Your ALS guide

Welcome to our educational guide on ALS respiratory challenges, support, intervention, and equipment. Find trusted information and helpful tips on the following topics.

Respiratory Symptoms - Many people don't notice breathing difficulties until after their diagnosis.

Respiratory Support - Respiratory specialists on your medical team can guide you every step of the way.

Monitoring Breathing - Breathing tests help inform the care and equipment you will receive.

Introduction to Respiratory Devices – Getting the right devices at the right time can make a significant difference.

Mask and Interface Options - You can choose from a variety of masks that will work with your devices.

Bilevel (BiPAP) Sleep Therapy - A bilevel machine can improve your quality of life and even help you live longer.

Portable Noninvasive Ventilation (NIV) - Portable ventilators can provide breathing support when needed.

Cough Assist Machine - This device can make you more comfortable by clearing your lungs and airway.

Suction Machine - This device clears saliva from your mouth when you're having trouble swallowing.

Invasive Mechanical Ventilation - After a tracheotomy, a ventilator attaches to a surgical opening in your windpipe.

Planning Ahead with Advance Directives - These documents let everyone know what medical treatment you would want.

Traveling with Your Devices - You can travel with your devices, but you'll need to be organized and plan ahead.

Respiratory Symptoms

ALS is a neurodegenerative disease that weakens voluntary muscles throughout the body. For the vast majority of people, ALS symptoms first appear in one of the limbs or in the muscles that control speaking and swallowing. Just a small percentage of people notice respiratory symptoms first.

Yet as ALS progresses, it will eventually affect the respiratory system. When ALS weakens the diaphragm (your primary breathing muscle), it becomes harder to fully inhale and exhale. This can lead to common ALS respiratory symptoms such as:

- Trouble breathing while lying down
- Shortness of breath
- General fatigue
- The need for more frequent naps
- Waking with morning headaches
- Feeling groggy or “out of it” in the morning
- Difficulty finishing a sentence without taking a breath
- A weaker cough

Whether or not you are experiencing any respiratory symptoms, it is important to attend an ALS clinic and have your breathing measured regularly. Oftentimes, breathing tests will detect a decline in breathing function before people living with ALS notice any symptoms. Getting respiratory support early can help you sleep better, have more energy, be more comfortable, live longer, and have a better quality of life.

Respiratory Support

When you're diagnosed with ALS, it is important to attend an ALS clinic or see a medical team that specializes in ALS. A respiratory therapist or nurse will monitor your breathing at each visit, suggest respiratory devices that may be helpful for you, and guide you every step of the way.

After you have received the diagnosis, your neurologist will schedule your first clinic visit. If you do not live near an ALS clinic or cannot attend one, contact your local ALS organization to ask if they can recommend a local neurologist who specializes in ALS or other medical professionals who can help.

If attending an ALS clinic is not possible, ask your primary care doctor or neurologist if they can either monitor your respiratory numbers or recommend a respiratory therapist who can do so. Once you qualify for a certain respiratory device, your doctor or neurologist will order the device through a local durable medical equipment company.

If any respiratory issues or concerns arise between visits, contact your medical team to ask questions or schedule a separate appointment.

Respiratory Equipment

Respiratory equipment is therapy for your respiratory system. It can help you sleep better, have more energy, and experience a better quality of life. Studies have shown that using respiratory equipment on a regular basis can help you live longer. Starting early can help you get used to the devices and begin feeling better sooner.

Most respiratory equipment is noninvasive, which means you can take the mask or interface on and off whenever you wish. If you get a tracheotomy at a later stage in the disease, however, you will become dependent on a mechanical ventilator to breathe for you 24/7.

Breathing Exercises for ALS

There are some daily breathing exercises that you can do early in the disease to help you open and expand your lungs, breathe more easily, and have a stronger cough.

"Breath stacking" involves taking a series of short breaths on top of each other without exhaling. After holding your breath for a few seconds, you fully exhale all at once. With expiratory and inspiratory muscle training, you use a small, handheld device to inhale and exhale against a resistance level that is set and adjusted according to your breathing test measurements. Other helpful breathing techniques include huffing and deep diaphragmatic breathing.

Ask your ALS respiratory therapist, pulmonologist, or neurologist about breathing exercises that might help you. Follow their guidance before trying these exercises on your own.

