

LIVING WITH ALS
RESOURCE GUIDE

10



Adapting to Changes in Breathing When You Have ALS



ADAPTING TO CHANGES IN BREATHING WHEN YOU HAVE ALS

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Living with ALS
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INTRODUCTION

The muscles of breathing become weaker as ALS progresses. In some people with ALS, difficulty breathing is their first symptom of the disease. 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 how to prepare for the decisions that will need to be made when the lungs need maximal assistance.

What we will cover in this resource guide:

- 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
- Bi-level Positive Airway Pressure (BiPAP)
- Diaphragm pacing system
- 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** (it separates your chest from your abdomen and does most of the work of breathing), the **intercostal muscles** (between the ribs), and **abdominal muscles** (especially important for coughing and exhaling maximally).

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

As breathing muscles weaken, the rib cage and chest wall become less elastic. Stiffer chest walls require more strength to allow air to flow in. You lose the reserves you once called upon during hard work or exercise. That is why healthcare providers 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 the 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 an 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.

Measurement of lung volume and airflow over time are recorded and displayed numerically and as a graph. The results are expressed as a percentage and are compared to a large sample of people of your age, gender, 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** (medical term: **supine**). Your healthcare provider measures the vital capacity of your lungs.

VC (Vital Capacity) measures the amount of air you can exhale after a maximum inhalation (in liters). The different measurements include:

- **Forced Vital Capacity (FVC)**, which measures the amount of air you can exhale forcefully after a full inhalation
- **Slow Vital Capacity (SVC)**, which measures the amount of air you can exhale slowly and steadily after a full inhalation
- **FEV₁** (Forced Expiratory Volume), which measures how much air you can forcibly exhale in one second (liters per second)
- **FEV₁/FVC**, which 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:

- **MIP** (Maximum Inspiratory Pressure) is a test of inspiratory muscle strength (internal intercostal and diaphragm).
- **MEP** (Maximum Expiratory Pressure) measures the strength of expiratory muscles (external intercostal and abdomen).
- **PCF** (Peak Cough Flow) measures expiratory flow and ability to protect the lungs by coughing forcefully.
- **SNIP** (Sniff Nasal Inspiratory Pressure) is another test of the diaphragm muscle strength which is measured through a probe in the nose. It may be useful if changes in the strength of mouth and throat muscles lead to inaccurate spirometry test results.

Passive Tests of Lung Function

Tests that involve normal exhalation without direct muscular effort include:

- **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 amount of oxygen in your blood stream.
- **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

Active tests (FVC, MIP, MEP, SNIP, PCF) 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.

An 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 the lower brain stem is disrupted, the muscles in your mouth and throat 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 the accuracy of lung function test results.**

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

- **Phrenic Nerve Conduction Study (PNCS):** The phrenic nerves stimulate your diaphragm. You may have had nerve conduction studies and electromyography, also known as an EMG, as part of your ALS diagnostic work up. This specialized test is done using the same technique. When the phrenic nerves — located on either side of your neck — are stimulated, your diaphragm involuntarily contracts. The contraction can be measured in the size of an electrical impulse (amplitude) from the diaphragm muscle. The lower the amplitude measured, the greater the loss of nerve fibers supplying the diaphragm.
- **Sniff test with fluoroscopy:** Fluoroscopy is a test of moving body parts. It is similar to an X-ray 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 moves. The radiologist can also see if one side of your diaphragm muscle contracts more than the other.

- **Nocturnal oximetry studies:** Your physician may want to measure your oxygen levels while you sleep at night. As your diaphragm becomes weaker, you are likely to breathe shallowly at night. Over time, shallow breathing may lead to impaired exchange of air with significant drops in your oxygen levels, which in turn will lead to frequent awakenings, awakening feeling tired and not refreshed, or with a headache. 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, then accessed and interpreted, and the results discussed with you.

