

Adapting to Changes in Breathing When You Have ALS



LIVING WITH ALS
RESOURCE GUIDE



ADAPTING TO CHANGES IN BREATHING WHEN YOU HAVE ALS

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A NOTE TO THE READER: The ALS Association has developed the *Living with ALS* resource guides for informational and educational purposes only. The information contained in these guides 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.



Introduction

The muscles of breathing are not spared as ALS progresses. This resource guide will teach you the basics of how the lungs function, the changes that will occur, how to be proactive in maintaining lung health and prepare you for the decisions that will need to be made when the lungs need maximal assistance.

The following topics will be covered:

- How the lungs work and the impact of ALS on breathing
- How lung function is measured and measurement definitions
- Symptoms of weakness in breathing muscles
- Maximizing lung function and respiratory health
- Positive airway pressure ventilation
- Clinical stages of respiratory decline in ALS
- Making advanced decisions about respiratory support



How the Lungs Work

ALS affects the **mechanical** function of the lungs. The major muscles of breathing are the **diaphragm** (which does most of the work), the intercostal muscles (those between the ribs), and **abdominal muscles** (necessary for coughing and exhaling maximally).

The respiratory muscles act like a pump, moving air in and out. When you breathe in, the diaphragm moves downward and the rib muscles expand outward, like an upside down umbrella opening. This natural movement allows air to flow easily into the lungs. Like a stretched rubber band, elastic recoil brings the lung muscles back to their resting position on exhalation.

As breathing muscles weaken, the lungs become less elastic. Stiffer lungs allow less air to flow in. You lose the reserves you once called upon during hard work or exercise. That is why we measure your lung volumes at regular intervals, ask about your breathing and make suggestions for maximizing your lung strength and avoiding lung infection.

Measuring Lung Function

Pulmonary function tests (PFTs) measure lung muscle strength and overall function. Lung function should be assessed at each clinic visit as part of an overall disease management strategy. Test results are used to determine need for breathing assistance and supportive therapies. Each clinic visit gives you the opportunity to discuss implications of test results and treatment options with your physician, nurse and respiratory therapist. In the outpatient clinic or office setting portable spirometers are used.

Spirometry

Spirometry measures how well you move air in and out of your lungs. A **spirometer** is a lightweight, portable and reliable microcomputer that measures, calculates, records and displays lung function test results. Measurements of lung volume and airflow over time are recorded and displayed numerically in a graph. The results are expressed as a percentage of predicted since your test results will be compared to a large sample of people your same age, sex and height.

Spirometry tests are done while you are sitting upright in a chair. However, **if you are having difficulty breathing when lying flat, or your physical symptoms of air hunger are not reflected in your lung function test results, you may be asked to repeat the breathing tests lying down (supine).**

- Vital capacity (**VC**) measures the amount of air you can exhale after a maximum inhalation (in liters).
 - Forced vital capacity (**FVC**) measures the amount of air you can exhale forcefully after a full inhalation.
 - Slow vital capacity (**SVC**) measures the amount of air you can exhale slowly and steadily after a full inhalation.
 - Forced expiratory volume (**FEV₁**) measures how much air you can forcibly exhale in one second (liters per second).
 - **FEV₁/FVC** is the ratio of forced expiratory volume in one second (FEV₁) compared to the forced vital capacity (FVC) expressed as a percentage (FEV₁%). It is used to classify your results.

Additional Lung Function Measures

Although spirometry tests tell us how our lung muscles work together (diaphragm, intercostal, abdominal), additional measures refine our knowledge. **Other lung function tests include:**

- Maximum inspiratory pressure (**MIP**) is a more sensitive test of inspiratory muscle strength (internal intercostal and diaphragm).
- Maximum expiratory pressure (**MEP**) measures the strength of expiratory muscles (external intercostal and abdomen).
- Peak cough flow (**PCF**) measures expiratory flow and ability to protect the lungs from saliva, food and liquids slipping into them.
- Sniff nasal inspiratory pressure (**SNIP**) is a sensitive test of global inspiratory muscle strength. It may be useful if changes in speech and swallowing muscles lead to inaccurate spirometry test results.

Passive Tests of Lung Function

- **Capnography** measures your exhaled carbon dioxide levels. As the lungs weaken it becomes more difficult to clear the lungs of this gas.
- **Pulse oximetry** measures the oxygen saturation level in your blood.
- **Respiratory rate** is the number of breaths you breathe in one minute. If your lung volumes decline, the body will naturally compensate by having you breathe more rapidly. You may not be aware of this, especially if the change in your lung volumes has been gradual or if you are less physically active than you once were.

