

Patient and Family Resource Guide to ALS

Section 8 Respiratory Care

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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 region of the brain degenerate, these muscles waste away. In some people with ALS, respiratory muscles may be affected late in the course of the disease; in others, these muscles are affected early in the disease process. The nerves that control movement of the diaphragm, the major muscle of breathing, are also affected.

Because of weakened respiratory muscles, people 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 person.

Problems

Early in the disease, sleep may be affected causing inefficient breathing during REM sleep. 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
- 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 your clinic nurse at the Lois Insolia ALS Clinic to discuss possible solutions.

How Is Breathing Monitored?

Breathing problems will be monitored during regular visits to your ALS Clinic by tests that measure the strength of respiratory muscles and how well the lungs are functioning. In the clinic, 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. A Maximum Inspiratory Pressure (MIP) will measure the ability to inhale forcefully. Other measures of breathing issues include a pulse oximeter, which is a device that noninvasively measures arterial blood oxygen saturation noninvasively using a comfortable finger sensor. An overnight pulse oximetry can detect changes in breathing during sleep. 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.

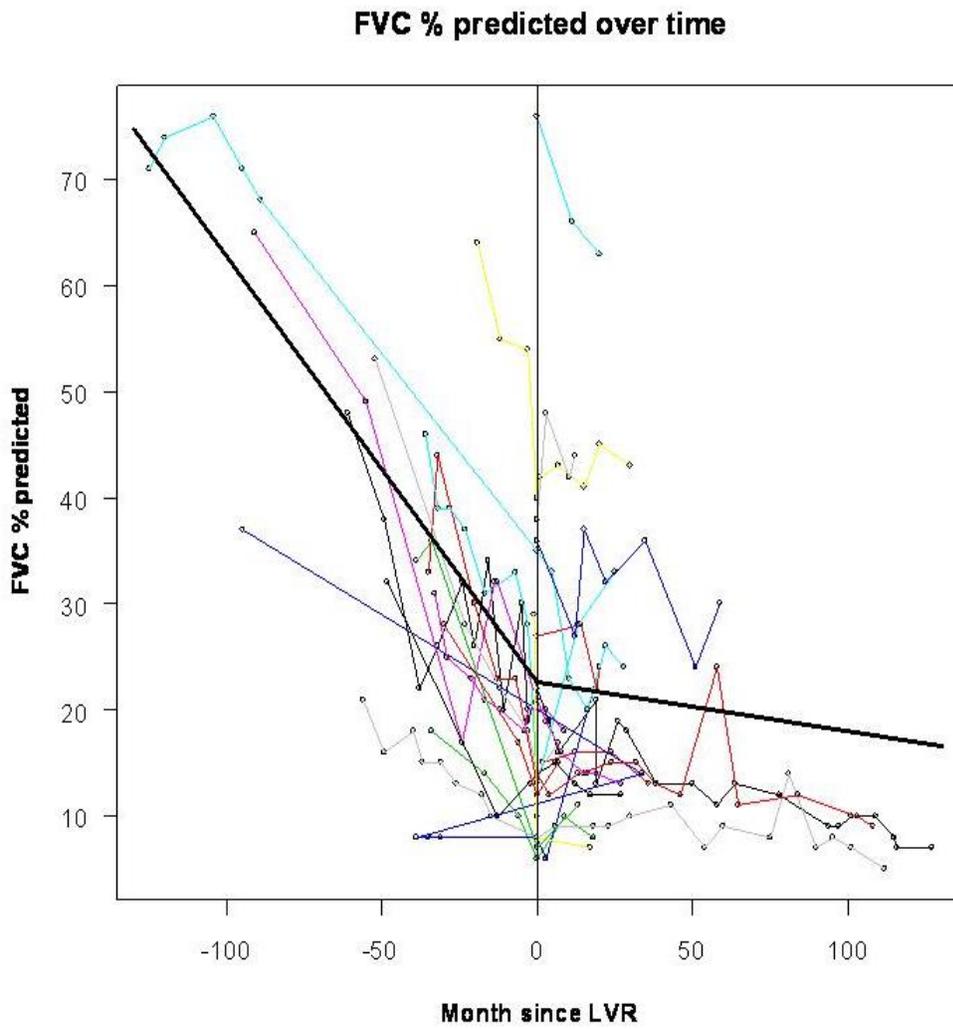
Noninvasive Solutions

Difficulty in breathing while lying flat may be relieved with the use of a wedge pillow or a hospital bed set so that the person with ALS 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. People who have a reclining chair may find relief by sleeping in that, as it allows you to lie at the appropriate angle for maximum breathing comfort.