Improving Your Sleep

Getting a good night's sleep is important for your health, well-being, and energy level. If you are finding it hard to get a solid night's sleep or are finding it harder to breathe when lying down, it may help to prop yourself up with pillows or a bed wedge. If you are still not getting the rest you need, it may be time to get a bilevel (BiPAP) machine or use a portable ventilator in bilevel mode to assist you with breathing while sleeping. This should improve your quality of sleep and help you feel more rested when you wake up.

Keep Your Primary Doctor

Even after you connect with a team of ALS specialists, you should continue to see your primary care doctor. If you get the flu, a cold, or an infection, it is especially important to see your doctor as soon as possible to get it cleared up.

Shortness of Breath

If you are experiencing shortness of breath, it is likely the result of a weakened diaphragm muscle. This makes it harder to expand the lungs enough to fully receive oxygen and fully exhale carbon dioxide. Typically, a bilevel (BiPAP) machine or a portable ventilator will help. Most people living with ALS do not need supplemental oxygen. The exception is when your oxygen levels drop below normal levels, which is usually from an underlying lung condition such as COPD or emphysema.

If you develop sudden shortness of breath, it is important to seek immediate medical attention, as the cause could be something other than ALS, such as a blood clot in the lungs.

Monitoring Your Breathing

Your medical team will begin monitoring your breathing even before you notice any respiratory symptoms. If you attend an ALS clinic, a respiratory therapist or nurse will ask about any changes you’ve experienced and perform one or more simple, noninvasive tests to measure your progression.

Forced Vital Capacity (FVC) is the primary test used to evaluate the breathing of ALS patients. It measures how quickly you can expel air from your lungs.

The Maximum Inspiratory Pressure (MIP) test measures the strength of your inhale through your mouth. The Sniff Nasal Inspiratory Pressure (SNIP) test is the same as the MIP, but it measures your inhale through your nose, which can be helpful for bulbar patients who are unable to form a good seal around the mouthpiece.

Your team might also recommend a capnography study, which measures your oxygen and carbon dioxide levels as you sleep. This can alert your team early on if your diaphragm function is impaired. This study takes place at home, with a clip on your finger and a thin tube in your nose.

The results of these tests, along with the observations you share, will give your team the information it needs to recommend the best care and respiratory equipment for you. The results from these breathing tests will also provide the documentation your neurologist will need for Medicare, Medicaid, and private insurance to cover your respiratory devices.

ALS Respiratory Devices

Your ALS clinic or medical team will monitor your breathing, provide respiratory support, and prescribe the right respiratory devices at the right time. Using respiratory devices can improve your quality of life and help you live longer.

These are the most common types of respiratory devices that help people living with ALS:

A bilevel (BiPAP) is a small breathing machine that delivers pressurized air through a face mask to assist with both inhaling and exhaling, primarily during sleep.

A portable ventilator (Trilogy) can help you breathe either noninvasively (via a mask) or invasively (via a trach). It has a bilevel mode for sleeping and daytime breathing support.

A cough assist machine helps your breathing by clearing phlegm and fluid-like secretions from your lungs. Using it daily can also help condition the diaphragm muscle, even if no secretions are present.

A suction machine is a simple device, similar to the wand at the dentist's office, that uses suction to remove excess mucus and saliva from your mouth.

Masks and Interfaces

When your bilevel (BiPAP) machine or portable ventilator (Trilogy) is delivered, a respiratory therapist from a durable medical equipment (DME) company will bring a variety of mask options and recommend the one that will be the most appropriate for you. This may depend on how you breathe at night and how well the different masks fit. Cough assist machines will arrive with the appropriate mask included.

These are the most common types of masks and interfaces for people living with ALS:

Nasal Pillows

With just two little cushions that seal at the base of your nostrils, this is the lightest and most compact interface option. Nasal pillows are generally good for people who breathe through their nose when sleeping. Because of its low profile, you can still wear glasses, read, watch TV, talk, and eat with nasal pillows in place. If you have a lot of facial hair, nasal pillows may help you get the tightest seal. If you tend to breathe through your mouth at night, nasal pillows may not be your best option, though you can ask about attaching a chin strap that can help keep your mouth closed.