“Non Volitional” Tests to Assess Diaphragm Strength

The active tests discussed above are “volitional” which means that lung muscle participation is voluntary. You choose to take a maximal breath in and forcefully blow it out for as long as you are able. It may require practice and repetition to achieve accurate and consistent results. Accurate results also are affected by the experience and skill of the nurse or respiratory therapist coaching you and the trust and rapport you establish with them.

But **the most important factor in obtaining accurate lung function test results is the strength of the muscles of your mouth and throat: the oropharynx.** These muscles include your vocal cords, lips, tongue, palate and jaw.

The muscles that control speech and swallowing are referred to as the **bulbar muscles.** Early anatomists thought that the lower brain stem—which is between your cerebral cortex and your spinal cord—resembled a flower bulb and named it bulbar. So, when the flow of information from your brain to muscles in your mouth and throat is disrupted, the muscles become weak or rigid. **When you are inhaling and exhaling air forcefully, weak and uncoordinated bulbar muscles reduce the open and smooth flow of air into your lungs. This negatively affects lung function test results.**

Your neurologist or pulmonologist may suggest other **measures of diaphragm strength that are non volitional (involuntary) and more easily observable.**

- **Phrenic nerve conduction study (PNCS):** The phrenic nerves stimulate your diaphragm. These tests are like the electromyography (EMG) of your muscles you might have had as part of your ALS diagnostic work up. When the phrenic nerves, located on either side of your neck, are activated, your diaphragm involuntarily contracts. The strength of the contraction is measured in amplitude. The larger the amplitude, the stronger your diaphragm strength.
- **Sniff test with fluoroscopy:** Fluoroscopy is a test of moving body parts. It is like an xray, but muscles and other structures are seen as a “movie” instead of a “still.” During the test you will be asked to “sniff” through your nose. This

will cause your diaphragm to contract quickly. The radiologist will be able to see how much your diaphragm drops. And because you have two diaphragm leaves—one on the right and one on the left—the radiologist can see if they contract the same or if one leaf is stronger than the other.



- **Nocturnal oximetry studies:**

Your physician may want to measure your oxygen levels while you sleep at night. If your diaphragm becomes weak, you are likely to breathe shallower at night. Over time, shallow breathing may lead to significant drops in your oxygen levels, which will lead to frequent awakenings, awakening feeling tired and not refreshed or headaches. Nocturnal oximetry studies can be done easily in your home by placing a clip on your finger and attaching it to a small recording device. The information will be stored, accessed and interpreted, and the results will be discussed with you.

Pregnancy in ALS and Its Impact on Breathing and Monitoring

Although pregnancy in women with ALS is not very common, it is generally considered a potentially risky event and requires special considerations. The course of pregnancy and the delivery may be normal. However, due to the increased minute ventilation requirements in pregnancy and the restrictive impact on the diaphragm caused by the enlarging uterus, **respiratory function should be carefully monitored**. Generally, ALS does not have harmful effects on fetal development and the neonatal outcome is good.

Symptoms of Lung Muscle Weakness

While changes in breathing function may vary from person to person, there is enough consistency among people with ALS to identify **milestones of respiratory decline**. They are listed here so you can plan and discuss treatment options with your physician, nurse and respiratory therapist:

- **Noticeable breathlessness** while walking, especially inclines and stairs
- Awakening in the morning **feeling unrested**, although getting an adequate number of hours of uninterrupted sleep
- Increasing **breathlessness with daily activities like dressing, eating and bathing**
- **Signs of sleep interruption or shallow breathing at night**
 - Waking up frequently to urinate
 - Nightmares
 - Waking with a choking sensation
 - Waking in the morning with a headache
 - Feeling sleepy during the day and needing more naps
 - Increasing overall fatigue
- **Breathing discomfort when lying flat in bed**
 - Needing to sleep only on your side
 - Needing to sleep elevated on two or more pillows
 - Needing the sleep upright in bed or a chair
- **Breathing discomfort while sitting or when speaking**

- **Need for noninvasive positive pressure breathing at night**
- **Shortness of breath at rest and intermittent use of noninvasive positive pressure breathing during the day** to rest lungs and improve energy levels
- **Need for manual and mechanical secretion mobilization** due to weak cough
- Need for **around the clock noninvasive positive pressure breathing**
- **Decision to have a breathing tube (tracheostomy)** placed surgically and breathing supported by a mechanical ventilator (life support), **OR decision to select hospice care** so you can be comfortable with bi level breathing and nutritional support while ALS continues to follow its natural course.

