

Living with neuromuscular disease

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Introduction

The goal for neuromuscular disease (NMD) patients living with a ventilator is to live full lives. Even though most neuromuscular diseases will not be cured, many of the symptoms resulting from neuromuscular disease can be managed in various ways, including respiratory support.

Choosing a ventilator that best meets your or your loved one's needs is key to creating a positive experience. It's important to consider both social and medical benefits as well as any physical barriers related to daily activities and caregiver access. Gathering information about your condition, consulting your doctor and learning how to enhance your quality of life will help set your expectations and build the foundation for acceptance and success.¹ There are many success stories and support groups at your disposal.²

When initially diagnosed with NMD, you will have concerns about how you will be able to maintain your quality of life. Some questions you may be asking yourself:

- How will I be able to communicate with my friends and family?
- How can I maintain my daily living activities?
- What should I do to remain independent and mobile for as long as I can?
- Will I be able to travel?
- What do I need to do to be comfortable and pain free?
- What decisions do I need to make if my disease worsens?

Overview of neuromuscular disease

Symptoms

The symptoms of NMD can be different for every person, but can include:

- Tingling
- Numbness
- Muscle weakness or cramping
- Muscle pain
- Shortness of breath
- Tightness in the chest
- Difficulty sleeping

Causes

When the nerve cells in the body are working correctly, they send messages to the muscles that a person is able to control. However, when the nerve cells become unhealthy or die, the messages between the muscles and nerve cells no longer work properly, causing the muscle to become weak or stop working. In the most serious cases, these weakened or nonworking muscles can affect your ability to breathe.

This quick guide is intended to walk you and your caregivers through the disease progression, treatment options, guidelines for improving quality of life and additional resources to support your ventilator management.

Types of neuromuscular disease

Three types of neuromuscular disease can affect a person's ability to breathe on his/her own, including: amyotrophic lateral sclerosis (ALS), myasthenia gravis (MG) and spinal muscular atrophy (SMA).

ALS

Also called Lou Gehrig's disease, ALS attacks nerve cells called neurons in your brain and spinal cord.³ When ALS first starts, it can cause mild muscle problems that interfere with walking or running, writing or speaking. Eventually, the person loses strength and cannot move. Medications can relieve symptoms and, sometimes, prolong survival. As this disease progresses, the muscles in your chest start to fail, causing periods of shortness of breath. Supplemental oxygen can temporarily help with the shortness of breath. However, when the chest muscles are no longer able to work, you will not be able to breathe on your own. A breathing machine called a ventilator can help, but most people with ALS die from respiratory failure, as there is no cure.

MG

Myasthenia gravis is an autoimmune disease that causes weakness in the muscles under your control. Your own immune system makes antibodies that block or change some of the nerve signals to your muscles, causing the muscles to become weaker.⁴

Common symptoms are trouble with eye and eyelid movement, facial expression and swallowing, but other muscles can also be affected. In most cases, the weakness gets worse with activity and better with rest.

Some medications help improve nerve-to-muscle messages and make muscles stronger. With such treatment, the muscle weakness often gets better. Some medications help prevent the body from making abnormal antibodies, while other treatments filter abnormal antibodies from the blood or add healthy antibodies from donated blood.

Sometimes a person may require surgery to remove the thymus gland, a lymphoid organ situated in the neck of vertebrates that produces T cells for the immune system.

For some individuals, MG can go into remission and no medications are required. The remission can be temporary or permanent.

SMA

Spinal muscular atrophy is an incurable genetic disease that attacks nerve cells in the spinal cord called motor neurons. These cells communicate with your voluntary muscles—the ones you can control, like those in your arms and legs. As the neurons die, the muscles weaken. This can affect walking, crawling, breathing, swallowing and head and neck control.⁵

There are many types of SMA, some fatal. Some individuals have a normal life expectancy, depending on the type of SMA and how it affects breathing. Treatments help with symptoms and prevent complications. These may include breathing machines to help with breathing, nutritional support, physical therapy and medication.