Nasal Mask/Interface

The nasal interface is larger than nasal pillows but less bulky than a full face mask. Triangular in shape, it sits on your face and covers your full nose. The nasal interface is generally good for people who breathe through their nose when sleeping. If you tend to breathe through your mouth at night, a nasal interface may not be your best option, though you can ask about attaching a chin strap that can help keep your mouth closed.

Full Face Mask

The traditional full face mask is more bulky than the other interfaces, but it does allow the option of breathing through your nose or mouth. This can be especially helpful if you tend to get congestion that makes it harder to breathe through your nose. If your air pressure settings are high, the full face mask will be the most comfortable option. This larger mask can make it hard to read, watch TV, or sleep on your side or stomach. There is also a higher chance of air leaking with a full face mask.

If your mask or interface is uncomfortable or doesn’t feel like it is fitting correctly, schedule another appointment with the respiratory therapist who set everything up. Making adjustments or trying a different mask or interface may solve the problem. It may also just take some time to get used to wearing a mask and using a new device. The benefits of using a bilevel or portable ventilator in bilevel mode are too great to give up because something doesn’t feel right at first.

Bilevel (BiPAP Sleep Therapy)

A bilevel machine, commonly referred to as a BiPAP, is a breathing machine primarily used during sleep that delivers pressurized air through a mask or interface to assist you with both inhaling and exhaling.

A bilevel (BiPAP) can help you sleep better, wake up feeling more rested, and have more energy during the day. In addition to improving your quality of life, research has shown that using a bilevel early and regularly can help you live longer.

How do bilevel machines work?

When the diaphragm muscles weaken and your body is unable to fully inhale oxygen and fully exhale carbon dioxide, your mind can begin to feel foggy, and you can get morning headaches. The bilevel machine assists you in being able to take a bigger breath and exhale more fully.

The bilevel is noninvasive, which means you can take it on and off and use it whenever you wish. You will not become dependent on it, but you will probably want to use it because you may feel better when you do. Bilevel machines do not have internal batteries, so they must be plugged in.

Your neurologist will prescribe the right pressure settings for you, and a respiratory therapist from a durable medical equipment (DME) company will visit to set up the machine and train you and your caregiver(s) how to use it. The respiratory therapist will try to find a face mask or interface that lets you sleep in your normal position, whether on your stomach, back, or side. The respiratory therapist will also show you and your caregiver how to use and refill the humidifier chamber, which humidifies the air to help prevent dryness in your mouth, nose, throat, and airway.

Can I use it during the day?

Yes. If you are having difficulty breathing during the day, or simply feel like you could use a little more breathing support, you can use your bilevel while you are awake with the same mask or interface you use when you sleep. This can give your diaphragm a break and increase your energy level. You can also use your bilevel during naps, just as you do when sleeping at night.

If you are finding that you need your bilevel more throughout the day, speak with your clinic team about options that might work better for you.

When should I get a bilevel?

Your ALS clinic team or neurologist should monitor your breathing every visit and let you know when you will qualify for and can benefit from a bilevel. Ideally, you will get a bilevel before you experience breathing difficulties so you can have time to get used to it.

How can I get one?

Your neurologist must submit your pulmonary function test results and documentation that demonstrate you need breathing support during sleep. The durable medical equipment company will process the order through your insurance, deliver the bilevel to you, and train you how to use it. Insurance companies do not usually require ALS patients to do a sleep study to qualify for a bilevel.

Using a Portable Ventilator in Bilevel Mode

Using a portable ventilator in bilevel mode provides the same therapy as a bilevel machine. Portable ventilators have more settings and uses, but they are more expensive. Your medical team will recommend the device they think is best for you based on your breathing measurements and symptom progression.

How much will I have to pay?

Bilevels are considered rentals for the first 13 months and are covered through Medicare, Medicaid, and most private insurance plans. After 13 months, the machine becomes yours and your supplies will continue to be covered by insurance.

Medicare will cover 80% of the cost of your monthly rental. Medicaid, supplemental plans, and secondary private insurance should pay for the remaining 20%. If you have Medicare but do not have Medicaid, a supplemental plan, or secondary private insurance, you will likely have to pay the remaining 20% out of your own pocket.