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 ahead 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 without a specific reason like using the bathroom
 - Nightmares
 - Waking with a choking sensation
 - Waking in the morning with a headache
 - Feeling sleepy during the day and needing more naps
 - Increasing overall sleepiness
- **Breathing discomfort when lying flat in bed**
 - Needing to sleep only on your side
 - Needing to sleep elevated on more than two pillows
 - Needing to sleep upright in bed or in a chair
- **Breathing discomfort while sitting or when speaking**
- **Need for noninvasive ventilation at night**
- **Shortness of breath at rest and intermittent use of bi-level positive pressure breathing during the day** to rest lungs and improve energy levels
- **Need for manual and mechanical secretion mobilization** due to a weak cough
- **Need for continuous around the clock noninvasive ventilation**

- **Decision to have a breathing tube** placed surgically into the wind pipe (tracheostomy) and breathing supported by a mechanical ventilator (life support), **OR, decision to select hospice care** so you can be comfortable with noninvasive ventilation 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 (such as asthma or chronic bronchitis), 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 lung 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
- Improve 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 and 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 a manual ventilation Ambu® bag
3. **“High-tech”** strategies include:
- Mechanical In-Exsufflation (M-IE) with a Cough Assist® (Philips Respironics) or Vital Cough® machine (Hill-Rom). 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 like the Trilogy® (Philips Respironics) or Astral® (ResMed).

Lung Health in ALS: Back to Basics

Your lungs have a natural, healthy ecology much like the healthy ecology of a salt marsh or estuary. They have innate immunity protection and adaptive immunity to harmful bacteria 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 airway lining.

The effectiveness of mucus depends, in part, on the balance of sodium, which has a positive charge, and chloride, which has a negative charge, 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.

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 white blood cells that engulf and digest foreign substances (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

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.** Please consult with your physician or healthcare provider prior to starting/receiving any medication.

- Get a pneumonia shot (medical term: **pneumococcal vaccine**) every 5 years.
- Get an annual flu shot (medical term: **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, or smoking).
- **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.)
- **Promote early detection of 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.)

BI-LEVEL POSITIVE AIRWAY PRESSURE (BI-LEVEL PAP)

BREATHING

Bi-level positive airway pressure breathing (bi-PAP) 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 a number of 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. It has become commonplace to use the term BiPAP when referring to this type of assistive device.

More accurate terms are Noninvasive Ventilation (NIV) and Noninvasive Positive Pressure Ventilation (NPPV). NIV or NPPV is a form of Pressure Support Ventilation (PSV).

Ventilation means moving air in and out of the lungs, expanding your airways and air sacs, and improving taking oxygen flow in and letting carbon dioxide out. Portable positive pressure generating devices support your breathing by replacing lung volumes that have been reduced because of a weakening diaphragm.

Pressure support is the difference between the pressure upon breathing in (Inspiratory Positive Airway Pressure or IPAP) and the pressure upon breathing out (Expiratory Positive Airway Pressure or EPAP). And that is why it is referred to as **bi-level positive airway pressure (two pressures:** a higher pressure set to adequately expand the lungs and a lower one to reduce the flow and pressure you must exhale against while maintaining some degree of air sac and lung expansion).

How Bi-Level Positive Airway Pressure Works

You will be fitted with a mask that covers your nose or your mouth and nose, or with nasal cushions that fit in your nose, or some combination. There are many options. A six-foot piece of corrugated plastic 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 to replace your individual lung capacity. As long as 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 bi-level positive pressure 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, or you report symptoms of nighttime shallow breathing (cited 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 and increased shortness of breath with activities calls for a discussion of assisted breathing.**

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 cm H₂O, or
 - PaCO₂ greater than 45 mmHg (increased levels of carbon dioxide), or
 - Oxygen levels (SpO₂) below 88% for 5 minutes while sleeping at night

Adjusting to Noninvasive Ventilation Breathing

Adjusting to assisted breathing may take time. That is why **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.

Initiating the use of breathing support devices may be a challenging decision because it is an acceptance or realization that the breathing muscles are getting weaker, and acknowledging this stage of ALS may be difficult for some people. Talking about these feelings is important—with your respiratory therapist, nurse, physician, partner, spouse, and friends. 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, 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 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: back, sides, flat or elevated.
- **Use your respiratory assistive device at night.** If 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. Table 1 shows some possible issues for first-time users and suggestions for managing them.

