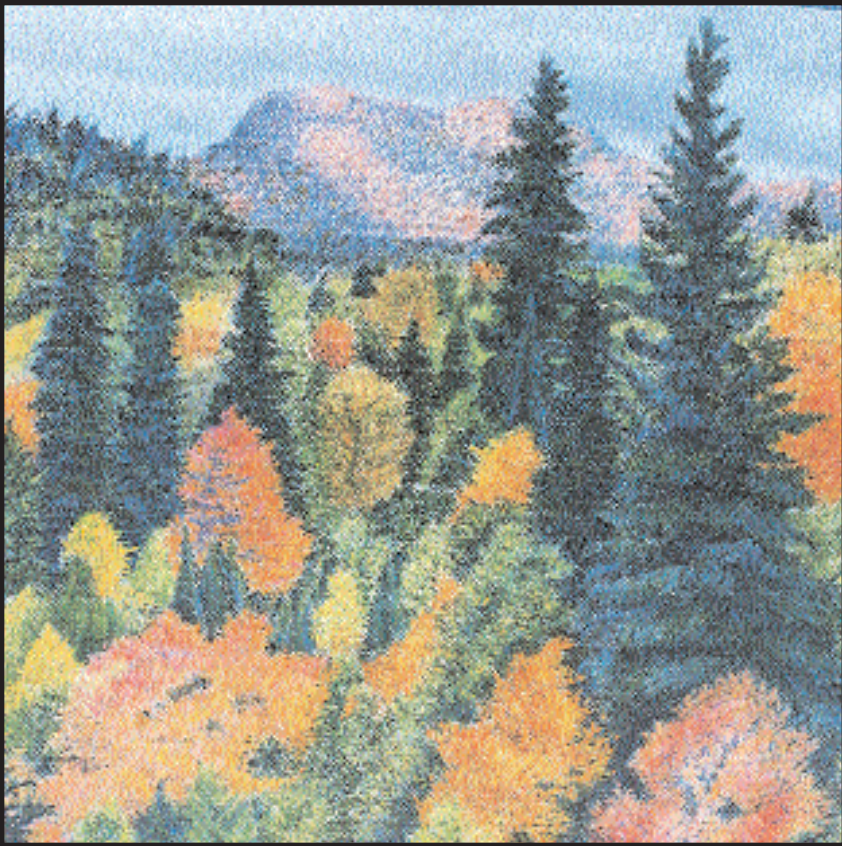


LIVING WITH ALS

# Adapting to Breathing Changes



**Cover:** *View from Agawa Canyon Train*

by Sharon Weiss.

The artwork on the covers of the *Living With ALS* Manuals was created by individuals with ALS or their family members.

*From the Artist*

**“This painting on the cover is a tribute to my father, Louis Bass, who suffered from ALS. My father was an enthusiastic traveler who made over 30 trips abroad with my mother. He would record his adventures with multiple cameras and numerous rolls of slide film. When he returned home, he would create beautiful oil paintings and black-and-white photographs from slides. He concentrated on capturing local people pursuing their everyday activities.**

**My father exhibited his paintings, photographs, and sculptures locally in Mission Viejo, California. He continued his travels and artwork until 1996 when he became too incapacitated. He lost his battle with ALS in January 1996, a few months short of his 73rd birthday.**

**I share my father’s interest in art, and obtained a B.A. degree in Pictorial Arts from UCLA. Like my father, I also enjoy traveling and recording my adventures on slides. The painting on the cover “View from Agawa Canyon Train,” was created after a fall foliage train ride through the Agawa Canyon in Ontario, Canada. The passengers spent all day on a slow train ride through a land carpeted with sugar maples and other colorful trees.”**

SHARON WEISS

*Loving daughter of Louis Bass and artist*

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*Manual 6*

**LIVING WITH ALS**

# Adapting to Breathing Changes

*Written by*

Edward A. Oppenheimer, MD, FACP, FCCP

*A note to the reader:* The ALS Association has developed the *Living With ALS* manuals for informational and educational purposes only. The information contained in these manuals is not intended to replace personalized medical assessment and management of ALS. Your doctor and other qualified health care providers must be consulted before beginning any treatment.

**LIVING WITH ALS**  
**Adapting to**  
**Breathing Changes**

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# Introduction

**T**

he medical care available for people with amyotrophic lateral sclerosis (ALS) is constantly improving. Further advances in treatment and survival look hopeful and exciting: Rilutek<sup>®</sup>, a medication for ALS, is approved by the Food and Drug Administration (FDA); impressive progress is occurring in molecular biology and genetic research related to ALS; specialists have a better understanding about the cause of this disease, and researchers and others in the biotech industry are developing treatments for the disease. The clinical management of ALS offers treatments to improve function and quality of life and increase survival.

This manual will discuss the information that is currently available on the breathing concerns that can occur with ALS. Specifically, it will cover the choices available to improve and assist with breathing. It will serve as a guide for you, your family, and friends on the following topics:

- Normal lung function;
- How ALS affects lung function;
- How to keep your lungs healthy;
- What your breathing options are, so you can choose what is best;
- What you can do when lung function decreases due to ALS.

Most persons with ALS and their families have more success, and feel more comfortable, when they understand this information and work through it. This manual will allow you to stay in control, to decide what is best for you, and to avoid “crisis

management” of respiratory complications. Some of these issues can be difficult to address; however, there are many encouraging developments.

This manual includes the terms that doctors and other health professionals use, so you can understand more easily what they are talking about. It is important for you to discuss your own situation and preferences with your doctor in order to obtain the best advice and to work out your plans. Be sure to write down questions and notes, and bring them with you when you have a doctor’s appointment.

# The Basics of Breathing

## H

### OW DO YOUR LUNGS NORMALLY FUNCTION?

When you take a breath of air, you use your diaphragm and sometimes your rib muscles. The **diaphragm** is a large dome-shaped muscle between your lungs and abdomen. As the diaphragm moves down, it expands your chest and causes air to move into your lungs. Your upper airway, mouth or nose, and throat need to be open.