If you do not qualify for Medicare—but have Medicaid and/or private insurance—find out what your durable medical equipment (DME) benefits will cover. If you have a Medicare Advantage Plan, be sure to ask about your DME benefits.

If you are facing out-of-pocket expenses that you cannot afford, talk with your DME provider and/or local ALS organization about financial assistance options. Respiratory equipment is not generally available from loan closets, but you can always ask.

When your bilevel machine is delivered, a respiratory therapist will bring a variety of mask options and recommend the one that will be the most appropriate for you. This may depend on how you breathe at night and how well the different masks fit.

What if I’m having trouble using my bilevel?

Some adjustments may be needed when you first start using a bilevel. If you are having trouble, schedule another visit with your respiratory therapist. Together, you should be able to resolve the issue. The pressure may need to be adjusted, your mask may be leaking, or you might simply need more time to get used to it. Whatever the problem, the benefits you’ll receive from your bilevel are too great to give up and stop using it.

Portable Noninvasive Ventilation

If you are experiencing respiratory symptoms like shortness of breath, noninvasive ventilation (NIV) can provide helpful breathing support through a mask that you can put on and take off as needed.

Studies have shown that noninvasive ventilation can alleviate respiratory symptoms, improve quality of life, and help you live longer. It is one of the most effective interventions available for people living with ALS. You can receive noninvasive ventilation either through a bilevel (BiPAP) machine or a portable ventilator (often referred to by the brand name Trilogy).

Bilevels (BiPAPs) are smaller and less expensive than portable ventilators, but they are not portable. Bilevels are primarily prescribed for nighttime use. Portable ventilators have more settings and functions and are more equipped to support your breathing during the day as well.

Portable ventilators have internal batteries, and you can buy backup batteries if needed. This means you can unplug the device and receive breathing support while moving around your home, going out, or traveling. Portable ventilators typically weigh under 12 pounds and come with travel bags that can hook onto the backs of wheelchairs.

Your medical team will recommend the device they think is best for you based on your breathing measurements and symptom progression. If you start with a bilevel, you can switch to a portable ventilator when needed.

Receiving noninvasive ventilation (NIV) is very different from invasive ventilation, which involves creating a surgical opening in your trachea and becoming dependent on a ventilator to breathe for you 24/7. Portable ventilators can be used for both invasive and noninvasive ventilation.

How can I get a portable ventilator?

Your ALS clinic team or neurologist will monitor your breathing every visit and let you know when you qualify for a portable ventilator.

You will need to have a face-to-face meeting with your neurologist, usually during a clinic visit, to get your portable ventilator prescribed for noninvasive use. Your neurologist will need to have pulmonary function test results and documentation that the durable medical equipment (DME) company can use to process the order through insurance. A respiratory therapist from the DME company will then deliver your ventilator and train you and your caregivers how to use it.

If you are using a bilevel and it is still considered a rental, you will need to return it when the portable ventilator is delivered.

How much will I have to pay?

Portable ventilators are considered long-term rentals and are covered
through Medicare, Medicaid, and most private insurance plans.

Portable ventilators are more expensive than bilevel (BiPAP) machines, so the monthly payment will likely be higher. Supplies such as masks, tubing, filters, and humidifiers, as well as periodic maintenance, will be included in your monthly rental fee.

Medicare will cover 80% of the cost of your monthly rental. Medicaid, supplemental plans, and secondary private insurance should pay for the remaining 20%. If you have Medicare but do not have Medicaid, a supplemental plan, or secondary private insurance, you will likely have to pay the remaining 20% out of your own pocket.

If you do not qualify for Medicare—but have Medicaid and/or private insurance—find out what your durable medical equipment (DME) benefits will cover. If you have a Medicare Advantage Plan, be sure to ask about your DME benefits.

If you are facing out-of-pocket expenses that you cannot afford, talk with your DME provider and/or local ALS organization about financial assistance options. Respiratory equipment is not generally available from loan closets, but you can always ask.

When your portable ventilator is delivered, a respiratory therapist will bring a variety of mask options and recommend the one that will be the most appropriate for you. This may depend on how you breathe at night and how well the different masks fit.

What if I'm having trouble using my ventilator?

