

## What are some assistive devices for helping me cough and clear secretions?

The muscles used to breathe are the same ones that help you cough. It may be necessary to use devices that help clear phlegm and secretions when you are unable to cough productively.

Mechanical insufflation-exsufflation machines, like the CoughAssist, are useful in clearing secretions by helping you cough. It is done by applying positive pressure then rapidly applying negative pressure to your airway. This quick change in pressures stimulates a cough. Air is delivered through a facemask, mouthpiece, or tracheostomy tube.

Suction machines are also used to remove excess secretions. A Yankauer tube, similar to what dental offices use, is attached to the suction machine and is inserted into your mouth to remove excess saliva secretions or mucus you cough up from your lungs. If you have a tracheostomy tube in place, a tiny catheter is inserted into the trachea to remove the lungs' mucus secretions. Caregivers must be taught how to perform this sterile technique.

Other devices, like The Vest, apply pressure to the chest wall and vibrate rapidly, which loosens mucus and moves it to larger airways, where it can be cleared by coughing or suctioning.

Medications can also be prescribed. Expectorants thin out secretions and make them easier to cough up. Anticholinergics decrease the amount of saliva produced.



A CoughAssist machine attached to a full face mask.

## Is a tracheotomy necessary?

When weaknesses of the mouth and throat muscles prevent you from breathing on your own and also prevent you from clearing mucus and respiratory secretions, a tracheotomy may be necessary. A hole is surgically created in the trachea (the windpipe) and air from a ventilator is delivered through a tracheostomy tube. Because it requires a surgical procedure, it is considered invasive ventilation.

## What are other ways to keep my lungs healthy?

To avoid infections, practice good hand hygiene techniques, like washing your hands with warm water and soap. Caregivers should also wash their hands or use an alcohol-based hand sanitizer before and after providing care.

It is recommended that you get an annual flu vaccine in the fall and a pneumonia vaccine. Also, contact your health care provider if you have symptoms of pneumonia, which include coughing, sputum production (may be clear, yellow, or green), chest discomfort with breathing, fever, and/or chills.

If you have difficulty with choking or coughing while eating or drinking, you should notify your health care providers. They may suggest using a thickener such as Thick-It or SimplyThick, to prevent food and liquid from leaking into your lungs.

For more information about ALS or to schedule an appointment with the ALS Pulmonary Clinic at The Johns Hopkins Hospital, please call: (410) 955-8511.

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# Caring for Your Lungs While Living with Amyotrophic Lateral Sclerosis (ALS)

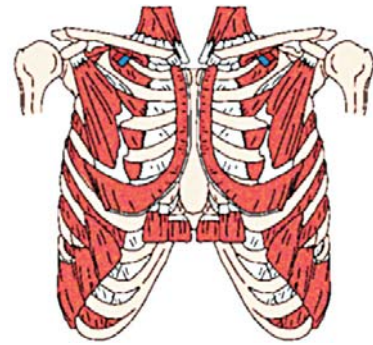


The Johns Hopkins University  
Amyotrophic Lateral Sclerosis Clinic

## Why is caring for my lungs important with ALS?

In ALS, the motor nerve cells die. When the motor nerve cells are no longer able to send impulses to the muscles, the muscles waste away, which causes muscle weakness.

The diaphragm (an arched muscle just below the lungs) and the intercostal muscles (muscles between the ribs) help us to breathe by assisting with air movement in and out of our lungs. Breathing is normally an easy and automatic action, but when the breathing muscles become weakened, it takes more energy. It may cause you to feel tired and will challenge your ability to fight off respiratory infections, like a cold, the flu, or pneumonia.



The breathing muscles.

## What tests are involved to determine how well my lungs are working?

Pulmonary Function Tests (PFTs) are done to determine how well your lungs can move air in and out of your lungs. Spirometry is a type of PFT that measures the amount and the speed of air you can forcefully exhale. It is tested by applying noseclips to prevent air leakage, and then blowing into a mouthpiece that is connected to a device that reads the air flow. The air flow is measured as the Forced Vital Capacity (FVC), and is commonly used as a reasonable overall measure of your respiratory function. Your FVC is expressed as a percent of the predicted value based on values from a healthy reference group.

You may also be asked to do spirometry while lying down, which is important to measure because it is more difficult to breathe while lying down. If your FVC is much lower while lying down (more than 10%), it usually means there is diaphragm weakness.

Peak cough flow is another test, which measures the force of a cough. You will be asked to cough into a peak flow meter, a handheld device that has a mouthpiece on one end and a scale with a moveable indicator on the other. A "normal" measurement depends on your height, age, gender, and stage of disease.

Maximum Inspiratory Pressure (MIP) and Maximum Expiratory Pressure (MEP) measure the strength of your respiratory muscles while inhaling and exhaling into a closed airway. You will be asked to inhale and exhale into the mouthpiece of a device called a manometer, which reads the pressures.

Pulse oximetry is a noninvasive measurement of the oxygen saturation of your blood and is measured by clipping a photoelectric device to your finger. Normal oxygen saturation is 95% or higher.

Capnography is a measurement of carbon dioxide at the end of exhalation. It is used to assess whether your lungs are adequate in removing carbon dioxide from your body. You will be asked to breathe normally into a nasal cannula or mask that is hooked to the capnography device. Normal values are 35-45 mmHg.



Spirometry being performed.

## What if my lungs are weak and I need help with breathing?

Based on the results of the tests and on how you feel, you and your health care provider may decide that noninvasive ventilation (NIV) is necessary. NIV will help you move air in and out of your lungs to reduce the amount of energy you have to exert. Bilevel positive airway pressure (BiPAP) machines are one type of NIV, which provide air at two pressures: one for inhalation and one for exhalation. How much you will need to use the BiPAP machine depends on the extent of your muscle weakness.



A BiPAP machine.

## What type of masks do I have to wear with the BiPAP machine?

Air from the BiPAP machine is delivered through various types of masks.

Nasal interfaces include nasal pillows and nasal masks. Nasal pillows fit into your nostrils and deliver air directly into your nose. It is compact, secured by headgear and chin straps that keep your forehead and vision area open, and enables side sleeping at night. Because it fits into the nostrils, irritation may develop in the nasal passages after long periods of use. To reduce irritation, warm humidification is also delivered with the BiPAP.

Nasal masks cover your entire nose. It is less compact, secured by headgear, and may block some areas of vision. However, it causes less irritation of the nasal passages.

Oral interfaces include mouthpieces, which deliver air through your mouth and can be useful during the daytime or during a cold with nasal congestion, when less air is able to be delivered through your nose.

Oronasal interfaces are considered full facemasks that fit over your nose and mouth. They are useful if you have air leakage through the mouth, especially with bulbar dysfunction (weakness of the muscles that causes difficulty with swallowing, speaking, keeping the mouth closed, and using the tongue). Because oronasal masks cover both your nose and mouth, it is important that you have use of your arms and hands to reduce the risk of aspiration of vomit or other injuries.

All interfaces are fit to your comfort. It is not unusual to have more than one interface to switch during the day to minimize irritation and to maximize comfort.



A full face mask, nasal pillows, and a nasal mask.