

ALS & BREATHING

A Les Turner ALS Foundation Guide for People Living with ALS

Contents

How ALS affects breathing	3
Symptoms of respiratory muscle weakness	4
Breathing evaluations	5
Oxygen and ALS	6
Mucus	6
Runny nose, oral care and extra saliva	7
Airway clearing devices	9
Cold or respiratory virus	10
Non-invasive therapies	10
Inclined support	10
Breath stacking	11
Non-invasive ventilation	11
Invasive ventilation (Tracheostomy)	13
Preparations for power outages	15
Travel	15
Palliative or hospice care	15
Learn more	16

Disclaimer Statement: The information in this guide is not medical advice. Talk to your ALS care team before making any decisions about your health or treatment. Together, you and your care team can find a treatment plan that works for you.



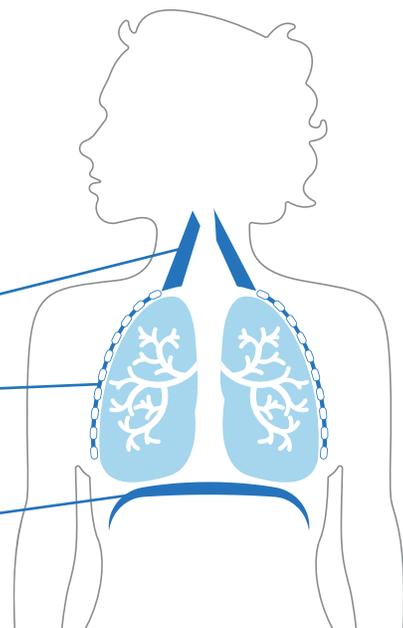
How ALS affects breathing

At some point, ALS will affect the muscles used for breathing. These are called “respiratory muscles” and they mainly include the:

muscles of the upper airway

intercostal muscles
(groups of muscles that run between the ribs)

diaphragm
(primary breathing muscle)



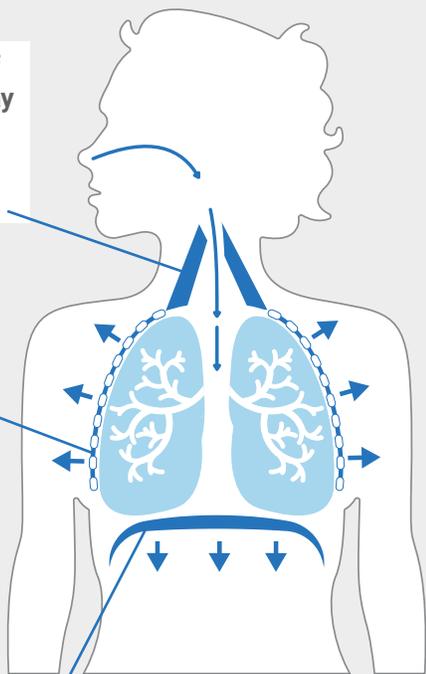
When you inhale...

(inspiration / breathing in)

The muscles of the upper airway contract to elevate the sternum.

The intercostal muscles contract to elevate and expand the ribcage.

The diaphragm contracts to move down and generate flow.



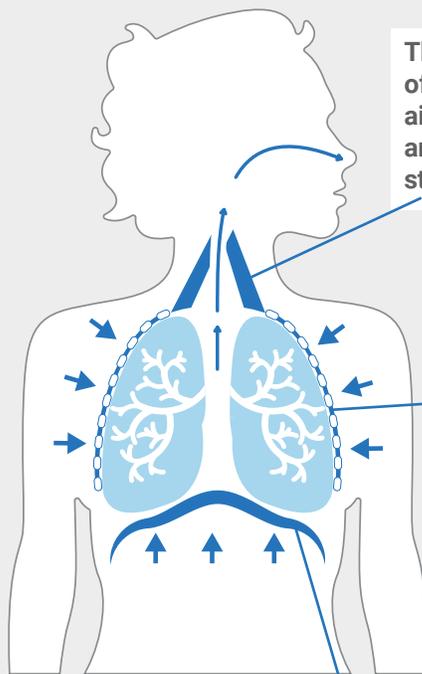
When you exhale...

(expiration / breathing out)

The muscles of the upper airway relax and depress the sternum.

The intercostal muscles relax to depress the ribcage.

The diaphragm relaxes to move up.



In some people with ALS, respiratory muscles may be affected late in the disease; in others, these muscles are affected early in the disease.