Maximizing Lung Function

You can maximize lung function at every stage of ALS, even before you experience symptoms of shallow breathing. In this section, we will explore a variety of techniques. The best way to learn how to perform breathing exercises is to see them and practice with a trained respiratory specialist. **There are links to resources that show you these techniques at the end of this resource guide.**

Goals of Lung Expansion Therapies

ALS is not a lung disease. Lung volume loss in ALS is due to pump (diaphragm) weakening. If you have not been diagnosed with lung disease (asthma, chronic bronchitis, emphysema, bronchiectasis or other diseases of the bronchioles, the airway lining or air sacs), the tissues and glands of your lungs are perfectly normal.

The goals of lung expansion and coughing therapies are to maximize normal lung function. Specifically, the goals are to:

- Improve cough effectiveness
- Improve voice projection
- Prevent air sac under inflation or collapse

- Improve suppleness of the lungs (compliance)
- Decrease the work required to breathe
- Reduce or prevent lung infection
- Allow for more time breathing independently without mechanical assistance

Sense of wellbeing and relaxation that accompanies slow, deep inhalation and full exhalation, reduced stress and lessened anxiety

Combined with energy conservation strategies, paced walking, exercise and stretching regimens suggested by a physical therapist experienced with neuromuscular disease, these breathing techniques will assist you as you move through the stages of ALS.

Breathing Techniques

1. “No tech” strategies include:

- Segmental and diaphragmatic breathing
- Breath stacking
- Active cycle breathing
- Pursed lip breathing
- Huff coughing
- Manual cough assistance (abdominal thrusts)

2. “Low tech” strategies include:

- Breath stacking with manual ventilation “Ambu”® bag

3. “High tech” strategies include:

- Mechanical inextufflation (M IE) with a cough assist device. M IE provides lung expansion and breath stacking in the “inhale/positive pressure” mode and cough assistance in the “exhale/negative pressure” mode.
- Breath stacking or lung hyper expansion (“sigh” breaths) with mouthpiece ventilation (MPV) mode available on portable mechanical ventilators.

Lung Health in ALS: Back to Basics

Your lungs have a natural, healthy ecosystem much like the healthy ecosystem of a salt marsh or estuary. They have innate immunity protection and adaptive immunity to microbial pathogens that might find their way in. How does this work?

- **Mucus**

The lower airways (bronchioles that look like descending tree roots) are lined with mucus. Mucus keeps the airways humidified and lubricated. Typically, most of the mucus is reabsorbed into the airway lining.

The effectiveness of mucus depends, in part, on the balance of positively charged sodium and negatively charged chloride in your lung vessels. This is why it is **important to stay well hydrated** so you have a healthy balance of these important electrolytes.

In addition, antimicrobial substances that are present naturally in the mucus lining of the airways (phagocytes and other professional scavengers) engulf and kill potentially harmful microorganisms.

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- **Lung transport system**

Mucus also acts as a type of sticky flypaper. Inhaled debris (microscopic particles) and dead and aging cells are trapped in mucus. Tiny hairs (cilia) that line the airways rhythmically beat upward and transport debris to the back of the throat. This debris is swallowed unknowingly or expectorated. The air sacs also contain macrophages that naturally clear bacteria and other organisms from the tiniest parts of your lungs. Cilia are hindered or paralyzed by inhaled smoke and fumes.

To maximize your lungs' natural self cleaning mechanisms, it is important to avoid inhaling harmful smoke and irritating substances from cigarettes, wood burning fireplaces, charcoal grills or in the workplace.

- **Prevention**

As they say, "an ounce of prevention is worth a pound of cure." By keeping your whole body healthy, you keep the lungs healthy. Below is a prevention checklist you can use to maximize lung health and avoid breathing complications.

