

GUIDE FOR CAREGIVERS



**A Guide for
Individuals
and Families**

DISCLAIMER: This document is meant to inform and educate the community. The information presented is not intended to replace discussions with your healthcare provider and is not and should not be considered to be medical advice. Please consult with your healthcare team and/or insurance company for information specific to you.

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About MDA

Muscular Dystrophy Association (MDA) is the #1 voluntary health organization in the United States for people living with muscular dystrophy, ALS, and related neuromuscular diseases. For over 70 years, MDA has led the way in accelerating research, advancing care, and advocating for the support of our families. MDA's mission is to empower the people we serve to live longer, more independent lives.



About this Guide

If you, or someone you love, has been diagnosed with muscular dystrophy, ALS, or a related neuromuscular disease, please know you're not alone. In this guide, we offer information, tips, and resources for finding caregiving help, as well as more specific information and considerations for providing care to a person with a neuromuscular disease. The information in this guide is designed to empower you and your family to take charge of your caregiving needs.

Support and guidance are also available through the MDA National Resource Center. Contact the MDA Resource Center at **833-ASK-MDA1 (275-6321)** or **ResourceCenter@mdausa.org**. MDA's trained resource specialists are available Monday through Friday, 9 am to 5 pm Central, to answer questions and provide one-on-one support and resources.



To learn more, visit mda.org and follow MDA on Instagram, Facebook, Twitter, TikTok, and LinkedIn.

About Neuromuscular Diseases

Neuromuscular diseases impact muscle function due to problems with the nerves or muscles. The spectrum of neuromuscular diseases is broad, with some occurring from birth and others appearing later in life. The impact of a neuromuscular disease on a person's mobility also ranges from mild to severe, and many of the diseases carry risks for respiratory and cardiac complications. Due to the progressive nature of neuromuscular diseases, many individuals will require assistance and support at some point in their journey.

Below is a breakdown of some of the various subtypes of neuromuscular diseases. This list is not exhaustive of all neuromuscular diseases. To learn more about a specific disease, visit mda.org.

Muscular dystrophies

The muscular dystrophies are a group of diseases that cause weakness and degeneration of the skeletal muscles. Some types include Duchenne muscular dystrophy (DMD), Becker muscular dystrophy (BMD), congenital muscular dystrophies (CMDs), limb-girdle muscular dystrophies (LGMD), and myotonic dystrophy (DM).

Motor neuron diseases

In these diseases, nerve cells, called motor neurons, progressively lose function, causing the muscles they control to become weak and then stop functioning. Amyotrophic lateral sclerosis (ALS), spinal muscular atrophy (SMA), and spinal-bulbar muscular atrophy (SBMA) are types of motor neuron diseases.

Ion channel diseases

Diseases associated with defects in proteins called ion channels typically are marked by muscular weakness, lack of muscle tone, or episodic muscle paralysis. Some types include myotonia congenita, hyperkalemic or hypokalemic periodic paralysis, and paramyotonia congenita.

Mitochondrial diseases

These diseases occur when structures that produce energy for a cell malfunction. Friedreich ataxia (FRDA or FA) and mitochondrial myopathies are types of mitochondrial diseases.

Myopathies

A myopathy is a disease of muscle in which the muscle fibers do not function properly, resulting in muscular weakness. Some types include congenital myopathies, such as central core disease and nemaline myopathies; distal myopathies, such as GNE myopathy; endocrine myopathies, such as hyperthyroid or hypothyroid myopathies; inflammatory myopathies, such as inclusion-body myositis and dermatomyositis; and metabolic myopathies, such as Pompe disease and McArdle disease.

Neuromuscular junction diseases

These diseases result from the destruction, malfunction or absence of one or more key proteins involved in the transmission of signals between muscles and nerves. Myasthenia gravis (MG), Lambert-Eaton myasthenic syndrome, and congenital myasthenic syndromes are types of neuromuscular junction diseases.

Peripheral nerve diseases

In peripheral nerve diseases, the motor and sensory nerves that connect the brain and spinal cord to the rest of the body are affected, causing impaired sensations, movement, or other functions. Charcot-Marie-Tooth disease (CMT) and Giant axonal neuropathy (GAN) are peripheral nerve diseases.

Because of continuing advances in research and medicine, this list is meant to serve as a guide. To find out if MDA covers a disease that is not listed here, contact the MDA Resource Center at **833-ASK-MDA1 (833-275-6321)** or **ResourceCenter@mdausa.org**.

Section One: Finding Caregiving Help

Introduction to caregiving

Caregiving is providing on-going assistance to someone who needs help with everyday tasks. It may include help with bathing or dressing, help with household tasks like laundry or cooking, support organizing and paying bills, or any combination of tasks. Caregiving can also involve emotional support for the individual.

Caregivers can be family members, friends, and/or hired through an agency or a combination of individuals.

As the needs of the individual change or increase, it may make sense to enlist the help of an outside caregiver. There are many benefits to finding a caregiver outside of the family. These benefits may include:

- Ensuring the safety and prevention of injury to the individual needing care and/or to the caregiver.
- Increasing efficiency in completing activities of daily living (ADLs).
- Fulfilling a need that cannot be done without help, particularly for persons living alone.
- Allowing individuals more autonomy in directing their care and routines, without impacting family roles.
- Giving family caregivers respite when care needs escalate or are needed 24/7.

Physical demands, emotional demands, other family responsibilities, jobs and more can take a toll on even the strongest, most devoted caregivers. Don't wait until stress becomes extreme. Rarely will someone provide the kind of care that the primary family caregiver can give a loved one, but many people, with the right training and direction, can do a lot of the tasks.

Although it may feel foreign to hire someone from outside your family to come into your home, keep in

mind that hired caregivers do this job because they enjoy what they do and want to be helpful.

Rewards of caregiving

Health research scientists are finding that — despite the tremendous burden that caregiving can present — people can benefit physically and emotionally from the act of caregiving.

It's believed that the positive emotions generated by helping behavior may bolster caregivers' overall sense of well-being, affecting life satisfaction and the ability to cope with stress. Positive emotions also have been shown to lead to increased longevity and faster recovery from cardiovascular stress.

These effects are available not only to the primary caregivers of individuals with neuromuscular disease or those who spend many hours a day caregiving — they also benefit volunteers and others who are asked to help. This “helper's high” can be found in the little moments such as a smile shared between caregiver and loved one. Some may find pride in overcoming difficulties or in knowing a loved one is receiving the best care possible.

Of course, these benefits arise for the caregiver only when he or she is given adequate support. So, inviting others to share the care not only helps the primary caregiver; it gives others a strong emotional boost — it's a win-win-win situation!



Positive emotions have been shown to lead to increased longevity and faster recovery from cardiovascular stress.

Types of Help

Agency help

Before you get started on finding a caregiver, it is important to understand the difference between home healthcare services and home care services.

Home healthcare services typically refer to care that needs to be performed by a **licensed medical professional**. These services are often short in duration, and the client usually needs to be homebound to qualify. There needs to be a medical reason for the service requested, and a licensed skilled provider such as a neurologist, registered nurse, physical therapist, occupational therapist, or speech therapist must be part of the care plan. For example, the individual may require wound care or intravenous medication in which a medical professional will need to monitor the symptoms or side effects.

Home care services are **non-medical** in nature, as they aid with daily care activities that do not require monitoring from a licensed provider like a physician or registered nurse. In many cases, receiving help to perform activities of daily living can mean the

difference between staying in one's home safely and needing to move to a community-based care facility or relying on a family member to provide care. These services may be offered for a few hours a week and all the way up to 24 hours a day. Home health agencies use personal care attendants (PCAs) to provide basic home care services. These are non-

licensed caregivers but are often certified as a medical assistant or nursing assistant. PCAs often benefit from training or direction about how best to perform tasks or healthcare interventions.

Home care assistance can include:

- Companionship
- Supervision
- Transportation for errands and appointments



- Household chores
- Meal preparation
- Medication setup and reminders
- Assistance with personal care (bathing, dressing, grooming)
- Assistance with mobility and transferring around the home

Some agencies can provide both “skilled nursing” and basic home care services. The level of care and need determines which services are enlisted. Often home care agencies also employ respiratory therapists and physical or occupational therapists and offer other healthcare services.

It is important to note that skilled nursing activities require a licensed nurse to perform the activity. Examples of skilled nursing activities include IV infusions, providing G-tube feedings, and some types of respiratory care, like suctioning a tracheostomy.

Talk to your MDA Care Center team, or your primary care provider (PCP) to discuss what types of care best meet your/your loved one's needs.

Working through an agency ensures that you have a partner in arranging and managing the care and the caregivers. The agency handles all hiring, training,



Some agencies can provide both “skilled nursing” and basic home care services.

supervision, payment, taxes, and other paperwork. The agency is responsible for finding a person that is qualified to perform the tasks that are needed. You are not responsible for any worker compensation injuries or damage to your home during their scheduled time. In addition, the agency will work with you to develop a backup plan if your regular caregiver is sick or has scheduled time off.

Private help

There may be times when you/your loved one would prefer to hire a caregiver privately and not work through an agency. Some families prefer to bypass the agencies and hire a person on their own to assist with caregiving. Someone with little or no medical training can be hired for a lower rate and can be trained in basic caregiving tasks such as lifting, bathing, and maintaining equipment. This type of aide can be a live-in assistant or someone who comes in a couple of afternoons a week to relieve the primary caregiver.

Agency vs. Private Help: Pros and cons

	Agency Help	Private Help
Pros	The agency handles hiring and letting go of employees.	There's no need to deal with agency guidelines, regulations, and applications.
	The agency performs background checks and keeps records such as timesheets.	You can possibly pay a lower rate to someone who doesn't come through an agency, especially a high school or college student.
	Agencies carry liability insurance.	In some states, Medicaid waivers offer funds to pay caregivers hired directly by the family.
	Agencies may offer backup staff if your staff member is out for any reason.	
Cons	You may have less input on the selection of aides since your aide(s) will come from the agency's pool of employees.	Some recordkeeping is involved when you hire private aides such as hours worked and payment history.
	If staff is not available, you may not have coverage that day.	There's no agency to send a backup aide when a caregiver is unavailable.
	Agency help may be more expensive than private help, unless insurance is covering it.	Health insurance may not cover the cost of an independently hired aide.
	You may live outside of the agency's service area.	You may be responsible for paying federal and state employment taxes if payment to the aide(s) is over a certain dollar threshold.
If the aide is a family member or friend, there can be awkward consequences if things don't work out or problems arise.		

Deciding What Help You Need

Before you look for an aide, **it is important** to know exactly what you need the person to do. Make a list of tasks and write a job description. Take a few days to jot down any chores that you could use help with as you go through your routine. Are you primarily interested in physical care, companionship, housekeeping, or a combination of all three? In the job description, include rules such as no smoking, no cooking, no loud music, etc. Do you need the person to have a car to do errands? Do you need someone to be on call or to work set hours? Is a male or female assistant preferred? Outlining your/ your loved one's preferences beforehand will make for a smoother experience.

Concerns about accepting outside help

Sometimes accepting outside help can be stressful, scary or create conflict when personal cares are to be carried out by someone other than the person's most trusted family member. Careful and clear communication about each other's needs may be helpful, as well as coming up with solutions to alleviate sources of stress and fear about the care plan. You may also enlist your medical team if anxiety of enlisting help is preventing delivery of essential cares.

If you/your loved one are hesitant to accept care from someone outside of the family, start slowly. Perhaps allow an outside caregiver to come into your home for just a few hours at first to help with a couple of tasks, then gradually build up the time and tasks

Finding an agency

You/your loved one's MDA Care Center team and/or primary care provider (PCP) can assist in identifying agencies in your area. Insurance and Medicaid case managers may also direct families to agencies contracted with their insurance or Medicaid plan. Home healthcare providers and home care providers may also be found through a quick internet search under "home care" or "home healthcare." Your Area Agency on Aging may also have a list. You can also



contact the MDA Resource Center at: **833-ASK-MDA1 (833-275-6321)** or **ResourceCenter@mdausa.org**. An MDA Resource Specialist can help you find agencies in your area.

Finding private help

To find private help (not through an agency), word-of-mouth is the best resource. Below are some ideas:

- Reach out to your faith community, friends, and neighbors to see if they know of someone.
- Use online ad boards, but be sure to research the website thoroughly before posting personal information to make sure it is a reputable website.
- Consider using your social media to post the opportunity in private community groups.
- Check with independent living centers or senior centers.
- Place classified ads in local or college e-newsletters or newspapers. College students, high schoolers or scouts may be able to fill a community service requirement by caregiving.
- Put announcements on bulletin boards or newsletters at hospitals or social service organizations.
- Ask at a community college or university career centers or departments of nursing, physical therapy, or social work. Students may be looking for part-time jobs or unpaid internships that offer



experience.

- Ask the social worker or other healthcare professionals at your MDA Care Center for other resources in your area.
- Call your community volunteer center.
- Ask at privately owned drugstores and medical clinics if they know of anyone.

Paying for Help

There are costs associated with in-home care. Some agencies will accept state and local funding sources, such as waivers and Medicaid. Services will be individually designed to consider the goals and needs of each client, as well as the family's budget. In-home care often can be integrated with other community-based services to increase one's options. Try not to get discouraged! There are several options to explore.¹

Medicaid waivers: Many states provide TEFRA or HCBS waivers under Medicaid. These provide funding for caregiving for children and adults with special healthcare needs. These are publicly funded services. See the sidebar to learn more about these programs.

Medicare: If ordered by a doctor as part of a larger plan of care, Medicare may cover assistance such

What are Medicaid Waivers?