Breathing therapies

Many NMDs advance significantly compared to others and may result in managing the disease with a combination of treatments, including noninvasive and invasive ventilation therapies. You can be managed at home with the assistance of your family and caregivers.

The severity and timing of respiratory complications vary, depending on your diagnosis and overall health. It is important to speak with your doctor to understand the risks for respiratory complications associated with your particular diagnosis.

To determine if you have ALS, MG or SMA, or if you have been diagnosed with an NMD that seems to be worsening, the doctor will need to perform various breathing assessments. Once you have completed the assessments, your doctor will prescribe medications and treatments to help relieve some of your symptoms.

Breathing assessments

Many types of breathing assessments can help the doctor evaluate your respiratory status, determine if your breathing is getting worse and provide valuable respiratory monitoring.

The three most common are a peak flowmeter, pulmonary function testing and pulse oximetry. These assessments will be performed when the doctor suspects the onset of neuromuscular disease. The doctor will then set up a testing schedule to monitor your respiratory status.

Peak flowmeter—A peak flowmeter is a portable, easy-to-use device. During your doctor visit, the doctor will have you blow into the device to measure how well your lungs are able to expel air and if they are becoming weaker. A peak flowmeter may also be used at home. Depending on the type of neuromuscular disease, you may need to perform peak flow measurements daily. These measurements document how quickly the respiratory muscles are weakening and may indicate a decreased ability to cough.

Pulmonary function testing (PFT)—PFTs are a group of tests that measure how well you can breathe and how effective your lungs are working. A PFT is usually performed at your respiratory doctor's office or in a hospital's outpatient respiratory therapy department. These usually take about an hour and consist of blowing into a tube or performing various breathing tests.

Pulse oximetry—Pulse oximetry uses a clip-like device placed on a finger or earlobe to measure the oxygen level (*oxygen saturation*) of the blood. This easy, painless test measures how well oxygen is being sent to parts of the body farthest from the heart, such as the arms and legs. In most cases, pulse oximetry is only used to test blood saturation in your doctor's office. However, if your NMD gets worse and you start having more difficulty breathing, a pulse oximeter may be given for home use.

Medications

At present, there's no cure for most NMDs. However, medication therapies can be very effective for treating or reversing the symptoms of neuromuscular disease. Although only a few medications are approved for use against the effects of neuromuscular disease, clinical trials are ongoing. In addition to medication therapies that target a specific neuromuscular disease, the medications used for most neuromuscular disease are steroids and pain medications, as well as bronchodilators for shortness of breath.

Steroids

One steroid that has proven effective in some neuromuscular diseases is prednisone, taken in pill form. It slows the loss of muscle function and increases muscle strength, providing a few more months to a few more years of leg and arm use.⁶

Although prednisone can offer some improvement in your neuromuscular disease, it has undesirable side effects which include weight gain, loss of bone mass, thinning of the skin, raised blood pressure and blood sugar, depression and difficulties with thinking, sleeping and controlling behavior. Discuss the side effects with your doctor.⁶

Pain medications

With NMDs, you may experience various types of pain, mostly muscular. There are many different types of pain medicines and each helps relieve a certain type of pain. Each medication offers varying relief and has its own advantages and risks. Pain medications can be delivered by various methods, such as an injection, time-released patches or pills. Discuss with your doctor which may be best for you.

Bronchodilators

Bronchodilators or bronchodilator medications are used to treat conditions where the airways have become irritated and narrow. Bronchodilators make breathing easier by relaxing the muscles in your lungs and widening the airways (*bronchi*).

The medication must be delivered through a medication delivery device, such as a metered dose inhaler (MDI) or a small-volume nebulizer.

Metered dose inhaler (MDI)—A metered dose inhaler is a small portable pressurized canister inside a plastic case with a mouthpiece that delivers a specific amount of medication in aerosol form. Because it is portable, you can use it anywhere or anytime.

Small-volume nebulizer—Small-volume nebulizers deliver a fine spray of bronchodilator medication directly into your lungs. This type of delivery system is not as portable as the MDI. An electric source, a small medical air compressor and a nebulizer are needed for treatment.