- √ Get the pneumococcal polysaccharide **pneumonia vaccine**.
- √ Get an annual **influenza vaccine**.
- √ **Treat upper respiratory tract infections and chronic or seasonal rhinitis.** (Use decongestants, antihistamines, sinus lavage or steroid inhalers to avoid sinus and nasal passage drainage into the lungs. Consult with your doctor before trying over the counter preparations.)
- √ **Reduce exposure to airway irritants/inflammatory agents** (dusts, fumes, aerosols).
- √ **Maximize nutrition.** (Malnutrition can lead to immunosuppression and reduce your germ fighting potential.)
- √ Maintain good **hydration.** (electrolyte balance)
- √ **Assess aspiration risk and avoid breathing in saliva, food or liquids.** (If you

have excessive saliva and have trouble swallowing thin liquids, you are at risk for having them enter your lungs and create an infection.)

- √ **Encourage good dental and oral care.** (An overgrowth of oral bacteria can find its way into the lungs inadvertently.)
- √ **Encourage adequate sleep** and address barriers to restorative sleep. (A good night's sleep is vital for healthy immune function.)
- √ **Encourage movement and safe exercise.** (Movement and exercise support immune function, increase lung expansion and help the lungs' natural cleaning functions.)
- √ **Create opportunities for early detection of a lower respiratory tract infection.** (See your doctor at the earliest signs of increased shortness of breath, cough or fever.)
- √ **Avoid infective agents.** (Practice good hand washing techniques and make sure those around you wash their hands too. If family or friends have a respiratory infection, have them keep their distance or wear a mask.)

Noninvasive Positive Airway Pressure Breathing

Noninvasive positive airway pressure breathing has been shown conclusively to **alleviate symptoms of diaphragm weakness and shallow breathing in ALS**. It has also been shown to prolong life expectancy if it is initiated early—when signs and symptoms of respiratory compromise occur—and if it is well tolerated and used consistently.

There are several terms and abbreviations that refer to respiratory assistive devices. The most common one is BiPAP®. BiPAP is a registered trademark of the Philips Respironics company and is used much as we use Kleenex® for tissues or Xerox® for a photocopier. Many clinicians also refer to noninvasive ventilation as Bi Level ventilation. The more accurate term is noninvasive ventilation (NIV) or noninvasive positive pressure ventilation (NPPV).

Ventilation means moving air in and out of the lungs, expanding your airways and air sacs and improving flow of oxygen in and carbon dioxide out. Portable positive pressure generating devices support your breathing by replacing lung volumes that have been reduced because of a weakening diaphragm. There are many modes of supporting your breathing efforts with NIV. Your clinician will work with you to find the best mode and settings to overcome the issues with shallow breathing as well as making it as comfortable as possible.

How NIV Works

You will be fitted with a mask that covers your nose, your mouth and nose, with nasal cushions that fit in your nose or some combination. (There are many options these days.) A six foot piece of plastic corrugated tubing will connect you to a small breathing device that may be as small as a bedside alarm clock or as big as a breadbox. (There are many options here too.)

When you take a breath in, the machine increases the flow of air to the lungs until it reaches a specified pressure or volume. This pressure or volume is set by a respiratory therapist or physician based on your individual lung capacity. If you are breathing on your own, the ventilator will only support your spontaneous breathing.

If, however, you breathe very shallowly or quit breathing for periods altogether, the machine will sense this and initiate a breath for you so you are never air hungry.

If the NIV device is set properly, your own breathing and the machine support should coordinate seamlessly giving you a restful night's sleep.

- **When should you start using breathing support?**

Your respiratory therapist, neurologist, nurse or pulmonologist will discuss with you the option of using noninvasive positive pressure breathing assistance when measures of lung function (spirometry tests) decline, when you report symptoms of nighttime shallow breathing (sited above) or a combination of the two.

Medical insurance providers have qualifying thresholds for lung function tests results that determine when they will financially reimburse for mechanical breathing assistance. Your healthcare providers will be aware of these standards and will take them into consideration when discussing noninvasive positive pressure breathing with you.