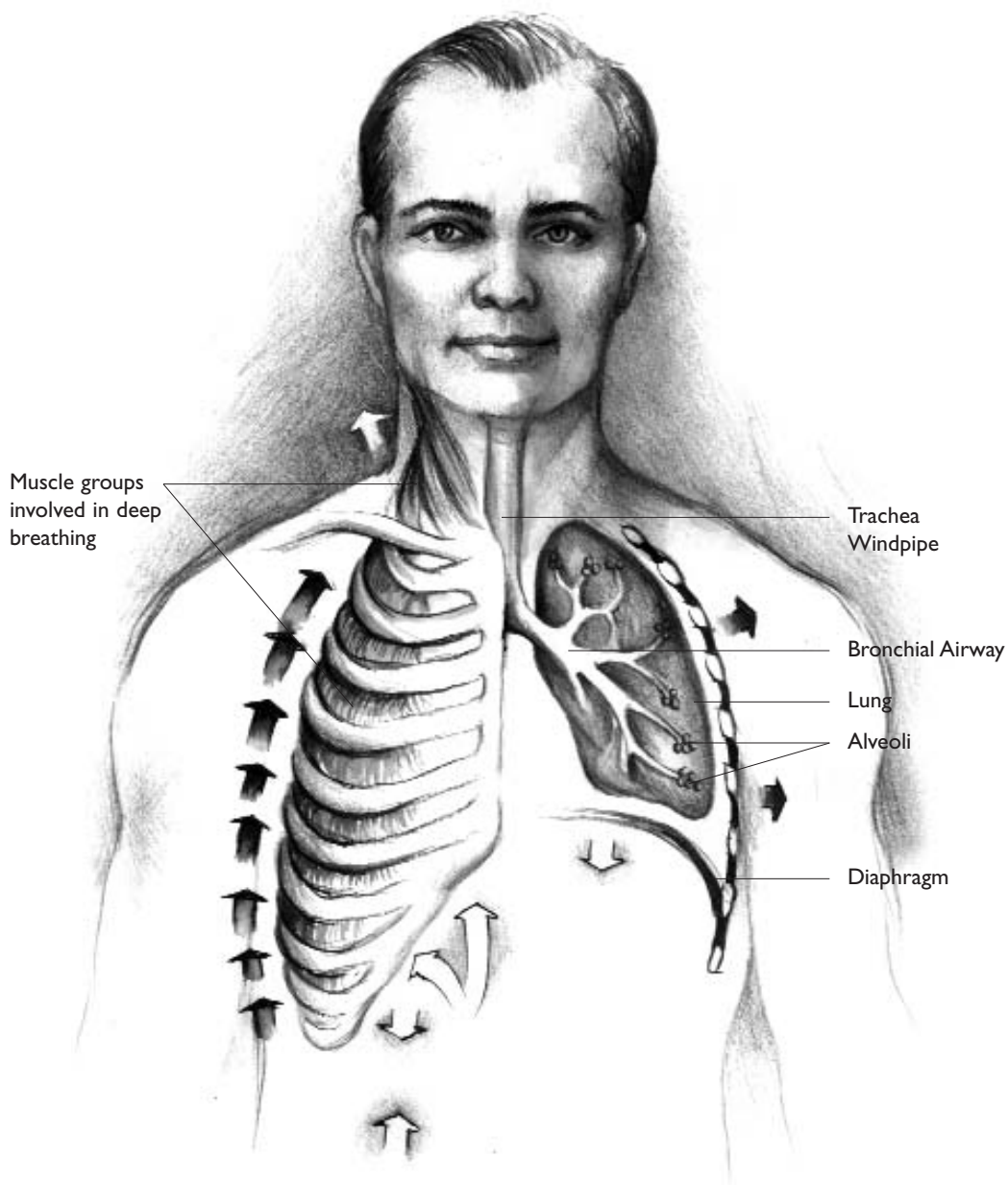
Air flows out of your lungs when you relax your breathing-in muscles. The air that you breathe in goes down the **bronchi** (the airways of the lung) into millions of **alveoli** (the small air sacs within the lungs); oxygen is taken up into the blood, and carbon dioxide (CO<sub>2</sub>) is eliminated. If there is not enough air exchange, due to decreased muscle function, then CO<sub>2</sub> and acid build up to abnormally high levels in the blood, and the oxygen level falls.

This breathing that moves air in and out of your lungs is called **ventilation**; too little is called **hypoventilation**, or under-ventilation. The gas exchange that occurs in the lungs, when oxygen is taken up into the blood and CO<sub>2</sub> is released from the blood into the air, is called **respiration**.

When you exercise, the muscles in your body need more oxygen, and the increased metabolism produces more CO<sub>2</sub> that then needs to be eliminated. The respiratory muscles work harder with exercise, helping you breathe in more deeply and take more breaths each minute; this process achieves just the right amount of ventilation to keep your CO<sub>2</sub> and oxygen saturation normal.

At night, when you sleep, many muscles relax; however, the continued muscle tone in your **pharynx** (passageway from nose and mouth to esophagus and trachea), tongue, and throat keeps your airway open to allow breathing. Your diaphragm must work harder with each breath when you are lying down, because it must push the abdominal organs down, as well as draw air into the lungs. When you are sitting or standing, gravity helps move the abdominal organs down as the diaphragm descends with each breath.

**FIGURE I**  
Lungs



*Cross-sectional view of the main air passages and the muscle groups involved in breathing: diaphragm, intercostal, neck and abdomen.*

# Your Lungs and ALS

## H

### **OW DOES ALS AFFECT THE FUNCTIONING OF YOUR LUNGS?**

ALS varies greatly from person to person and symptoms related to your breathing can start early or much later in the course of the disease. In some people the progressive loss of motor neuron function is very slow, in others faster. The motor nerves for the respiratory muscles are often affected later in the disease after weakness occurs in the arms or legs. But sometimes the respiratory muscles are affected very early in ALS, when minimal weakness exists elsewhere. The complications of respiratory muscle weakness can include difficulty breathing and clearing secretions. There are a number of devices and techniques commonly used by many people with ALS that can support your breathing if your respiratory muscles are affected.

#### **Breathing Muscles Become Weak**

When ALS affects the nerves that control the respiratory muscles, breathing becomes strained and weak. This condition may develop gradually over weeks or months or, in very few circumstances, it occurs suddenly with little warning. Often the first sign is recognized during exercise or while breathing at night. You may be aware of shortness of breath with exercise or the inability to do some activities, but such sensations might not be noticed if your arms or legs are already too weak for exercise or fatiguing activities. When you are in bed, you may sense “breathlessness” when lying flat. This symptom is an important indication of trouble; you may need to sleep with pillows or even try sitting up in a

chair to sleep. Treatment can help, but if it is delayed too long or not desired, then complications will occur.

In the past, severely diminished breathing was the usual cause for death in ALS; today, problems can be prevented through ventilation options available to ALS patients.

### **Decreased Cough Effectiveness**

When the motor nerve function of the respiratory muscles is affected, cough strength decreases and breathing becomes weak. Decreased cough strength increases the risk of pneumonia and aspiration, because an effective cough is needed to keep the airways clear.

### **Loss of Bulbar Function and Breathing Difficulty**

**Bulbar** refers to the part of the brain known as the medulla oblongata, which is at the top of the spinal column. **Bulbar impairment** indicates loss of motor nerve function to the muscles controlling speech, swallowing, and the upper airway. This condition results in problems with speech, swallowing, the ability to maintain an open upper airway during sleep, and the ability to clear away saliva as well as a tendency to **aspirate**. In the latter, food or liquids, including saliva, can drop down in the bronchial airways and alveoli of the lungs (**aspiration**). When coughing is ineffective or secretions increase, aspiration can cause parts of the lung to fill up resulting in **pneumonia**, an infection or inflammation of the alveoli and the very small airways of the lung.

When swallowing becomes a problem, nutrition can be affected. A feeding tube can help to ensure good nutritional intake. If you have excess saliva, try using tissues, a machine with a suction tip, or various medications to decrease its formation. Signs of bulbar problems include thickened speech, swallowing difficulty, a tendency to gag, an accumulation of saliva, and weight loss. (See Manual 5.)

