

Chapter 5

Respiratory Issues



HOW DOES ALS AFFECT BREATHING?

Respiratory problems are perhaps the most serious of medical complications in ALS. Breathing difficulties occur from the gradual deterioration of muscles involved with breathing: the *diaphragm* and the *intercostals*.

The diaphragm is an arched muscle located just beneath the lungs

that moves up and down and allows air to come in and move out. The intercostals are muscles between the ribs that contract and relax and also assist with air movement.

As ALS weakens these muscles, you'll become conscious of the act of breathing, which is normally automatic, and it will consume additional energy.

Weakening respiratory muscles may increase your fatigue levels and deplete your energy. The effort to breathe, when these muscles aren't functioning well, is hard work.

Weakening of the respiratory muscles can also challenge your ability to respond to the stresses of colds, flu or pneumonia — illnesses that are caused by bacterial or viral infections. Pneumonia also may be caused by aspiration of food or fluid into the lungs, which can be caused by weakened muscles.

But, as in many other areas, new knowledge and technology make it possible for you to work, travel and continue with normal activities while obtaining the help you need with breathing.

MDA health care experts recommend that people with ALS get a flu vaccine in the fall, get a pneumonia vaccine, and be familiar with the symptoms of pneumonia:

- coughing
- sputum production (may be clear, yellow or green)
- chest discomfort with breathing
- fever/chills

People who are having difficulty with liquids (choking or coughing) should use a thickener (such as ThickIT or SimplyThick) in their diets to prevent aspiration of fluids into the lungs.

SPECIAL NOTE: As your respiratory muscles weaken, your neurologist may refer you to you a pulmonologist, a doctor who specializes in issues related to the lungs and respiratory system. Be sure this specialist understands that

your breathing problems aren't caused by problems with your lungs; they're caused by weakness of the muscles that operate the lungs. Treatments for these two types of conditions are quite different.

Your best approach is to have your ALS physician communicate with the pulmonary specialist, and to work closely with a *respiratory therapist* who's familiar with ALS.

THE ROLE OF THE RESPIRATORY THERAPIST

The respiratory therapist on your ALS health care team is responsible for obtaining measurements of your respiratory function and instructing you and your family in the use



of therapeutic measures and equipment prescribed by the pulmonary specialist. The importance of careful monitoring of respiratory function and proper instruction in therapeutic measures is essential in the overall care of people with ALS.

Your RT or pulmonary specialist will measure the *forced vital capacity (FVC)* (total amount of air that can be moved in or out of the lung). This can easily be accomplished by exhaling into a spirometer. The FVC is easy to perform and is a meaningful indicator of changing respiratory status in the person with ALS.

Evidence of respiratory involvement might include:

- shortness of breath with or without exertion
- increased lethargy or loss of energy
- poor cough
- difficulty breathing while lying flat
- headaches, especially when you awaken

If any of these symptoms occur frequently, you need a medical examination of your respiratory status.

The RT assists in the instruction of therapeutic measures ordered by the pulmonary specialist such as incentive spirometry, assistive coughing and breathing exercises, suctioning, intermittent positive pressure breathing, and postural drainage.

The therapy of respiratory dysfunction in ALS is primarily aimed at general supportive measures. Considerations may include ventilatory maneuvers (voluntary or positive pressure) to prevent atelectasis (lung collapse), a cessation of smoking program, instruction in maintaining nutrition and prevention of aspiration, flu vaccinations, and medications to

decrease the work of breathing.

If oral or pharyngeal secretions (from the mouth or nose) become excessive, drugs that decrease saliva production or suction devices to remove secretions may be beneficial. If low blood oxygen levels are documented, supplemental oxygen may be given. All infections should be promptly treated.

ASSISTED VENTILATION

Breathing, meaning the exchange of oxygen and carbon dioxide that normally occurs, may become less effective for you in advanced stages of ALS. The result may be respiratory distress, which has many symptoms:

- the inability to sing or shout
- the inability to cough or sniff hard
- the ability to speak only in short sentences
- apparently labored breathing



- use of muscles in the neck or abdomen to compensate for a weakened diaphragm
- headaches at waking
- excessive daytime sleepiness
- exhausted appearance or weight loss owing to retained excess carbon dioxide

Before these respiratory complications emerge, your doctor will probably begin to discuss various methods and steps of respiratory support — *ventilation*. There are more options for assisted breathing today than ever before, some that can prolong life for several years.