Medicaid waivers were established to waive rules that usually apply to accessing Medicaid programs. The goal is to allow states to expand coverage or improve access to home-based care for children and adults with disabilities who might otherwise require care in a facility or go without appropriate help in their home. There are several types of waiver programs. Two of the most common types of waivers are the Home and Community-Based Waivers (HCBS) and the Tax Equity and Fiscal Responsibility Act (TEFRA), also known as the Katie Beckett option. These waivers give states the option to extend Medicaid to children with disabilities by counting only the child's income for eligibility, rather than the income of the parents/family.²

Please note that waiver programs and eligibility vary from state to state. Contact your state's **Medicaid office** to find out what waiver programs might be available for you or your loved one.

² "What is a Waiver?". Kidswaivers.org, Complex child magazine, Retrieved on February 22, 2022, <https://www.kidswaivers.org/>.

as bathing, dressing, and basic medical care, but only on a part-time or intermittent basis. Some Medicare Advantage plans may offer broader coverage of care even if skilled nursing is not needed.

Private insurance: Skilled nursing and home health care may be paid by some private insurance if the level of care is determined to require a licensed nurse.

Long-term care insurance: Some long-term care insurance plans will pay for home care assistance depending on the policy coverage.

Veterans programs: If you/your loved one is a former service member, there are programs through

¹ <https://www.aarp.org/caregiving/financial-legal/info-2017/afford-a-homecare-worker.html>

the Department of Veteran's Affairs (VA) that may be able to assist with the costs of home care. To learn more, contact your local [VA benefits office](#).

Older Americans Act: Some in-home assistance for low-income people over age 60 with a disability may be covered under the [Older Americans Act](#), with funds administered by the state.

Private pay: Paying out-of-pocket for caregiving through an agency can be expensive. It is important to consider your/your loved one's finances before considering this option. Some sources of income to consider may include annuities, investments and savings, and life insurance policies that can be used for qualified home-care expenses. Federal tax deductions may be available for those that pay out-of-pocket for home care.

A few states fund the hiring and paying of a family member as a caregiver. To find out if this applies to your state, check with your county's [human services office](#), [Area Agency on Aging](#), or state department of social services.

Hiring, Training, and Managing Caregivers

Once an agency has been identified, below are some questions to ask before working with them:

- What type of training, supervision and monitoring does the agency provide its staff?
- Is there specific training provided related to neuromuscular diseases? If no, is the agency open to learning from the MDA Care Center team or other primary care provider (PCP) about my/my loved one's specific disease?
- Does the agency develop a plan of care, and how often is it reviewed and by whom? (Typically, a licensed medical provider has to sign-off on the plan. This could be your/your loved one's PCP, neurologist, or palliative/hospice care provider.)
- Will the agency be in communication with my/my loved one's neurologist and/or PCP?
- Is there a 24-hour emergency contact for questions or concerns?
- How does the agency ensure that staff are

available for the requested times?

- How does the agency ensure consistency of care providers?
- Is there a charge for canceling a scheduled day?
- How does the agency screen their staff? Are criminal background checks completed?
- What happens if I don't feel my caregiver and I are compatible?
- Is there a minimum number of hours of service I must receive?
- Are the agency's caregivers employees or independent contractors (1099 tax form)?

Often, an agency requires a referral from a doctor or other licensed healthcare provider to start providing services. Below is an overview of the typical process that will occur:

Step 1: Referral

Typically an agency requires a doctor/other licensed healthcare provider to make a referral.



Step 2: Benefits Investigation

The agency does a benefits investigation to determine if insurance will cover the costs associated with their services.



Step 3: Interview

The agency typically interviews the person receiving care as part of the enrollment process. Together they determine how to best accomplish the referral goals.



Step 4: Identifying a Caregiver

After understanding the needs and preferences of the person receiving care, the agency seeks out and hires a caregiver for the tasks.



Step 5: Provider Review

The plan of care is reviewed and signed by the referring provider. The plan of care needs to be

reviewed, updated, and certified periodically.

Interviewing and hiring private help

Hiring your own caregiving assistant means you have to screen applicants for criminal records, review their background and training, follow-up with references, etc. Take caution to avoid dishonest, incompetent, or even dangerous applicants from entering your home by requesting a criminal background check. You may need to pay for the background check as part of the hiring process.

Initial communication: When someone calls in response to the ad or inquiry, ask what they'd like to know. For example, it's a bad sign if their first question is "How much does the job pay?" or "How much time off do I get?" It's better if they ask what some of the duties are or how many hours they'd be needed. Explain the job briefly, but you do not need to describe the details of your/your loved one's exact diagnosis or physical condition or the number of people in the household. You can ask a bit about their experience or background over the phone or via a video conference; the initial screening should let you weed out inappropriate applicants. If you get a good feeling from the phone call, ask the person to meet for an interview. This could be an in-person interview of you could conduct an initial interview via video conferencing and then, if that goes well, you can set-up an in-person meeting. For in-person meetings, consider safety and invite another friend or family member to be present with you or meet at a public place.

Interviewing: Expect interviewees to dress appropriately (that doesn't necessarily mean suits or heels, but they should be well-kempt and presentable) and to arrive on time or call if they must be late. Expect someone who listens, makes good eye contact, and asks relevant questions. Applicants should bring a résumé and should be able to provide professional and personal references. You can also print out a simple job application form from the internet or create your own and ask them to complete it prior to the interview.



After the interview: When you've narrowed the field to one or two leading candidates, it's time to check their background. This is imperative, especially if the person hasn't been referred by someone you know. You can't be too careful about exposing your home and your loved ones to someone you don't really know. Make sure they give you a permanent address, date of birth, Social Security number and driver's license number. Run a criminal background check and driving records check (if they will be driving you/your loved one). There are several websites that will do this for a fee or will explain ways to go about searching; see [lexisnexis.com](https://www.lexisnexis.com); [virtualchase.com](https://www.virtualchase.com); [searchsystems.net](https://www.searchsystems.net). Watch out for scams. Reputable websites include [ReverseRecords.org](https://www.ReverseRecords.org) and [CourtRecords.org](https://www.CourtRecords.org). You can also check with your state's Bureau of Criminal Apprehension, as they offer free services.

For all applicants, ask for names and contact information of several people you can call for references. Former employers are best; students also may list teachers or people from church or the neighborhood for character references. If an applicant was not chosen for the job, make sure to let them know. This can be done over the phone or via email. You do not need to provide specific details as to why they were not selected; simply let them know you decided to go with another candidate and

that you appreciated their time and interest in the position.

Offering the position: For caregivers you hire directly (not through an agency), you may wish to draw up an informal contract with the caregiver that spells out hours, pay rates and other details of the job. You each should sign two copies, and each keep a signed copy.

Additional considerations when hiring privately: Check with an accountant to be sure you're keeping the correct records. Record hours and payments. Tell aides that, as contractors, they're responsible for paying their own Social Security taxes. Some people have the caregiver sign a time sheet or pay sheet every week or two weeks and give them a copy of the record to keep.

At the end of the year, send a W-2 form to the IRS indicating how much you've paid for income tax purposes; the caregiver also gets a copy. Your homeowner's insurance should cover liability for injury. If you're concerned, ask the person to sign a no-fault agreement..

Hiring a live-in caregiver

If you are seeking a live-in caregiver, be specific about expectations: Can anyone move in with them? Can they bring a pet? What's included —rent,

utilities, food, phone, internet, furniture? Are days off set or negotiable? Can they have friends over for dinner or overnight? Ask if the applicant is in a relationship, and make clear whether it's okay for the partner to spend time at your home. If so, how often? It is also important to consider legal issues, including whether or not adding a live-in caregiver will violate rental agreements, occupancy issues and more. For example, if the caregiver is injured while caring for you in your home, will your renter's or homeowner's insurance cover medical costs?

Setting expectations

Once you have decided to move forward with an agency or a private caregiver it is important to clarify requirements and preferences at the beginning of the working relationship. Below are some suggestions to get off to a great start:

Be completely honest about needs. Agencies, as well as private aides, should be familiar with almost every type of assistance required; you/your loved one does not need to feel embarrassed or concerned that you are asking too much.

State preferences from the start. Be specific about hours, daily routines, and household rules such as "no smoking" and off-limits areas of the home. Don't assume the aide will know your exact wishes.

Make it clear who in the family is the authority or decision-maker. Is it the person with neuromuscular disease or the primary family caregiver or both? If everyone in the family gives orders to the aide, the result can be confusion and failure to give proper care.

Give feedback to the employee or agency on a timely basis. If there's a problem, no matter how small, address it immediately to be sure the agency and employee are clear on your wishes. Inviting someone to spend many hours in your home isn't easy. First, all those involved must accept that the assistance is needed enough to sacrifice a bit of privacy. Eventually everyone, including the person with a neuromuscular disease and the primary caregiver, will come to accept that this helper can



improve the family's quality of life.

Training

What Should I Ask?

Asking the right questions will help you learn about the applicant's communication style, problem-solving skills, understanding of the job, and qualifications. Ask open-ended questions about their interest in the position, background, work ethic, and ability to meet the requirements of the job. It is important to be respectful in the interview and stick to questions that pertain to the job. Personal questions not related to the job should be off-limits.



Be specific about your expectations for a caregiver.

Now that your caregiver is on board, it is important to provide them with information and resources about your/your loved one's neuromuscular disease and your/their specific needs. Below are some tips on how to support your caregiver as they learn the

job:

- Prepare a written summary and/or share print-ready materials from reputable medical websites or non-profit organizations, such as MDA's educational materials (mda.org/services/education-materials).
- Make copies of any instructions or advice you've received from healthcare professionals, such as those regarding activities of daily living. Consider creating a training video of each expected intervention. Videos are easily stored on a tablet, phone, or cloud storage via a link.
- Go through each new caregiving task carefully; demonstrate several times and show the new person how to recognize your/your loved one's reactions. Watch as the aide performs the task the first few times and give helpful hints. Be patient with the learning curve.
- Post a daily schedule, listing times for getting up,

bathing, meals, meds, etc. Be very specific about dosages of medications.

- Provide a monthly or weekly calendar that lists times of therapy and doctor appointments. If the individual requiring care has speech difficulties, these lists will help new caregivers understand the needs they may be describing. Explain any other keys to assist with communication.
- If you are managing the caregiver for your loved one, be sure to ask your loved one for feedback on the caregiver and share that feedback with the caregiver.

Other Sources of Help

Most of us find it difficult to ask for help; we like to show the world, and ourselves, that we can handle it, whatever it may be. Caregiving is a personal, round-the-clock experience; sometimes it can be difficult to know how to parcel out tasks. There are many ways others can help. Friends, neighbors, church members, co-workers, social acquaintances, and family members may have offered to help, but they may not know how. People usually will help if they receive specific directions.

Using volunteer help from those who care about your family has many advantages:

- It saves money.
- It draws friends and family closer together and makes everyone feel needed.
- It gives the primary caregiver a little respite, which will benefit his or her physical and mental health.
- Knowing there's help can make it easier to keep a loved one at home, avoiding or delaying placement in a care facility.

Start by making a list of all the people who have offered to help, and all those who haven't offered but who might be willing if asked. Develop a list of needs, including frequency, and start matching needs with available people. Friends and neighbors don't need to get involved in the most intimate aspects of caregiving. Household chores like cleaning or meal

preparation, yardwork, running errands, providing childcare, etc. may be more comfortable and appropriate for them, and they'll allow the primary caregiver more time for hands-on, one-on-one care.

Here are some other tips for getting friends, family members and neighbors involved:

Be clear about what you need. Avoid vague requests: "Maybe you could stop by to visit with AI for a few minutes some time?" Instead, be direct and specific: "Could you come by Sunday and watch the game with AI on our TV, so I can take a nap upstairs?"

Brainstorm sources of help and expertise. Be careful not to ask the same people over and over. Think about neighbors and acquaintances who might be glad to do simple repairs to your house or stop by the dry cleaners. Ask a knowledgeable relative to help sort out bills or insurance papers.

Get help getting help. Widen your list of helpers by asking a close friend or relative to ask people on your behalf. Using an intermediary often makes the process more comfortable for everyone.

Set up a care calendar using a website such as Lotsa Helping Hands (lotsahelpinghands.com) which provides an efficient, effective way to recruit and schedule volunteer help from family and friends.

Keep a list of tasks and needs on your phone or in a notebook, so when well-wishers ask, "Is there anything I can do to help?" you can tell them. Or ask if you can call them when you need something from the store or a short respite. If they say yes, call them.

Expect some hesitation or refusal. Your request simply may come at an inconvenient time. Consider giving them a second chance, but don't spend time trying to convince someone who's refused two or three times or doesn't keep a promise.

Accept gifts. Some people may not have time to give of their time but would be happy to offer financial support. If asked, suggest a takeout meal,

a professional housecleaning, a gift card to buy supplies, or a movie or audiobook.

Don't get bossy. If you find you're ordering your friends around and expecting them to do things in certain ways or take on additional chores, stop and think. Either let the person do the task in his or her own way, or realize that if you need that much control, it would be better to hire someone who expects to take orders.

Children as helpers

Even very young children like to feel they're helping their loved one. Children have helped parents and siblings with suctioning, feeding tubes, changing trach tubes and catheters, or household chores. They can learn by watching and can be given simple tasks — such as "bring Mommy the formula for Daddy's dinner" — at first.



It is important not to over tax a young child, as this could cause role confusion.

Children involved in caregiving learn compassion, cooperation, patience, and responsibility. Most siblings or children of a person with a neuromuscular disease consider caregiving as part of the family routine.

It is important not to over tax a young child though, as this could cause role confusion and lead to resentment if the child's needs go unmet. Children/teens who take on excessive responsibility for caregiving may develop depression, anxiety, insomnia, loss of interest in school, or delay becoming independent. Children/teen caregivers should be encouraged to get in touch with the social worker at the MDA Care Center or a school guidance counselor to find support.