Symptoms of respiratory muscle weakness

There are several signs and symptoms of respiratory muscle weakness. These include:



Waking up tired and feeling groggy



Shortness of breath when lying flat



Daytime sleepiness or tiredness



Changes in voice (tone and volume)



Morning headaches



Restless sleep (tossing and turning)



Weak cough



Not feeling hungry



Acid reflux (a burning feeling in the chest or throat caused by stomach acid)



Feeling more irritable and forgetful than usual



Having more saliva than usual



Feeling less interested in things



Nausea or bloated stomach



When ALS affects respiratory muscles, it results in “respiratory muscle weakness.”

People who have “respiratory muscle weakness” have lost strength in the muscles that help them breathe.

If you experience any of these symptoms, tell your ALS care team.

Breathing evaluations

You may have your breathing checked at ALS clinic visits. To do this, there are several tests health care providers might use.

Breathing test	What the breathing test measures	How the breathing test is done
 <p>Forced Vital Capacity (FVC)</p>	<p>The strength of the diaphragm muscle.</p>	<p>You are asked to breathe out into a machine called a spirometer.</p>
 <p>Maximum inspiratory pressure (MIP)</p>	<p>The strength of the diaphragm and the intercostal muscles (groups of muscles that run between the ribs).</p>	<p>You are asked to breathe into a spirometer.</p>
 <p>Overnight oximetry</p>	<p>The amount of oxygen in your blood. It can also check for changes in breathing during sleep.</p>	<p>This is a test that is done at home. You are asked to wear a comfortable sensor (provided by a respiratory company) on your finger while you sleep.</p>
 <p>Arterial blood gas (ABG)</p>	<p>How much oxygen and carbon dioxide is in the blood.</p>	<p>Health care providers take a blood sample from you.</p>
 <p>Sleep study</p>	<p>Breathing, oxygen levels, leg movement and sleep cycles while you are sleeping.</p>	<p>In some cases, a sleep study might be needed. Health care providers use these studies to help them find out what is causing the problem and how best to treat it. A sleep study can be done at home or in the hospital.</p>

Oxygen and ALS

When people living with ALS have low oxygen levels, it is usually because they have respiratory muscle weakness.

The muscles that control your breathing may be too weak to breathe in enough oxygen and breathe out enough carbon dioxide. Because of this, you may have higher than normal levels of carbon dioxide in your blood.

Your care team will usually recommend a recommend “Non-invasive ventilation” or NIV (special breathing equipment). **These machines help the respiratory muscles breathe in oxygen and breathe out carbon dioxide.** If oxygen levels are still low, supplemental oxygen may be added to these machines with an order from a pulmonologist.



Using “supplemental” or extra oxygen will usually not help people living with ALS.

In fact, it can be very dangerous.

Supplemental oxygen can trick the brain into thinking that there is no need to breathe.

Mucus

You may have difficulty clearing “thick secretions,” or mucus, from the back of your throat. This is because your respiratory muscles are not strong enough to cough it out. Yet the buildup of thick secretions can lead to even more problems with swallowing and breathing. There are several ways to prevent this:



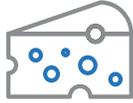
Hydration

Drinking more water – even just a glass or two – can help make secretions thinner and easier to handle. Water is the best option, though anything that does **not** contain alcohol or caffeine may also help.



Humidity

A room humidifier, either hot or cold, adds extra moisture to the air. The extra moisture helps reduce secretions and also helps prevent dry mouth. The humidifier should run 24 hours a day in the room where you spend most of your time. If there are pets in the household, a humidifier with a HEPA filter is recommended.



Reduce Dairy Intake

The protein in dairy products can increase thick secretions in the throat. Because of this, it may be helpful to reduce dairy intake.



Medications

Some oral or nebulized medications can help thin secretions if the suggestions listed above are not effective.



Nebulized Saline and Medication

A nebulizer is a machine that turns a liquid into a mist that can be inhaled through a mouthpiece or face mask. Nebulized saline or saltwater may be prescribed first to thin secretions enough for them to be coughed or suctioned out. Saline is not commonly covered by insurance, but it can be made at home or ordered online. If saline isn't effective, medications may be requested to help make the airways bigger so secretions can move.

Runny nose, oral care and extra saliva

You may find it hard to blow your nose, keep your mouth clean, or remove extra saliva. These problems can lead to further trouble with breathing. There are several ways to help.

How to reduce thin secretions in your nose



Sinus Rinse Products

Certain products, such as a WaterPik™ System, can help keep sinuses and nasal passages open. A bulb syringe or nasal suction may also help.



Medications

Medications, such as antihistamines, can help dry-up nasal passages.