Some adjustments may be needed when you first start using a portable ventilator. If you are having trouble, schedule another visit with your respiratory therapist. Together, you should be able to resolve the issue. The pressures may need to be adjusted, your mask may be leaking, or you might simply need more time to get used to it. Whatever the problem, the benefits you'll receive from your ventilator are too great to give up and stop using it.

Cough Assist Machine

A cough assist is an airway clearance device that helps your breathing by moving phlegm and fluid-like secretions from your lungs. Most people living with ALS use cough assist machines and feel more comfortable as a result. Regular use can help maintain the strength of your diaphragm and help prevent infections like pneumonia and bronchitis.

How does a cough assist work?

When your respiratory muscles are weakened, it can become difficult to cough strongly enough to draw phlegm from your lungs up to your mouth. The cough assist machine simulates a natural cough by applying positive pressure to inflate your lungs before using a vacuum-like suction to pull everything up.

Once secretions are in your mouth, you can use a suction machine to clear everything out.

How often should I use it?

You should use your cough assist regularly, even when you don't feel you have anything in your lungs to clear. Regular use, such as once in the morning and once in the evening, serves as therapy to strengthen your diaphragm, keep your lungs clear, and reduce the chance of infection, which can be more difficult to recover from for people living with ALS. Each session produces multiple coughs and typically lasts between five and ten minutes.

When should I get one?

If you are experiencing a weak or impaired cough, your neurologist will order you a cough assist machine, often during a clinic or doctor visit. A durable medical equipment (DME) company will process the order through insurance and deliver the cough assist to you at home or at your ALS clinic. A respiratory therapist will train you and your caregivers how to use it and adjust your settings if needed.

How much will I have to pay?

Cough assist machines are considered rentals for the first 13 months and are covered through Medicare, Medicaid, and most private insurance plans. After 13 months, the machine becomes yours and your supplies will continue to be covered by your insurance. You may need to be proactive and ask your insurance for replacement masks, tubing, and filters ahead of time.

Medicare will cover 80% of the cost of your monthly rental. Medicaid, supplemental plans, and secondary private insurance should pay for the remaining 20%. If you have Medicare but do not have Medicaid, a supplemental plan, or secondary private insurance, you will likely have to pay the remaining 20% out of your own pocket.

If you do not qualify for Medicare—but have Medicaid and/or private insurance—find out what your durable medical equipment (DME) benefits will cover. If you have a Medicare Advantage Plan, be sure to ask about your DME benefits.

If you are facing out-of-pocket expenses that you cannot afford, talk with your DME provider and/or local ALS organization about financial assistance options. Respiratory equipment is not generally available from loan closets, but you can always ask.

Suction Machine

A suction machine is used to remove excess mucus and saliva that can accumulate in your mouth when your mouth and throat muscles become weaker and your swallowing may be impaired.

Clearing these secretions can help you breathe more easily, choke less when eating and drinking, and generally feel more comfortable. The suction machine is often used in conjunction with the cough assist machine, which pulls secretions from your lungs and throat up into your mouth for removal. The suction machine has an internal battery, so you can take it wherever needed.

How can I get one?

If you are having trouble managing secretions, your neurologist will order you a suction machine. No test is needed, and you do not need to meet any other specific criteria. A durable medical equipment (DME) company will process the order through insurance and deliver the suction machine to you at home or at your ALS clinic. A respiratory therapist will train you how to use and maintain it. Suction machines are often ordered in conjunction with bilevel machines.

How much will I have to pay?

Suction machines are fairly inexpensive. They are considered rentals for the first 13 months and are covered through Medicare, Medicaid, and most private insurance plans. After 13 months, the machine becomes yours and your supplies will continue to be covered by your insurance.

Medicare will cover 80% of the cost of your monthly rental. Medicaid, supplemental plans, and secondary private insurance should pay for the remaining 20%. If you have Medicare but do not have Medicaid, a supplemental plan, or secondary private insurance, you will likely have to pay the remaining 20% out of your own pocket.

If you do not qualify for Medicare—but have Medicaid and/or private insurance—find out what your durable medical equipment (DME) benefits will cover. If you have a Medicare Advantage Plan, be sure to ask about your DME benefits.

If you are facing out-of-pocket expenses that you cannot afford, talk with your DME provider and/or local ALS organization about financial assistance options. Respiratory equipment is not generally available from loan closets, but you can always ask.