Long-distance caregivers

Today, family members are spread across the world, and it isn't always possible for them to relocate to care for a parent or sibling. Despite the physical distance though, there are many things that long-distance secondary caregivers can do to support their loved one. For example, they can:

- Manage the loved one’s budget
- Make phone calls and research resources online
- Participate in telehealth visits
- Provide emotional support through regular phone calls, video chats, text messages, and more
- Offer financial support if their budget allows. If a loved one is reluctant to accept money, offer to pay for a specific need such as food, equipment repairs, medical supplies, etc.
- Coordinate communication between family members, providing updates on the loved one.

Long-distance secondary caregivers should do their best to stay informed, but also respect the primary caregiver’s leadership by offering helpful suggestions gently.

“Share the care” circles

A unique method of organizing helpers to share the load is known as a Share the Care circle. Share the Care circles can start with a small group, such as a prayer group at church, a handful of co-workers or a bunch of longtime friends. This core group takes on responsibility for organizing needs and finding people to fill them, removing the burden from the primary family caregiver of recruiting and organizing volunteers.

A circle eventually includes dozens of people and reaches far beyond the personal acquaintances of the individual needing care. Long-distance caregivers can play important roles in the circle. Groups often give themselves names: Jeff’s Angel Group, David’s Circle, Friends of Cindy. Information about needs and schedules can be posted on a care coordination calendar website such as Lotsa Helping Hands, Caring Bridge or even a shared Google Calendar, so that volunteers can offer specific help.

In care circles, a few individuals take responsibility for organizing areas such as laundry, shopping, childcare, communication, etc. The circles ripple outward to include all kinds of specialists: attorneys, accountants, fundraisers, building contractors, handymen, equipment donors. As each person is

contacted for help, that person reaches out to others. The circle organization assures that no one person has too much responsibility, and everyone has a manageable workload.

Other Assistance

Online and delivery help

Many services and businesses offer delivery and pickup. These include pharmacies, grocery stores, laundry and dry-cleaning services, restaurants, and more. Online ordering makes it easy to order groceries and have them delivered to your home or pick them up without leaving your car. Using these efficiently can save tremendous amounts of time (and gasoline) that make it worth any extra cost. Explain your situation to businesses such as car repair shops, etc., and they may be willing to pick up and deliver.

Respite care

Respite care allows the primary caregiver time to rest and recharge. Primary caregivers should call on their network of paid or volunteer helpers to stay with their loved one so they can get a haircut, go to a ball game, shop, enjoy a hobby, or just sit in the park and watch birds.

Every community has respite facilities such as adult day care centers that will care for the loved one for a few hours or sometimes even days to give the caregiver a break. In addition, a loved one who qualifies for hospice can spend a night or two at a hospice facility.

Schedule respite regularly — it will help everyone in the family, including your loved one. At first it may feel uncomfortable or “selfish” to take time for yourself, but remember you can’t care for others if you aren’t taking care of yourself.



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Support groups and counseling

Individuals living with a neuromuscular disease and their caregivers credit support groups with new friendships, important advice on equipment and more. Individuals and families who are farther along on the journey can help new members come to the realization that they can get through this. Some support groups invite knowledgeable speakers or have casual social time. There often are no boundaries on what things can be discussed. Toileting, child-rearing problems, intimacy, family conflicts, breakdowns, death — all are parts of the experience that can be brought to a support group.

Private or family psychological counseling may be helpful for a time and often is covered by insurance. Search for psychologists or social workers online, ask for a referral at the MDA Care Center, or contact the MDA Resource Center for community resource referrals.

MDA's Community Groups also offer a safe space for members of the neuromuscular disease community to share resources, experiences, and support. To join, visit mda.org/care/community-groups.



Section Two: Daily Care for Individuals With Neuromuscular Diseases

This section offers suggestions and resources for handling some topics that might arise in your/your loved one's daily care. There is no way to anticipate each individual's specific needs, so we encourage you to speak with your doctor or another medical professional for more in-depth information that pertains to your loved one.



DISCLAIMER: This document is meant to inform and educate the community. The information presented is not intended to replace discussions with your healthcare provider and is not and should not be considered to be medical advice. Please consult with your healthcare team and/or insurance company for information specific to you.

Young Adult Caregiver Considerations

Teaching children self-advocacy and directing others to provide their caregiving promotes confidence and leads to greater autonomy as they get older. Children can begin learning about their condition and participating in decision-making from a very young age. It is developmentally appropriate for children to notice and point out differences between themselves and their peers. Support your child as they learn to tell their peers and adults about their neuromuscular condition, or use of braces or a mobility device. Role play is a great way to help children practice these conversations. Ask your child about their conversations with friends and classmates to ensure they have the tools to talk about their differences positively. Allow your child to ask questions and encourage open conversations. Some helpful books to read with your child include; *I Have Muscular Dystrophy* and *It's Okay!* by Dr. William M. Bauer & Mallory Bauer, *I'll Walk with You* by Carol Lynn Pearson and *The Abilities in Me* by Gemma Keir.

Often, school provides the first self-advocacy opportunities. Check with your library or bookstore to find children's books specific to the diagnosis or books addressing disability. School teachers or guidance counselors can also use these books to open classroom discussions about unique differences between classmates. Peers can learn how to support one another both emotionally and physically.

Adult caregivers can encourage and answer questions using age-appropriate language. If you are unsure of the language to use with your child, talk with your care team's psychosocial support staff, such as child life specialists, social workers or nurses. As a general rule, children need honest and simple answers to their questions. Use of metaphors can be difficult for children to interpret and apply to their own situation. Avoidance of sensitive topics instills a sense of taboo or worry that the topic is worse than reality.



Puberty can be a difficult transition for anybody. Adolescence is a time to develop a sense of identity, develop friendships and gain autonomy or independence from parents. Adolescents with disabilities may face inequities or discrimination that can lead to insecurities. There may also be physical barriers to accessing recreation and socialization opportunities, such as lack of accessible transportation, accommodations and help to complete physical tasks (which to this point may have been done by family members). Teens with disabilities need to learn how to ask for help and direct others who offer help. Sometimes this is with a friend when there is already a good rapport. Other times, it may be with a stranger (e.g. opening a door or pushing an elevator button).

Encourage your adolescent to pursue their interests, identifying activities or skills that promote self-esteem. Adults should strategize with the youth to identify and remove access barriers, but also teach people without disabilities acceptance and how to offer help. Examples of help include showing interest in including them and outright asking if they would like help. For the teen with a disability, there may be a lack of self-confidence or actual bullying preventing full participation in their community. Consider finding a mental health provider during this

time. This may help your adolescent develop coping or communication skills and provide an outlet to discuss frustrations related to their diagnosis or other aspects of life. **MDA's Virtual Learning Program: Recognizing and Addressing Mental Health Concerns in Children** can serve as a guide for navigating neuromuscular disease and mental health in youth.

Provide opportunities for your child to ask for and receive caregiving help from other individuals in their lives. Parents should move from an active role to a more supportive role. This can be tough for parents, but young adults need to develop skills to express their own needs, coordinate services and be flexible in establishing accommodations. Support your teen while allowing them to take charge of their caregiving. Encourage them to set up and manage their caregiving schedule, participate in interviewing outside caregivers, and fully participate in planning meetings pertaining to caregiving at school. Participating in the process is the best way to learn how to direct cares

MDA offers a variety of programming and educational materials for youth to explore including Summer Camp, Mentorship Programs, print

resources, Access Workshops, and other young adult programming.

ALS Special Considerations

Caring for a loved one with ALS can bring some unique challenges. Patients with ALS may experience much more rapid changes in function than patients with other neuromuscular conditions. Frequent visits to your ALS clinic (typically every 3-6 months, or as often as recommended by your neurology team) will be helpful in navigating these changes. Often it may be necessary to re-institute care with physical therapists, occupational therapists, or other professionals due to a change in condition.

ALS usually starts out in one of two ways: bulbar onset or limb onset. When the motor neurons in the brain stem are affected first, then symptoms start in the face, mouth, throat and tongue, called the bulbar region.

More commonly, motor neurons in the spinal cord are affected first and symptoms start in the limbs — arms, legs, hands or feet. Rarely, the respiratory muscles are affected first. In all types of onset, symptoms eventually spread to other areas of the body.

ALS doesn't unfold in a predictable pattern. Some people have slow progression of symptoms, while others experience a rapid loss of ability. Sometimes there's a big loss followed by a plateau. Although each individual follows his or her own course, it's possible to make broad generalizations about the progression of the disease.

ALS caregivers move from helping with buttons and balance to providing a steadying arm, feeding a meal bite-by-bite, helping with more intimate personal tasks, learning to operate medical equipment and supervising a 24-hour care operation that includes other sources of help. Caregivers of ALS patients also have to deal with the emotional stress of watching their loved one decline rapidly. One positive result of the COVID pandemic is that



mental health resources such as counselors is now available by video consultation.

Early modifications include taking up anything that can cause tripping, such as scatter rugs or pet toys, and installing handrails in the shower/tub. Later modifications include ramps to the outdoors and increased bathroom space.

In two-story houses, a downstairs bedroom space may be necessary. Locations range from simple to elaborate: moving furniture, changing to wider-opening door hinges, installing an electric door opener, remodeling a bathroom, installing overhead tracks for an electric lift system. Modifications may be tax deductible to some degree.

“Get it before you need it” is the mantra of ALS. Some people see using assistive equipment as giving in to the disease, but in fact the opposite is true. Adaptive equipment is like a weapon in the battle against the disease. Canes, braces, walkers, wheelchairs, communication devices, coughing and suction machines, feeding tubes, lift chairs, hospital beds, alternating pressure mattresses, mechanical lifts and assisted ventilation make it possible to thrive as ALS progresses. Assistive equipment enables greater independence and safety for your loved one, while making it easier for you to be an effective caregiver. Not using this equipment can result in injuries to the person with ALS and the caregiver. Another reason to get equipment early is that it takes time to order and get approval from insurance or Medicare. If a move to assisted living, nursing home or hospice is being considered down the line, be sure to get all essential equipment first, as Medicare may not pay for it afterward.

Patients with ALS may also have the added challenge of fronto-temporal dementia (FTD) to contend with. FTD is a change in cognition that often manifests as personality change, loss of inhibition, or anger and lashing out at caregivers. It can be difficult to differentiate these changes from stress or situational depression, anger or anxiety, so reach out to your ALS Care Team if you notice anything concerning.

Activities of Daily Living Basics

Accessibility at home

Home modifications can make it easier on everyone. Plan ahead and make modifications before they're needed. Regardless of your budget, there are things that can be done to improve the safety and accessibility of your home. There may also be financial resources available in your local area. Talk with your MDA Care Center social worker or contact the MDA Resource Center to see what programs might be available in your area.

Modifications that you can start right away include:

- Clearing clutter from walkways in the home.
- Removing anything that can cause tripping, such as scatter rugs or pet toys.
- Rearranging furniture to make the environment more accessible.
- Installing handrails in the shower/tub.
- Adding non-stick decals or mats to the shower or tub floor.
- Adding equipment such as bedside commodes or urinals, shower chairs, lift chairs, etc., to improve safety and reduce the amount of energy expended on physical tasks.



Modifications that might happen down the road depending on disease-progression may include:

- Installing ramps or stair lifts.
- Creating a sleeping area on the main floor of the home.
- Utilizing smart home technology for lights, door locks, television sets, etc.
- Bathroom modifications.
- Total lift systems including portable lifts such as a Hoyer, or an overhead lift system.

Your/your loved one's primary care provider (PCP) or MDA Care Center team can help you arrange a "home safety assessment," where a physical and/or occupational therapist can evaluate your home and make specific recommendations on assistive devices and home modifications.

Falling

Even when a person can walk, trips and falls can occur without warning, and it may be impossible to use the arms to brace for a fall. Injuries from falls range from minor to major and the recovery period can lead to additional loss of physical ability — the last thing anyone with a neuromuscular disease wants.

The best way to avoid falling is to use assistive equipment; however, many people will consent to using a cane or a folding shopping cart but feel embarrassed about using more "medical" devices like a walker or wheelchair. Assistive equipment actually allows more independence and fights the disease by preserving precious strength and energy.

Lifts

As neuromuscular diseases progress and muscles become weaker accessibility needs can increase. Mechanical lifts can save caregivers' backs, necks, and shoulders from injury. Lifts are useful in transfers, such as from bed to wheelchair, or from wheelchair to toilet or bath, and in getting someone up off the floor after a fall. Lifts are operated mechanically or with motors and have a sling of sturdy material on which the individual sits. There are several types of slings, geared for different uses.

Besides freestanding lifts, some operate on ceiling tracks.

Sometimes people with neuromuscular disease are leery of lifts, feeling insecure and vulnerable swinging in this new contraption. Practice on other family members to get a feel for it and to allow your loved one to see it in action. Once the mechanics have been mastered, this is a terrific piece of assistive equipment.

Tips for Preventing and Handling Falls

Follow these tips for loved ones and caregivers:

- Ensure the home environment is safe by eliminating tripping hazards and adding sturdy handholds at strategic locations. Shield sharp corners on tables and counters and add a shower chair in the bathroom.
- Helping someone up off the floor after a fall can be dangerous for both of you. When possible, use a mechanical lift or recruit a neighbor or family member to help. If you can't get the lift to the person, roll the person onto the lift sling or a sheet and pull to a location where the lift can be used.
- Be careful to use proper body mechanics when lifting. If no help is available, call 911. If you ask the rescuers not to use their sirens, they usually won't.
- Ensure your loved one has a cell phone, emergency assistance pager or other signaling device for falls that happen when you're not around.
- If the individual stays alone for periods of time, ensure rescuers (neighbors or emergency personnel) can get into the house by hiding a key outside.
- Protective equipment like knee and elbow pads and a helmet can make falls less injurious. Lighter weight shoes are easier to negotiate.