### **Breathing Problems during Sleep**

The following are some breathing difficulties that can occur during sleep:

- If the bulbar muscles weaken, noisy breathing, snoring, or even the closing off of the upper airway results; this intermittent, blocked or stopped breathing is called **obstructive sleep apnea**. A decrease in the oxygen saturation of the blood can result.
  
- During sleep, nerve and muscle functions relax, and, as a result, nighttime under-ventilation problems may occur causing the amount of carbon dioxide to rise. Symptoms include morning headaches, lethargy, shortness of breath, disturbed sleep, daytime sleepiness, and poor appetite. Because obstructive sleep apnea and under-ventilation often occur together, a sleep study may be advised. It is usually done overnight in a sleep laboratory, where oxygen, CO<sub>2</sub>, muscle movement, airflow, brain electrical activity, and heart function are measured. An alternative would be an overnight oximetry study at home that records arterial (relating to the arteries) oxygen saturation and heart rate.

### **Progressive Muscle Weakness Heading to Respiratory Failure**

Respiratory failure tends to occur much later with the progressive loss of respiratory muscle strength. Inability to breathe independently may happen quickly under some circumstances, resulting in a crisis. A common pattern of this respiratory problem is a weak cough leading to pneumonia, sometimes with fever. In this situation, aggressive treatment with antibiotics and, if needed, a mechanical ventilator for a short time may allow recovery to independent breathing again.

Progressive respiratory muscle weakness usually is identified in one of the following three ways:

- A **temporary problem**, often due to a chest infection, which, when treated (sometimes in a hospital), allows independent breathing again for weeks, months, or longer.
- A **weak cough** that can lead to serious problems, even though breathing is still strong enough; this condition can be effectively improved.
- **Permanent or life-threatening respiratory failure**, caused when the breathing muscles become too weak to expand the lungs and to provide an adequate cough, thus requiring assisted ventilation for survival.

If you are alert to the warning symptoms of respiratory muscle weakness, you can identify the decreasing muscle strength months before life-threatening respiratory failure occurs. Be sure to stay in control and plan your strategy, so you can avoid an emergency or panic situation.

## COMMON RESPIRATORY DISEASES AND LUNG CONDITIONS

Be aware of the following **common respiratory illnesses** and other factors can cause complications in persons with ALS.

- **Bronchitis** is an inflammation and irritation of the bronchial airways (tubes) in the lungs, characterized by a cough. It is often caused by viruses, bacteria, irritants such as cigarette smoke, or allergies. The inflamed airways secrete a sticky mucus which causes productive coughing; if the lungs are not cleared, pneumonia may occur.
- **Pneumonia** causes the very small airways, the **alveoli** (sacs) and surrounding tissues to fill up with pus and mucus which prevents airflow. Proper airflow is necessary for oxygen to be taken up through the alveoli into the blood, and for CO<sub>2</sub> to be released from the blood and ventilated out of the body. Pneumonia can be caused by bacteria or viruses, and may be a complication of a cold, flu, aspiration, or bronchitis.
- **Aspiration** (liquids or solids in the airways and parts of the lungs) can block airways and gas exchange, cause irritation, lead to damage when stomach acids are involved, or carry infection from the mouth and gums. Small amounts of aspirated material can be cleared by the airways or coughed out. If aspiration is frequent or involves large amounts of material, then a medical evaluation is needed, especially by a speech and swallowing specialist, and a pulmonary consultant.

# Keeping Your Lungs Healthy

**I**f you have ALS and take proper care of yourself, you can stay healthy much longer and avoid many breathing complications. The following are some simple guidelines:

- Try not to become underweight because it will weaken your body, muscles, and immune defense system. Avoid becoming overweight, because it can increase the work of the muscles used for inhaling and exhaling and the likelihood of breathing problems during sleep.
- Try to control constipation which can lead to uncomfortable stomach problems and interfere with breathing; this condition usually responds to added fruit, vegetables, fiber, and liquids in your diet, or to drinking a full glass of warm water in the morning. A stool softener may be used and, occasionally, a mild laxative if needed. (Please read Manual 5 for more information on this topic.)
- Breathing and coughing techniques can help maintain healthy lung function. **Deep breaths** help to fully expand the lungs; take five-to-10 deep breaths, with a short rest in between, several times a day. **Effective coughing** is needed to clear secretions. Also, **assisted cough** methods can be very helpful, particularly if you have a cold or chest infection. These techniques will be discussed later.

- Obtain a complete baseline **health examination** from your primary care physician, which should include an office review of your health and any symptoms you are experiencing. Remember to make good use of the time by preparing notes ahead. If you can, bring along a family member or friend to listen to what the doctor says. In addition to your history and physical examination, this visit also may include blood and urine tests and a chest x-ray.

Having a disease such as ALS can make you more likely to develop infections in your lungs and problems with breathing. Some specific suggestions to prevent common respiratory conditions include:

- Try to keep a distance of 30 feet or more from people with a viral cold or bronchitis, who show such symptoms as a runny nose, sneezing, coughing, and/or fever. **Viral respiratory illnesses** are quite contagious, often difficult and may lead to a bacterial infection of the chest or sinuses. To reduce your chances of catching a cold from a person in your home, wash your hands after handling common-use items and avoid touching your eyes and nose.
- Symptoms that suggest a **possible bacterial infection** and should be treated by a doctor include: a fever of over 101° that does not respond within two hours to home treatment; yellow or green nasal or bronchial secretions persisting for several hours; continuous sinus pain; and chest congestion that causes difficulty in clearing secretions or breathing. Prompt use of an antibiotic is recommended for a bacterial respiratory infection, as well as methods to ensure effective coughing. Some people with a tendency to wheeze may be prescribed medication such as Albuterol®, which can be used with a simple metered dose inhaler and a spacer (such as the Aerochamber®).

- Avoid exposure to **dust** and **fumes**. If you smoke cigarettes, stop, and try not to inhale “secondhand smoke.” Stay as *active* as possible, and exercise within your limits.
- Obtain a **Pneumovax immunization** which should decrease your risk for one type of common bacterial pneumonia (**pneumococcal**). An **influenza immunization** should be obtained in October or November every year; while it does not protect you from all colds and flu, it does provide some protection against influenza viral infections which commonly occur in the winter.

### **WHAT ARE SOME EFFECTIVE COUGHING TECHNIQUES?**

When respiratory muscle strength decreases, methods to achieve an **effective cough** can be critical in keeping your airways clear. An effective cough requires a big breath in, coordination of the throat (bulbar) muscles, and rapid airflow out. A good volume of air is needed to push the secretions up the airways. The coordination of the bulbar muscles is also needed to hold the air in, and then suddenly release it, so that peak airflow occurs as you breathe out rapidly. Then, your respiratory muscles need to produce rapid air flow.

