

# Maintaining Pulmonary Function With Amyotrophic Lateral Sclerosis

Pulmonary function is how well your lungs work. With amyotrophic lateral sclerosis (ALS), muscles weaken over time. This can affect the respiratory muscles that maintain pulmonary function.

Individuals with ALS typically have healthy lungs, unless they have an underlying lung disease, such as chronic obstructive pulmonary disease (COPD). If you have a pre-existing lung disease and/or use inhalers, it is important to discuss this with your physician and/or pulmonologist, so they can perform breathing tests. When performing a breathing test, such as spirometry or pulmonary function test, your forced vital capacity (FVC) or slow vital capacity (SVC) may be lower than expected due to weakened muscles.

It is important to see a pulmonologist even before you notice any loss of strength, so the doctor can follow you over time. It is good to get out of bed daily, stay active, and do activities that involve taking deep breaths, such as yoga or meditation.



## How ALS affects breathing

ALS commonly affects the respiratory system. The nerves that control the breathing muscles do not work correctly, and the muscles become weak. Weak breathing muscles can make it challenging to take a deep breath. Individuals with ALS start to breathe shallowly as a result.

When you take shallow breaths, you do not exchange the air in your lungs well. The “bad” air, carbon dioxide (CO<sub>2</sub>), tends to stay in your lungs, and the amount of CO<sub>2</sub> increases over time. This prevents the “good” air, oxygen (O<sub>2</sub>), from getting into your lungs. The body compensates by breathing faster. When your body is working hard to breathe fast, you feel tired and your respiratory muscles get tired. This can lead to respiratory failure.



## Keeping lungs healthy

It is important to have good hygiene and a healthy lifestyle, which can help prevent lung infections and slow down the decline in respiratory function.

Follow these tips to maintain lung health:

- ▶ Wash your hands often with soap and water. Alcohol-based hand sanitizer is a good alternative if you can't wash hands.
- ▶ Avoid crowds during cold and flu season.
- ▶ Avoid cigarette smoking, vaping, and other chemical inhalants. These can damage your lungs, worsen your airway clearance, and increase the chance of respiratory infections.
- ▶ Avoid exposure to secondhand smoke, chemicals in the home and workplace, and radon. These can worsen lung disease.
- ▶ Make your home and car smoke-free zones.



# Medical Equipment You Can Use at Home

Some respiratory devices can be helpful with ALS. Using a device for your breathing will help you take deeper breaths and rest your breathing muscles.

Equipment needs should be determined in consultation with your doctors. Breathing devices are typically prescribed when your FVC/SVC is less than 50% (a Medicare guideline) or if a sleep study shows a need for nighttime breathing support. This section provides examples of types of equipment an individual with ALS may use.

**Breathing machines:** Also called ventilators, these machines can deliver bilevel positive airway pressure (BiPAP) or volume-targeted ventilation. When the ventilator is used with a nasal mask or face mask, it is called noninvasive ventilation. A BiPAP device is typically a rent-to-own machine, while a volume-targeted machine usually is rented continuously for a lifetime. A volume-targeted machine generally is supported by a respiratory therapist who can come to your home to check on the machine and you.



A BiPAP machine has two pressures, one for inhalation and one for exhalation.



A cough assist device can help you clear your airways and keep mucus and germs out of your lungs.

**Cough assist devices:** Having weak respiratory muscles can affect your ability to cough and sneeze. This can make it hard to clear mucous out of your airways and can lead to pneumonia or respiratory infections. To prevent infections, you may need to use a cough assist device. This device will help you cough effectively and move the mucous out of your lungs or the back of your throat.



Portable suction machines are lightweight and easy to move.

**Suction machines:** If you experience drooling or the inability to clear mucous from your mouth, a suction machine can help. This does not stop the drool from happening, but it helps to remove it from the mouth.

## Insurance coverage

Although some devices need to meet certain criteria, respiratory devices are covered by almost all insurance companies. Depending on your insurance coverage, you may be responsible for a deductible or copay. Your pulmonary care team will help with obtaining insurance coverage when breathing devices are needed.

## Learning to use pulmonary equipment at home

When you begin using a new type of respiratory device, your providers will offer training in clinic. In addition, a respiratory therapist from a home care company may come to your home to help set up your pulmonary equipment and train you and your caregivers on using it. Companies that supply pulmonary equipment have on-call providers to help with training and troubleshooting the devices. You can always reach out to your healthcare providers if you have any questions.

# Preparing for an MDA Care Center Visit With Your Pulmonary Team

People living with ALS generally are in the clinic every few months. Here's what you need to know to be ready for pulmonary visits.

## What to expect

During your visit, providers will take a medical history, perform a physical examination, measure lung function (pulmonary function tests), and perform other studies as needed.

## Common tests

If your pulmonologist suspects your breathing may be too shallow at night (nocturnal hypoventilation) or you have sleep apnea, they may order a sleep study. Sometimes, the pulmonary team will perform other tests, including lung imaging, blood tests, oxygen/carbon dioxide measurements, swallowing/feeding evaluation, and microbiological studies. The most common symptoms indicating insufficient breathing (underventilation or hypoventilation) are fatigue, poor sleep, nightmares or night terrors, and headaches, especially right after waking. Tell your pulmonary team if you experience any of these symptoms, if your cough gets weaker, or if you have any questions/concerns about your medication, medical equipment, or treatment plan.