Motorized lift chairs and uplift seat cushions (with a spring-powered seat) can boost a person upright when arms are too weak to help push up. Be sure the chair is correct for the person's height. These aren't likely to be covered by insurance policies, while mechanical lifts usually are. Check your policy. Elevators can also be installed especially in newly constructed buildings. To go upstairs and downstairs, a chair lift can be installed on the home's major staircase.

Safety

In addition to being prepared for the unexpected, there are some precautions to keep in mind for everyday safety.

- Keep the house free of obstacles. Remember that weak arms make it difficult or impossible to brace for a fall, and know that knees abruptly can give way without warning. Be available to lend an arm during walking, and encourage the use of assistive equipment.
- Using a wheelchair is much safer than attempting to walk when each step is a struggle. Some individuals may resist the wheelchair but may be willing to use it when a lot of walking is required. Keep the seat belt on whenever the person is in the wheelchair — at home, in public, in the car, etc. A slight bump, a steep ramp, or uneven terrain can make someone tumble out of the chair.
- The caregiver and individual also should carry cell phones with important and emergency numbers programmed in. A medical alert bracelet or medallion can provide first responders with important information about your/your loved one's health.

Many cities maintain voluntary registries for people with disabilities to self-identify to receive assistance during emergencies and disasters. Contact your local emergency management office to find out more.

MDA also offers fillable Emergency Alert Cards that you/your loved one can keep in their wallet or purse in case of an emergency. To download an ER Alert Card, visit mda.org/education.

Personal Hygiene

Bathing

Personal hygiene is very important and ensuring that you/your loved one is receiving proper hygiene care can make a huge difference in how you/your loved one feels. As a neuromuscular condition progresses, bathing techniques may change.

For some, handrails, shower chairs and handheld showerheads make it possible for people to continue bathing themselves. Others may need more assistance with shampooing their hair, brushing their teeth, or shaving. In some cases, it may be necessary for the caregiver to completely oversee personal hygiene, including tooth brushing, hair combing, shaving and nail trimming. Mechanical lifts and a shower chair with back and arms make the task easier. A bathing sling is used on a standard lift and has a cutout for easier transfer into the tub.



Mechanical lifts can save caregivers' backs, necks, and shoulders from injury.

Individuals who have difficulty sitting independently may need extra support in the bathtub. There are many different types of shower and bath equipment ranging from simple positioning support chairs to fully adjustable reclining sling chairs. Work with your/your loved one's occupational therapist to help find the best solution for you/your loved one.

Some bathing tips:

- Too-hot water and too-vigorous toweling off can cause dry, itchy skin.
- Hot tubs and spas may be inadvisable because the heat can overtax the breathing system, and jets can damage weak muscles.
- Dry shampoo can help you/your loved one go longer between showers or baths, without feeling like your/their hair is getting dirty or greasy. There are also dry shampoo caps, which look like shower caps, that let you clean your/your loved one's hair without getting dry shampoo on your shoulders.

- Use a hand mitt for scrubbing. Maneuvering a washcloth can be tricky, but an exfoliating glove makes it easier. When you're done, drop the mitt on the floor of the shower and rub your feet on it to get your feet clean without having to bend over.
- For sponge baths, a height-adjustable bed, such as a hospital bed, eases the strain on caregivers' backs.
- Foam grip tubes that look like tiny pool noodles, can be cut to size and put around the handles of items like razors or toothbrushes for better grip.
- If it's difficult to brush with a traditional toothbrush, try an electric toothbrush shaped like a bite guard. It vibrates to cleanse teeth without having to move it around your mouth.



Dressing

While the individual may still be able to dress without assistance, some techniques can help with weakened hands and arms. Attach a pull, like a key chain, to the ends of zippers. A reacher can help pull up pants or move clothing around. Hooks and loops can be sewn onto skirts or pants. The person should sit for as much of the dressing process as possible; when standing, support should be nearby. Thanks to a number of disability advocates, clothing manufacturers are producing more accessible clothing lines. Some brands include Target Kids Adaptive, Zappos Adaptive, and Tommy Hilfiger Adaptive.

Eventually a caregiver's help may be needed. Larger sizes are easier to manipulate, and looser underwear makes toileting easier. Your loved one can wear pants or skirts with the back seam opened, sitting on a towel in lieu of underwear. When the wearer is seated, the clothing looks normal. This makes toileting easier as no undressing is required.

Teeth

Dental care for a person with a neuromuscular disease may require caregiver assistance. As it gets harder to hold a toothbrush or hold the mouth open, the first step is an electric toothbrush, sometimes with a Waterpik. A child's toothbrush or foam-tipped swabs such as Toothette plus Oral Swabs with

Mouth Refresh Solution can be used without water and are easier to get into a tight mouth. These are available at many pharmacies and medical supply stores. Biotene toothpaste foams less than others and is easier to swallow or spit out.

Some people with neuromuscular diseases bite their cheeks, lips, and tongues. A strong mouthwash can help heal the sores. A variety of mouth guards, such as SleepRight night guards or plastic sports mouth guards, can be found in drugstores. A dentist can order a rubber bite block or a custom mouth guard. These guards can hold the mouth open while teeth are brushed and be worn at night to prevent teeth grinding.

Massage may help relax a clenched jaw. Be careful not to force anything into the jaw, because the person may inadvertently bite down hard on a toothbrush or a finger.

Getting teeth professionally cleaned can be tricky when the individual has excess saliva, a tight jaw, or trouble sitting up or lying back. Any serious dental work should be done as early as possible after the diagnosis to minimize these complications. In most communities there are dentists who specialize in helping people with disabilities; the regular dentist

or county dental society may be able to recommend someone.

During a dental checkup or cleaning, strongly remind the hygienist to suction constantly and thoroughly to avoid choking on saliva. Sometimes caregivers take over the suctioning because they're more familiar with signs from the patient and have to suction the mouth frequently at home.

Medicare and some private health insurance may cover dental care. A prescription or letter from the neuromuscular disease specialist can help ensure the coverage, though it's not a guarantee.

Toileting

Naturally, people with neuromuscular diseases prefer to take care of their own bathroom needs for as long as possible. This can be a touchy subject and is best handled step by step as needs increase – and with humor and as much privacy as possible.

- **Remodel.** At home the bathroom should have a door wide enough for a wheelchair to enter, and enough space for it to turn around, i.e., a 5-foot radius. Removing the door and hanging a curtain instead may be all that's needed. Special hinges also enable wider door openings. Add grab bars around the tub and toilet.
- **Stay regular.** As managing the bathroom becomes more difficult, people are sometimes tempted to cut back on their liquid intake or "hold it in" as long as possible to minimize trips. This is a bad idea. Dehydration can lead to constipation and swallowing problems, which are uncomfortable and unhealthy. Most people with neuromuscular disease have bladder and bowel control and know when they need to use the restroom. Persons with incontinence may use incontinent briefs or pads.
- **Raise the seat height.** Even for those who can transfer onto and off the toilet independently, getting up from a seated position can be a problem. Many raised toilets are available, including seats to be added to an existing toilet, raising the height by 2 to 13 inches. This becomes more important as leg, hip and trunk muscles

become weaker. A new toilet with a very high seat can be installed; there are self-contained units that don't require drilling into plumbing. A less expensive option is a portable, adjustable-height bedside commode chair that can be placed over the toilet. Some commode/ shower chairs have wheels, allowing transfers to take place in another room.

- **Lift with a sling.** A hygiene sling has a large cutout that makes toileting possible from the lift.
- **Say hey to the bidet.** Arm weakness may make it difficult to use toilet paper effectively (hand-held toilet paper extenders are available). One independence strategy is the bidet. These provide a gentle warm water wash and warm air drying. Some have remote controls that can be operated by hand or foot, or by someone outside the door. Bidets run from \$400 to \$800 and usually can be installed by the homeowner. Bidet wands are good for traveling or public restrooms.
- **Try portable urinals.** Portable urinals can save several bathroom trips a day. A bottle shaped urinal is a good solution for men. Portable urinals for women have larger openings and handles. These can be used independently or with a caregiver's help, in standing, lying, or seated positions. It's best to put down a waterproof pad first.
- **Consider a catheter.** Men can use external condom catheters; some experimenting may be necessary to find the right type. Foley catheters involve inserting a tube into the urethra and connecting to a drainage bag. These indwelling catheters are used by men and women, and don't interfere with movement or transfer. The bag has to be checked and emptied, and the insertion site needs to be cleaned daily. If the catheter becomes clogged, painful, and leaky or shows signs of infection, the doctor may replace it. The drainage bag should be cleaned periodically with two parts vinegar or chlorine bleach and three parts water. A surgically placed suprapubic catheter may be used if the Foley is too uncomfortable or causes continued infections. It's inserted directly into the bladder through the abdomen wall.
- **Toilet training for kids.** Toilet training a toddler

or young child with a neuromuscular disease may take longer. Remain supportive and encourage them throughout the process. Some children continue to use a brief for longer periods. If your child is becoming frustrated, consult with your physician.

- **Keep it clean.** The time may come when your loved one is unable to transfer to the toilet at all and must perform bladder and bowel movements while in bed. Adult diapers (which also may be worn under clothes when going out) or disposable bed pads under the person in bed are the usual solution. Thorough, gentle cleaning is important after a bowel movement to protect against bedsores. Wearing rubber gloves, turn the person onto one side and wash gently with baby wipes and a very mild soap such as Johnson's Bedtime Bath. Dry carefully and use some Desitin or other powder. A mix of Desitin with zinc oxide is comfortable and protective. To clean sensitive areas in the shower, install a hand-held shower head or use a make-shift shower with two enema bags, one containing liquid soap and warm water, and one with warm water. Use one to clean the area, then the other to rinse.
- **Using public restrooms.** In public places, there's usually a toilet stall big enough for manipulating a wheelchair. Still, your loved one may need assistance to transfer to the toilet and adjust clothing; a bidet wand may be convenient to carry. If the caregiver is the opposite sex from the person with neuromuscular disease, ensure privacy by asking a bystander to stand guard or announcing "man (or woman) in the room" whenever someone enters. Or carry along a sign saying "Handicapped — opposite sex using restroom," and attach it to the door.

Health Monitoring and Interventions

Ears

Some individuals report a feeling of the ears being plugged, as if they have water in them or need to

"pop." Check with a doctor to rule out infection, sinus problems, wax buildup, or other medical causes. Hearing loss is associated with some neuromuscular diseases. In the absence of these problems, it's possible the cause may be weakness of the muscles that maintain tone in the Eustachian tubes connecting the mouth and ear canals. In addition, weakness in the muscles that elevate the nostrils (thus opening airways) can lead to a chronic feeling of being stuffed up.

Possible remedies include "nose openers" used to stop snoring, such as Breathe Right nasal strips. These strips of tape go across the nose and hold open the nostrils. Antihistamines and decongestants sometimes help, and some say they can get their ears to pop by using their Cough Assist machine. For some, the ears become painfully folded during side-lying. Some caregivers create an "ear pillow" to prevent this problem. Using a piece of foam of the proper thickness to keep the head in alignment (not tilted up too high), cut out a 4-inch hole in the center for the ear, and cover the foam with a pillowcase.

Eye care

Some individuals may have burning or dry eyes, or — just the opposite — watery eyes. Either case can be irritating and even painful. This could be caused by lack of blinking or by air blowing from respiratory equipment. Saline eyedrops, or a warm wet washcloth or cold washcloth compress, may help. An antibiotic may be needed if irritation persists.

Itchy scalp

Some people with neuromuscular disease seem to experience greater-than normal scalp itchiness. There are a variety of possible causes, including dryness, yeast or fungus overgrowth, and stress. Check with a doctor and experiment to find the right remedy. Some things that have worked for others:

- Use gentle hypoallergenic hair care products.
- Shampoo hair in lukewarm water — hot water strips away protective scalp oils. Towel dry gently, not vigorously. Don't wash too frequently or too infrequently.
- Use a humidifier.
- Over-the-counter remedies include Benadryl,

Absorbine Jr. (applied liberally to the scalp), shampoos containing ketoconazole (such as Nizoral) or coal tar (such as T-Gel). Scalpicin Anti-Itch Scalp Treatment is a spray that can be used several times a day.

Learn more hygiene tips in these Quest Media articles:

- [Tips for Better Hygiene with a Disability—And Why it Matters](#)
- [Easy Clean: Bathing Products for People with Disabilities](#)
- [Back-to-School Beauty Hacks with Wheelchair Barbie](#)

Jaw clenching

An uncomfortable tightening or chattering of the jaw in response to cold, anxiety or pain may occur with neuromuscular disease. There are many different medications that can help with relaxation; we encourage you to speak with your physician to discuss different medical interventions.

Pressure sores

Whenever skin is under prolonged pressure, tiny blood vessels are compressed, the supply of oxygen and water is interrupted, and skin starts to die. Pressure sores (decubiti) are caused by staying too long in one position and by medical equipment such as a ventilator mask or neck brace. Sores can lead to infection, long and inconvenient recovery periods, and in extreme cases, death.

Some cautions to observe:

- Nutritional or respiratory problems can cause skin to become very fragile.
- People who are immobile should have their skin checked thoroughly at least once a day. For light-colored skin, an area that stays red longer than 15 minutes after pressure is removed is the beginning of a pressure sore. For dark-colored skin, look for areas that are darker or purplish-blue in color.
- A developing pressure sore causes a burning, itching pain. Even if there is little or no redness at the spot, treat the area as a pressure sore by

reducing pressure and cleaning the area. Prompt response to this early sign can make a huge difference in just a few days.