Some experts say that assisted ventilation is the single most significant factor in the increased life expectancy of people with ALS in recent years.

You should give the question of ventilatory support serious thought in advance and put your wishes in writing so they'll be known to your caregivers and medical team if you have a respiratory emergency. You may want to state your wishes formally in a *medical directive*; you can alter this document any time you change your mind.

Ventilators are now small, portable and quiet, but maintaining a person with one at home can be very expensive and taxing on caregivers.

Remember that your health care team and other professionals at the MDA clinic are there to help you understand your options and answer your questions as you make difficult decisions about ventilation. Be sure to discuss this subject in detail, including the options described here, so you can make your choices clear to your doctor and loved ones.

SPECIAL NOTE: It's wise to think ahead about your choices and needs for respiratory help. Without planning, you could experience a respiratory crisis and have to make important decisions in an emergency situation.

TYPES OF VENTILATION AND EQUIPMENT

Noninvasive

This form of ventilation has seen several advances and wider use in recent years. Noninvasive devices don't involve surgical invasion of the body. Many times, ventilators can be used for several hours a day or just during sleeping hours. People with ALS whose bulbar (mouth and throat) muscles are extensively weakened, however, may need more extensive ventilation solutions.

There are several forms of noninvasive ventilation. One is *pressure-cycled* vent machines, which deliver air at a set



pressure level with a variable volume of air on a timed cycle.

A CPAP (continuous positive airway pressure) machine is *not* indicated in ALS. CPAP increases the work of breathing by forcing the user to exhale against resistance. This can be dangerous for those with ALS.

A BiPAP machine, on the other hand, is often prescribed in ALS. BiPAP (the trademark name of a machine distributed by Respironics) is short for *bilevel positive airway pressure* and delivers air at two pressures, one for inspiration and one for expiration (inhalation and exhalation). A number of nasal or face masks and attachments are available, and can be customized for the best fit.

Volume-cycled ventilators deliver a pre-set amount of air. Volume vents can deliver air in far greater pressures and volumes than pressure vents can. Although these machines were traditionally used only with invasive interfaces



(such as a tracheostomy, see below), some doctors now prescribe them for use with a mouthpiece, nasal or face mask.



Another noninvasive form of ventilation uses *negative pressure* through a corset-like device that wraps around the chest and creates negative pressure, allowing the lungs to expand. With negative pressure applied on a timed cycle, the lungs inflate and deflate alternately, as in regular breathing. This is the same technique that was used years ago in “iron lungs.”

Invasive

Invasive ventilation is delivered via a *tracheostomy*, a surgically created hole in the trachea (windpipe) through which air is forced. The tube through which the



air is delivered also is called a *tracheostomy (trach) tube*.

A ventilator delivers air on a timed cycle through the trach, and ensures that you'll take a mini-

imum number of breaths per minute. Many ventilators can then be adjusted to respond to the person's own efforts to breathe, or to completely override these efforts. The decision to start tracheostomy-delivered ventilation is often a permanent one because it's usually impossible for people with ALS to recover the ability to breathe on their own.

Many people with ALS ultimately need a trach because of the weakness of the mouth and throat (bulbar) muscles. Invasive ventilation is thought to be a more reliable means of delivering air to the lungs when the disease is advanced. But one important drawback of a trach is that it interferes with the body's normal mechanisms for clearing the respiratory tract of mucus. Various solutions can



combat this problem.

Not everyone with ALS will need or choose to have a tracheostomy, but there may come a time when it's necessary for continued breathing.

In addition, most people relying on invasive ventilation will need humidification because the nose, through which air is normally moisturized, has been bypassed.

COUGHING AND CLEARING SECRETIONS

Coughing and clearing secretions from the lungs are activities that most people do automatically.

Normally, the lungs constantly move excess mucus and inhaled particles up toward the mouth to a spot where they can be coughed up.

As ALS advances you may eventually have difficulty coughing and clearing your throat because of weakened abdominal and throat muscles. Material that would normally be coughed up instead will fall back down into your lungs where it can cause respiratory irritation and infection. This may occur whether or not you're using assisted ventilation.