## What to bring

Bring any breathing equipment you use at home with you to your clinic visit. Your pulmonary team might adjust the settings of these devices while you are at the clinic and troubleshoot any possible issues.

## How to prepare

Before your clinic visit:

- ▶ Confirm your visit with the clinic.
- ▶ Write a list of your questions and concerns, and bring them with you.
- ▶ Gather any medical documents you may need.
- ▶ Prepare any medicines, durable medical equipment, and emergency supplies you need to bring with you.
- ▶ Arrange your transportation.
- ▶ Request an interpreter if you need one.



# Maintaining Your Pulmonary Function at Home

Breathing consists of two steps: inhalation and exhalation. Each step requires several different muscles. Breathing therapies, exercising, healthy eating, and infection prevention are all important factors in maintaining respiratory function for people with ALS.

Your doctor may refer you to outpatient physical therapy (PT), a nutritionist, or other healthcare providers who can help with these factors. But there also is a lot you can do at home to promote good breathing capacity:



**1** Your provider might recommend breath stacking therapy, also called lung volume recruitment. The idea is to stack one small breath on top of another until the lungs are filled. Filling your lungs to their maximum capacity can increase lung volume, cough effectiveness, lung and chest suppleness, and speaking volume. You can practice breath stacking with a mouthpiece or self-inflating bag and mask (manual resuscitator bag).

**2** It is important for an individual with ALS to be aware of their weight. You can lose weight when your body is working hard to breathe. Using noninvasive ventilation can create bloating. If either is occurring, discuss the issue with your healthcare team so they can help determine the cause and adjust ventilation settings if needed.

**3** Eating a large meal can put pressure on the diaphragm, which can make it harder to breathe. Eating small meals more frequently can help alleviate this. It is also important to make sure you do not eat while using noninvasive ventilation, as this can allow food to enter the airways and lead to aspiration pneumonia.

**4** Try to get out of bed every day, as sitting and standing up can help your lungs expand better and help prevent pneumonia.

**5** Staying as active as you can is important. Get some fresh air, enjoy your family and friends, sing, and try assisted yoga.

**6** A good night's sleep can give you more energy and improve concentration.

**7** It is important to keep up with your vaccinations, including the pneumococcal and annual flu vaccines. Talk with your doctor about you and your family getting the flu shot every year.

**8** Oral hygiene is important to prevent bacteria in the mouth that can cause chest infections. In addition to brushing and flossing your teeth twice a day, visit your dentist regularly.

**9** In case of a respiratory emergency, it is important to know what to do and who to contact. Make an emergency plan with your family and pulmonary team, and make sure your family members and caregivers have a copy.

# Frequently Asked Questions

## How do I know if I'm experiencing respiratory complications?

It is essential to let your doctor and/or respiratory therapist know if you feel short of breath. Sometimes you'll notice this first when you lie down at night. Elevating your head can help. Some individuals with ALS feel short of breath when talking, showering, or bending over.

Daytime sleepiness can be a sign that you are breathing too shallowly at night and not getting restful sleep. A weak cough or sneeze can also indicate respiratory muscle weakness.

## How do I know if I need ventilation?

You need ventilation if your FVC/SVC is low. Other signs of needing ventilation are shortness of breath, daytime sleepiness, or having to use your neck, shoulders, or belly muscles to breathe. Medicare guidelines for needing ventilation are FVC/SVC less than 50% or a positive sleep study.

## Can I travel with ventilation?

Yes, you can. Make sure you and your caregivers know what to bring with you and what to do in an emergency. Long trips and air travel are possible with careful

planning involving you, your caregivers, your medical team, and airlines or transportation providers.

## How do I eat while using ventilation?

It is important to remove your mask or nose piece before eating. The ventilator could be harmful if you were to swallow something the wrong way or choke. The air pressure coming from the machine can push the food or drink into your lungs, which could lead to pneumonia.

## How can I prepare for an emergency?

The more prepared you are for emergencies, the better you can handle them. Keep records of your recent visits to the doctor, test results, medications you take, doctors' names and contact information, and insurance information. It is a good idea to have all this information stored as hard copies in a file and digitally on your smartphone or portable device.

Determine your emergency contacts. During visits with your healthcare team, make a plan to handle respiratory emergencies. Learn the emergency phone numbers of the clinic and hospital. Prepare an emergency bag of

clothes, medical supplies, and medications, and bring your respiratory devices with you to the hospital if you can. If you have a hospice team, you can contact them for support. Call 911 if you experience significant respiratory symptoms.

## What should I know about anesthesia?

If you need to undergo anesthesia, your doctor should be made aware of any breathing difficulties you have. You should also let them know if you use any breathing devices. Most people who require anesthesia will need to be able to lie flat. Discuss this with your pulmonary team for clearance prior to any procedures that require anesthesia. Using noninvasive ventilation after anesthesia can be helpful if you feel groggy and weak.

This resource was developed with the expertise and knowledge of Kristina Dzwonchyk, regional director of clinical services at AdaptHealth and member of the Board of Directors, Connecticut Society for Respiratory Care.

**To learn more about ALS, visit [mda.org](http://mda.org) or contact the MDA National Resource Center at 833-ASK-MDA1 (275-6321).**

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