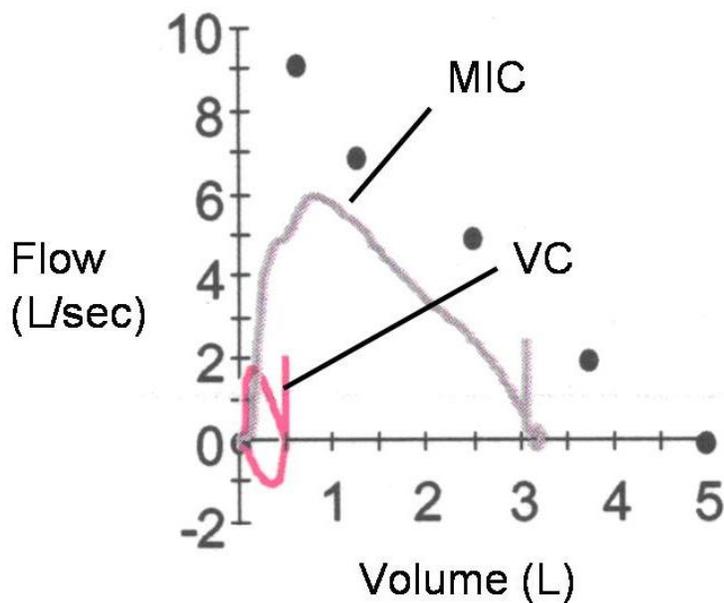
Lung Volume Recruitment

Lung volume recruitment or breathstacking can help to keep lungs open and chest muscles flexible. A daily routine of sitting tall and taking 5-10 slow, deep breaths twice a day will also help open those airways. This technique should be assisted with the use of an Ambu™ bag and mask if deep breathing becomes difficult. It is easy to do at home or on the go. This can be taught to PALS and caregivers during a clinic visit.





McKim, DA. (2011). *American Thoracic Society*, Presentation.



Lung volume recruitment is helpful because it assists the PALS with taking a bigger breath than they can take on their own. The example above shows a red line where the person with ALS is on their own and the gray line shows where they should be. The assisted breaths with the Ambu™ bag helps you achieve a much fuller breath. Data presented to the American Thoracic Society shows that patients were able to decrease the progression of their forced vital capacity by regularly performing lung volume recruitment.

Positive Air Pressure (PAP) Devices

The most commonly used device to assist nighttime breathing in PALS is a PAP, which stands for Positive Airway Pressure. There are specific versions of these devices now designed for ALS patients.

- VPAP ST
- iVAPS
- Astral
- AVAPS
- Trilogy

PAP is not a ventilator or “respirator,” and it is not invasive. It may stimulate a breath, but it does not “breathe” for the person with ALS as a ventilator does. PAP is a supportive device that can help ease breathing difficulty and its symptoms. Use of a PAP can improve nighttime sleep, resulting in decreased daytime fatigue and sleepiness, and increased energy levels. It is not limited to nighttime use; many people use this device during the day, particularly while napping.

PAP is a machine that works by providing pressure through a mask worn over the nose and mouth or through nasal pillows. The mask is similar to the kind one might use to receive oxygen in the hospital. The machine provides pressure when the person inhales and less pressure when the person exhales. This helps the person draw in oxygen and expels carbon dioxide. The machine also has a *back-up rate*. If it does not count a certain number of breaths taken by the user in one minute, it will stimulate the user to take “extra” breaths. Some PAP devices are set by pressure and some are set to guarantee a certain volume. Your ALS team will work to help determine the best device and settings for you.

Does Oxygen Help in ALS?

Oxygen is used to treat conditions where oxygen levels are low. People 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, people 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.

People with ALS need the simulation of inhaling a proper breath provided by a PAP device. Oxygen levels should be measured while the person with ALS is on PAP. If the levels are low with PAP, supplemental oxygen may be required.

What Can Help with Thick Secretions, or Phlegm?

Many persons 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.

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 PALS 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 alcohol or caffeine may also help. Use fluids of substance like Gatorade (G2 is low in sugar), popsicles, broths or fruit juices.

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.

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.

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

- **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.

What about thin secretions or drooling?

Nasal Secretions: Some people may find it hard to simply blow their nose. This impairment creates the need for an agent to dry up those nasal passages such as antihistamines. Other ways to keep the sinuses open are to use sinus rinse products or a WaterPik™ System for the nasal passages.

Oral Care: Oral hygiene is a very simple task to prevent buildup of bacteria in the mouth. Diluted mouthwash, especially the varieties with baking soda, is very effective to swish and spit. For those who cannot coordinate those motions, using mouth swabs to wipe out the oral mucosa will do the trick. This care should be done at least twice a day, in addition to normal brushing of the teeth and the tongue. Waterpik™ has systems for ultrasonic cleaners and suctioning.

