

## Section H

### RESPIRATORY CARE

At some time in the course of the disease, ALS may affect the bulbar nerves. These nerves control the movement of the muscles related to swallowing, speaking, coughing, and keeping the airway open for breathing. As the nerve cells of the bulbar nerves degenerate, the muscles waste away. In some patients, respiratory muscles may be affected late in the course of the disease; in others, these muscles are affected early in the disease process.

Because of weakened respiratory muscles, patients with ALS have problems inhaling enough air and exhaling enough carbon dioxide. The carbon dioxide level builds up in the blood, and the oxygen level drops. This problem may not be obvious, but a variety of symptoms can indicate that breathing is affected. Treatment for breathing problems depends upon the cause of the problem. Additional testing may be needed to determine the underlying cause in a particular patient.

#### **PROBLEMS**

Signs of breathing problems include morning headaches, daytime sleepiness, sleeping poorly, waking frequently, snoring, noisy breathing, poor appetite, shortness of breath when moving or when lying flat, and nausea without such other gastrointestinal symptoms as vomiting or diarrhea. Other symptoms of poor breathing at night can include increased irritability, forgetfulness, and apathy. If any of these symptoms appear, notify the Clinical Nurse Coordinator for the Lois Insolia ALS Center to discuss possible solutions.

#### **How Is Breathing Monitored?**

Breathing problems can be monitored during regular Center visits by tests that measure the strength of respiratory muscles and how well the lungs are functioning. The patient can exhale into a spirometer, a device that indicates how the ability to exhale, or Forced Vital Capacity (FVC), compares with that of most people of the same sex, height, and age. This test is performed on a regular basis at the Lois Insolia ALS Center visit. Other measures of breathing issues include an oximeter, which is a device that noninvasively measures arterial blood oxygen saturation noninvasively using a comfortable finger sensor. The measurement of arterial blood gases (ABG) is another test that can determine the severity of breathing problems. A small amount of

blood is taken from an artery, and the amount of oxygen and of carbon dioxide in the blood is compared to the normal ranges.

For severe symptoms of breathing difficulty, especially in nighttime breathing, a sleep study might be needed to help find out the specific cause of the problem. In a sleep study, breathing, oxygen levels, leg movement, and sleep cycles are measured while the patient is asleep. This information can help determine the best course of treatment, whether it is medication or the use of a noninvasive assistive breathing device. If the ALS physician determines that a sleep study is appropriate, the Clinical Nurse Coordinator can assist in arranging to have this study performed at a hospital sleep lab or at home.

### **NONINVASIVE SOLUTIONS**

Difficulty in breathing while lying flat may be relieved with the use of a wedge pillow or a hospital bed set so that the patient lies at a 30–45 degree angle. At this angle, the main muscle of breathing, the diaphragm, does not have to work against gravity, as it does when lying flat. Patients who have a reclining chair may find relief by sleeping in that, as it allows a patient to lie at the appropriate angle for maximum breathing comfort.

### **BiPAP**

The most commonly used device to assist nighttime breathing in ALS patients is a BiPAP, which stands for Bi-level Positive Airway Pressure. BiPAP is not a ventilator or “respirator,” and it is not invasive. It may stimulate a breath, but it does not “breathe” for the patient as a ventilator does. BiPAP is a supportive device that can help ease breathing difficulty and its symptoms. Use of a BiPAP can improve nighttime sleep, resulting in decreased daytime fatigue and sleepiness, and increased energy levels. It is not limited to nighttime use; many patients use this device during the day, particularly while napping.

BiPAP is a machine that works by providing pressure through a mask worn over the nose and mouth or through a nasal cannula. The mask is similar to the kind one might use to receive oxygen in the hospital. The machine provides pressure when the patient inhales and less pressure when the patient exhales. This helps the patient draw in oxygen and expel carbon dioxide. The machine also has a *back-up rate*. If it does not count a certain number of breaths taken by the patient in one minute, it will stimulate the patient to take

an “extra” breath. The amount of pressure the BiPAP is set to provide is determined by the doctor and is based on body size. A Respiratory Therapist will help make the use of the machine comfortable.