Other treatments

To lessen some symptoms of your NMD and help improve your quality of life, other medical treatments may be prescribed:

- Nutritional support
- Assistive equipment
- Mobility program, which includes positioning, energy conservation, breathing exercises and quality of sleep
- Airway clearance
 - Airway clearance techniques
 - Airway clearance devices
- Suctioning

Nutritional support

When the muscles used in swallowing and chewing are weakened, there's a risk of dehydration, malnutrition, choking or respiratory infections caused by inhaling food or liquid into the lungs (*aspiration*).

A gastrostomy tube (*g-tube* or *feeding tube*) is permanently placed through your stomach wall, so a liquefied diet can be fed directly into your stomach. You can have food and drink by mouth for pleasure and extra nutrition. However, if the g-tube has been placed because of choking, no food or drink should be given by mouth.

Assistive equipment

There is a variety of different assistive equipment. If your muscles become too weak, communication devices allow you to convey your needs and thoughts using special computer adaptation and software. In addition, assistive equipment can help with everyday tasks, such as special feeding utensils, cups and straws. Transfer boards and mechanical lifts make it easier and safer for family and caregivers to move you if needed. Finally, walkers, wheelchairs and foot, ankle and leg braces can help keep you safe during physical or occupational therapy, or doing daily living activities.

Mobility program

A mobility program, which includes various physical and occupational routines, as well as proper positioning, energy conservation, breathing exercise and quality of sleep can help lessen some of your symptoms.

Physical and occupational therapy helps keep your body flexible and mobile and can help lessen some of the side effects of NMD. Therapies such as range-of-motion exercises and stretches help prevent freezing of the joints of your knees, hips, feet, elbows, wrists and fingers.⁷

Positioning can also be helpful. By elevating the head of the bed, you may experience less shortness of breath. The head of your bed can be raised by using extra pillows under your head, neck and chest, or placing pillows or blankets under the mattress, or between the mattress and box spring.

Each day, set aside time to rest between performing daily living activities. Your daily living activities (*e.g., bathing, dressing and eating*) should be spaced apart. When performing these activities, sit down whenever possible to reduce unnecessary steps. If needed, you should ask someone to help if the activity causes you to become short of breath. In addition, the time of day you perform various activities should be taken into consideration, because you may have more energy in the morning than later in the day.⁷

Breathing and coughing techniques can help maintain healthy lung function. These can be done several times a day by taking five to 10 deep breaths with a short rest in between, to strengthen the lungs and help them expand fully.⁸

Finally, your quality of sleep can be improved by using medical equipment correctly. In addition, adjustment of the room light, temperature and noise levels can enhance overall quality of sleep.⁸

Other treatments (*cont'd*)

Airway clearance

Coughing and clearing the airway of secretions is normally taken for granted. However, an NMD can weaken the respiratory muscles, making both coughing and the ability to cough up secretions in the airway difficult.

At times, your cough may need some assistance. You can give your cough a boost or, if you are too weak to cough, the doctor may order a medical device to help you bring up the secretions or sputum. An airway clearance device may loosen the secretions, but you still may need some assistance getting the secretions or sputum all the way out of your airway. If this occurs, you may need to be suctioned. It is essential to use an airway clearance device before you cannot cough at all and a respiratory infection develops.

Airway clearance techniques

Your doctor and healthcare providers will discuss which airway clearance technique or device is best for you. This quick guide will review a few of the most common types.

Self-assisted cough—You can perform this procedure if you have the ability to cough, but it feels as if your cough needs a boost or it becomes difficult to bring up sputum. To perform this procedure, you need good muscle strength and the ability to hold your breath and cough without someone helping you.