Table 1: Breathing Support Issues and Solutions

PROBLEM	CAUSES	SOLUTIONS
Runny nose or nasal congestion	<ul style="list-style-type: none"> • Humidity deficit in nasal and sinus passages due to cool, dry air from NIV 	<ul style="list-style-type: none"> • Increase temperature setting on NIV humidifier • Allow humidifier to warm up before using NIV for the night
Dryness or burning sensation in throat or nose	<ul style="list-style-type: none"> • The body's normal response to inadequate humidity is to increase blood flow to nasal passages (for warmth) and increased mucus production (for moisture). This results in narrowed nasal passages and symptoms of a "cold" or "allergies." 	<ul style="list-style-type: none"> • Irrigate nasal passages before using NIV with a saline moisturizing spray to remove irritating allergens that have collected in the nose during the day. • Saline moisturizing gels with aloe vera may be used inside the nose to increase comfort. • Over-the-counter nasal decongestants (spray or pills) may be used IF you do not have high blood pressure and your physician approves.
Dry mouth upon awakening	<ul style="list-style-type: none"> • Mouth breathing 	<ul style="list-style-type: none"> • If mouth breathing is due to nasal passage blockage, try the solutions above. • If the jaw needs support, request a chin strap or use a mask that covers both mouth and nose. • If mouth breathing occurs only while sleeping on your back, try side sleeping.

PROBLEM	CAUSES	SOLUTIONS
Excess water collects in mask or tubing	<ul style="list-style-type: none"> Condensation forms if there is a large difference between the temperature of air coming from NIV and of the bedroom. 	<ul style="list-style-type: none"> Place NIV tubing under bed covers. Cover NIV tubing with fabric or socks (with feet cut off). Increase the temperature of the bedroom.
Sore or dry eyes	<ul style="list-style-type: none"> Air leaking from mask into eyes 	<ul style="list-style-type: none"> Adjust mask cushion so it seats better on the bridge of the nose.
Mask or nasal cushion leaks	<ul style="list-style-type: none"> Mask or straps not positioned properly 	<ul style="list-style-type: none"> Review manufacturer's suggestion for mask or nasal cushion fitting. Re-seat mask and adjust straps. Do not over-tighten straps. Lie down with the mask and NIV blower on. Loosen Velcro straps until you feel an air leak. Slowly tighten straps equally on both sides just until leak stops. Do this with upper and lower straps.
Soreness or abrasion on bridge of nose	<ul style="list-style-type: none"> Mask cushion too tight or improper fit 	<ul style="list-style-type: none"> Loosen head straps or adjust cushion so it is not pressing on the bridge of the nose. Use a gel cushion or Duoderm to pad bridge of the nose. Switch to alternative mask or nasal cushions and alternate them so nose can heal.
Redness or marks on face when waking up in the morning	<ul style="list-style-type: none"> Skin does not breathe well where the mask cushion sits on the skin. Increased warmth from heated NIV air may cause temporary redness. Mask/head straps are too tight. Sensitivity to mask materials. 	<ul style="list-style-type: none"> Wash face. Redness and marks should go away within 20 to 30 minutes after removing the mask. Be patient. Older skin may not bounce back as quickly due to decreased collagen production. Make sure the mask is not too tight. (See tips above.) Use one of the alternative masks or nasal cushions.

If you have problems not covered here or the suggestions do not work, please contact your homecare respiratory therapist or, the Registered Respiratory Therapist (RRT) on your ALS clinical care team.

ADDITIONAL BREATHING ASSISTANCE DEVICE OPTIONS

Additional Noninvasive Ventilation Options

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



Another type of NIV used for ALS is an **Average-Volume Assured Pressure (AVAP)** device. Like a BiPAP machine, it delivers higher pressures of air to help you inflate your lungs, and lower levels of air while you exhale. The difference between the two devices is that a BiPAP device delivers set pressures, while an AVAP device delivers a certain volume of air (based on your body's needs) and will adjust the pressure to make sure the right volume is achieved. Because the pressure varies based on how much your own muscles are active in breathing, the pressure is generally lower while you are awake. Some people find this more comfortable and easier to get used to.