Certain techniques and medical equipment can enable you and your caregivers to create or assist a cough, and then clear mucus secretions from your airway. Talk with your doctor and your RT about these interventions.

Breath Stacking

An RT can show you this technique, which involves closing the throat after each breath taken in through a mouthpiece, and then coughing.

Abdominal Thrust

Caregivers can be taught how to increase coughing efficiency by pressing on your abdomen.

Assisted Coughing Devices

An In-Exsufflator machine delivers a large volume of air into the lungs and then quickly reverses the air flow to pull out secretions, just as a cough would.

An example is the CoughAssist. Used with a facemask, with a mouthpiece or with an adapter to a patient's endotracheal or tracheostomy tube, this machine can be used to clear your airway as needed. This type of machine is often recommended for use in conjunction with invasive ventilation, and can be



demonstrated by your RT.

Another option that's been recently studied for its effectiveness for people with ALS is The Vest. This system uses a technology called *high-frequency chest wall oscillation*. During therapy, The Vest inflates and deflates rapidly, applying gentle pressure to the chest wall. This loosens and thins mucus and moves it toward the larger airways, where it can be cleared by coughing or suctioning.

OTHER EQUIPMENT

Suction Machines

Portable or stationary suction machines can provide appropriate suction for removing the lungs' mucus secretions.

Caregivers must be instructed in sterile techniques for suctioning, which usually involves inserting a tiny tube (catheter) several inches into the trachea via a trach tube. In the absence of a trach, suction devices similar to those seen in dentists' offices can be directly inserted into the mouth. Others can be inserted through the nose via a tube with a soft catheter tip. The tube is attached to a suction machine.

In addition, most people relying on suction machines will need humidification because the nose, through which air is normally moisturized, has been bypassed.

Pulse Oximeter

Oximeters are electronic devices about the size of a small cell phone that measure the amount of oxygen in the blood through a painless sensor that can be clipped to a finger or earlobe. If your RT or pulmonologist finds that your oxygen level is normal (at least 95 percent "saturation") without any supplemental oxygen, it's likely that air exchange (of oxygen for carbon dioxide) is adequate.

If saturation levels dip below normal, you and your doctor have to decide whether the problem is chronic under-ventilation because air exchange isn't adequate, or whether there's mucus plugging the airways.

MEDICATIONS

Bronchodilators

These medications, such as those commonly used to treat asthma (albuterol, Proventil, etc.), dilate (open) the airway and are sometimes prescribed for people with ALS. However, many doctors don't think this type of drug is helpful because it won't improve the muscle weakness that's the source of respiratory problems in ALS.

Expectorants

This type of drug, commonly found in some over-the-counter cough medicines, can thin secretions and make them easier to cough up. Doctors sometimes prescribe them for people with ALS, as well as recommend an increase in fluid intake.

Saliva Management

Weakened breathing muscles, along with weaker muscles in the mouth, in ALS may result in drooling or *sialorrhea*. This isn't a case of excessive saliva production; it occurs when you're unable to swallow saliva as well as before. There are several medications and other treatments that your physician may suggest to control drooling.

Spotlight on Feeding Tubes

Insertion of a percutaneous endoscopic gastrostomy tube, commonly called a PEG tube or a feeding tube, sometimes coincides with respiratory support in ALS. This intervention may be necessary to prevent choking episodes and to enhance the ease of feeding if swallowing problems are present. A PEG tube delivers food directly into the stomach from the outside and not down the throat.

As a result of bypassing the mouth, a PEG tube can reduce, but may not eliminate completely, the danger of aspirating food or liquid into the lungs. With better nutrition and less energy used for eating, weight loss may stabilize, or even partially reverse, and respiratory function often improves. Ideally, such a tube should be placed before serious respiratory complications arise, and some doctors insist that it's "never too early" to consider a PEG tube in ALS.

Placement of a PEG tube is another personal and important decision that should be made with the help of your ALS health care team. For resources on the topic, see Chapter 10.

Some doctors advocate proactive placement of a feeding tube to prevent any weight loss, choking episodes or nutritional deficits.

For more detailed information about nutrition in ALS, see the *MDA ALS Caregiver's Guide*.