The self-assisted cough is performed while you are sitting and folding your arms below your rib cage, over your stomach. Then take a deep breath and hold it until you have to cough. When you begin to cough, lean your upper body forward and downward against your hands. This technique helps give the diaphragm a slight push to help force the air out of the lungs. A family member or caregiver can assist if your arms are weak. Before trying this treatment, please contact your doctor or healthcare provider.⁹

Chest physiotherapy (CPT)—CPT is the standard treatment for mobilizing airway secretions, but is very labor intensive and time consuming for both you and your caregiver. Your caregiver rhythmically strikes your chest wall with cupped hands or with a mechanical vibrator over your lung fields.

In recent years, less labor-intensive and time-consuming airway clearance devices are available, offering alternatives to standard CPT and are in most cases preferred. For example, a bronchodilator treatment with a small volume nebulizer may be given while using airway clearance devices, such as the ones below, if needed.

Types of airway clearance devices

Various devices help clear the secretions in your airway when the respiratory muscles become weaker. The most common devices for the home include: an insufflator-exsufflator, high frequency chest wall oscillation, a flutter and the acapella[®] device. With all airway clearance devices, a healthcare provider must provide training to you, your family members and/or caregivers.

Insufflator-exsufflator—There are numerous types of insufflator-exsufflator devices available. The goal is to help with airway clearance, as well as inflating the areas of the lungs that may not be inflated and not exchanging air. This type of device is usually used daily in cycles of three to five times. A cycle consists of the device pushing air into the lungs at a set pressure, then forcing the air out of the lungs with a vacuum. After a pause, the cycle repeats. The most common device used is the CoughAssist.

High-frequency chest wall oscillation (HFCWO)—HFCWO is performed with a mechanical device, usually with a patient inflatable vest, that applies positive pressure air pulses to the chest wall. These oscillate the chest and the vibrations reportedly cause flow to increase in the airways, loosening secretions and producing a cough.

Other treatments (*cont'd*)

Flutter—A flutter device is a portable device designed to help clear mucus, by combining positive expiratory pressure therapy with high-frequency oscillations within the airway. The principle behind this device is that when you exhale into the flutter device, a steel ball bearing fluctuates back and forth at a high frequency. Effectiveness is dependent on position. The device results in positive pressure and vibration in the airways that help with the removal of secretions in the airway.

Acapella—The acapella is a handheld airway-clearance device that operates on the same principle as the flutter device. It uses a counterweighted plug and magnet to cause the valve to close; however, it is not position dependent like the flutter. The portable acapella comes in three models and can be used with a mask or mouthpiece, as well as in-line with a small-volume nebulizer.

Suctioning

Suctioning may be needed to clear your airway of secretions if you cannot get them out by yourself. Suctioning requires additional equipment and supplies, which include a suction source and catheter.⁹

The type of suctioning depends on the portion of your airway that requires suctioning and whether there is an artificial airway, such as a tracheostomy tube. The most common types of suctioning are oropharyngeal (*a suction catheter leading through the mouth to the lower airways*); nasopharyngeal (*a catheter through the nose to the upper airway*); nasotracheal (*a catheter through one nose nostril to the lower airways*) and through an artificial airway.

If you can cough effectively but cannot swallow or cough out the secretions or sputum, a Yankauer (*tonsil tip*) suction catheter helps clear secretions from the mouth. If you cannot cough and do not have a tracheostomy tube, then nasopharyngeal and nasotracheal suctioning is performed to help remove secretions from the lower airway.

Ventilation

Ventilation is either noninvasive or invasive. Both types require a medical device at some level to assist your breathing by allowing your respiratory muscles to rest.

Ventilation therapy will begin if your breathing is progressively getting worse and will be based on your breathing assessments. Usually if you are experiencing symptoms of respiratory complications, you may start using a noninvasive ventilation device, then progress to a bi-level, positive airway pressure device and eventually an invasive ventilator.

Noninvasive ventilation

If your NMD is not severe, you can use noninvasive ventilation devices that provide pressure during your inspiration and expiration.

You will breathe through an external mask, nasal prongs or a sipper tube. Nasal masks also will allow you to talk, but your voice may sound different than usual. Oral or full-face masks interfere with speech, so these are typically used at night or intermittently throughout the day.