The most common **qualifying tests** are:

- √ Your forced vital capacity (FVC—a measure of overall lung strength)
- √ Your maximal inspiratory pressure (MIP—a measure of diaphragm and abdominal muscle strength)
- √ Your carbon dioxide levels (PaCO₂ or ETCO₂—signs of shallow breathing)
- √ Your oxygen levels during nighttime sleeping (SpO₂—a sign of nighttime shallow breathing)

The symptoms you experience are also an important factor when discussing breathing assistance. **Inability to breathe comfortably while lying flat or increasing shortness of breath with activities call for a discussion of assisted breathing.**

The following is a **summary of factors that lead to recommending treatment with bi level positive pressure breathing:**

- **Your symptoms of impaired lung function** at night or during the day
 - Dyspnea (shortness of breath)
 - Orthopnea (inability to breathe comfortably while lying flat on your back)
- **Observed signs of respiratory insufficiency**
 - Tachypnea (rapid breathing rate, greater than 20 breaths per minute)
 - Decreased chest expansion (shallow breathing)
 - Use of accessory muscles (muscles in the neck and upper chest) to assist a weakened diaphragm

- **Objective measures** of significant chest wall restriction due to lung muscle weakness
 - FVC less than 50% of predicted or
 - MIP less than -60 cmH₂O or
 - PaCO₂ greater than 45 mmHg (increased levels of carbon dioxide) or
 - Oxygen levels (SpO₂) below 88% for five minutes while sleeping at night

Adjusting to Bi Level Positive Airway Pressure Breathing

Adjusting to assisted breathing may take time. **It is a good idea to start earlier rather than wait until your breathing is very uncomfortable.** It is a process and may require both psychological and physical adjustments.

Accepting breathing support may be difficult for some because it is an acceptance of or realization that the lungs are getting weaker. Talking about these feelings with your respiratory therapist, nurse, physician, partner, spouse and friends is important. The goal is for you to be comfortable and have the best quality of life, as defined by you.

Here are some steps that might help:

- ✓ **Wear the mask or nasal cushions for short periods during the day without the breathing machine.** Distract yourself by reading, watching TV, listening to music or talking to friends or family. Guided imagery or meditation can help.
- ✓ **Attach the mask or nasal cushions with the tubing to the respiratory assistive device.** Turn on the blower and use it for short periods during the day using one or more of the distractions listed above.
- ✓ **Try using the device for naps during the day.** Try different sleep positions, such as back, sides, flat or elevated.
- ✓ **Use your respiratory assistive device at night.** If you are unable to fall asleep or awakening frequently, a prescription or over the counter sleep aid may be useful for the first week or two.

- √ **If you awaken and cannot fall back asleep with the device, remove it and try again the next night.** Do not skip a night. The more you use it the easier it will become.

Once you have relief of symptoms of shallow breathing, the more motivated you will become to use the support.

Additional Breathing Assistance Device Options

Additional Noninvasive Positive Pressure Ventilation Options

There are more options than ever before for receiving breathing assistance (noninvasive positive pressure ventilation) in the home. Ventilation options that only could only be delivered in the hospital a few years ago can now be found in outpatient settings. This is primarily due to the advancement of microprocessor technology and to the commitment of major manufacturers to treat people with neuromuscular diseases.

Ventilators initially designed for people with a tracheostomy (breathing tube placed in the trachea) can now be used with a mask or nasal cushions for supported breathing. These ventilators have more options than the smaller bi level positive pressure ventilation devices, including dual modes: one for nighttime mask assisted breathing and a second for daytime breathing with a mouthpiece (also referred to as "on demand ventilation," sip ventilation, or MPV).

These ventilators also have internal and built in external batteries that allow for mobility if you are using a wheelchair in or out of the house.

No matter what initial ventilation options, settings or machines are chosen it is essential that your respiratory therapist or pulmonologist download and review the data stored in the machine on a regular basis. They will determine if you are being ventilated well and, if not, use the information to make needed changes. ALS is a progressive disease, and the initial settings may not be sufficient to support your breathing as time goes on.

Advanced Life Support

When bi level noninvasive positive pressure breathing with a mask is no longer effective at providing adequate lung expansion or if you have not been able to use NIV/NPPV successfully, you will **need to choose between tracheostomy and mechanical ventilation or hospice care**. Choosing tracheostomy and mechanical ventilation (**invasive breathing support**) depends on your goals for care and requires first considering a checklist of factors that must be in place to help you plan and make the transition from hospital to home a smooth one. **See the checklist “Considerations for Home Care with Mechanical Life Support” later in this resource guide.**

Challenges To Noninvasive Mechanical Assisted Breathing: Bulbar Muscle Weakness

If you have moderate to severe impairment of the muscles of speech and swallowing, the bulbar muscles, you may find it harder to adjust to treatment with noninvasive positive pressure ventilation. **Loss of muscle tone or rigidity of muscles in the oropharynx contributes to airflow restriction and turbulent airflow in the back of the throat.**

If excessive saliva pools in the mouth and back of the throat, positive pressure breathing with high flow inspiratory pressures may predispose you to aspirate fluids into the lungs. There are a number of treatments to reduce excessive saliva accumulation and drooling. These are covered in detail in The ALS Association’s resource guide, *Managing Symptoms of ALS*.