If peak cough flow is low, **assisted cough** techniques may be needed, particularly when you have a respiratory infection. If vital capacity (vital capacity refers to the total amount of air that you breathe out, after you have breathed in as much as possible) is low, the cough may be too weak. Some people with ALS can learn to gulp in several breaths, stacking one on top of another, to increase the volume available for an effective cough. Individuals living with polio have learned to use a technique called **frog breathing** to gulp in additional air; if your therapist is not familiar with this technique, you can contact a nearby polio support group, or the International Ventilator Users Network

(I.V.U.N.): call 1(314) 534-0475 or visit their web site at [www.post-polio.org/IVUN.html](http://www.post-polio.org/IVUN.html).

You also can use the **huffing** or **squeeze cough** technique to clear secretions. Huffing is done by taking two deep breaths, and then forcing all the air out of your lungs with a series of huffs, with your mouth and vocal cords open. Repeat this procedure several times to move phlegm from deep down in your lungs upward so you can cough it out. Another person can assist you by applying pressure inward, to your lower rib cage or abdomen, with each cough.

There are additional coughing techniques that you can learn which can be extremely important for preventing pulmonary complications such as pneumonia. These methods assist and augment coughing for people with neuromuscular disease and should be taught by an experienced physical or respiratory therapist.

As respiratory muscle weakness increases, you may find a mechanical device like a cough machine helpful. The Cough Assist<sup>®</sup> device (sold by Respironics) has a mouthpiece and cycles between positive pressure to load your lungs with a deep breath, and then a rapid negative pressure to assist rapid airflow out. The cost of this device is covered by Medicare.

If you and your caregivers learn these techniques, you may be able to continue independent breathing longer, and may be able to avoid pneumonia or postpone the need for tracheostomy (invasive ventilation explained later).

## **HOW CAN YOU DETERMINE HOW WELL YOUR LUNGS ARE FUNCTIONING?**

There is no single correct way to assess lung function for people with ALS. Physicians vary in their approach, depending on the needs of each patient. Since care is complex and time in the doctor's office may be limited, remember to bring a family member along to ensure that all of your concerns are being addressed. Bring notes, take notes, keep a list, and work closely with your doctor.

The following are some specific methods for determining how well your lungs are functioning:

- Arrange for baseline **pulmonary function tests (PFTs)** as part of the initial evaluation. A PFT consists of putting a mouthpiece in your mouth, and nose clips on your nose, and breathing into a computerized machine or simple hand-held device. It is usually performed in a pulmonary function laboratory, in a neurologist's office, or at an ALS clinic. After the baseline tests are done, they can be repeated, as needed, every three-to-six months. These tests are used to determine the strength of your respiratory muscles and how well your lungs are functioning overall. If your lips are weak, a lipseal mouthpiece or mask can be used so your breath is accurately measured.
- Review and discuss the findings with your doctor and neurologist, and pay attention to any other medical problems that might affect your breathing. These include respiratory allergies, asthma, chronic obstructive lung disease sometimes related to smoking, or residual lung changes from past pneumonia. Sinus or dental disease also can increase the chance of lung infections; therefore, have a dental checkup and regularly scheduled cleanings.

- Have your **vital capacity** (VC) checked regularly. Your VC lying down may be less than when you are sitting or standing. If your VC falls to 50% or less of “normal” based on your age, sex, and height, you and your doctor should be concerned; review your options and take action. The American Academy of Neurology’s practice guidelines suggest that if the forced vital capacity reaches 50%, use of noninvasive positive pressure ventilation (NIPPV) should be considered. At 30-35% or less, acute respiratory failure and a life-threatening crisis can occur without any additional warning. However, some patients live for months at this level of respiratory functioning or below it.
- Respiratory muscle strength should be monitored regularly. This procedure requires measuring **maximum inspiratory force** (MIF) and **maximum expiratory force** (MEF) during an optimal effort to breathe in or to breathe out against a closed tube. MIF is related to the strength of your inspiratory (breathe-in) muscles, and MEF to the strength of your cough, which requires a sudden forceful breath out. When these measurements decrease significantly, then an unsafe situation exists.
- A change in **arterial blood gas** (ABG) measurements or in **oximetry** (a simple noninvasive oxygen saturation test) is another way to assess lung function; however, this change usually occurs very late, often when you are in crisis. If there is no other lung disease, a fall in **arterial blood oxygen saturation** (SaO<sub>2</sub>) usually indicates weak respiratory muscles and decreased ventilation.

- **Peak expiratory flow rate (PEFR)** is used to measure the maximum airflow you can generate after taking a deep breath; you blow out quickly and as hard as possible for one-to-two seconds. It tests lung function, coordination, and your muscle power. **Peak cough flow** is done to measure cough. It is done both with your cough and using manual assisted cough methods.
  
- A significant abnormality in **nighttime breathing function** can cause a fall in oxygen saturation and a change in heart rate and rhythm. Overnight measurements at home, using a portable oximeter with an eight-to-10-hour memory (**nocturnal oximetry**), can give very helpful information. A drop of oxygen saturation during sleep, to less than 88% for at least five continuous minutes reflects muscle weakness and is an indication for action. A complete study at a sleep laboratory is sometimes advised. However, a good clinical evaluation, nocturnal oximetry plus the simple tests of lung function, will often be satisfactory.
  
- **Home monitoring of pulmonary function** can be done with either of two simple, inexpensive devices: an **incentive spirometer** and a **peak flow meter**. Incentive spirometers are usually used by hospital patients to make sure they breathe deeply in order to keep their lungs well-expanded and secretions cleared out after surgery; there is an indicator attached to see if your breathing-in ability is steady or decreasing. A peak flow meter, which measures PEFR, is frequently used in medical offices and emergency rooms as a simple measure of breathing and cough effectiveness, and is often used for home monitoring by people with asthma as well.

# Adapting to Breathing Problems

T

his section addresses the general process of making decisions about your life and health and then, more specifically, discusses the decisions related to ALS respiratory complications and breathing options.

## **PLANNING IS A “MUST”**

It is normal to become worried after you learn of the ALS diagnosis. At first, many people find it hard to stay organized and in control of their lives. A difficult period of adjustment may occur; however, before long, most people can focus again on living. Learn how to live with your medical problems and to stay mentally and vigorously engaged. Do not give in or give up. If you feel depressed or anxious, counseling or psychotherapy and/or the use of medications to control your depression may help you. If you feel overwhelmed, you may fail to get enough information and then neglect to plan your life adequately. It is best to plan ahead so that you do not have to make plans during a crisis.

The *first step* is to get information about assisted breathing options and discuss it. Understanding something new usually requires hearing about it several times, especially when it concerns your own health. So, involve a friend or family member to help you remember the details better. Also, talk to other persons with ALS and their families, or to an ALS support group leader. You may want to read as much as possible, review the very good educational videotapes available, and check reliable Internet sites about ALS.

The *second step* is to be sure you consider all the options for ventilation and understand what is involved—the risks, advantages, costs, and burdens. Carefully consider the advice of your doctor and other health professionals.