Bi-level device

You may be placed on a bi-level positive airway pressure device, if your breathing worsens and you need additional support. A bi-level device is a relatively small, quiet machine that creates air pressure and airflow coordinated with your breathing. It delivers inspiratory positive airway pressure (IPAP) when you breathe in, pushing air into the lungs. This is followed by a significantly lower expiratory positive airway pressure (EPAP) that allows you to exhale.

Bi-level devices usually work well in the earlier stages of ventilatory problems, when ventilatory support is only required for part of the day or night. If you require continuous invasive ventilatory support, however, you may need an invasive ventilation device.

Invasive ventilation

For invasive ventilation, you will breathe through a tracheostomy tube or ET tube that is inserted directly into your airway to deliver air to your lungs. These ventilators have features that are more advanced than those found on many noninvasive ventilation or bi-level devices. It can be used to partially or fully help your lungs function when your NMD gets worse.

When beginning invasive ventilation, consider your medical needs and your desired quality of life. The idea of a ventilator being large, bulky, taking up a lot of space and limiting mobility is in the past. Home-care ventilators are small, portable and light weight, allowing you to have a more normal quality of life. The small nature of the ventilator will allow you to maintain your daily living activities, including going to the grocery store, doctor's office, visiting family and friends, and traveling.

Many ventilator users describe themselves as healthy and enjoying a high quality of life. They report that ventilatory assistance has enhanced their independence, energy and overall health.

Ventilation (*cont'd*)

Tracheostomy tube

A tracheostomy is a surgical opening in the windpipe (*trachea*), made with a surgical incision below the vocal chords or Adam's apple. A tracheostomy tube is placed in the opening in the neck and goes directly into your airway. When you breathe, the air goes in and out through the tube instead of through the mouth and nose. A tracheostomy tube is also referred to as an artificial airway or a trach.

If you need to be on noninvasive or invasive ventilation at all times—or if you find wearing a mask is too difficult or uncomfortable—you may prefer a tracheostomy tube. Moreover, a tracheostomy tube is also preferred if you have trouble swallowing.

For some, a tracheostomy is short term and for others it may be permanent. If you require invasive ventilatory support, the tracheostomy tube will remain in place to allow the delivery of air from the ventilator.

If your neuromuscular disease is getting worse, and if at the time of your tracheostomy procedure you continue to have difficulty breathing on your own, you may wake up in the recovery room with the tracheostomy tube connected to a ventilator. Once all the sedation medications wear off, you will realize that the ventilator is breathing for you.

Your doctor and healthcare team will carefully monitor your progress and make any necessary adjustments to the ventilator. In most cases, your home ventilator will be used while you are in the hospital to confirm you, your family members and/or caregivers are comfortable with it before you go home.

Many people admitted to the hospital for a tracheostomy and invasive ventilation remain in the hospital from several weeks to a month. During the hospital stay, you, your family members and caregivers will receive training on the equipment you will need at home.¹⁰

Before leaving the hospital, your home-care company will ensure you and your caregivers are given training and information on how to do the following:

- Operate the ventilator
- Perform routine tracheostomy care
- Respond to a ventilation, tracheostomy or airway emergency
- Perform suctioning via a tracheostomy tube
- Troubleshoot and perform daily maintenance and inspections of the ventilator
- Perform manual ventilation with a resuscitation bag

Having a tracheostomy and starting invasive ventilation at the same time can be extremely overwhelming, not only for you, but for your family and caregivers. Don't forget it will take time to become comfortable with the procedures needed to operate the equipment and to maintain your new daily care routine.

Considerations and recommendations

Speaking while on noninvasive or invasive ventilation

You may wonder if you will be able to speak when you are on a ventilator. The answer to this question is complex, and it depends on your current ability to speak and communicate, as well as your degree of muscle weakness.

The type of ventilator interface used can affect your speech. If using a sip and straw interface, your speech may not be affected. When receiving noninvasive ventilation, your speech may not be affected if you use nasal pillows and have good muscle control in the back of your mouth and around the voice box. However, if you are using a full-face mask interface, your speech may be muffled or difficult to understand.