Drying agents to control saliva production pose their own problems. **Dry mouth with positive pressure ventilation is uncomfortable.** Weak jaw muscles contribute to mouth breathing which aggravates oral dryness. A **heated humidifier, a must for everyone** who uses positive pressure ventilation, may help alleviate dry mouth, as can a chinstrap to support the jaw and keep the mouth closed. You and your respiratory therapist will need to be patient, creative and persistent to find solutions

You and your respiratory therapist will need to be patient, creative and persistent to find solutions and settings that work.

and settings that work. Even if you are not able to gradually increase the inspiratory pressure or volume settings for optimal lung expansion and lung volume replacement, you may find use of assisted breathing gives you some support and lessens your shortness of breath.

Helpful tips and tricks for adjusting to noninvasive positive pressure ventilation can be found at the end of this resource guide.

A Word About Oxygen

We talk about low levels of oxygen in ALS, especially at night, and the body's natural mechanisms for maintaining normal oxygen levels during the day (by having you breathe faster or your heart beating more rapidly). So, you would think that providing supplemental oxygen through a small tube into the nose would do the trick. Sadly, it is not so simple.

In ALS, the main respiratory system issue is a weakened diaphragm causing the inability to take large enough breaths to adequately eliminate CO₂. Low oxygen levels may occur, but that is mainly due to shallow breathing and inability to take large breaths, causing collapse of the lower airways. This collapse of lower airways affects the ability to ventilate and carry oxygen to the rest of the body. To minimize this issue, NIV is the preferred treatment. Supplemental oxygen may be necessary in severe respiratory failure or if there is an underlying secondary pulmonary disease like COPD. However, generally, supplemental O₂ is not needed in ALS. In fact, supplemental O₂ can have a deleterious effect in ALS patients. As CO₂ rises due to hypoventilation, chemoreceptors in your brain tell you to increase your breathing to blow off CO₂. However, because of the weakened diaphragm, often you will not have the ability to take larger or faster breaths. You may show a low blood oxygen level as a low PaO₂ or SpO₂ if checked by a clinician. If the clinician gives supplemental O₂, you may get an increase in the PaO₂ or SpO₂ levels due to

sending more O₂ to the already ventilated areas of the lungs, but they are just “masking” the real problem of shallow breathing. If the SpO₂ is being monitored by a clinician, they may think you are now doing better. However, your chemoreceptors in your brain telling you to increase ventilation may be blunted due to the “increase” in oxygen levels, potentially worsening the ability to eliminate CO₂ and breathe faster or deeper.

Lastly, oxygen is classified as a medical gas and is regulated by government agencies. Like any medication, physicians must document a physiological need for it, show it is not harmful and demonstrate its benefit. **In ALS, oxygen administration may be detrimental** if it is used in place of bi level positive pressure breathing support. Over time, if you have gotten used to higher than normal levels of carbon dioxide in your lungs and bloodstream, supplemental oxygen may result in impaired breathing, profound sleepiness and lack of clear thinking or disorientation.

Make sure those who suggest and prescribe oxygen are very knowledgeable about ALS and the national and international recommendations for safe and effective treatment.

Advanced Decision Making About Respiratory Support

ALS is a devastating diagnosis. Physicians and staff members of ALS multidisciplinary clinics know this and want to support you and your loved ones at every stage of this disease. One of the ways to do this is to be honest with you about what they know about ALS and assist you in preparing for what lies ahead. They cannot give you an exact timeframe but can discuss treatment options with you at each stage—the clinical forks in the road.

They do this not to take away hope, but to give you reassurance, realistic expectations and hope for the best possible life with ALS. **As ALS progresses, their goals change from maximizing physical function to providing effective and compassionate supportive care.**

The focus is on **YOU** and all decisions are **YOURS**. It is your medical team’s responsibility to ensure that your decisions are based on accurate information about your breathing

status in terms that you fully understand. **Making decisions about accepting or foregoing invasive mechanical breathing support and communicating them in words and writing can be a positive experience.** It allows you to be proactive, knowing uncomfortable symptoms of breathlessness or air hunger can be well controlled no matter your choice. But again, it is **your decision**.