Invasive Mechanical Ventilation

When noninvasive ventilation no longer provides enough breathing support, invasive mechanical ventilation will be discussed as a way to extend your life. This requires tracheostomy surgery, which creates a circular hole in your windpipe at the base of your neck where the ventilator tube will connect. After that, the portable ventilator will breathe for you.

What would this mean for me?

Going on invasive mechanical ventilation means that you will need to be connected to your ventilator full-time and will need 24/7 care. You will receive nutrition through a feeding tube and will need to communicate through a speech-generating device.

What type of care would I need?

Once you are dependent on a ventilator, you will need 24/7 care. If you hire caregivers, they must be skilled, vent-trained professionals. This type of care is expensive, and is not covered by Medicare. Medicaid and some private insurance plans may pay for some level of care.

Family members can be trained to care for you. They will need to be fully trained before you return home from the hospital, which can take two to four weeks. Once at home, family caregivers will need to care for the trach site, maintain supplies, and suction secretions from the airway multiple times an hour.

How should I make this decision?

Getting a tracheotomy is a very personal decision. Talk with your neurologist, ALS clinic team, and a representative from your local ALS organization about the advantages and disadvantages of mechanical ventilation.

Talking about this with your loved ones is not easy, but it is extremely important. You may want to ask the social worker or nurse at your ALS clinic to facilitate the conversation, answer questions, and educate everyone on the realities of each option.

Though it is ultimately your decision, consider and discuss the impact it will have on everyone involved. Your loved ones may have strong feelings and differing opinions and may even disagree with what you want. Try to explain your thinking and wishes as clearly as possible.

Once you make your decision, be sure to fill out advance directives to make your wishes very clear to your loved ones and medical professionals. This will provide the gift of clarity for everyone and help ensure that your wishes are followed.

Advance Directives for ALS

Advance directives are written documents that you can complete to let everyone know what type of medical treatment you would—or wouldn't—want in the event of a medical emergency. If you become unable to make decisions, advance directives will make your wishes clear to your loved ones and to the medical professionals caring for you.

Why should I do this?

If you do not create advance directives ahead of time, your loved ones may find themselves in the challenging position of trying to make a decision for you. This can lead to confusion, guilt, and animosity. Advance directives can provide peace of mind for you and clarity for your loved ones.

It is very important to have a discussion with your loved ones and decide in advance whether or not you would want to have a tracheostomy procedure and be placed on invasive mechanical ventilation. If you do not discuss or write down your decision, the decision could be made for you in an emergency situation—and it may not be what you want.

Unwanted tracheotomies can and do happen. Medical professionals are obligated to do everything in their power to keep you alive—unless they have instructions from you stating otherwise.

Advance directives are simple forms. You do not need a lawyer to fill them out, though you can always seek legal advice if you'd like. You can change your advance directives whenever you'd like. The most important thing is to put your current decisions in writing so there is never any doubt about what you want.

Which forms should I fill out?

The three main types of advance directives for people living with ALS are a living will, a medical power of attorney, and a do not resuscitate (DNR) order. The official names of these documents can vary between states.

A living will is the core document that describes the type of life-prolonging support you would or wouldn't want to receive, such as CPR, ventilation, a feeding tube, and pain management. A living will is different from the type of will that assigns property and money to loved ones upon death.

A medical power of attorney (POA), also called a health care proxy or durable power of attorney for health care, is a legal document that designates the person or persons you would like to make medical decisions on your behalf should you become unconscious or unable to make decisions. This person may be called a proxy, agent, surrogate, or representative.

Designating a medical power of attorney in addition to filling out a living will allows your proxy to communicate with medical professionals and make decisions in situations that may not be spelled out directly in your living will. If you have clearly communicated with your proxy, he or she should have a good idea of what you would or wouldn't want.

It is important to choose a proxy you trust and who will follow your wishes, even if your loved ones strongly disagree. Ideally, this person can remain emotionally calm and stable in the event of an emergency. You do not have to choose a family member, and you may not designate a person who is on your medical team. You can choose more than one person, but if you do, you will need to designate which person will be in charge if both are present at the same time.

If you do not want first responders or medical professionals to attempt life-saving measures should you stop breathing or your heart stop beating, your doctor will need to sign a do not resuscitate (DNR) order for you. Your DNR will not be valid unless your doctor completes it and signs it.