- Ensure your loved one always is properly positioned in bed, chair, and wheelchair. Reposition at least every two hours or whenever asked by the person due to discomfort.
- Protect body parts that sustain a lot of pressure (especially skin over bony areas like elbows, shoulders, heels, tailbone, shoulder blades, ears, and the back of the head) with pillows, pads, specially designed cushions, or cushioned skin dressings such as Duoderm.
- When making a foam pad to sit on, don't cut out the center (as is done for corns or bunions) as this can further decrease circulation.
- When people no longer can reposition themselves in a chair or wheelchair, get a pressure relief cushion, like a ROHO or Jay cushion, as ordinary foam or air cushions aren't adequate to prevent pressure sores.
- Experiment to find the most comfortable and effective pressure reduction in bed, such as standard or alternating pressure air mattresses, real or synthetic sheepskin, foam mattress overlays, and automatic turning mattresses.
- Beware of recliners, where people with neuromuscular diseases tend to spend a lot of time. Outfit the recliner with a pressure cushion or sheepskin padding, protect elbows and heels, and keep a close eye out for problems.
- When respiratory equipment masks and neck supports cause skin breakdown, alternate with different masks or head support systems to give skin a rest.

Shoulders

Immobility can cause a painful condition called frozen shoulder. Although the shoulder may move, it's stiff and movement causes pain. Range-of-motion exercises can prevent this condition and are a component in its treatment.

Shoulder pain also occurs when the weight of the arms isn't supported, causing a constant pull on weakened muscles and the shoulder joints. Carrying heavy objects can cause the arm to be subluxed

(dislocated). Ensure proper positioning by supporting arms with armrests or pillows. Caregivers also need to protect the loved one's shoulders during transfers, by not pulling on their arms to move them.

Skin

Some skin changes have been noted in neuromuscular diseases, such as changes in the biochemical properties of collagen and elastin which run through the dermis, or middle layer of skin. Blood vessels in the dermis also display irregularities and protein deposits as neuromuscular disease progresses. Poor nutrition and respiration can make skin more fragile.

Caregivers can take several steps to ease skin woes for loved ones with these diagnoses.

For dry itchy skin:

- Moisturize the air with a humidifier.
- Hot water strips the skin of moisture; use lukewarm water for baths and showers. If a hot soak is a must, use bath oil.
- From the standpoint of cleanliness, it's usually not necessary to take a full bath every day; a "parts" cleanup with a sponge usually is sufficient.
- Use mild or soap-free cleansers. Avoid deodorant soap.
- Pat skin dry with a towel; don't rub.
- Immediately after patting dry, seal in moisture with the greasiest lotion tolerated.
- Shave with lotion instead of foam.
- Try over-the-counter itchy skin remedies, such as those containing cortisone.
- In some cases of chronic itching or skin sensitivity, a physician may prescribe an anti-epileptic drug.

Fungal infections like "jock itch" can be caused by being seated all day, which creates warm, damp pockets in underarms, groin, and skin folds.

To combat skin infections:

- Dry skin completely after washing, using a hair dryer on the cool setting.

- Buy clothing that wicks moisture away from the skin.
- Use over-the-counter antifungal sprays, powders, and creams; avoid cornstarch, talcum, or other nonmedicated powders.

Healthy-skin nutrition includes:

- Drinking eight to 10 glasses of water a day.
- Taking a daily multivitamin-mineral supplement.
- Eating daily servings of foods rich in linoleic and essential fatty acids such as safflower oil, nuts, avocado, seeds, soybeans, salmon, tuna, shrimp, and corn oils.
- Getting extra protein, zinc, and vitamins A, C, and K for skin damaged by pressure sores.

Be sure to consult with a nutritionist and/or dietitian before making any changes in your diet.

Gastrointestinal (GI)

Constipation

This is a very common and frustrating problem. It may be the result of general immobility; a side effect of some medications; the need for more fiber and liquid in the diet; and/or muscle weakness that makes it hard to bear down to expel the stool.

Constipation isn't defined by how often a person has a bowel movement, but by whether the stools are hard, dry, and difficult to pass. It's not necessary to have a daily bowel movement, so long as the task can be accomplished without straining.

Common remedies:

- Gentle dietary fiber is found in raw fruits and vegetables, bran, seeds, or high-fiber cereal bars.
- Bulk or fiber laxatives are a concentrated form of dietary fiber.



People who are immobile should have their skin checked thoroughly at least once a day.

- Stool softeners keep stools moist and lubricated.
- Stimulants increase involuntary muscle contractions, moving the stool along more quickly.

A daily regimen can pull water into the intestines and soften stool. Others swear by mini-enemas or suppositories. With a physician's guidance, keep trying until finding the solution that works best for your situation.

Things to consider:

- Always respond promptly to the urge to defecate.
- Establish a regular bowel routine, where defecation occurs on a somewhat predictable schedule.
- Consult your doctor or MDA Care Center for suggestions on how to establish a bowel routine.
- Fiber and fluids must be taken together. Without adequate fluid, fiber isn't effective and in fact can aggravate the problem. This also is true for fiber taken through a feeding tube.
- Although there's a danger of becoming dependent on laxatives, stimulants, suppositories or enemas, this issue isn't as acute for people with muscle disease. Regular and predictable bowel movements that follow the person's normal pattern make life more comfortable and easier for everyone. One caveat: With regular laxative use, increase fluids and be alert for signs of dehydration, such as irregular heartbeat, disorientation, extreme thirst, and headache.
- Proper positioning helps move things along. A squatting position is more effective than sitting upright. Carefully elevate your loved one's feet on a low stool and have him or her lean forward. Provide stability and support as needed. For additional force, a pillow can be hugged to the abdomen or gentle abdominal massage provided. If using a lift sling, ensure the buttocks are the lowest part of the body.
- Assist weak muscles by gently but firmly massaging downward on the lower abdomen for several minutes. Some have reported success by squeezing together the person's buttocks and massaging downward for several minutes, or by lightly circling the skin around the anus (on the outside) to trigger the colon to move.

When constipation is chronic, fecal impaction may result. This is a large mass of dry, hard stool that can't be expelled. Symptoms include abdominal cramping and discomfort. Watery stool may move around the mass and leak out, soiling clothes. Do not treat your loved one with an anti-diarrheal product without consulting a physician. Left untreated, impaction can be life threatening and require emergency surgery. Laxatives won't resolve fecal impaction. If suppositories or enemas don't work, the mass may have to be manually removed by a healthcare provider. Suppositories may be given between manual removal attempts to help clear the bowel. If you have any doubts, see your healthcare provider for a diagnosis.

Drooling

This has been called one of the most annoying symptoms in some forms of neuromuscular disease. The problem isn't making more saliva, but having less ability to swallow it. This can cause choking, as well as skin irritation, frustration, and social isolation due to embarrassment.

For some individuals, drooling (sialorrhea) can be controlled or at least brought to a tolerable level with medications. In some cases, a speech language pathologist can demonstrate saliva management strategies, such as head posture, using facial muscles and achieving more frequent swallowing. Physicians may prescribe medications to reduce (not eliminate) the flow of saliva.

Reducing saliva flow can cause another problem — too-thick saliva, which can be difficult to swallow or cough out. A dry mouth also can hasten tooth decay. Remember that drooling and medications that reduce secretions can increase the body's need for water.

Caregivers have tried various creative ways to handle the flow of saliva:

- To catch the saliva, fold a terry cloth towel or washcloth three or four times, and secure it below the lower lip, tucking it behind the jaw or fastening it with an elastic band behind the head.

- Make or buy rolled gauze pads (like those used by dentists) and tuck them between the gum and cheek. Those with arm mobility can wear terry cloth sweat bands on the wrists, for frequent mouth wiping.
- Buy face masks (such as those used by transplant patients or people with allergies) and line the chin area with a strip of absorbent cloth. Cut out the portion that goes over the nose if it causes discomfort.
- Cover bed pillows with a waterproof pad and a folded towel to catch nighttime drool. Suck up saliva with a portable suction machine, similar to those used by dentists.
- The person can grip a folded cloth or paper towel between the teeth to absorb saliva.

Natural saliva reduction strategies include:

- Before bed, gargle and swish with warm salt water, especially under the tongue (some recommend kosher salt).
- Reduce or eliminate dairy, sugar (including artificial), and grain/gluten products from the diet.
- Use a few drops of anise oil in the mouth (available at health food stores).
- Drink warm tea.

Dry mouth

When using medication to dry up saliva, or when using noninvasive pressure ventilation mouth dryness and nose bleeds may result. Remedies include increased fluid intake and attaching a humidifier to the ventilator or in the room. Try an over-the-counter dry mouth spray (available in the oral hygiene section) or moisturizing mouthwash.

Swallowing concerns

Swallowing can be a concern in some forms of neuromuscular disease. Assessment may include consultation with a dietitian or a speech-language pathologist to determine best food textures and medications to help avoid choking or aspiration into the lungs. Many medications may be difficult to swallow. Talk with your pharmacist and providers to determine if medications can be crushed and put in a bite of easily swallowed food and which medication

may come in liquid forms.

Individuals with swallowing difficulties are at risk for choking and may be advised to eat more slowly and take drinks frequently during the meal to aid in food moving smoothly down the throat. Caregivers can ensure that they are not alone during mealtimes and provide some companionship during these extended mealtimes.

Caregivers can ensure that food is prepared in a way to limit swallowing or choking risk.

Genitourinary (Genital and Urinary Organs)

Pregnancy

With neuromuscular disease, there are many factors to consider when contemplating having a child. For women who are considering pregnancy, these factors should be taken into account:

- The enlarging uterus will push up against the diaphragm, squeezing the lungs, which can exacerbate weak breathing.
- Some neuromuscular diseases involve the heart and can be associated with pregnancy-related cardiomyopathy.
- If a Caesarean section is required, certain types of anesthesia may be a problem because of the risk to respiration.
- Vaginal delivery requires strong muscles in the lower body.
- Prolonged bed rest can lead to significant loss of muscle mass, which may not be regained in those with a neuromuscular disease. However, many mothers who have neuromuscular weakness report regaining their strength, especially if they have worked with a physical therapist to regain strength.
- Genetic testing is a type of medical test that identifies changes in chromosomes, genes, or proteins. The results of a genetic test can confirm or rule out a suspected genetic condition or help determine a person's chance of passing on a genetic disease. However, many neuromuscular

diseases are hereditary, or caused by changes in genes which code for how muscle or nerves are formed or function.

- Some couples choose to meet with a genetic counselor to learn about preconception or prenatal testing options. The options allow prospective parents to learn if their offspring are at risk to develop neuromuscular symptoms which may present at birth (requiring early medical care) or later in life. Often during this time, a plan to oversee and care for the mother with neuromuscular weakness throughout the pregnancy is solidified.
- Mothers with neuromuscular weakness may benefit from an extended stay in the hospital to recover, or enlisting a home health agency to help the parents and the newborn adjust to the new routines at home that include feeding and caring for the infant, as well as mother's recovery.

Urinary urgency

There are studies to show incontinence, hesitancy and frequency are common in some NM disorders, possibly related to changes in nerve or the smooth muscle. The sphincter that relaxes to release urine may spasm, causing an urge to urinate. Some prescribed medications may contribute to urinary symptoms and symptoms may reflect an unrelated health problem. Consult your providers to determine the cause of symptoms and to determine a treatment option. Other muscles and nerves are involved, and they may be weakened, making it harder to tighten the sphincter. At times the bladder may have a spasm, causing a powerful urge to urinate. Some medications may lead to more frequent urination.

Urinary urgency may prove temporary. Medications are often prescribed to relax the bladder and lessen urinary symptoms — watch for side effects including headache, dry mouth, constipation, or diarrhea. Urinary urgency also can be a sign of a urinary tract infection, prostate trouble, fibroids in the uterus, or other causes, so be sure to have a thorough medical checkup.

Muscles, Joints, and Mobility

Blood clots

Some people with neuromuscular disease may be at risk for deep vein thrombosis (DVT), a type of blood clot that forms when people are immobile. DVTs are extremely dangerous because they can break off and travel to the lungs, causing a life-threatening pulmonary embolism. Range-of-motion exercises can help to prevent DVT. Your doctor may provide recommendations based on the needs of your loved one.

Signs of a clot in the legs include:

- Redness, heat, or discomfort in one leg.
- One leg that's more swollen than the other.
- Swelling that doesn't go down overnight.
- Pain upon standing or stretching calf muscles.

Symptoms of a clot that's traveled to the lungs include sudden-onset chest pain and shortness of breath. If you suspect a blood clot, don't massage the area. Contact a doctor immediately.

Contractures

To remain healthy, joints must be moved through their range of motion on a regular basis. When joints aren't moved fully and regularly, as happens with some neuromuscular diseases, a contracture may develop. This abnormal tightening of muscles and other tissues around a joint immobilizes the joint, causing pain when it's moved. Because of the pain, the person moves the joint even less, further aggravating the problem. Contractures can develop very quickly as muscles become weakened and, in some cases, paralyzed. Physical therapy and range-of motion exercises are key to preventing contractures. Massage, splints, braces and proper positioning in a bed or chair may help prevent or ease discomfort. Check with your doctor or physical therapist for more information.

Equipment

“Get it before you need it” is the mantra for individuals and families living with neuromuscular disease. Some people see using assistive equipment as

“giving in” to the disease, but in fact the opposite is true. Adaptive equipment is like a weapon in the battle against the disease. Canes, braces, walkers, scooters, wheelchairs, communication devices, coughing and suction machines, feeding tubes, lift chairs, hospital beds, alternating pressure mattresses, mechanical lifts and assisted ventilation make it possible to thrive as neuromuscular diseases progress. Assistive equipment enables greater independence and safety for your loved one, while making it easier for you to be an effective caregiver. Not using this equipment can result in injuries to your loved one or to you.

Another reason to be evaluated early for durable medical equipment (DME) is that it takes time to order and get approval from insurance or Medicare. If a move to assisted living, nursing home or hospice is being considered down the line, be sure to get all essential equipment first, as Medicare may not pay for it afterward.