Excess Saliva: Sialorrhea or drooling occurs when muscles such as the tongue and those controlling swallow begin to fail. It is necessary to remove as much excess saliva to prevent choking, drooling and aspirating saliva into the airways. To decrease the saliva, people with ALS can use red grape juice to swab out their mouth or papaya enzymes to thin out the saliva to make it easier to clear. Use of an oral suction wand may also be necessary. These devices come in small and portable packages. Some people with ALS can be prescribed medications such as amitriptyline (Elavil), glycopyrrolate (Robinal) or scopolamine patches that have side effects of dry mouth when used in small doses. Botox™ or Myobloc™ is also an option for consideration to block out the saliva glands. However, it is important to maintain a certain amount of moisture and clearance of the mucosa to prevent infection.

Airway Clearance Devices

Chest Physiotherapy is a technique used by respiratory therapists to mobilize secretions from the outer airways by cupping their hands and lightly pounding over the lung fields. This technique can be very exhausting for both the person with ALS and the caregiver. Similar motions can be created with vibrating massage wands available online and in department stores.

High Frequency Chest Wall Oscillator: This very important airway clearance device has recently been described as effective for airway clearance in people with ALS when therapy is started with the Forced Vital Capacity (FVC) at 70% to 40%. This device creates tiny bursts of air movement that helps mobilize the mucous in the outer airways to the central, bigger airways. This mobility is lacking in people with ALS when physical mobility decreases and it becomes harder to take deep breaths on their own.

Cough Devices: Moving mucous into the airways allows persons with ALS to cough up those secretions. However, if the diaphragm and other respiratory muscles are too weak, they may need assistance to cough effectively. A CoughAssist™ or VitalCough™ device is available to simulate the cough mechanism. It creates a negative pressure, then a positive pressure in a cycle simulating a cough. A suction device may also aid in the final clearance of those secretions from the mouth.

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.

What if an ALS patient catches a cold?

The ALS patient is not any more susceptible to colds and viruses than anyone else. However, when the person with ALS has an increase in nasal, oral or chest secretions, dealing with those secretions can be a lot of work. It is important to contact the ALS physicians at the Lois Insolia ALS Clinic for assistance in handling a cold before it becomes a more serious respiratory infection.

Power Considerations for Respiratory Devices

People living with ALS may require the use of devices throughout their daily activities. There are mobile power devices that will help provide access to these devices. This is a good idea in case of severe weather or other electrical failure.

Battery Geek, Cpapman, Cpap.com and Battery Power Solutions have support people available by phone. We suggest calling and providing the specific model PAP you use to get direction on which batteries would work best with your specific PAP device.

It may be helpful to have a car inverter should you like to travel using your devices in an automobile.

Here are some contacts/ websites you might find useful:

www.cpap.com	800-356-5221
www.cpapman.com	855-235-7126
www.thebatterygeeks.com/default.asp	512-692-6837
www.batterypowersolutions.net	877-445-5228

Local power companies also allow people requiring breathing devices to list with their emergency services in case of power failure.

Air Travel

People with ALS traveling with medical devices are encouraged to contact their airline before making a reservation. Devices may be carried on the aircraft and are not considered a carry-on item. The airlines recommend having enough battery power for devices to consist of one and one-half the flight time. This will ensure a safe amount of time for arrival.

The following websites are helpful for traveling:

http://www.faa.gov/passengers/prepare_fly/

<http://www.tsa.gov>

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. People with ALS are faced with the decision of whether or not to use a ventilator. This is a difficult decision to make. The person with ALS 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 person with ALS 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 person 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 Guide.

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

Persons 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 person who is dependent on a ventilator and later has it removed will not be able to breathe adequately enough to sustain life. However, one 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 to determine the appropriateness of the request and to discuss the implications of the decision. Removal from the ventilator may occur in the hospital or at an in-patient hospice unit. The patient would be sedated during that time to ease the discomfort associated with the difficulty breathing, and to make the final moments as comfortable as possible. It may be minutes to many hours or even a few days before breathing ceases completely after the ventilator is discontinued.

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 Clinic. The ventilator impacts both the person with ALS and family in many ways, including financially and emotionally. The patient and family can also view a video that

demonstrates the impact of ventilator support. Although the Clinic's staff members feel it is best to view the video with a staff member present for support and additional information, the video is available on the Les Turner ALS Foundation's www.lesturnerals.org

Disclaimer: All care has been taken in preparing this document. This information is of a general nature and should be used as a guide only. Always consult your health care team before starting any treatments.