### **Does Oxygen Help in ALS?**

Oxygen is used to treat conditions where oxygen levels are low. Patients with ALS do not usually have significantly low oxygen levels in the blood, unless they have a lung or heart problem in addition to ALS. However, as the muscles that assist with breathing weaken, it is more difficult to push carbon dioxide out than it is to take the oxygen in. Therefore, patients with ALS do develop higher than normal levels of carbon dioxide in the blood because of poor gas exchange caused by the disease. Over time, this causes the respiratory center, the part of the brain that controls involuntary breathing, to function improperly.

The use of high levels of oxygen in ALS can actually cause breathing to slow and even stop. When the level of carbon dioxide in the blood is high, control of breathing shifts from the brain to alternative chemical receptors, which are cells that are sensitive to the presence of certain chemicals. These cells are located in the carotid artery in the neck and in the aorta, a blood vessel in the chest. The alternative receptors become accustomed to telling the body to breathe under the condition of high levels of carbon dioxide. If high levels of oxygen are then introduced, the alternative receptors stop working. Then neither the brain nor the alternative receptors tell the body to breathe. ALS patients don't usually get extra oxygen because it isn't needed unless there is a heart or lung problem and because it can knock out the body's involuntary drive to breathe.

### **What Can Help with Thick Secretions, or Phlegm?**

Many patients with ALS develop a problem with thick secretions, or phlegm, in the back of the throat. This can cause some increase in swallowing and breathing problems. The problems can develop for several reasons.

**Dairy products** The protein in dairy products causes an increase in thick secretions in the throat. If someone with ALS is regularly ingesting large quantities of dairy-based products, decreasing such intake may help reduce the thickness of mucus in the back of the throat.

**Dehydration** Hardly anyone actually takes in enough fluids during the day to maintain adequate hydration of the body. The basic recommendation is 8 cups (64 ounces), or 2 liters (2000 cc) of fluid a day. It is usually difficult for anyone to drink that much fluid, and it is especially difficult when there are problems swallowing or getting to the bathroom. Many ALS patients take in only 400-500 cc of fluid a day. Increasing the daily fluid intake by even a glass or two can help make secretions thinner and easier to handle. Water is the preferred fluid, although anything that does not contain salt or caffeine may also help.

**Low humidity** Home heating and air conditioning rob the air of moisture. Even if the furnace has a built-in humidifier, it is not usually adequate for the needs of someone with ALS. To help reduce secretions and relieve dry mouth, use a room humidifier, either hot or cold, to add extra moisture to the air. Place the humidifier in the room where the person with ALS spends the majority of his or her time, and run it 24 hours a day.

**Medications** Some medications can help to thin the secretions if the suggestions listed above are not effective.

**Robitussin** (plain, no initials) One to two teaspoons may be taken every six hours as needed to reduce or thin secretions. If eating is a problem because of the secretions, Robitussin taken one to one-and-a-half-hours before eating can help thin the mucus. If secretions are a problem during sleep, the medication can be taken one-and-a-half hours before bedtime.

**Nebulized saline** A Nebulizer is a device that turns a liquid into small particles that can be inhaled through a mouthpiece or face mask. Nebulized saline, or salt water, can thin secretions enough for them to be coughed or suctioned out. This can be done every four hours or before meals or bedtime. The pharmacy will require a prescription for the saline and for the Nebulizer.

**Nebulized medication** Saline is tried first because it does not cause any side effects, but sometimes it becomes ineffective. Then a medication must be used to thin the secretions. The medication that is prescribed is called Mucomyst. It is used in the same way as the Nebulized saline.

**Suction** A suction machine can be used in the home. A battery-operated portable suction machine can be obtained for use when traveling. Suction machines use a catheter, a tube similar to those used at the dentist's office, to remove secretions from the mouth and the back of the throat. If a person with ALS cannot cough effectively or spit out secretions, a suction machine should be ordered to help clear the secretions. The Lois Insolia ALS Center Clinical Nurse Coordinator can help obtain the suction items.

## **VENTILATORS**

A ventilator, or respirator, is a mechanical device that provides artificial breathing for a person who can no longer breathe effectively on his or her own. In ALS, the muscles necessary for breathing, that is for adequate inhalation of oxygen and exhalation of carbon dioxide, eventually become severely impaired. Without intervention, respiratory failure can result. Patients with ALS are faced with the decision of whether or not to use a ventilator. This is a difficult decision to make. The patient and family must be fully aware of all the consequences of the decision, whether it is for or against the ventilator. Using a ventilator will significantly alter the manner in which a patient lives and the way he or she is cared for in the home or a nursing home.