Speaking valve

A special valve, known as a speaking valve, is used so you can talk to your family, friends and caregivers. If you have a tracheostomy, the speaking valve attaches to your tracheostomy tube and the ventilator tubing. If using invasive ventilation, you may find it is easier to speak in cycles with the ventilator.

Quality of life

Your quality of life may change a little or in many cases greatly when you are diagnosed with neuromuscular disease. You must learn how to adapt to your home environment, as well as your ever-changing health condition.

"Many healthcare providers and individuals regard the use of invasive ventilation, as an unpleasant burden, as well as place a stigma and assumption regarding someone's disability. In contrast, for the individuals who participate in quality of life interviews, mechanical ventilation is regarded as a form of assistive equipment, similar to a wheelchair. In these participants' opinion, mechanical ventilation is perceived as a benefit to independent living, enhancing energy and overall health, and considers himself or herself to be healthy and enjoy a high quality of life."¹⁰

To improve your quality of life on invasive ventilation, your doctor and healthcare providers need to evaluate the following measures:

- Physical function
- Emotional state
- Social interaction

Physical function—Refers to your ability to perform tasks you were previously able to perform. As your neuromuscular disease gets worse and more severe, it may be difficult for you to have the same physical function. Pain may be a factor on your overall physical function. By controlling your pain, your overall physical function may improve. Over time, you will become comfortable using the ventilator and may feel positive about its benefits.

A majority of individuals on invasive ventilation reported an overall improvement to quality of life as a result of how they felt physically.¹⁰

"My energy was back, I was renewed, It was wonderful...it was just,...it was noisy because the air had to escape, it went *kshhhhh* with every breath, but that didn't bother me because I was so glad to have this wonderful thing that was making me breathe," said one.¹⁰

"I discovered it just gave me so much more energy to work throughout the day I figured what the heck, you know, why struggle when I don't have to," said another.¹⁰

The benefits associated with the use of invasive ventilation appear to help with the adjustment process as these participants found that their overall health and energy improved and the occurrence of health problems, such as respiratory infections and headaches, decreased, resulting in fewer admissions to the hospital for respiratory conditions.¹⁰

Considerations and recommendations (*cont'd*)

Emotional state—In most cases, the emotional adjustment to using invasive ventilation starts as soon as being placed on a ventilator. Although adjustment is both physically and emotionally demanding during the initial weeks, months and years of ventilator dependence, adjustment and acceptance do occur over time as you become familiar with the ventilator, your overall health improves, and daily life and routines become established.¹¹

Typically, there are three phases to adjusting to invasive ventilation:

1. Deciding to start using invasive ventilation
2. Introduction to invasive ventilation
3. Ongoing adaptation and learning how to live using invasive ventilation

However, an emergency introduction may not involve any decision-making phase.

In many neuromuscular diseases, as you become more fatigued and tired, depression may be prevalent. However, in many cases, the use of medication treatment and psychological support may be helpful and improve your quality of life.¹⁰

To improve your quality of life on invasive ventilation, your doctor and healthcare providers need to evaluate the following measures:

- Physical function
- Emotional state
- Social interaction

Individuals with a positive perception of life and personal philosophy are better able to move forward and enjoy their lives.

Social Interaction—Some individuals initially felt "embarrassed and stigmatized by the visibility of their equipment or tracheotomy," according to the CHEST study. Lacking knowledge or experience, and being influenced by common public and media perceptions that being on a ventilator is an "ICU life-supported" phenomenon, it was difficult for them to believe they had any kind of quality of life ahead of them.¹⁰

Some individuals experienced the termination of their employment at the same time they were started on invasive ventilation. They felt that their dependence on the ventilator seriously inhibited their quality of life, at least in the early years, if not on an ongoing basis.¹⁰

However, many who had felt their quality of life was initially affected later developed positive coping skills that enabled them to surmount, incorporate and adjust in a positive way to yet handle and deal with another encumbrance.