The most difficult situations arise when you don't have a plan. You don't want to find yourself in the emergency room with diaphragm failure having to make an immediate decision about accepting or foregoing life support. And you may not want to put your loved ones in that emotionally painful situation either. Not making a decision about your life is in fact making a decision: to have unknown medical personnel make the decision for you.

A [video series](#) on respiratory decision making in ALS is available on the ALS Association website. Viewing the series with your family and loved ones and discussing your reactions with your clinic team can be very useful. **In the videos people with ALS and their caregivers speak directly and honestly about the decisions they made to accept or forego different levels of breathing assistance and why.**

Choosing Advanced Life Support (Invasive Mechanical Ventilation)

You may decide that life is worth living in any way possible, at least for some time and within certain boundaries. You may have a specific goal in mind—the birth of a child or grandchild, an adult child's wedding, your own wedding anniversary, a family reunion or other significant life events. **It is your choice.** It can be a long term or short term goal. If you have made detailed plans to prepare for surgery and advanced life support at home, your ALS care team will support you in coordinating the care you need.

Before deciding on a tracheostomy and home mechanical ventilation, it is important to discuss with your physician and loved ones the possibility of withdrawing life support and under what conditions you would want to do so.

Mechanical ventilation treats respiratory failure but will not stop ALS from progressing. You will want to discuss if there are any physical or emotional conditions you would find intolerable. You need to make this clear while you can still communicate your wishes.

Having a checklist and reviewing it in detail with your family, physician, nurse, respiratory therapist and clinic social worker will help you plan ahead and make the transition from hospital to home a smooth one.

Checklist: Considerations for Home Care with Mechanical Life Support

Who will care for you in your home?

- √ Will your medical insurance cover the cost of a mechanical ventilator, portable suction unit, secretion mobilization devices and all disposable supplies in the home?
- √ Who will provide 24 hour care for you in the home?
- √ If family members, will they be comfortable managing a mechanical ventilator and your tracheostomy care and suctioning secretions from your lungs?
- √ Will family care providers have the physical and emotional support systems to help them help you?
- √ Will your medical insurance pay for in home help to assist with ventilator, tracheostomy, suctioning, bathing, feeding, dressing and other necessities of daily life?
- √ If insurance does not cover this cost and you do not have enough family members to provide 24 hour care, do you have the financial resources to pay for nursing care?
- √ Is there a medical home care equipment company near your home that will provide the mechanical ventilators and respiratory therapists to train family and home caregivers in ventilator management, cleaning and maintenance?

Home Safety Checklist:

- ✓ Is your home suited for full time mechanical ventilator support?
- ✓ Is there a bedroom or other room large enough for you and your medical equipment on the first floor of your house?
- ✓ Is your electrical system safe and does it meet the local codes?
- ✓ Is there adequate amperage to accommodate all medical equipment and household appliances?
- ✓ Are there enough electrical outlets for multiple electrical medical devices in the bedroom?
- ✓ Do you have working smoke alarms and fire extinguishers?
- ✓ Is your house clean and uncluttered?
- ✓ Is there sufficient room in the hallways to accommodate a wheelchair and gurney if emergency medical personnel are called to the home or you must be evacuated quickly?
- ✓ Can your home be easily located and accessed by emergency vehicles and personnel?
- ✓ Is there space in the home to clean, disinfect and dry non disposable medical equipment?
- ✓ Is there adequate storage space for disposable medical equipment?
- ✓ Is there air conditioning in warm climates?
- ✓ Are there safe and efficient heating systems for cold climates?
- ✓ Are there provisions for prolonged electrical power outages like generators or large batteries?
- ✓ How quickly can emergency personnel arrive at your home after a 911 call?

This list is not meant to be daunting. Your safety is of most concern. Again, planning ahead is key. **If it appears your home is not the safest place for you, or if there are insufficient financial resources or home caregiving support, living in a sub acute unit or other long term care facility is an option.** Your clinic social worker or ALS Association care manager can assist you in thinking through your options and finding a workable solution.



Foregoing Advanced Life Support and Choosing Hospice Care

At this point there is no cure for ALS, so in essence, all care is supportive—to relieve distressing symptoms, improve comfort and maximize

quality of life as defined by you. If you choose to forego mechanical and invasive life prolonging measures, hospice care is the best alternative.