The *final step* is deciding whether or not to use mechanical ventilation. Repeated discussions should provide a good understanding of the options, the resources available, and the pros and cons. When planning is done carefully, the decisions tend to be more valid and appropriate. However, it is important to review them from time to time because your needs or wishes may change as time goes on.

If you choose to use mechanical ventilation, there is an additional important step: You and your family, or other caregivers, need to learn the needed skills of managing the mechanical ventilation, so that you can do the procedures competently, especially when there are problems or during an emergency. Even better, if possible, would be a trial period at home.

Note that in the United States, it is ethical and legal for an adult to use a ventilator, and then stop its use, if it is no longer desired. Instead of a ventilator, you can choose to have medications available at home to take as needed for comfort, when breathing becomes very weak.

## **COMMUNICATE YOUR DECISIONS**

Once your plans and wishes are in order, be sure your doctors know and agree to honor them by completing an advance directive. A form designed to tell your doctor who can make decisions for you if you cannot make them for yourself is the **Durable Power of Attorney for Health Care (DPAHC)**. The DPAHC designates someone to represent you if you are ever unable to express your wishes. The selected individual(s) must know your wishes and agree to support them.

# Ventilation Options

**M**

echanical ventilation involves using a portable breathing device that assists your lungs and helps you to breathe. It does not prevent progression of ALS, but it does allow you the freedom and flexibility to move around and not worry about your ability to breathe. This type of equipment can be mounted on a shelf of a wheelchair, with a battery, so you can go almost anywhere. Mechanical ventilation can be used non-invasively or via a tracheostomy, depending on your needs.

## **NONINVASIVE POSITIVE PRESSURE VENTILATION (NIPPV)**

**Noninvasive positive pressure ventilation (NIPPV)** involves the use of interfaces such as a nasal mask or nasal pillows (explained under Nasal Interfaces) with a small portable ventilator. Interfaces are the devices that fit over your nose, mouth and face and connect to the mechanical ventilation. The advantage of noninvasive ventilation is that you have the opportunity to try this type of assisted breathing without committing to a more permanent option. It is important to work with an experienced respiratory therapist, which gives you the opportunity to try assisted ventilation, get hands-on experience, and then make further decisions based on the trial at home. You may stop or continue its use at this point.

The **negative pressure ventilation** method started with the **iron lung** that saved many lives during the polio epidemic of the 1950s; ventilation occurs by pumping air out of the tank and back into it, in a preset cycle, which creates a partial vacuum (negative pressure) in the space around the person and causes the chest and lungs to expand.

There are centers in the United States that specialize in pulmonary rehabilitation and mechanical ventilation and are experienced in using negative pressure ventilators; however, these devices are not used frequently today for people with ALS. Newer **positive pressure ventilation** (PPV) methods for NIPPV have been developed that are very effective and allow more mobility.

Assisted ventilation using a mouthpiece, nasal mask, or nasal pillows can be provided with a small portable ventilator, such as a **volume ventilator** or a **bilevel ventilator** (such as the BiPAP® used frequently by people with ALS). Some new ventilators include both the volume and the bilevel options, and improved safety and portability features.

NIPPV should be the first consideration, whenever possible. It is easier to arrange, care is less complex, there are fewer complications, and overall cost is much lower than tracheostomy-PPV. (This option will be discussed later.) Start NIPPV early, before life-support ventilation is needed. The most frequently used method is nasal-PPV.

## **Nasal Interfaces**

**Nasal interfaces** are specially designed nasal masks, or devices that fit into the nostrils called nasal pillows. Many types of masks are now available, including those that are custom-fitted. They were first developed for people with obstructive sleep apnea, and were then used or adapted for assisted ventilation. These methods can be successful alternatives to invasive (tracheostomy) ventilation. Nasal-PPV is a major advance that is continually being improved and utilized more often, as medical centers and respiratory home care companies gain experience. Try different masks to find which are comfortable, do not irritate your skin, and allow effective ventilation without excessive air leaks. The technique of nasal-PPV requires some practice for regular use, and many people with ALS alternate between a nasal mask and the nasal pillows. Some people need to try different bilevel and volume home ventilators (as well as various nasal masks) until they find the system that works best for them. The optimal system should be comfortable, relieve respiratory symptoms and maintain your oxygen saturation (checked with oximeter) at 95% or higher without any added oxygen.

## **Oral Interfaces**

**Mouth interfaces** use a small portable ventilator to deliver a set volume of air through a simple mouthpiece or a lipseal (mouth-seal) held securely between the lips. The mouthpiece can be strapped to your head, especially if used at night, so it stays in place. You may combine daytime use of a mouth interface with another noninvasive method of ventilation at night. A mouthpiece is very useful in the case of nasal congestion due to a cold or nasal allergy.

It is important that you select the best device for you. There are many models available that allow you to be at home, in the community, and at work and to travel. Nasal or mouth interfaces can use a small volume or bilevel ventilator or one which has both options. Your respiratory therapist can review the details related to equipment options with you.

The benefits of using NIPPV are:

- the ability to cough effectively;
- little or no problem with aspiration;
- the ability to communicate effectively (by any effective method, speech is not necessary); and
- the ability to breathe without the ventilator for at least an hour at a time (some people are completely ventilator dependent 24 hours/day).

NIPPV improves daytime breathing, improves sleep, and alleviates respiratory symptoms following nighttime use. Today, there is a variety of excellent nasal masks available, in addition to the nasal pillows. The advantages of this treatment are:

- the nose and mouth offer convenient routes for the delivery of NIPPV;
- it is more acceptable and easier to use than a tracheostomy; and
- it is noninvasive and portable.

The challenges in using NIPPV are:

- finding the best mask, fit, and strap adjustment requires the therapist and patient to work together over time;
- custom-fitted silicone-molded nasal masks, which may provide a better seal at higher ventilator pressures, require additional time and expertise;
- some people with ALS are unable to adapt;
- if bulbar impairment is severe, NIPPV may be more of a challenge to use; and
- not all medical centers have had experience with these devices.

Side effects of nasal-PPV include skin irritation or abrasions, dryness or inflammation of the nose, inflammation of the eyelids due to air leaks, swallowed air traveling down into and distending the stomach. Some people have excessive mouth air leakage during sleep, which can result in inadequate nighttime ventilation. This situation may be improved by using chin straps. Monitoring overnight oximetry can be done to be sure ventilation is effective: oximetry oxygen saturation should be 95% or greater (without using added oxygen).

### **BiPAP-S/T<sup>®</sup>**

The **BiPAP-S/T<sup>®</sup> Ventilatory Support System**, used for nasal-PPV, was developed specifically for mask or nasal pillow application for non-life-support ventilation of adult patients. This was the first bilevel device. There are now other models and other brands available; some have different features. BiPAP-S/T<sup>®</sup>, and other bilevel equipment, deliver two different levels of pressure: one during inspiration (IPAP) and a lower pressure during expiration (EPAP). “IPAP” means inspiratory positive airway pressure. “EPAP” means expiratory positive airway pressure. This bilevel setup may enhance comfort by lowering the average airway pressure delivered, as well as compensating for most mask leaks. These systems are small, lightweight, portable, and easy to

operate and maintain. The disadvantages of some of the bilevel ventilators are that they lack alarms, do not have an internal battery, and may cause uncomfortable nasal and oral dryness. Some new bilevel ventilators have more advanced features, have better safety features and are approved for 24-hour use.