Feet

In addition to exercises recommended by a physical therapist, and/or the use of orthotics (leg braces), feet need protection in bed, as heels can develop pressure sores and the weight of blankets and foot drop (inability to turn the ankle or toes upward) can cause pain. “Float” your loved one’s heels above the bed using small pillows at the ankles. Prevent feet from flopping down or to the side by bracing them with more pillows. Support the weight of sheets and blankets with a blanket lift at the end of the bed. Blanket lifts, which fit between the mattress and box springs and extend upward to support the bedding, can be purchased from medical supply outlets, or made at home. Sheepskin pads, knee or elbow pads under the heels, foam boots and heel pads also are helpful.

For foot drop see the medical care team about leg braces, some of which the loved one can sleep in. By the way, caregivers should protect their feet and knees also. A thick rubber kneeling pad can help a lot if you’re frequently up and down. And watch for repeated banging of knees against furniture when turning or lifting the individual. Knee pads or a new technique may be in order.



Hands

As hands, arms and shoulders weaken, caregivers can seek out various aids to extend dexterity. These include:

- Wraparound lap desks to support the arms
- Mobile arm supports to allow both horizontal and vertical motion
- Lightweight wrist splints
- Eating utensil holders or specially designed eating utensils such as lightweight large-handled cups and plate guards
- Key holders, doorknob extenders, and light switch extension levers
- Lightweight reachers
- Card holders (for playing cards)
- Button and zipper hooks
- Long-handled sponges
- Velcro fasteners on pants and shirts
- Pencil grips
- Book holders
- Speaker phones

Consult an occupational therapist for more aids and to help solve specific problems. Hand-curling can be managed with hand splints, or by putting a rolled washcloth inside the hand, to keep it in a more natural position and prevent fingernails from digging

into palms. Holding small, heated rice bags in the hands for 10 to 20 minutes can make them more comfortable.

Some other techniques to help adapt to changes in hand and arm strength include:

- In addition to range-of-motion exercises for the shoulders and elbows, try the “prayer position” exercise. Put the hands together as if praying and point the fingers up and down; this helps stretch fingers and wrists and releases stiffness in forearms.
- A sling supported with a strap across the back and over the opposite shoulder cradles the arm and can help protect the shoulder joint from uncomfortable stretching.
- Arm rests on chairs may need to be padded to be high enough to support weakened shoulder muscles. This is especially true if a pressure relief cushion is put on the seat.

Neck

Neck muscles may be weak in some forms of neuromuscular disease, which can make it hard to turn or hold the head upright. This can lead to strain or pain, especially when turning the head, raising and lowering into bed, or rolling over. Poor neck posture also can impair breathing, swallowing and communication. Effective management of neck weakness can prevent or treat pain and injury. The key is to find the best methods of supporting the head.

Be aware that neck weakness makes the person more vulnerable to injury during transfers and when riding in a vehicle. If the head suddenly drops down, back, or to the

side, muscles and ligaments in the neck can tear, or the cervical spinal cord or neck vertebrae can be seriously injured. For some, even a slight jerk is all it takes to tear muscles.

Encourage and assist neck-stretching exercises (after consulting a physical therapist).

Two simple exercises:

1. Slowly turn the head to each side as far as possible without pain; hold and return to the center.

2. Tilt the head sideways on each side, so the ear points down toward the shoulder; hold and return to center.
3. Use a lumbar roll or cushion behind the lower back to prevent slumping when sitting, and to prevent the head and shoulders from tipping forward. Pillows under each arm also promote upright head position when sitting.

Work with a physical therapist to investigate different types of soft collars, neck braces and head supports such as a band around the forehead that attaches to a headrest. Alternating the use of collars and head support systems helps reduce pressure points and skin breakdown. Thin-cushioned skin dressings (e.g., Duoderm) also protect the skin.

Ensure the person’s bed pillow isn’t too thick, as that can cause neck strain. Try placing a rolled towel under the back of the neck, coupled with a thin pillow for the head.

Swollen extremities

In some people with neuromuscular disease, fingers, hands, arms, ankles, feet, or legs may appear swollen, sometimes extremely so. Edema is the retention of fluid and has several causes.

Most commonly in neuromuscular diseases, edema is due to being immobile. Medications used to control drooling can also cause the body to retain fluids, leading to edema. Muscle activity helps push blood through the veins to the heart. Without muscle tension and support, blood pools in the veins, and the pressure causes water in the blood to leak out into surrounding tissue, causing swelling. Edema can be very uncomfortable. While the person is in bed or wheelchair with tilt/recline features, elevating the swollen parts above the level of the heart can reduce edema; prop the hands, knees, and feet on pillows. If using an adjustable hospital bed, note that it raises

the knees, not the feet. It’s necessary to put a pillow at the foot of the mattress to bring the feet up level with the knees.

When the person is out of bed, correct positioning

is critical. Recliner-type chairs can contribute to swelling of feet and ankles because, with the footrest up, all the weight of the legs is on the calves, reducing circulation. Adding pillows in the gap between the chair and leg rest can help distribute the weight and improve circulation.

Whenever the individual is sitting upright, allowing the legs to hang without support will cause severe swelling and should be avoided. Adjust the height of wheelchair footrests or put a box or cushion under the feet to reduce pressure at the thigh and back of the knee.

Swelling of the hands can be minimized by using a lap tray to support the arms. The best lap trays (easily cut from plywood or plexiglass) surround the waist and extend back at the sides to support the elbows. Elbow pads will be needed, and the hands can be raised on a pillow.

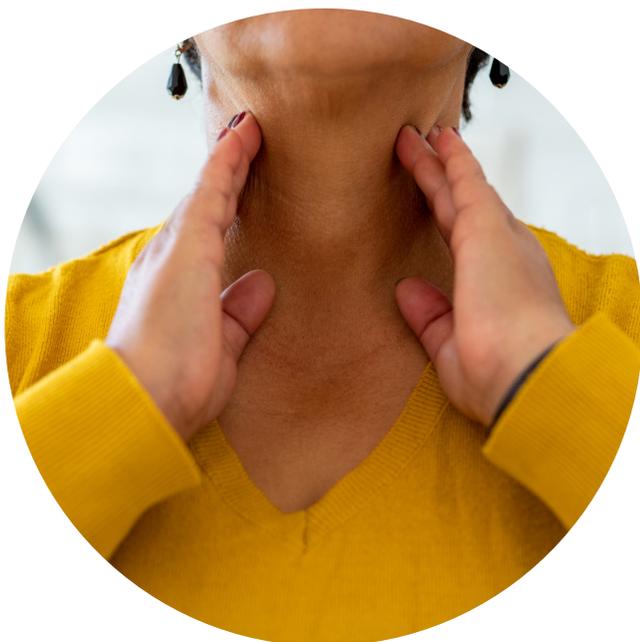
Even a small amount of exercise, such as assisted walking, isometric tightening of muscles, or range-of-motion exercise, can improve circulation. Wearing compression stockings and shoes that lace or fit snugly are better at controlling swelling than lose fitting slippers. Keeping the legs cool also is more comfortable. Reducing salt intake sometimes is recommended, although it's important to keep up

fluid intake.

Diuretics, which increase the production of urine, often are prescribed but should be used only if other methods aren't enough to relieve discomfort or swelling. Diuretics can counteract medications used to control drooling, can deplete fluids, and require additional trips to the bathroom — all problems for people with neuromuscular diseases. The family and medical team need to find a balance between controlling edema and managing other symptoms.

Edema also can be a sign of other medical conditions, such as congestive heart condition, cirrhosis, or kidney problems. Be sure your medical team is aware of swelling that worsens, or occurs in unexpected locations, such as around the eyes.

Edema in a single leg can be a sign of a blood clot.



Exercise

Energy/fatigue

Some forms of neuromuscular disease lead to muscle fatigue which may manifest as general fatigue. Mental exertion also may be fatiguing, plan activities with energy conservation in mind, saving energy for the most important or enjoyable aspects of the day, then rest when needed. Whatever activity a person can do is fine. Whatever activity a person can do is fine; they should rest when fatigue sets in. Consult with your physician about durable medical equipment which may reduce physical exertion. Extreme fatigue may indicate breathing problems, which may need to be addressed with assisted ventilation. In later stages, fatigue may occur even after a passive activity such as being bathed. Some individuals may sleep more hours than usual, even 12 or more hours a day. Be sure daytime naps don't keep the person awake at night. Additionally, some forms of neuromuscular disease have excessive daytime sleepiness as a symptom. Certain medications can help manage these symptoms. Talk to the physician about any concerns about sleepiness and alertness of your loved one.

Exercise for individuals with neuromuscular disease

It's not known how much exercise is valuable for people with neuromuscular disease, and the answer is sure to vary among the different diseases. Before beginning an exercise program, consult the doctor or physical therapist about frequency, duration, and level of intensity. The goal of gentle exercise is to help maintain mobility, improve endurance, and minimize pain from the effects of muscle wasting. When exercise isn't possible, physical therapy and range-of-motion can help serve the same purposes.

Important exercise guidelines include:

- **Don't overdo it.** Soreness after exercise might be an indication that you're damaging muscle, which isn't good.
- **Think aerobic.** This helps improve heart health and circulation. Possibilities include a stationary bicycle, upper body ergometer, elliptical

machines or aquatics, or multiple repetitions of gentle resistance strengthening and stretching exercises.

- **With weights, start small.** Increase only when appropriate and by a very small percentage.
- **Just say no to pain.** Forget the motto "no pain, no gain." This doesn't apply to people with muscle disease. Focus on maintaining functional strength, endurance, and independence.
- **Be flexible.** Don't hold to a rigid program. The person should listen to his or her own body and use common sense.

Exercise for caregivers

Caregiving, especially lifting, is hard on the back, neck, and shoulders. Good body mechanics (like lifting with the legs, not the back) and assistance (using a mechanical lift or helper) significantly reduce the risk of injury and should always be observed. But without proper flexibility and strength in legs, hips, shoulders, abdominal, and buttock muscles, an injury can be just one wrong move away. Regular exercise — say an hour at the gym or a daily walk — is a very good idea for caregivers, with both physical and emotional benefits. Talk with a trainer or doctor about a good program for you. Because it can be hard to find the time for lengthy exercise breaks, consider incorporating several quick exercise periods into daily routines. The following 30-second stretching and strengthening exercises, performed several times throughout the day, can have a positive cumulative effect. They can be fit into odd moments, like while waiting at a traffic light, cooking, or standing in line. Pairing an activity with an exercise — say doing partial squats while waiting for the toast to pop, hamstring stretches during a TV commercial and core muscle strengthening while blow-drying hair — can allow caregivers to improve without even realizing it.

NOTE: Check with your doctor before undertaking any new exercise program.

- **Flexibility:** This is critical to preventing injury during the many caregiving tasks that put the body into unaccustomed positions. Stretch to the point of discomfort but not beyond. Pain or



distance isn't the point. Hold stretches about 15 to 30 seconds, repeating several times. Never bounce to get a greater stretch.

- » **Lower back stretch:** Tight hamstrings contribute to lower back problems. Stretch hamstrings by sitting in a chair with your feet on a footrest. Keeping knees straight, lean forward until feeling an easy, not painful stretch in the back of the thighs. Or bend your knees just enough to allow you to touch your toes.
- » **Shoulder stretch:** Scratch your back. Point your elbow in the air and scratch the top of your spine. Next, curl each arm around the front of your neck and scratch over your opposite shoulder. Finally, hold your elbow down by your side and twist your hand back up between your shoulder blades.
- » **Neck stretch:** Lower your ear to your shoulder on either side.
- **Strengthening:** Muscle strength is important for lifting and for stabilizing the lower back region. To strengthen without using weights, tighten muscles to their maximum, hold for a count of 10, relax for a count of 10 and do several repetitions a day. Be sure you're stable, either by taking a wide stance or holding onto a secure object.
 - » **Legs and hips:** Do partial squats while holding

a stable object like a countertop. Keeping your back straight, slowly lower as if sitting down. Go down as far as comfortable then back up. For greater effect, use only one leg at a time. When rising from a chair, use only one leg. Hold onto an object for stability but don't pull yourself up.

- » **Lower back:** Squeeze buttocks and/or abdominal muscles as tight as possible and hold for a count of 10. To strengthen core muscles, pull your navel, waist, and buttocks toward the middle of your body, as if trying to pull your body away from your clothes. Hold, release, repeat.
- » **Shoulders and arms:** (a) Take a weight such as a shoe or soup can and lift it straight up in the front and to the side, from the waist to over the head. Hold arms straight out to the sides at shoulder height, then bring one arm across in front of the chest, keeping the elbow straight. (b) Force shoulder blades together, then relax, repeating several times. Shrug shoulders up toward the ears then relax.
- » **Neck:** Place your palm in the middle of your forehead and gently press, resisting the push with your neck muscles. Repeat on each side and the back of your head.

Massage

Regular massage by a professional or a caregiver is physically and emotionally therapeutic for people with neuromuscular diseases and can help them relax or even sleep. Gentle massage is preferred to deep muscle massage. Caregivers also will find a massage for themselves is a great way to ease stress.

Pain

Although muscular dystrophy typically doesn't directly cause pain, it often leads to some painful secondary conditions. It's estimated that about two thirds of people with neuromuscular diseases experience chronic discomfort or pain, primarily due to muscle cramps, pressure sores, stiff joints, overstretched muscles, and spasticity (jerky movements caused by rigid muscles).