Choosing hospice care is not “giving up.” In fact, it can be seen as affirming life and accepting dying as a normal process, neither postponing nor hastening death.

Hospice care can be provided at home, in a specialized hospice facility or in a long term care facility. Hospice care is well suited for ALS as it utilizes a holistic, integrated approach that addresses all aspects of life, among them:

- **Physical** – comfort and symptom control (controlling pain, oral secretions and breathlessness)

- **Emotional** – addressing fears and concerns, yours and those of your loved ones
- **Intellectual** – ongoing information and education by hospice staff
- **Spiritual** – facilitating life review, meaning of life discussions and ways to be at peace at the end of life
- **Practical** – support teams to assist caregivers at home

In the U.S., Medicare has disease specific requirements for people with ALS who seek hospice care. You must have:

- √ Rejected invasive mechanical ventilation
- √ Critically impaired respiratory function (with or without use of bi level noninvasive positive pressure breathing assistance) and/or
- √ Severe nutritional insufficiency (with or without a feeding tube)

Critically impaired respiratory function is defined by Medicare as:

- √ FVC equal to or less than 40% of predicted and two or more of the following symptoms:
 - Dyspnea (shortness of breath)
 - Orthopnea (inability to breathe comfortably while lying flat)
 - Use of accessory muscles (neck and upper chest muscles that assist when the major muscle of breathing—the diaphragm—is very weak)
 - Paradoxical abdominal movement (the belly moves inward involuntarily when you take a breath instead of outward—a sign the abdominal muscles must support a weak diaphragm)
 - Breathing rate greater than 20 breaths per minute while resting

- Weak cough
- Reduced vocal volume (a sign of shallow breathing and weak diaphragm)
- Symptoms of sleep disordered breathing (frequent awakenings, morning headaches, increasing fatigue and lack of refreshing sleep that are likely due to shallow breathing and reduced oxygen levels at night)

Noninvasive Positive Pressure Ventilation (NPPV) At the End of Life

At the end of life in ALS, noninvasive assisted breathing support with mask or nasal cushions provides supportive, comfort care and should be included in the hospice benefit. Its use will not alter the progression of the disease. The majority of people with ALS die peacefully due to increasing levels of carbon dioxide that accumulates in the lungs and bloodstream in spite of continuous or nocturnal use of breathing assistance.

Summary Statement

We all hope a definitive cause and a cure for ALS is found. The future looks brighter than ever before. In the meantime, respiratory therapists, along with multidisciplinary team members and care managers, will work with you, your family and loved ones to help you manage respiratory symptoms and be proactive in your decision making process every step of the way.

Resources

Books

Respiratory Management of ALS. Lee Guion MA, RRT, FAARC. Jones and Bartlett Publishers, 2010.

- Comprehensive textbook on the assessment and management of respiratory symptoms in ALS. Written in a style and language accessible to non clinicians.

Respiratory Therapy: Natural history, measurements, and treatments. James B. Caress, MD and Connie C. Paladenech, RRT, RCP. In **Amyotrophic Lateral Sclerosis: A Patient Guide for Clinicians.** Edited by Richard S. Bedlack and Hiroshi Mitsumoto, Demos Medical Publishers, 2013.

- Accessible coverage of all aspects of ALS and useful resources appropriate for non clinician readers.

Videos

How ALS Affects Breathing

Being aware of symptoms and how you can prepare for and manage them is key to quality of life and, often, to peace of mind. These videotaped conversations with physicians, healthcare providers and families living with ALS have been developed to introduce and explain the complicated topic of breathing changes in ALS.

www.als.org/navigating-als/living-with-als/therapiescareaddressing-respiratory-changes

Website

AmyandpALS.com

- Web site hosted by Amy Roman, SLP focusing on speech and communication solutions for people with ALS. The site includes information on ALS and breathing. Lee Guion MA, RRT, FAARC is a regular contributor and author of the interactive “Breathing Lessons” blog which covers breathing solutions and links to videos on breathing exercises such as breath stacking.



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About The ALS Association

The ALS Association is the largest philanthropic funder of ALS research in the world. The Association funds global research collaborations, assists people with ALS and their families through its nationwide network of care and certified clinical care centers, and advocates for better public policies for people with ALS. The ALS Association is working to make ALS a livable disease while urgently searching for new treatments and a cure.