How to keep your mouth clean



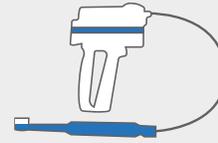
Diluted mouthwash

Mouthwash can help keep bacteria from building up in the mouth. A mouthwash made with baking soda is especially effective.



Mouth swabs

Mouth swabs can be used to clean out mucus from the mouth. This should be done at least twice a day, in addition to routine brushing of the teeth and tongue. This should be done even if the person is not eating or drinking by mouth.



Suction toothbrushes

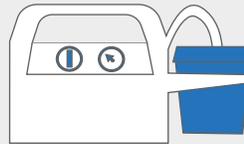
Products such as Waterpik™ and Plak-vac suction toothbrushes can help clean and suction out the mouth.

How to remove extra saliva



Juice

Use purple grape juice to swab out your mouth. You can also use papaya or pineapple juice or take supplements to thin out the saliva. This can help prevent choking or drooling.



Oral suction wand

Oral suction wands can make it easy to clear out saliva. The wand should be used in the mouth, not in the throat.



Medications

There are several medications that might help, but they can have the side effect of dry mouth.



Blocking saliva glands

To prevent the body from making too much saliva, some people will choose an option that requires a doctor's visit. These options include Botox™ or Myobloc™ or radiotherapy.

Airway clearing devices

Airway clearance devices are equipment that can be used to help remove secretions and make it easier to breathe. These airway clearance devices are listed below and would need to be ordered by your physician. Cost will depend on your insurance coverage.



Chest Physiotherapy

Respiratory therapists use this method to move secretions out of the outer airways. They cup their hands and lightly pound over the chest and back.



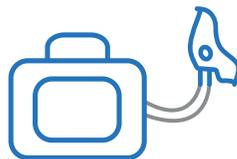
High-Frequency Chest Wall Oscillator

This device makes tiny bursts of air that move mucus out of the airways. But it is sometimes hard for people when they cannot move as much or breathe as deeply on their own.



Suction

Suction machines use a tube, similar to those used at the dentist's office, to remove secretions from the mouth and the back of the throat. For deeper suctioning, a different device may be recommended by a physician. If you cannot cough or spit out secretions, these machines can help. Suction machines can be used in the home, and there are also battery-operated ones for use when traveling.



Cough Devices

Cough devices help people cough when their muscles are too weak to cough on their own. CoughAssist™ or VitalCough™ are two available types. A suction machine may also help remove any secretions missed by the cough device.

Your ALS Guide has short educational respiratory videos for you to learn more about equipment and managing respiratory symptoms.

YouTube Videos:
bit.ly/als-respiration
or youralsguide.com

Colds or respiratory viruses

It is really hard for people living with ALS to get rid of the extra mucus that comes with a cold or virus. If you have a cold but do not have an airway clearance device, you should contact your ALS care team.



It is also recommended to get a flu shot every year and stay up to date on all recommended vaccines.



When someone living with ALS catches a cold, it is important to use airway clearance devices. These devices can help keep a cold or virus from turning into something more serious like pneumonia.

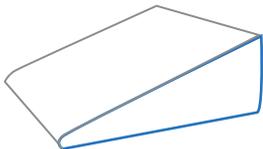
Non-invasive therapies

A number of “non-invasive” therapies can be used to help you breathe comfortably.

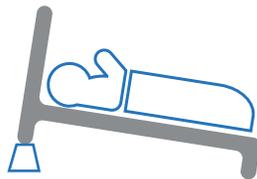
Inclined support

People living with ALS may have trouble breathing while lying down flat. It might be helpful to use inclined support.

These could include:



Using a wedge pillow



Raising the head of the bed



Sleeping in an electric reclining chair or power wheelchair

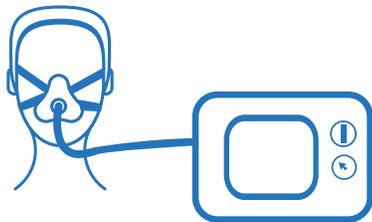
Breath Stacking

- Breath stacking is a daily exercise that you can do to help open and expand your lungs.
- It is a way to fill your lungs with more air than you usually take in.
- It can be done with or without a device called an Ambu™ bag with a one-way valve.

Why is it important?

- When breathing muscles weaken, the lungs get smaller. That makes it hard to breathe in enough oxygen and breathe out enough carbon dioxide.
- Breath stacking helps to keep the lungs open and the chest muscles flexible so you can breathe better.
- It also helps keep the lungs big and strong enough to remove secretions.

Non-invasive ventilation



“Non-invasive ventilation” or NIV is the use of special equipment to support breathing. Non-invasive means that it does not break the skin or require any surgery. It is also the preferred and most used form of ventilation in ALS care.

