing invasive ventilation, they can temporarily continue to speak by using a “Passy-Muir®” valve; this valve is attached to the tracheostomy and augments airflow on exhalation through the vocal cords and upper airway to enable speech.

Most ALS patients using invasive ventilation have to be hooked up to the ventilator 24 hours a day, although there are some cases in which the person can come off the ventilator for periods of time during the day. Invasive ventilation does not slow down the weakness and eventual death of the voluntary muscles involved in ALS. The disease continues to progress despite the ventilation.

The ALS patient on invasive ventilation will require skilled care 24 hours a day, 7 days a week. Care can be provided by LPN’s, RN’s or Respiratory Therapists or by family/friends who have been trained. Insurance policies vary regarding what is covered under home care. Some ALS patients utilizing invasive ventilation are cared for in nursing homes, while many are able to remain at home. Your family and friends must decide whether or not they can devote the amount of time needed, to be sure that someone is always with you. Those who support your care at home have to learn the details of what is really involved; often they must put off many other personal plans. Family members generally need to make their own decision to assist with care, especially since the burdens are heavy. Home care should be arranged so that excess responsibilities are not carried by any one person.

If the care works out well, people with ALS who use invasive ventilation, and all other appropriate treatments, can live for years. Many individuals who chose this option are satisfied with the quality of their life. You may want to consider invasive ventilation if:

- You are highly motivated and engaged in living.
- Your medical problems, including ALS, are stable and progressing slowly.
- Your level of independent functioning allows some ADL (activities of daily living).
- Your ability to communicate is good (e.g., using a communication device), and you can help direct the care needed.
- You and your family understand the options, both the pros and cons.
- Your family is able and wants to participate, if needed.
- You have access to an experienced, multidisciplinary professional team.

You may want to not chose invasive ventilation if:

- Your disability is advanced.
- Your ability to communicate effectively is very limited.
- Your interest in or motivation for living is low.
- Your are not mentally alert or self-directed.
- You do not have caregiver support available from family or friends and do not want to live in a nursing home.
Potential Problems with Invasive Ventilation:

- Quality of life can become marginal with a perceived loss of dignity.
- 5-10% of people with ALS develop a totally locked-in state (inability to communicate in any way).
- There is a possibility of being institutionalized in a Nursing Home.
- Family members/friend’s lives can become disrupted and deferred, leading to emotional strain, anger and feelings of entrapment.
- Personal and family financial resources can be consumed.
- A burden is placed on medical care and community resources.

The decision to choose invasive ventilation is a very personal one. It may be based on practical realities, a desire to avoid being a burden to others, or your perception of a reduced quality of life. You will not live as long without a ventilator, but many people with ALS can arrange to have drugs available to keep them comfortable if respiratory distress occurs. Usually, your local hospice can provide services to help you and your family at home.

It is important for you to decide what is best for you; do not let an emergency decide for you. In the past, most people used invasive ventilation without advance planning as a result of emergency hospitalization. Stay in control so you and your family can choose what is best for you. Speak with your neurologist, social worker and mental health worker regarding this issue and others related to your Advanced Directives.