### **Nasal CPAP**

**Nasal CPAP** is *not* usually helpful for people with ALS, but it is considered the most effective noninvasive treatment for obstructive sleep apnea. CPAP helps to keep the airways in the lungs open, acting like an air splint, but not directly assisting respiratory muscle function. When there is respiratory failure with increased CO<sub>2</sub> in the blood, the use of CPAP alone is usually inadequate. A CPAP machine requires fairly strong respiratory muscles and could aggravate your ability to breathe. Physicians who specialize in neurology, pulmonary medicine, or respiratory care are familiar with nasal CPAP equipment. (Nasal-PPV is true assisted ventilation, *not* just continuous airway pressure, such as nasal CPAP.)

### **Other Noninvasive Methods for Assisting with Ventilation**

There are many other helpful devices that are not used very often nowadays except at centers that specialize in pulmonary rehabilitation. Although they were popular during the previous 30 years, now nasal-PPV is relatively easy and very effective in comparison. Thus, the other methods are only briefly covered below:

- **Tank ventilator** (iron lung) applies intermittent negative pressure to the body below the neck.
  
- **Porta-lung** is a smaller fiberglass version of the iron lung.

- **Raincoat ventilator**, pneumowrap, or Emerson wrap works on the same principle as the iron lung. It encloses most of the body in a plastic wrap envelope and is connected to a negative pressure ventilator which cycles air out of and into the plastic wrap.
- **Chest cuirass** (chest shell) is a fiberglass “turtle shell” fitted across the chest and upper abdomen with two Velcro® straps running around the back. When air is sucked from the shell through a hose attached to a small negative pressure ventilator, the chest expands and air is inhaled.
- **Exsufflation belt** (pneumobelt) works by intermittently pressing on the abdomen to augment the breathing-out phase. The patient must be in the sitting or standing position.
- **Rocking bed** tilts your body back and forth 13-to-19 times per minute. The abdominal organs and diaphragm slide back and forth, as your body tilts, assisting ventilation.
- **Frog breathing** (glossopharyngeal breathing) is a technique discovered in the 1950s by people with respiratory muscle weakness due to poliomyelitis. Larger breaths are achieved by gulping air into the lungs using the tongue and throat muscles. This can improve breathing and also improve effective cough. It is a useful technique to learn if there is no bulbar impairment.

## TRACHEOSTOMY

Tracheostomy-ventilation offers an effective method of breathing for people with ALS. A tracheostomy is a surgical opening (a tubular passageway) into the trachea (the windpipe, located in the lower front part of the throat) into which a plastic or metal tracheostomy tube is placed. This method of ventilation is considered invasive because it involves a surgical opening which assists in breathing. When necessary, a small suction tube can be inserted into this opening to remove secretions (suctioning), or a mechanical ventilator can be connected to the tube. Eating and speaking can continue, if there is no pre-existing speech or swallowing problem, but often they may require extra effort. A tracheostomy speaking device, such as the **Passy-Muir**<sup>®</sup> valve, can improve the abilities to talk and swallow.

Many individuals who choose this option are satisfied with their quality of life, particularly if they can live at home. Some have used a ventilator for over 15 years at home, and would have lived even longer except for developing other serious medical problems. Family members usually support this choice, even though the burden of caregiving is heavy.

In the United States, some 2% of people with ALS select mechanical ventilation following advance planning. When their doctors have had experience with this treatment option, the percent is higher. In the past, most people used long-term invasive ventilation without advance planning as a result of emergency hospitalization. "It just happened."

Stay in control so you and your family can choose what is best for you. **Do not let an emergency decide for you.** Life-support mechanical ventilation indicates that it is used almost 24 hours a day, that independent breathing is possible for only very short periods or not at all, and that the person would not be able to live without it.

When it is no longer desired, any treatment including a ventilator can be removed, and medications can be taken to ensure comfort. Many people use the ventilator only as long as their ability to use noninvasive ventilation is possible. Others want to continue on mechanical ventilation permanently.

**What are the advantages of a tracheostomy?**

- It provides a small, secure connection to your airway for suctioning and for mechanical ventilation if needed.
- Nebulized medications (sprays or mists) and oxygen can be delivered through it.
- It is the method of choice for mechanical ventilation, when noninvasive ventilation is no longer adequate, secretions cannot be well managed, and when life-support mechanical ventilation is desired.
- It leaves the face free and avoids headgear, straps, and skin pressure problems. If desired it can be covered with a scarf so it is not obvious.
- Doctors, nurses, and respiratory therapists are very familiar with tracheostomy care.
- For people who have little independent ability to breathe, tracheostomy-ventilation may be a safer system and has been found to provide much longer survival.

### **What are the disadvantages of a tracheostomy?**

- Some people feel it is too invasive, increases their disability and dependence, makes them look less “normal,” and might prevent them from wearing certain articles of clothing (collar or tie).
- Secretions require suctioning, often at night as well as during the day.
- Coughing to clear the smaller airways is difficult.
- The tracheostomy site (**stoma**) can become infected, bleed, or develop inflammatory tissue (**granulations**) that needs to be removed.
- It requires more skill and makes care more complicated.
- Some people have resulting difficulty with speech and swallowing.

### **Tracheostomy Positive Pressure Ventilation**

Tracheostomy positive pressure ventilation (tracheostomy-PPV) is often best for people who need nearly 24-hour-per-day life-support ventilation. Some people use tracheostomy PPV for only eight-to-12 hours a day. Indications for a permanent tracheostomy-PPV include: inability to use noninvasive ventilation, poor control of symptoms or persistent elevated levels of CO<sub>2</sub> when using noninvasive ventilatory assistance; poor cough and inability to clear bronchial secretions; problems with aspiration; and deterioration due to neuromuscular disease, so that independent, spontaneous ventilation becomes intolerably brief.

Tracheostomy-PPV can be used at night, and/or during the day, or continuously, usually with one of the small portable volume ventilators. The rate (number of breaths per minute) can be either set or triggered by the person. The advantages of tracheostomy-PPV include its effectiveness; the simple and convenient

equipment; ventilators that attach to wheelchairs or other mobile devices (e.g., carts, pedestals); safety factors; improved long-term survival; and its familiarity to health care professionals trained in hospital intensive respiratory care. The disadvantages are related to the tracheostomy itself.

### **Tracheostomy and Ventilator Speaking Valves**

A tracheostomy can affect your speaking ability. Yet, you can have satisfactory speech, if a small air leak is created around the tracheostomy tube cuff by partially or completely deflating the cuff, or by using an uncuffed or fenestrated (with openings) tracheostomy tube. Then air moving up around the tracheostomy tube, to the vocal cords and mouth, enables speech. When a cuffless tracheostomy tube is used, or when the cuff is kept deflated, check that nighttime ventilation is adequate. Occasional monitoring with overnight oximetry can be done at home.