There are several avenues to pain relief:

- **Rest and sleep.** Lack of sleep can make anyone especially sensitive to discomfort. A good night's sleep or a few days' rest from overexertion sometimes can relieve aching and cramps. See "Sleep" on page 39 for ideas on more comfortable sleep.
- **Check equipment.** As the person becomes less mobile, pain can arise from ill-fitting equipment, such as incorrect wheelchair seating. Any prolonged position can contribute to pressure sores. Ensure your loved one always is positioned correctly in the bed, chair or wheelchair. Experiment with cushions, mattresses and pillows until the right support is found. If it's impossible to get comfortable, investigate a change in bed or wheelchair.
- **Relieve coughing.** Prolonged coughing from the flu or from weakened respiratory muscles can become exhausting and make the chest muscles sore and achy.
- **Movement and touch.** Stretching, exercise and light hand pressure can help reduce pain. Every day, encourage, assist, or perform range-of-motion exercises — these are critical for preventing or easing pain. Range-of-motion, if not too strenuous, can feel like a gentle massage. Applying heat, such as microwaveable moist heat pads, directly to the area of discomfort can provide relief. Warm baths or showers may work.
- **Don't forget massage.** Gently massage the painful area until it relaxes, or have the person sit facing the back of the chair or lie down for a relaxing back rub.
- **Medicate.** Sometimes it's necessary to consult a doctor about medications for pain, spasticity, and cramps. If pain is persistent and your primary care provider and MDA care center team has not been able to provide relief, you may be referred for palliative care or a pain clinic for advanced or additional types of pain management.

Positioning

Proper body alignment and support can forestall a variety of problems, like pressure sores and

joint pain. In general, the head, shoulders, hips, and feet should be aligned and not too flexed or overextended.

Caregivers have several strategies to achieve proper positioning:

- Lots of pillows, in different sizes and firmness, stabilize a person in comfortable positions. When side-lying, put a pillow between the person's knees to keep hips in alignment. For back-lying, put a pillow under the knees and elevate the arms slightly with pillows. Use more pillows to "float" heels above the bed and keep feet from flopping to the sides or downward. Large body pillows, rolled towels, and air cushions work for some.
- There's no such thing as a one-size-fits-all wheelchair. Consult an OT or PT to customize a chair to your loved one's unique body proportions. Important measurements include depth of the seat, position and height of the headrest, distance from seat to footrest, height of armrests, and backrest trunk support. Finding and fitting the proper wheelchair cushion is a crucial element.
- A wheelchair wraparound lap desk helps support the trunk, arms, and hands.
- In bed, a drawsheet is a key positioning aid. The drawsheet is placed on top of the bottom sheet, extending from the person's shoulder to buttocks with at least 6 inches of sheet remaining on each side. Grasping and pulling on the sheet (alone or with a helper) allows even a large person to be rolled to the side or hoisted higher up on the mattress. Ask an OT or PT for a demonstration.
- Silky sheets and pajamas can make positioning easier.
- No one should have to sit in the same position all day or lie in one spot all night. When your loved one no longer can change positions, an important part of caregiving is helping shift their limbs or entire body.
- If the person keeps sliding out of the wheelchair, try a seat belt or rubbery shelf liner on the seat and foot supports.

Range-of-motion (ROM) exercises

Stretching and moving muscles and joints is essential to maximizing movement and minimizing pain. Blood clots, pressure sores, discomfort, sleeplessness, and contractures are some of the potential consequences of immobility. Caregivers can learn ROM from a physical therapist. If a loved one can perform exercises alone, encourage daily practice. When assistance is needed, caregivers should assist only to the point at which the person can do it alone. Once voluntary movement is gone, passive ROM should be performed by a caregiver every day.

Social Concerns

Socializing

Isolation is a risk for those with neuromuscular diseases. Symptoms such as drooling, immobility, and difficulty in communicating may make people reluctant to see friends or go out. But it's important to keep up social activity and be part of the world and the community, and it's possible to find ways to adapt in almost any circumstance. Socializing helps fight off depression and enables the individual to make a contribution. It also can help relieve caregiver burnout when others can keep your loved one company.

Sometimes friends are reluctant to stay in touch if the disease makes them uncomfortable. If friends or family members seem uncertain how to relate to the person with a neuromuscular disease, remind them he or she is still the same person, and encourage them to talk about things other than the disease. Your loved one will want to talk about the same interests as their peers — sports, politics, movies, etc. Friends can come over to watch a ball game or concert on TV as a way of simply being together.

Friends also can be caregivers at times so caregivers can take a break or get other things done. Make it clear that they aren't being asked to provide personal or medical care, but rather simply to be there to talk or call for help if there's an emergency.



Going to public events is feasible and enjoyable. Most public places are accessible to wheelchair users, thanks to the Americans with Disabilities Act. With an adapted vehicle or public transportation, people with neuromuscular diseases can continue to work, go to movies, ball games, kids' activities, church, family events, and restaurants.

Driving

Caregivers sometimes worry that their loved ones should no longer be driving. Although it's possible to install hand-operated driving controls to compensate for leg and arm weakness, there is no guarantee that adaptations to the vehicle will be safe for use if muscle weakness and vision worsens. Set up a driving evaluation with an occupational therapist or the Association for Driver Rehabilitation Specialists to assess needs and abilities.

Caregivers can help ease the transition to nondriver status by rounding up a supply of readily available drivers, investigating taxi cabs and accessible public transportation, and scheduling trips that accomplish several things at once. If you're concerned that your loved one is unsafe on the road but won't give up the car keys, talk to his or her doctor.

Independence

Independence fuels a “fighting spirit.” Help your loved one maintain the highest possible degree of independence, changing strategies as the disease progresses. Figuring out ways around the limitations of neuromuscular disease is a creative game in which caregiver and loved one are on the same team and every victory for independence is a blow against the disease. (Note: The use of assistive equipment like wheelchairs is a move toward independence, not away from it.)

- **Be patient.** Independence for a person with a neuromuscular disease can be slow and painstaking. Sometimes it seems faster and easier for caregivers to do it themselves. Breathe, relax and be grateful for whatever degree of ability the person has.
- **Be collaborative.** Remember that people affected by neuromuscular disease are capable individuals even if they can’t communicate clearly. Through whatever means possible, discuss choices, make joint decisions, and defer to the loved one’s wishes in decisions regarding their medical care.
- **Ask if they want help before helping.** Don’t take over tasks that still can be performed if the person is given adaptive devices and time.
- **Let your loved one use your hands.** When a person with a neuromuscular disease needs help with something, it can be very frustrating to have a helper take over the task and do it their way. The result is seldom what the person envisioned. Your loved one doesn’t need a caregiver’s brain to plan things, just a pair of willing hands. Suggestions are fine, but ignoring the person’s ideas is demeaning.
- **Help set up your loved one with a computer and internet access.** Help locate and install adaptive technologies that enable computer use when movement is limited or absent. Computers provide entertainment and social interaction and can enable adults with neuromuscular disease to help with household chores such as paying bills, tracking down information, hiring services and grocery shopping.
- **Use adaptive devices and strategies.** Consult a physical or occupational therapist for

suggestions. Adaptations include rearranging household objects or furniture and changing the way a task is done (i.e., sliding something rather than carrying it).

- **Don’t sacrifice safety for independence.** It can be dangerous to leave a loved one unattended when they require assistance with mobility and communication. Families have found several ways to monitor loved ones, such as cell phones, baby monitors, emergency response buttons, friendly neighbors, etc.

Socializing For Children

Children spend hours at school socializing with their peers. When mobility becomes more challenging, sleepovers with friends and attending physical activities can present potential barriers to budding friendships. Parents can often troubleshoot these barriers, or have the youth work out ways to invite friends over for activities at their own home or a public place, like attending a movie or sporting event. If an accessible vehicle is needed for the activity, offer to drive or lend the accessible vehicle to reliable drivers in the friend group. When young people graduate from high school, there can be a shrinking of natural friend circles. This is true with or without a disability. It is important to spend time and effort to maintain or foster new friendships to avoid isolation. This may include joining book clubs, a fantasy football league, a gaming community, or other groups who meet regularly.

Travel

People with all kinds of disabilities travel the world. To get around in town, investigate local public transportation and services for those with disabilities, or look into buying/renting a van that can accommodate a wheelchair.

Airlines, hotels, and tourist sites have accommodations. Definitions of “handicapped

accessible” vary widely, however, so it’s best to call in advance and find out exactly what’s available. Ask about the ability to use a portable Hoyer lift in the room. Many hotels install boxes under the bed to prevent items from rolling under the bed. These boxes block Hoyer bases.

Cruises are a particularly comfortable way for people with disabilities to travel.

Airlines allow transportation of respiratory equipment, wheelchairs, and other equipment, but usually as luggage. Damage to power chairs is not uncommon.

If you’d rather leave your equipment at home, you may be able to get a loaner wheelchair or other equipment in the city you’re visiting. Contact the MDA Resource Center at **833-ASK-MDA1 (833-275-6321)** or by email at **ResourceCenter@mdausa.org** to ask about loan closet resources at your destination.

Many families assume that they need to get an accessible vehicle in order to maintain



Set up a driving evaluation with an occupational therapist or the Association for Driver Rehabilitation Specialists to assess needs and abilities.

independence. While the use of a modified vehicle may be helpful for some, there are other options available to ensure your loved one’s needs are met without needing to take on the financial cost. The average accessible vehicle can cost well over \$70,000. Should you family be interested in an accessible vehicle, talk to your MDA Care Center’s social worker

for additional resources and information on low-interest loans, grants or applications for advocacy organization funding.

Sleep

Sleeping well

Helping the person with a neuromuscular disease

sleep well also helps the caregiver sleep well. Taking extra time to ensure comfort when a loved one goes to bed can cut down on repositioning later in the night. Some caregivers create a checklist of bedtime adjustments to make sure nothing is forgotten.

Although comfort is an individual thing, common elements are pillows for stability and to prevent pressure sores, a blanket lifter to keep weight off the feet, and blankets that don’t restrict weakened movements by being too heavy or tight.

A comfortable mattress is essential. Some people prefer “memory foam” mattresses or mattress toppers; others use air mattress toppers or invest in automatic turning or alternating pressure mattresses. Automatic hospital beds allow people who can operate a remote to reposition themselves; the height-adjustable feature protects the caregiver’s back and makes caregiving a little easier. When sleep is difficult, be sure to contact your physician to seek assistance. To deal with nighttime saliva, try elevating the head off the bed.

Sleep deprivation (for caregivers)

Caregivers may get up numerous times a night to reposition or help their loved ones, leading to chronic sleep deprivation. Often the problem isn’t getting up, but the inability to fall back to sleep afterwards.

Researchers say chronic sleep deprivation can cause depression, fatigue, forgetfulness, lowered alertness, reduced creativity, inability to speak and write clearly, lowered resistance to disease, weight gain and increased risk of stroke, heart attack and adult-onset diabetes. Sleep-deprived people are more prone to falling asleep while driving. It’s a problem that needs to be solved quickly.

Strategies to get more rest while still providing nighttime care include:

- **Make it quick and quiet.** When getting up, don’t turn on the lights (use a low-level nightlight if necessary), don’t have a conversation or do anything mentally stimulating, and stay up the minimum amount of time necessary.
- **Don’t try too hard.** If you can’t fall back to sleep

within 10 or 15 minutes, get up and do something relaxing, then return to bed as you feel yourself getting drowsy. Performing a good all-over body stretch can add in relaxation.

- **Decrease caffeine, alcohol, and nicotine.** Especially avoid caffeine in the afternoon, as it can contribute to sleeplessness at night. Although some people find that an alcoholic drink before bed helps them fall asleep, alcohol increases the likelihood of waking later in the night.
- **Power nap.** Aim for a short (15- to 30-minute) nap sometime during the “midday trough” between 1 pm and 3 pm, when your body naturally wants to rest. A longer nap may leave you groggy and unable to sleep at night. If you can’t fall asleep, just rest quietly with eyes closed for a brief period.
- **Go to bed.** A simple way to get more sleep is to go to bed a little earlier. It sounds obvious, but tired people often stay up to watch the late show rather than turn in earlier. Record favorite late-night shows and try to go to bed and get up about the same time every day.
- **Check out your sleep space.** An uncomfortable mattress, snoring spouse, too much light, being too hot or too cold, pets who jump on and off the bed, and outside noises all are subtle distractions that make it hard to settle back down. Try eyeshades, putting pets elsewhere or buying a more comfortable mattress.
- **Three on, three off.** If possible, share nighttime caregiving duties. For optimum benefits, sleep experts recommend a three-nights-on, three nights-off schedule, rather than switching with someone every other night.
- **Talk to your doctor.** Not all caregiver sleep problems are caused by stress or getting up in the night. Schedule an appointment if nothing else is working.

Nightmares

Bizarre dreams or nightmares, coupled with morning headaches and mental fuzziness, may indicate respiratory problems in neuromuscular disease or be a side effect of some medications. We encourage you to speak with your pulmonologist or other

medical providers for further testing that may be available.

Medical Care and Emergency Preparedness

Emergency medical information

Collect important medical information in one spot to facilitate doctor and emergency room visits. Keep this information current, and post a copy on the refrigerator, where ambulance crews have been trained to look in an emergency. You can also provide copies of this information to your doctor’s office proactively.

Information should include:

- The person’s name, birthdate, diagnosis, and special information (i.e., can’t speak, can’t move legs, uses letter board, uses BiPAP, etc.)
- Emergency contacts and phone numbers (note if someone holds durable or healthcare proxy and/or power of attorney)
- Insurance information, including identification numbers, addresses and phone numbers
 - » Names of doctors and phone numbers
 - » Daily medications: times and dosages
 - » Adaptive and supportive equipment
 - » Allergies
 - » Special diets
 - » Advance directives and living will
 - » An updated history of surgeries and major medical events
 - » Prominently note any special orders such as “Do Not Resuscitate” or “No Tracheostomy.”

Emergency preparation

Hurricanes, earthquakes, fires, and power outages — muscle disease brings special challenges to any kind of emergency. Some planning will help the family be prepared. Before an emergency occurs, contact your state and local government’s office of emergency management or local fire department to find out what emergency relief assistance is offered. They’ll tell you whom to contact in event of a disaster, where to go and what to have ready. Some of these offices will

have special arrangements for people with disabilities. You may need to register in advance for these services.