"I just accepted it, because I went through my life having kind of everything normal and then a major change would happen, and I'd go from there, so at that point I was kind of used to having events change things, and I knew it was out of my control in a sense so I just kind of accepted it," said one person.¹⁰

These individuals held perceptions of life and a personal philosophy, which enabled them get on with their lives and enjoy them completely.¹⁰

Resources

Support groups

- **Amyotrophic Lateral Sclerosis Association**
The association provides a forum to share information and practical experience, a safe place to allow your emotions to speak for you, an educational gathering spot where speakers and caregivers address subjects of major interest and exchange, and a place to witness firsthand the constant miracle of people continuing to live productive, fulfilling lives in spite of having ALS. See:
 - About ALS: <http://www.alsa.org/about-als/>
 - ALS Association Chapter Support Groups: <http://www.alsa.org/community/support-groups/>
- **Myasthenia Gravis Foundation of America**
The Myasthenia Gravis Foundation helps those affected by MG by providing information on the diagnosis and treatment of MG. It also provides support groups, and can assist in locating community resources. See:
 - Foundation Homepage: <http://www.myasthenia.org>
 - Community Support: <http://www.myasthenia.org/CommunitySupport.aspx>
- **Muscular Dystrophy Association (MDA)**
MDA provides a wealth of information and support to those affected by any of the 43 neuromuscular diseases covered under the organization. Over 200 offices across the country provide support groups, education, equipment assistance and other resources. See:
 - Association Homepage: <http://www.mdaua.org>
 - Help Through Services and Support: <https://www.mda.org/services/support-groups>

Online resources

- Johns Hopkins Medicine: Types of Muscular Dystrophy and Neuromuscular Diseases: http://www.hopkinsmedicine.org/healthlibrary/conditions/nervous_system_disorders/types_of_muscular_dystrophy_and_neuromuscular_diseases_85,P00792/
- MDA: Learn About Neuromuscular Diseases: <https://www.mda.org/disease>
- National Institutes of Health: Neuromuscular Disorders: <https://www.nlm.nih.gov/medlineplus/neuromusculardisorders.html>
- University of Pittsburgh Department of Neurology: What is Neuromuscular Disease?: http://www.neurology.upmc.edu/neuromuscular/patient_info/what.html

Important end-of-life decisions

In many instances, you will be able to attain the quality of life you desire even though you may be continuously on a mechanical ventilator. However, if you become close to the end of your life, the ventilator often just prolongs the dying process until another body organ fails.

Life support replaces or supports a failing bodily function. If the neuromuscular disease is curable or treatable, life support is used temporarily until the illness or disease can be stabilized and the body can function normally. However, if the neuromuscular disease is incurable, the body will never regain the ability to function without life support.

Questions to consider

- Do you wish to receive noninvasive ventilation before being placed on invasive mechanical ventilation?
- Do you wish to receive invasive ventilation through a tracheostomy tube?
- Do you wish to be resuscitated via chest compressions, emergency ventilation or drugs to keep your heart pumping, should your heart stop working?

To prepare for the possibility of impending respiratory failure and the inability to breathe on your own, you need to consider your desired quality of life and goals. Once you have done this, discuss your wishes with your doctor, who can explain the types of interventions available and the subsequent consequences of each.

After carefully considering and discussing your choices, communicate your wishes to your doctor, family members and caregivers.

When discussing end-of-life wishes, you need to make clear to your loved ones and your doctor whether you would want to be placed on a ventilator—even if you would never regain the ability to breathe on your own or return to an acceptable quality of life. We also encourage you to seek expert help and advice. Be your own advocate! As a patient, you need to decide the level of care that you wish to receive.