The Passy-Muir<sup>®</sup> speaking valve makes use of a one-way valve attached to a cuffless tracheostomy. It augments airflow on exhalation through the vocal cords and upper airway to enable speech and also clears secretions. The Passy-Muir<sup>®</sup> valve, which must be prescribed by a doctor, is the only one that can be used in-line with the ventilator. Benefits include spontaneous speech, a louder voice with more control for sentence structuring, hands-free speech, better hygiene at the site of the stoma, good management of secretions, and improved swallowing. A respiratory care practitioner familiar with this valve needs to assess the safety of the system and adjust the ventilator settings to compensate for any air leak. Consultation by a speech and communication specialist, who is experienced with a tracheostomy or ventilator, is recommended.

There are other talking tracheostomy tubes and speaking valves available, but they are not used in-line with the ventilator. These alternatives also should be discussed with your respiratory care practitioner. In some cases, when the muscle weakness of ALS limits speech, an **augmentative or alternative communication (AAC)** system may be needed. Since many are available, you should obtain an evaluation by a speech-language pathologist familiar with these systems. There are simple, inexpensive devices, as well as very sophisticated portable communication devices, with high-quality synthesized speech (discussed at length in Manual 5).

## **THE NEED FOR OXYGEN**

Oxygen is generally used when the lungs are diseased, which is in contrast to using mechanical ventilation for respiratory muscle weakness due to ALS. Usually if the oxygen saturation is low, a person with ALS is having under-ventilation from respiratory muscle weakness and needs properly adjusted assisted ventilation rather than oxygen. Sometimes with bilevel ventilators the IPAP is set too low to achieve optimal ventilation. Oxygen may be needed if there is also chronic lung disease, pneumonia, aspiration, or for comfort when long-term survival is not the goal. Oxygen may be delivered with devices such as nasal prongs, or connected to a tracheostomy site, oxygen mask, or any type of ventilator. Home oxygen equipment includes electric oxygen concentrators, compressed tank oxygen, and liquid oxygen systems (which are easily portable for outside use). Oxygen may be prescribed by a physician if it is medically necessary, based on ABG (arterial blood gas) measurements made when a person is medically stable. People with ALS who develop pneumonia may need oxygen, and it may sometimes be part of hospice care.

Oxygen is usually administered continuously, 24 hours per day; the amount needs to be carefully regulated at a specified flow rate liters per minute (lpm) or concentration (percent). Too little will not provide the oxygen needed by body tissues; too much can sometimes depress the breathing control center and lead to respiratory failure and collapse. Oxygen should not be used for shortness of breath or respiratory symptoms when the ABG level is satisfactory. When there is respiratory muscle weakness due to ALS, and too little ventilation as a result, the CO<sub>2</sub> level increases, and the oxygen level drops reciprocally; in other words, the low oxygen is due to under-ventilation. The treatment for under-ventilation due to weak muscles is *not oxygen*, but instead assisted ventilation to bring down the CO<sub>2</sub>—which brings up the oxygen level.

### **Safety**

Anyone using equipment to assist their breathing should review safety issues carefully. There can be equipment problems or problems due to a mistake by the user, or caregiver. You should have, and know how to use, a back-up Ambu-type resuscitator bag. If you require the ventilator for 20 to 24 hours per day, and have little ability to breathe independently, you should have a second complete ventilator system available for safety and for mobility. Every possible problem, and the required action to correct the problem, should be identified, written down, and used for regular safety drills. Some ventilators have more safety features as well as an internal battery plus easy connection to an external battery. You will need to balance convenience and safety. For example, sometimes a ventilator's warning alarm can be life-saving, but sometimes it produces nuisance false alarm signals. Remember, if you have very little ability to breathe independently be extra careful; regular safety drills are essential for everyone involved in your care. When safe long-term survival is your goal, and you need to use the ventilator for 20 to 24 hours per day, discuss with your doctor the pros and cons of noninvasive versus tracheostomy ventilation.

# Making a Decision about Ventilation

**M**

aking a decision about ventilation can be difficult, but if you plan well and consider all of your options in advance, you will feel prepared and in control. In this section the various options and treatment strategies for assisted breathing will be discussed. The following is a list of options for you to consider regarding mechanical ventilation.

- **Decide not to use mechanical ventilation at all.** Instead, use 1) any treatment for comfort and/or 2) medication to relieve respiratory distress or other problems, as needed. Consider requesting hospice care when respiratory symptoms begin.
- **Decide to use mechanical ventilation only on a temporary basis,** if there is a good chance of resuming independent breathing. An example is when you are doing well but develop a chest cold, and then pneumonia. If intensive treatment can return you to your level of function and quality of life before the respiratory infection, then spending a few days in an intensive care unit (ICU) with a ventilator might be worthwhile, especially if afterwards you would likely resume fully independent breathing.

■ **Decide to use mechanical ventilation on a regular basis.**

Deciding to use NIPPV is a different level of decision-making than is deciding to have a tracheostomy and long-term mechanical ventilation. With NIPPV, you can choose when you use it and you can take the interface on and off yourself (unless hand and arm weakness prevents this). The need and costs for in-home caregiving are significantly less than are required with tracheostomy ventilation.

Tracheostomy ventilation might be especially appropriate if you are very active and have few limitations due to ALS. However, after discussions with their doctors, most people want to establish some special *limitations*, such as under certain conditions (permanent coma) discontinuing all life support (including mechanical ventilation and tube feeding). Many patients with ALS choose to use only NIV (noninvasive ventilation) and decide not to have a tracheostomy for mechanical ventilation or suctioning. Many people can use a nasal mask, for example, for assisted ventilation over months or even years, depending on individual factors. Other people with ALS decide they want to use tracheostomy-ventilation, particularly when nasal-PPV is not possible. If long-term survival is desired, and the caregiving and other resources are available, tracheostomy-ventilation should be planned.

## **WHAT FACTORS SHOULD BE CONSIDERED WHEN DECIDING ABOUT A TRACHEOSTOMY?**

After reviewing the information you need and the advice from your doctor, you may be ready to make a decision about mechanical ventilation. However, you should consider family and friends, available resources, and the risk of long-term institutional care.

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 mechanical ventilation, and all other appropriate treatments, can live for years. An example is Jack, who lived seventeen years with home mechanical ventilation (HMV). His 24-hour care was an extraordinary effort for the family, even though paid caregivers were also available to help. Jack was satisfied with his quality of life and his decision.

With assistance from a social worker, evaluate the resources available to you and how to apply for disability and other benefits. You can determine what you need for home care, and then examine your medical benefits, community resources, and personal and family assets. When the caregivers or resources are not available, using a mechanical ventilator at home may not be practical. However continued innovative efforts to find assistance may make it possible. Moreover, if home care is not an option,

some people find long-term institutional care acceptable, such as a skilled nursing facility, while others would rather not plan on mechanical ventilation if they cannot be at home.