Even if you already stated in your living will that you don't want CPR or other life-saving measures, you should still fill out a DNR. In the event of an emergency, first responders and hospital staff will be obligated to try to save your life—unless they immediately see your DNR. They will not have time to look for and read your living will. Your DNR form should be posted in a visible place at home and ideally filed in your medical records.

Note: You may want to ask if you need a do not intubate (DNI) order in your state as well. Intubation is when a medical team puts a tube through your nose or throat if you are unable to breathe on your own. Intubation can lead to a tracheostomy, in which case you may wake up trached and vented, whether you wanted it or not. Clearly stating in your DNI or elsewhere that you don't want intubation or mechanical ventilation will help avoid this scenario.

Where can I find these forms?

The social worker at your ALS clinic, local ALS organization, or local hospice should be able to give you the forms, help you fill them out, and answer your questions. Your hospital should have forms available, too.

You can also search online for advance directives in your state. The National Hospice and Palliative Care Organization makes it easy to find and download your state's advance directive forms. They also provide state-specific information and step-by-step instructions that walk you through the process. The website e-forms has downloadable DNRs and downloadable living wills for each state.

Another option is called Five Wishes, which is a simple living will that gives you the freedom to express your most important wishes in a variety of areas, from medical and legal to personal and spiritual. It is a legal document that is accepted in most states. You can ask your local ALS organization or ALS clinic for a copy, or you can buy a copy from their website or complete it online.

If you spend a significant amount of time in another state, you may want to find out it if each state honors the other's forms. If not, you may want to fill out advanced directives for both states.

You can also create your own advance directives form, but you will need to make sure you comply with your state's requirements in order for it to be considered a legal document.

Every state has its own forms and laws about advance directives.

What else do I need to do?

We recommend starting with the social worker at your ALS clinic, local ALS organization, or local hospice. Review the forms together, ask questions, and discuss your options. You may also want to meet with your neurologist and ask what could happen in various medical scenarios.

Though this can be a difficult topic to discuss with your loved ones, it is important to explain your decisions and make sure that everyone understands your wishes, even if not everyone agrees. If you are worried about initiating this conversation, ask your social worker to facilitate, answer questions, and educate everyone.

Once you have made your decisions, you will need to fill out the forms and sign them with a witness present, and possibly a notary, depending on your state's laws. If you comply with your state's requirements, your forms will become legal documents when you sign them.

Give copies to your proxy, loved ones, medical teams, and the hospital where you would go in the case of an emergency. Keep your original in a safe place that others know about and can access in the case of an emergency. Other options for making sure your wishes are shared include emailing copies, storing them on the cloud, keeping a card in your wallet, traveling with your documents, and having a flashdrive with you.

Also be sure to tell your loved ones who you have designated as your proxy. Explain that this person's role is to carry out your wishes.

Traveling with Respiratory Devices

Traveling with respiratory equipment is possible, though it does require advance planning. Make sure you leave home with everything you need, such as extra batteries, power converters, adapters, extension cords, masks, and tubing.

Can I travel by plane with my equipment?

Yes. Most respiratory devices are FAA-approved. Check your device manual or consult with your respiratory therapist to make sure. Contact your airline—ideally a few weeks in advance—to ask if you need approval to bring your device(s) on the plane. Bring all related paperwork, such as your neurologist’s prescription for each device.

Your respiratory devices should not be considered carry-ons and can be stored in the overhead bin. You can use your respiratory devices as needed while on the plane. Make sure you have enough battery-backup time for unexpected delays. You can ask ahead of time if there will be power outlets on the plane.

Talk with your respiratory therapist ahead of time about your travel plans. Ask for tips, ideas, and concerns regarding traveling with your specific devices.

Keep in mind that you will need to be able to sit in a standard airplane seat for the duration of the flight.

How portable are all the devices?

Bilevel machines, used primarily for sleeping, must be plugged into an outlet—unless you purchase an external battery out of pocket. Portable ventilators, multifunction ventilators, cough assists, and suction machines all have internal batteries, which means you can use them while you’re on the move, whether in a car, bus, train, or plane. If you have a power wheelchair, your portable ventilator can plug into your wheelchair battery, if needed.