Advance healthcare directive (*living will*)

It is advisable to prepare a legal document that clearly states your wishes for treatment and care. An advance healthcare directive or living will contains your instructions regarding treatment and care. For instance, you can indicate whether or not you wish to receive basic life support, such as cardiopulmonary resuscitation or advanced life support (*which generally includes mechanical ventilation*). You also may specify the type of medical response you desire—such as ventilation only, medication only or no resuscitation efforts—in the event you suffer cardiac arrest or respiratory failure. Prior to making these decisions, it is important to consult with your doctor who can explain the treatments and potential consequences of your choices. Moreover, the laws regarding advance healthcare directives vary with each state, so it is important to consult a lawyer in your state.¹¹

You can revoke or change your directives at any time, provided you are mentally competent to do so. Review and update your healthcare directive regularly.

Durable power of attorney

A durable power of attorney is a legal document that gives someone you trust the power to make decisions on your behalf should you become unable to do so. The person you have chosen should have a copy of this document and also family members and caregivers, to give to first responders and emergency room staff.¹¹

Palliative care

Palliative care, according to the World Health Organization, is “an approach to care that improves the quality of life of patients who are facing the problems associated with life threatening illness through the prevention and relief of suffering by means of early identification and correct assessment of pain or other problems, whether physical, psychosocial or spiritual.”¹²

Palliative care respects your choice of medical care and helps your family to deal with practical coping issues and grief throughout the disease process and in cases of bereavement.

Important end-of-life decisions (*cont'd*)

Commonly used life-support measures

As your neuromuscular disease progresses, you and/or your family members will need to make some decisions about what types of life support measures should be implemented. The most commonly used life support measures include artificial nutrition and hydration, mechanical ventilation, cardiopulmonary resuscitation and the starting or stopping of treatment.

Artificial nutrition and hydration—Artificial nutrition and hydration, or tube feeding, supplements or replaces ordinary eating and drinking by giving a chemically balanced mix of nutrients and fluids through a tube placed directly into the stomach, the upper intestine or a vein.

Artificial nutrition and hydration can save lives when used until the body heals. Long-term artificial nutrition and hydration may be given for serious intestinal disorders that impair the ability to digest food, thereby helping you to enjoy a quality of life that is important to you. Long-term use of tube feeding frequently is given for end-stage conditions. Often, the treatment will not reverse the course of the disease itself or improve your quality of life. Some healthcare facilities and doctors may not agree with stopping or withdrawing a feeding tube. Therefore, you need to explore this issue with your doctor and clearly identify your wishes about artificial nutrition and hydration in your advance directive.¹¹

Mechanical ventilation—For those at the end stage of their neuromuscular disease, mechanical ventilation often merely prolongs the dying process until some other body system fails. It may supply oxygen, but it cannot improve the underlying condition. When discussing end-of-life wishes, make clear to your loved ones and your doctor whether mechanical ventilation should be started if you would never regain the ability to breathe on your own or return to an acceptable quality of life.

Cardiopulmonary resuscitation (CPR)—CPR is a group of treatments employed when breathing and/or the heart stops. It is used in an attempt to restart the heart and breathing and can be performed either by a family member at home or in a hospital. Electric shock and medications also are used frequently to stimulate the heart, but are only available in a hospital. When used quickly in response to a sudden event like a heart attack or drowning, CPR can be lifesaving. But the success rate is extremely low if you are at the end of a terminal disease process, as CPR only offers a small chance of recovering and/or leaving the hospital.

Stopping and starting treatment—A distinction often is made between not starting treatment and stopping treatment. However, no legal or ethical difference exists between withholding and withdrawing a medical treatment in accord with your wishes. If such a distinction existed in the clinical setting, you might forgo treatment that could be beneficial out of fear that once started it could not be stopped. It is legally and ethically appropriate to discontinue medical treatments that no longer are beneficial. It is the underlying disease—not the act of withdrawing treatment—that causes death.

Final thoughts

When making decisions about palliative care and commonly used, life-support measures, ensure you have gathered all of the facts and understand how it will benefit you. It is essential to understand the advantages and the disadvantages of each type of treatment. A treatment may be advantageous if it helps suffering, restores function or enhances your quality of life. However, the same treatment can be considered burdensome if it causes pain, prolongs the dying process without offering benefit or adds to the perception of a diminished quality of life.

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