Some questions which may help you decide on tracheostomy long-term mechanical ventilation include:

- Is it advised by your doctors?
- Is it desired by you and your family?
- Do you have the necessary resources available, including caregivers' help?
- Are you, your family, and/or friends able to learn the needed skills?
- Can you afford the cost of long-term mechanical ventilation?
- Would you be able to live at home, or need to be in a nursing home?

This decision is very personal. It may be based on practical realities, consideration of how your decision will affect the lives of your family and friends, or your perception of a reduced quality of life. Your health care team and hospice can help you and your family plan, so that as respiratory function declines, you will be comfortable, without pain or suffering.

## **HOME MECHANICAL VENTILATION (HMV)**

You may want to consider the following issues in deciding if home mechanical ventilation is a choice that you want to make.

- You are highly motivated and engaged in living.
- Your medical problems are stable or progressing slowly.
- Your level of independent functioning allows some ADL (activities of daily living).
- Your ability to communicate is good, 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 in your care, if needed.
- You have financial resources available for equipment and caregivers.
- You have access to an experienced, multidisciplinary, professional healthcare team.
- You are able to use NIPPV on a trial basis first.

Many people with ALS who have chosen tracheostomy mechanical ventilation report that they have a very good quality of life. There is no right or wrong decision – it is a personal choice.

Some people evaluate the following factors in making a decision about tracheostomy ventilation:

- Your disability is advanced.
- Your ability to communicate effectively is very limited.
- Your interest in or motivation for living is low.
- You are not mentally alert or self-directed.
- You do not have caregiver support available from family or friends.
- You do not have the resources needed to be at home.
- Your health care professionals have little experience with HMV.
- You are unable to use NIPPV on a trial basis.

### **Potential Challenges with HMV**

- Quality of life can become marginal with a perceived loss of dignity.
- About 10% of people with ALS eventually will develop a totally locked-in state (inability to communicate in any way) on long-term tracheostomy ventilation.
- There is a possibility of being institutionalized in a nursing home.
- Family members' 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.

### **WHAT ARE THE EXPENSES INVOLVED?**

Some or most of the costs may be absorbed by health insurance coverage, but some costs may be your responsibility. Know what your out-of-pocket expenses will be. Costs vary considerably depending on where you live; therefore, you should review the following information with a medical social worker who can help you evaluate your options.

- Equipment rental is required for one or two ventilator systems, external battery, wheelchair adapted for a portable ventilator, suction equipment, hospital bed, and a lifting device for transfers. Although in the long run renting is more expensive than buying, most companies will include 24-hour emergency response, maintenance, retraining, and other important services in the rental cost.

- **Long-term mechanical ventilation** is rarely provided in a hospital. The main alternatives are to be at home or in a skilled nursing facility. The cost and intensity are much less for a person with ALS using noninvasive ventilation compared to tracheostomy-PPV. With NIPPV, the major expense is renting equipment.
- Professional “intermittent” visits from a home health agency are usually covered initially. In the past, health care insurance coverage usually stated that nurses may qualify as a covered benefit; this is no longer standard practice. Paid caregivers, particularly nurses, are the most expensive part of home care. Private duty nursing care, a practical/vocational level nurse, is costly and may be required 16 to 24 hours a day (while family members work or sleep). More commonly personal attendants are hired directly by a family (not from an agency).

An important option for home health care is **personal attendant services** (or assistance provided at home by **unlicensed caregivers**) which is less expensive than licensed health care providers. In the past, insurance benefits usually did not cover unlicensed caregivers, but recent changes permit some use of trained attendants as caregivers. Yet, overall, most health care benefits do not cover the cost of paid caregivers—nurses or attendants.

Many residential living facilities have limited experience caring for people with ALS using ventilator support. Some nursing homes do care for people using mechanical ventilation, often in subacute care units. Nursing home costs are usually paid for a limited period of time by Medicare or private insurance; or on an on-going basis by Medicaid.

## The Last Word

This manual has reviewed how to stay healthy and avoid breathing problems as long as possible. Each person with ALS is unique with many different possible patterns of progression of the disease and differences in preferences and living situations. There is not one single choice that is best for everyone. You need to go over this information a number of times to become familiar and comfortable with the details and the options. Talk to your doctors and to others with experience. Carefully consider your physician's advice and then determine what you and your family prefer, and whether or not the needed resources are available. Try not to procrastinate; you and your family must consider all aspects of assisted ventilation carefully *before* it is needed.

A major advance in ALS care is using a home trial of NIPPV before a crisis occurs. It is best to start when some respiratory symptoms are present and either the VC is decreasing toward 50% or there is a drop of oxygen saturation during nighttime sleep to less than 88% for at least five continuous minutes. This type of noninvasive ventilatory support can be stopped if it is not desired. In some people bulbar impairment may become severe enough so that an invasive tracheostomy will need to be considered. Several alternatives to support ventilation are available. Persons with ALS now can continue living "well" for years after respiratory weakness if they wish, with planning and resources. Remember, it is *your* choice.

# Notes

The following is a list of the topics covered in the *Living With ALS* manuals:

*Manual 1*

**What's It All About?**

This manual provides an overview of ALS, what it is, and how it affects your body. It also provides information on what kinds of resources are available to help you deal with ALS more effectively.

*Manual 2*

**Coping with Change**

This manual addresses the psychological, emotional, and social issues that you must face when your life is affected by ALS. It provides information on how to cope with the many lifestyle changes and adjustments that occur when you live with ALS.

*Manual 3*

**Managing Your Symptoms and Treatment**

This manual discusses the symptoms that affect you when you have ALS and how to treat them. It also covers the most recent breakthroughs in medications and how these treatments can improve the quality and duration of your life.

*Manual 4*

**Functioning When Your Mobility Is Affected**

This manual covers the full range of mobility issues that occur with ALS. It specifically discusses exercises to maximize your mobility, as well as how to adapt your home and activities of daily living to help you function more effectively.

*Manual 5*

**Adjusting to Swallowing and Speaking Difficulties**

This manual addresses swallowing difficulties and how to maintain a balanced diet with ALS. It also covers how speech can be affected by ALS and the specific techniques and devices available for improving communication.

*Manual 6*

**Adapting to Breathing Changes**

This manual explains how normal breathing is affected by ALS. Specifically, it explains how to determine if you have breathing problems and what options are available to assist you as your breathing capacity changes.

*The information contained in this manual can be very valuable to people living and dealing with ALS. Please donate this manual to your local library if you no longer need it.*

## A Reason for Hope



The Amyotrophic Lateral  
Sclerosis Association

DC/MD/VA Chapter  
615 S. Frederick Ave., Suite 308  
Gaithersburg, MD 20877  
TEL: 301/978-9855 (Outside DC Metro area: 1-866-348-3257)

Fax: 301/978-9854  
[www.ALSinfo.org](http://www.ALSinfo.org)

The ALS Association is the only national not-for-profit voluntary health organization dedicated solely to the fight against amyotrophic lateral sclerosis (often called Lou Gehrig's disease) through research, patient and community services, advocacy, professional education and public awareness.

Member of the National Health Council and Community Health Charities (CHC)