People sometimes refer to non-invasive ventilation by other names. The most common ones are:

- **BiPAP** (Bi-level Positive Airway Pressure) machine
- **VPAP** (Variable Positive Airway Pressure) machine
- **Home ventilator** (portable with internal battery)

Why is NIV important?

- When breathing muscles get weak and tired, NIV can help.
- The goal is to rest the respiratory muscles so that they don't get too tired.



How to stack breath

To do breath stacking, **sit tall and take 3-5 slow, deep breaths from the mouth without exhaling.** This should be done twice a day.

A nose clip can help make sure air doesn't escape from the nose while you are breath-stacking.

If it is hard to breathe, an Ambu™ bag with a mask can help.



Ambu™ bag

Watch this video, bit.ly/ambu-bag for instructions or ask your pulmonologist during a clinic visit about breath stacking.



To learn about NIV, answer a few questions to help you think through what is most important to you and get ready to talk with your ALS care team about your options.

alsdecisions.org/breathing

- Resting these muscles helps improve sleep, energy and communication.
- NIV may also help keep the body from collecting too much carbon dioxide if you don't take enough breaths while sleeping.
- NIV may help people live longer with ALS.

How does NIV work?

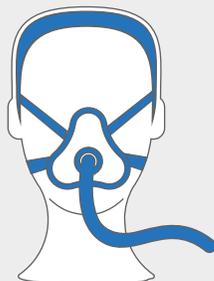
NIV uses air pressure to help you breathe. Here's how it works:

- You'll wear a mask that's attached to a device. There are a few different types of masks, so you can choose the one that's most comfortable for you. Some masks cover just your nose, and others cover your mouth and your nose.
- When you wear the NIV device, it helps you get more air into your lungs, so you can breathe more easily.
- Some people use NIV all the time. Some people only use it at night or for short periods during the day. As your needs change, your ALS care team may recommend that you use NIV more often. Your care team may also change the amount of air pressure that you get through the NIV device.

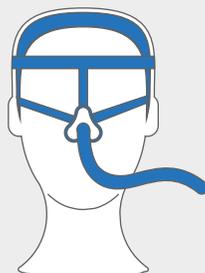
Feeling uncomfortable while wearing the NIV device

Wearing the NIV device may be uncomfortable at first. Some people find that the mask irritates their nose or makes their throat feel dry. Some people feel bloated when they wear the device. Your care team can help you make adjustments so that it's more comfortable for you to wear the NIV device.

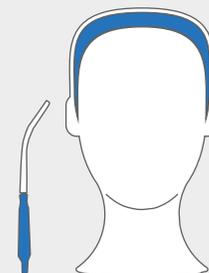
Some of the available masks or interface options for NIV



Full face mask



Nasal mask



Sip and puff handheld

Talk with your ALS care team to choose the best option for you.

Invasive ventilation

You have the option to choose between non-invasive and invasive ventilation.

“Invasive” means that a doctor will do surgery to place a breathing tube inside your windpipe (trachea). This is called a tracheostomy.

Why is invasive ventilation important?

Not everyone with ALS will need or choose to have a tracheostomy, but there may come a time when it’s necessary for continued breathing. You may have respiratory weakness that can no longer be treated with a mask and non-invasive ventilation. You may also need a tracheostomy to help remove secretions from your airways.

Tracheostomy

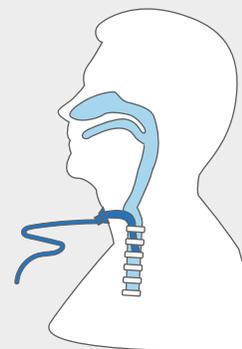
- A tracheostomy is a surgical opening in the neck. A short tube is inserted into the throat. The tube keeps the airway open. Additional tubing connects the tracheostomy to a machine. A portable ventilator will breathe for you.
- The breathing machine blows air thru the tube and into the lungs.
- Air from the lungs flows out through the tracheostomy opening in the throat, instead of through the nose or mouth.



You will most likely need to have a feeding tube placed. To learn more about feeding tubes visit, alsdecisions.org/nutrition.

How to communicate when you have a tracheostomy

- The tracheostomy is located below the vocal cords. Air movement across the vocal cords is required for speech. If possible, your doctor can adjust your machine settings to facilitate speech.
- You may not be able to speak or communicate in the same way that you did before the surgery.
- With today’s technology, you have options to communicate with the people who are most important to you.
- Your ALS care team can customize the best way for you to meet your communication needs.