Some ventilators can be used in an AVAP setting and offer additional features such as a back-up battery (in the event that your power goes out) and a "sip and puff" option. "Sip and puff" refers to a second type of mouthpiece that can be used if you get short of breath with activities like talking or eating. Instead of using a mask (which is still used when sleeping or you need the support for longer periods), your machine can be switched to a straw-like connection. When you need help getting a full breath, you "sip" on the straw and the

machine delivers a full breath of air. Many people with ALS like using this to get full breaths between bites of food while eating or words and sentences while talking. In order to use the sip and puff option (medical term: **open mouthpiece ventilation**), you need to have good facial strength and be able to purse and seal your lips around a large straw.

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 ahead 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 the 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 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 the 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.

Hand Weakness

Individuals with profound hand weakness are less able to tolerate NIV because of the challenge associated with independently putting on and taking off the mask. This can be remedied by having assistance from a caregiver, but calling for help with a mask on is difficult. One solution is to have a switch that can be operated with your head or legs to activate an alert when help is needed.

DIAPHRAGM PACING SYSTEM (DPS)

The **Diaphragm Pacing System (DPS)** was approved for use as a Humanitarian Device by the FDA in 2011. The safety and potential benefits of this technology are still being investigated through clinical trials. DPS may have the potential to reduce the loss of vital capacity in ALS through therapeutic electro-stimulation of the diaphragm in some patients. If you are interested in learning more about DPS, please speak to a knowledgeable physician at your clinic.

How DPS Works

- Four electrodes are surgically placed on the diaphragm, attached like little suction cups.
- Wires attached to these suction cups are fed through a small incision in your abdominal area and attached to a box that provides the electrical stimulation.
- When an electrical impulse stimulates the phrenic nerve, the diaphragm contracts and moves downward, allowing air to flow into the lungs.
- The goal of therapeutic DPS is to preserve diaphragm muscle mass and delay respiratory failure. **It is not a substitute for bi-level positive pressure breathing, although it may reduce the amount of pressure.**

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 the absence of a lung disease that affects your air sacs (alveoli), the reason for decreased oxygen levels in ALS is lung muscle weakness. If you breathe shallowly, you are not able to exchange air and get rid of carbon dioxide effectively. Rising carbon dioxide levels replace oxygen and impair access to adequate levels of oxygen from the air around you.

That is why the treatment for shallow breathing (and its resulting decreased oxygen) is to give you a bigger breath, through bi-level noninvasive positive pressure breathing at night and breath stacking, mouthpiece ventilation, or manual or mechanical sigh breaths during the day, as needed. This way oxygen, a component of the air we breathe, will find its way to all areas of the lungs, especially the smallest bronchioles and air sacs at the bottom and sides of the lungs.

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 benefit. **In ALS, oxygen administration may be detrimental** if it is used in place of bi-level positive pressure breathing support. Normally, your body takes high levels of carbon dioxide as a signal to initiate a breath. Over time, if you have gotten used to higher-than-normal levels of carbon dioxide in your lungs and blood stream, your body may rely on a low level of oxygen to know when to take a breath. Supplemental oxygen may get rid of this signal for breathing and result in impaired breathing. The result can be profound sleepiness, and lack of clear thinking or disorientation.

Make sure those who suggest and prescribe oxygen are very knowledgeable about ALS and familiar with guidelines for safe and effective treatment.

ADVANCED DECISION MAKING ABOUT RESPIRATORY SUPPORT

ALS is a devastating diagnosis. Physicians and all staff members of ALS multidisciplinary clinics know this and will support you and your loved ones at every stage of this disease. Healthcare providers will discuss all your medical and treatment options and assist you in preparing for challenges associated with the progression of the disease. They will give you reassurance, realistic expectations, and hope for the best possible life with ALS. **As ALS progresses, the healthcare providers' 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, communicated in terms that you fully understand and comprehend. **Making decisions about accepting or forgoing invasive mechanical breathing support and communicating your wishes 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 controlled no matter which decision **you** make.

The most difficult situations arise when you don't have a plan. You do not want to find yourself in the emergency department 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 3-part video series on respiratory decision making in ALS is available through your regional ALS Association chapter or the national ALS Association website. Viewing it with your family and loved ones and discussing your reactions with your clinic team can be very useful. **In the video, people with ALS 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, your ALS care team will support you in coordinating the care you need.

Before going into surgery and accepting 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.

Checklist: Considerations for Home Care with Mechanical Life Support

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.

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 9-1-1 call?