### **Tracheostomy**

Anyone who needs a ventilator for respiratory system support for longer than about two weeks will require a surgical opening of the airway called a tracheostomy. A tracheostomy is a surgical incision through which a short tube is passed into the throat. The tube keeps the airway open and connects an external ventilator to the lungs. Air from the lungs flows out through the tracheostomy opening in the throat, instead of through the nose or mouth.

The tracheostomy is located below the vocal cords. Because air movement across the vocal cords is required for speech, a tracheostomy alters the ability to communicate in a "normal" fashion. A patient who could speak before the surgery, will find his or her ability to communicate greatly altered. Speech is not impossible with a tracheostomy, but it may be difficult if the ventilator is required constantly. Speech requires a special type of tracheostomy tube adapter, called a "speaking valve," that may not be appropriate for all patients. Alphabet boards, alternative speech devices or adaptive methods of communication may be required in order for a patient using a ventilator to make his or her needs and wishes known. These devices

and methods are described in detail in Section G, Speech, of this Resource Book.

Daily care is required to keep the tracheostomy site clean and to prevent infection. To keep the airway open, secretions from the lungs must be regularly cleared from the tube with a suction catheter. Equipment in contact with the tracheostomy must be kept clean to prevent lung infections.

### **The Ventilator and Other Equipment**

The ventilator is an external piece of equipment that provides respiratory support and/or additional oxygen. Ventilators are available in different types and sizes, including portable ventilators. The type of ventilator required depends on the individual patient's needs and is determined by the surgeon and/or the pulmonary or respiratory specialist involved in the patient's care. The family and other caregivers must be instructed in how to operate the ventilator and what to do if it malfunctions. Tubing that runs from the machine to the tracheostomy site carries air to and from the lungs. Humidifiers are usually required to provide moisture to the lungs and to aid in thinning secretions. Drainage systems to collect excess moisture are usually present in at least one place along the tubing. They need to be emptied periodically throughout the day. A suction machine and disposable catheters are needed to clear the tracheostomy of secretions. The frequency of suctioning depends on the patient's needs and the amount of secretions that the lungs produce.

A generator, or emergency power source, is usually recommended as a backup in case of a power failure in the home's electrical system. A portable oxygen tank and a special device called an ambu-bag can be used to provide oxygen and artificial breathing for the patient in case of a power outage or ventilator failure. The ambu-bag is connected to the oxygen tank and then squeezed to provide a breath. The family and caregivers in the home will need to be trained in the use of this device.

A patient on a ventilator requires 24-hour care because of the need for suctioning and for monitoring for alarm warnings. Occasionally, an insurance plan may provide care by an outside agency for some portion of these hours. Medicare does not pay for such services. If a patient is insured and is considering ventilator support, it is prudent as part of the decision-making process to check with the insurance company regarding the extent of

coverage for home care. Some patients may not have a caregiver or a home environment in which care with a ventilator is possible. If desired, these patients can go to an extended care facility that is capable of caring for them and the ventilator.

Patients with ALS usually cannot be removed from dependence on the ventilator for breathing. The muscles necessary for breathing become so severely impaired that they eventually do not function at all. A patient who is dependent on a ventilator and later has it removed will not be able to breathe adequately enough to sustain life. However, a patient may decide to be removed from the ventilator, even though he or she probably cannot survive without it. If that decision is made, an evaluation is normally done by a physician and a psychiatrist to determine the appropriateness of the request and to discuss the implications of the decision. It may be minutes to many hours or even a few days before breathing ceases completely after the ventilator is discontinued. The patient can be sedated during that time to ease the discomfort associated with the difficulty breathing, and to make the final moments as comfortable as possible.

Ventilator support alters the manner in which a patient is cared for, communicates, and lives. This issue may be brought up by the physician during one of your clinic visits at the Lois Insolia ALS Center. The ventilator impacts both the patient and family in many ways, including financially and emotionally. The Center's Home Liaison Nurse and Social Worker are available to make home visits to discuss ventilator support issues. The patient and family can also view a video that demonstrates the impact of ventilator support. Although the Center's staff members feel it is best to view the video with a staff member present for support and additional information, the video may be obtained from the Les Turner ALS Foundation.