Tracheostomy



Alphabet boards



Alternative speech generating devices



Adaptive methods of communication



You can learn more about ALS & Communication by visiting lesturnerals.org/communication.

Implications of invasive ventilation

Invasive ventilation support may drastically change how you are cared for and communicate. Therefore, this very personal decision should be discussed and considered thoroughly.

Give yourself time to consider how you want to live and experience the rest of your life. This decision impacts both you and your family in many ways, including financially and emotionally. Before a decision is made, advance directives should be in place.

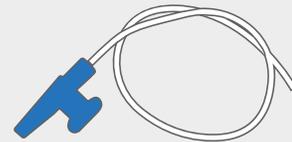


Does insurance cover invasive ventilation?

If you have private health insurance and are considering invasive ventilation support, it is important to first contact your insurance company.

A person with invasive ventilation support requires 24-hour care from a caregiver, family member or nursing facility. Some insurance plans pay for an outside agency to provide part of this care.

Medicare does not pay for such services.



How to care for a tracheostomy

The tracheostomy site and equipment needs to be cleaned every day. This will help prevent lung infections.

Secretions from the lungs must be regularly cleared from the tube with a disposable suction catheter. This will help keep the airways open.

To find out more, visit youralsguide.com/invasive-mechanical-ventilation

Removal of invasive ventilation

Although the use of invasive ventilation will keep you alive, it will not cure or stop ALS symptoms. At some point you may decide that you no longer want to prolong your life with the use of a ventilator. Before you have surgery, you'll want to have a discussion with your doctor and family about your wishes regarding removal of ventilation.

To be taken off invasive ventilation, you must first be evaluated by a physician. You will then be admitted to hospice for removal of ventilation and end-of life-care.

Removal from invasive ventilation may take place in the hospital or at an in-patient hospice unit. You would be given medications to ease any discomfort and make the final moments as peaceful as possible. After the invasive ventilation is disconnected, it may be minutes to hours before breathing ends completely.

Preparations for power outages



A generator, or emergency power source, is required as a backup in case of a power failure. Battery packs can help in case of severe weather or power outages.



Some devices include internal batteries.



Local power companies also allow people who need NIV to register with emergency services in case of power outages. The restoration of their service will be prioritized.

Travel



If traveling by air, the airline should be contacted before making a reservation.

- Non-invasive ventilation and other breathing equipment may be carried on the airplane. It does not count as a “carry-on” item.
- Airlines recommend having enough battery power to last for nearly double the flight time.
- If the use of a BiPAP/VPAP or home ventilator is required in flight, a back-up battery will be needed.



A car inverter can help when traveling or using respiratory devices in a car.



Palliative or Hospice Care

With or without the use of NIV or invasive ventilation, your ALS care team may recommend palliative or hospice care. Palliative care won't help you live longer, but it can provide care and support to help you stay comfortable. Hospice care provides compassionate care and comfort at the end of life.

A palliative care provider or ALS care team can give you medicine to help with your symptoms, other equipment and recommend exercises to help with your breathing, posture and cough.

Talk to your ALS care team to figure out the right treatment plan for you.

Learn more

The Les Turner ALS Foundation exists to guide you to answers, support you and your loved ones and advance scientific research. To learn more about living with ALS visit, lesturnerals.org/resources.

My ALS Decision Tool™

If you have ALS, you will need to make some important decisions about your health care. As your disease progresses, your ALS care team may recommend different care options. You can use this tool to learn about some common ALS treatments, answer a few questions to help you think through what is most important to you and get ready to talk with your ALS care team about your options. To learn more, visit: alsdecisions.org.

**MY ALS
DECISION
TOOL**

ALS Learning Series

Our online ALS Learning Series aims to empower the ALS community through the latest information and insights. Educational webinars and interactive Q&A's covering a diverse array of topics, from nutrition to respiratory care, are offered monthly featuring members of the Foundation's Support Services team, our Lois Insolia ALS Clinic at Northwestern Medicine and other national ALS experts. To learn more about ALS care and research, visit: alslearningseries.org.

**ALS
LEARNING
SERIES**

My ALS Communication Passport to Quality Care

My ALS Communication Passport to Quality Care was created to make your life easier. You will be able to share health information and care preferences with caregivers. You have a lot of information to keep track of, and this tool will help you do that. To find out more, visit: lesturnerals.org/passport.

**MY ALS
COMMUNICATION
PASSPORT
TO QUALITY CARE**

Support Groups

We facilitate support groups to provide people living with ALS, caregivers, and family the opportunity to share their experiences, give encouragement and help each other navigate their journey with ALS. To find out more, visit: lesturnerals.org/support-groups.