This list is not meant to be daunting. Your safety is of most importance. 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 may be 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 time 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:** on-going 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
- Have critically impaired respiratory function (with or without the use of noninvasive breathing assistance)
- Have severe nutritional insufficiency (with or without a feeding tube)

Medicare defines critically-impaired respiratory function 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 Ventilation (NIV) at the End of Life

At the end of life in ALS, noninvasive assisted breathing 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 blood stream in spite of continuous or nocturnal use of breathing assistance. In the last weeks or months of life, your oxygen needs may not be met by using NIV. The addition of oxygen to your bi-level positive pressure device may help you be more comfortable.

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 interdisciplinary team members and care managers, will work with you, your family, and other 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

By: Lee Guion MA, RRT, FAARC

Publisher: 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.

Amyotrophic Lateral Sclerosis: A Patient Guide for Clinicians

Edited by: Richard S. Bedlack and Hiroshi Mitsumoto

Chapter: Respiratory Therapy: Natural History, Measurements, and Treatments

By: James B. Caress, MD and Connie C. Paladenech, RRT, RCP

Publisher: Demos Medical Publishers, 2013

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

Video

Respiratory Decisions in ALS

Three-part video developed in cooperation with the ALS Association, Forbes Norris MDA/ALS Research and Treatment Center, and Les Turner ALS Foundation.

Covers developing a care plan, respiratory support options, and making decisions for your quality of life. (available through The ALS Association National Office or local chapters)

Website

Army and pALS

www.AmyandpALS.com

Website 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 who authors the interactive "Breathing Lessons" blog which covers breathing solutions and links to videos on breathing exercises such as breath stacking.



The following is a list of topics covered in the *Living with ALS* resource guides:

Resource Guide 1

What is ALS? An Introductory Resource Guide for Living with ALS

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

Resource Guide 2

After the ALS Diagnosis: Coping with the “New Normal”

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

Resource Guide 3

Changes in Thinking and Behavior in ALS

This resource guide addresses how thinking and behavior may be affected by ALS and how these changes can impact disease course, symptom management, and decision making.

Resource Guide 4

Living with ALS: Planning and Making Decisions

This resource guide reviews areas where careful planning and decision making will be required and will provide you with resources to help you and your family plan for the future.

Resource Guide 5

Understanding Insurance and Benefits When You Have ALS

This resource guide provides strategies and helpful hints to better navigate health insurance and benefits. While understanding insurance and benefits may feel overwhelming, the guidelines outlined here should help simplify the process for you.

Resource Guide 6

Managing Symptoms of ALS

This resource guide discusses a variety of symptoms that may affect you when you have ALS. As the disease progresses, various functions may become affected and it is helpful to understand potential changes so that you know what to expect and how to manage these new changes and symptoms.



Resource Guide 7

Functioning When Mobility is Affected by ALS

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

Resource Guide 8

Adjusting to Swallowing Changes and Nutritional Management in ALS

This resource guide will help you understand how swallowing is affected by ALS and what you can do to maintain nutrition for energy and strength and to keep your airway open.

Resource Guide 9

Changes in Speech and Communication Solutions

This resource guide covers how speech can be affected by ALS and explores a variety of techniques, technologies, and devices available for improving communication. By maintaining communication with others, you continue to make a significant difference in their lives, while retaining control of your own.

Resource Guide 10

Adapting to Changes in Breathing When You Have ALS

This resource guide explains how breathing is affected by ALS. Specifically, it will teach you the basics of how the lungs function, the changes that will occur, and how to prepare for the decisions that will need to be made when the lungs need maximal assistance.

Resource Guide 11

Approaching End of Life in ALS

This resource guide examines thoughts and feelings about dying and end of life. Approaching end of life is difficult and support is critical to help sort out feelings, expectations, and plans. By talking to friends, family, professionals, and planning and communicating your wishes, you can help prepare for the best possible end-of-life phase.



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About The ALS Association

The ALS Association is the only national non-profit organization fighting Lou Gehrig's Disease on every front. By leading the way in global research, providing assistance for people with ALS through a nationwide network of chapters, coordinating multidisciplinary care through certified clinical care centers and fostering government partnerships, The Association builds hope and enhances quality of life while aggressively searching for new treatments and a cure.

For more information about The ALS Association, visit our website at www.alsa.org.