Emergency checklist for people with disabilities:

1. Prepare a medical information list.
2. Send copies of important documents to an out-of-town contact person in case the originals are lost or destroyed in a disaster.
3. Know which shelters are best prepared to accept people with disabilities and special medical needs.
4. Have a pet care plan. Shelters don't always accept pets or service animals. Contact your local animal shelter.
5. Make an equipment plan.
6. Have an extra battery for a motorized wheelchair or scooter. Know how to recharge the battery.
7. Have a patch kit, can of seal-in-air product, and inner tubes for a wheelchair with inflatable tires.
8. Have a lightweight manual wheelchair for backup.
9. Have a converter for your communication device.
10. Pack a low-tech communication board and preprinted key phrases in your disaster kit
11. Having a small generator at home to use while power is out can be life-saving for individuals

dependent on ventilators.

12. Order an adapter kit for your ventilator so it can be plugged into your car or a marine battery.
13. Contact your electricity provider to see if they offer a "priority reconnection service" that will help ensure your power is restored as soon as possible. Even with this service, power could still be out for a long time following a disaster.
14. Have a list of the style and serial numbers of medical devices.
15. Create a disaster supply kit and escape plan.

A comprehensive checklist for people with disabilities is available from the American Red Cross. In case you must get out of the house quickly, determine the best escape routes and practice them with the family. Remember that you'll need alternatives to some traditional plans. For example, emergency experts advise heading for the basement in a tornado, but these aren't usually wheelchair accessible. Will you be able to carry the individual and/or their equipment downstairs or should you go somewhere else? It's also a good idea to discuss a finalized evacuation plan with other family members who may not live with you, as well as with neighbors, friends, and home care aides in case anyone other than the primary caregiver needs to assist.

Medical care

It's wise to have a primary care physician for care that doesn't involve the individual's neuromuscular disease, and to have this doctor consult with your



Emergency room alert cards

MDA offers emergency room alert cards for DMD, ALS, MG and general Neuromuscular Disease available on MDA.org. Print and complete the appropriate card to keep in your wallet in case of an emergency so medical staff are aware of any conditions and concerns you may have.

MDA Care Center physician when necessary. Regular medical checkups, flu shots and pneumonia vaccinations are essential to preventing respiratory complications. Persons with neuromuscular diseases are still at risk to have diabetes, cancer, hypertension, etc. A primary care provider should monitor for typical age-related medical conditions. Having a NM disease or any other health condition may impact risks associated with anesthesia or surgery. For example, a respiratory system weakened by NMD may make surgery more difficult.

Keep your MDA Care Center doctor and other doctors informed about all your loved one's medications and conditions. Knowing as much as possible about the neuromuscular disease will help a caregiver determine whether a symptom is related to the disease or has some other cause. When in doubt, call someone at your MDA Care Center.

Medication

To date, a handful of medications have been approved by the US Food and Drug Administration to treat neuromuscular diseases, and others that may be used "offlabel." We encourage you to work with your MDA Care Center physician and the drug companies to learn about the treatments that may be available to you and the patient assistance programs that may be available through the drug company.

Temperature

Cold extremities — feet and hands — affect some



people with neuromuscular disease. If these problems persist, practical solutions such as extra socks and gloves, room heaters, massage, range-of-motion, hot water bottles, or hot packs can help.



Finalized evacuation plan

Discuss a finalized evacuation plan with other family members who may not live with you, as well as with neighbors, friends, and home care aides in case anyone other than the primary caregiver needs to assist.

Keep emergency phone numbers in your wallet and near telephones where they're available to everyone involved in caregiving. Caregivers who work outside the home should check with supervisors about any emergency plans in effect at the workplace. For example, some places won't let employees leave for home until an "all clear" has been given by local authorities. Find out whether your home health agency has special provisions during an emergency. Will they continue to provide care and services at another location if your loved one needs to be evaluated?

Respiratory Care in Neuromuscular Disease

Respiratory health is a vital issue for children and adults who have neuromuscular diseases. These conditions progressively weaken muscles, including those you use in breathing and coughing.

Fortunately, MDA Care Centers include pulmonary experts who are experienced in monitoring and making recommendations highly specific to neuromuscular weakness. The breathing risks associated with NM disorders tends to be very different than other common respiratory conditions. The breathing problems are not a result of abnormal lung tissue or function, but are related to the muscles of breathing becoming weaker over time. This guide will help you to also learn about respiratory health and strategies to reduce complications commonly associated with neuromuscular weakness.

Breathing is accomplished with the effort of respiratory muscles. A dome-shaped muscle called the diaphragm, located below the lungs, and muscles between the ribs called the intercostals, pull air into the lungs. This inspiration, or inhalation, is followed by a relaxation of these muscles as air leaves the lungs. The process of breathing in and out, taking in oxygen and exhaling carbon dioxide, is called air (or gas) exchange, or ventilation. The pathway that air follows from the nose, through the trachea and into the lungs is the airway.

When you forcefully inhale or exhale, you use extra, or accessory, muscles of breathing. Accessory respiratory muscles include those in the abdomen, chest wall and neck. Over time, neuromuscular disorders can weaken all muscles involved in breathing. When these muscles don't move, air can't move in and out of the lungs effectively. Respiratory failure, often in association with an infection (pneumonia), can cause frequent hospitalizations and can be a cause of death for people with neuromuscular disorders. Sometimes a decline in respiratory function can be so gradual that it isn't noticed at all or is attributed to some other cause,



such as general fatigue, depression, or sleep problems.

In this portion of the guide, you'll learn about monitoring respiratory functions, and risks that may be associated with a neuromuscular disease.

Risks of hypoventilation

The causes of hypoventilation are reduced muscle strength for ventilation, reduced lung volume over time, and sometimes restrictions to take in air due to spine or ribcage deformities, like scoliosis.

When the lungs cannot fully expand, the lungs may not have access to enough air containing oxygen and may not be able to expel or exhale carbon dioxide. This results in abnormal "blood gases". Blood gases will be further described under respiratory tests.

For people who have chronic hypoventilation, they may not develop or recognize subtle changes in blood gas values and can go undetected for months or years if pulmonary testing is not completed. For others, or for someone who has a sudden illness on top of the chronic/long term hypoventilation, they may be rapidly symptomatic. (See signs of respiratory inefficiency list)

Hypoventilation can cause atelectasis, or collapse of small areas within the lung where gases exchange. The collapsed areas offer a moist environment where a virus may have an opportunity to grow and cause infection, commonly referred to as pneumonia. Pneumonia can also be caused by inhaling or aspirating foreign matter into the lungs, most notably food particles from uncoordinated swallowing.

Ultimately, the greatest risk of hypoventilation is respiratory insufficiency that leads to respiratory failure. It is most common for individuals with NM weakness to initially benefit from maximizing the expansion of lungs during the day with a cough assist device, and treatment of hypoventilation during sleep. We all tend to breathe more shallowly during our sleep than when we're awake, moving about and talking. If daytime breathing is low, then nighttime breathing is lower yet. Without treatment, blood gases can become very abnormal and lead to loss of consciousness or even death.

Many specialists recommend getting a breathing test shortly after diagnosis. This gives baseline measures against which later tests of breathing can be compared. As respiration needs more attention, the physician will recommend regular visits to a pulmonologist and respiratory therapist (RT) for interventions to support breathing.

Pulmonary function tests usually involve breathing into a computerized machine through a mouthpiece. One of the most important functions being measured is vital capacity (VC), or how much air a person can expel after taking a deep breath.

It is important to see a respiratory specialist who works with neuromuscular disorders. Pulmonologists and respiratory therapists usually deal with lung disorders such as asthma or chronic obstructive pulmonary disease (COPD); and may be unfamiliar with your loved one's diagnosis. You need the respiratory team to understand that the breathing problems in muscular dystrophy and related diseases aren't a result of lung disorders, but of muscle weakness. Treatment for problems with the lungs won't correctly address these needs and can

be detrimental or delay the correct care.

NOTE: Supplemental oxygen, via an oxygen tank, is not a solution to breathing problems in neuromuscular disease and may even be harmful. Supplemental oxygen is for people with lung diseases.

Coughing

The cough reflex is an explosive expulsion of a volume of air from the lungs. The high pressure and speed of this air propels irritants, such as mucus, up and out of the lungs. Neuromuscular disorders can weaken the ability to cough, but coughing is needed to remove mucus, especially during an acute respiratory infection. Weakened abdominal and throat muscles in some forms of neuromuscular disease diminish the ability to cough. When a person can't cough up mucus and inhaled particles, these can fall back into the lungs, where they can cause irritation and infection. Weakened swallowing muscles also make aspirating (inhaling) food and liquids into the lungs more likely, which can irritate the lungs and bronchial tree. This can lead to pneumonia or choking.

If your loved one's ability to cough isn't sufficient to clear secretions from the throat, lungs and trachea, you can learn manual or mechanical assisted coughing techniques. When coughing is weakened, it's important for caregivers to provide daily help in keeping lungs clear.

Assistive coughing (quad cough)

The simplest method is to place your hands or arms over the person's stomach, just below the ribs and under the diaphragm, while he or she is lying down or sitting. After they take one or two deep breaths, firmly push in and upward toward the ribs as they cough. This will force air from the lungs more quickly and help push any secretions out through the mouth. Have a tissue or portable suction device available to collect any mucus coughed up.

This is best done on an empty stomach. If the individual gets nauseous or throws up during the process, try gentler pushing. An increase or other

change in mucus may indicate a respiratory infection needing medical attention.

NOTE: Let a respiratory therapist demonstrate these techniques so you'll be sure to perform them correctly. If the individual has any discomfort or bad reaction, consult the therapist or physician.

For mechanical assistance an insufflator exsufflator is a very handy and effective device. A mask is put over the mouth, and the machine blows air into the lungs, then reverses the flow, simulating a cough. After either a manual or mechanical assisted cough, you may need to use a portable suction device to remove secretions from the mouth through a small tube. You can rent or buy a suction device (similar to those dentists use) from a medical equipment vendor.

Doctors also may suggest these cough assist methods:

- Expectorants — prescription medications that thin secretions, making it easier to cough and clear the secretions
- Breath stacking — closing the throat after each inhaled breath taken (without exhaling) through a mouthpiece to increase the amount of air in the lungs and then coughing
- Oscillation vests or airway clearance systems to “shake up” mucus in the chest

Signs of respiratory insufficiency

Watch for changes in breathing, which may develop very gradually. Apparent fatigue, depression or sleep problems often are associated with developing respiratory weakness.

Symptoms of respiratory muscle weakness and chronic underventilation include:

- General fatigue, drowsiness, lethargy
- Sleep disturbances such as nightmares, night terrors, sleep apnea (interrupted breathing during sleep), or sudden awakening
- Morning headaches
- Daytime sleepiness
- Confusion, disorientation, anxiety
- Poor appetite, weight loss

- Excessive yawning or hiccups
- More labored breathing, especially when lying down
- Rapid, shallow respirations with increased heart rate
- Weakened or softened voice, speaking in short phrases, inability to sing or shout
- Difficulty coughing and clearing the throat (weakened abdominal and throat muscles also contribute to this)

Any of these symptoms, seen regularly, should be reported to a physician.

NOTE: People with neuromuscular disease may not show the kind of heavy labored breathing associated with overexertion.

Simple ways to minimize respiratory problems:

- Most important is to avoid exposing your loved one to any type of cold, flu, or virus. (A weaker respiratory system can't effectively fight off infections.) This can push weakening lungs to pneumonia and respiratory crisis. If the person's respiratory system has shown any weakness, avoid close contact with anyone who is sick or who has been exposed to someone with a contagious illness. It's also important that caregivers protect themselves from cold and flu — and no hugs and kisses with your loved one if you have a cold.
- Raise the head of the bed while the person is lying down, either awake or asleep. You may want to purchase or rent an adjustable hospital bed.
- Place more fans in the house to keep air circulating and fresh.
- Have the individual and primary caregivers get a flu shot and a pneumonia vaccination.
- Increase the intake of fluids, as long as the person doesn't have trouble swallowing them.
- Help your loved one perform respiratory exercises, only if prescribed by a doctor or respiratory therapist.
- Include respiratory equipment or supplies as part of your preparation for emergencies.
- Review with the pulmonary care team a list of medications being taken. Some drugs can have

an adverse effect on respiratory function and can be switched to others with fewer respiratory side effects.

- Consider inserting a feeding tube to reduce the danger of aspirating food into the lungs.
- Good hygiene is important. Wash hands thoroughly and regularly to prevent the spread of germs.

Scoliosis and respiratory health

Scoliosis — a sideways curvature of the spine — is a common complication in neuromuscular disorders. Scoliosis prevents full expansion of the chest and can interfere with breathing.

Scoliosis in these diseases occurs because of weakening of the muscles that normally support the spine. At MDA Care Center visits, especially in growing children, the spine will be physically examined for curvature, and the degree of the curve may be measured by X-ray. A curvature may progress surprisingly quickly in a child. Keep a close eye on a progressing spinal curvature, which may crowd the lungs, and consider spine-straightening surgery on your doctor's recommendation.

Sleep apnea and muscle disease

Some studies have shown a high incidence of sleep disorders in people with neuromuscular diseases. But the cause of these problems may be different than for those without muscle diseases, and the treatment is different as well.

Among the general population, common causes of respiratory problems during sleep include pauses in breathing (apnea) either caused by brain abnormalities (called central apnea), or by collapse of the upper airway, blocking breathing (called obstructive apnea). But for those with neuromuscular diseases, breathing problems during sleep may be caused or complicated by the fact that the muscles that aid respiration — the diaphragm and intercostals — have been weakened by muscle disease. Weak respiratory muscles can lead to nocturnal hypoventilation (ineffective breathing during sleep) or nocturnal apnea (periodic cessation of breathing during sleep), when gravity, body position, and

neurological factors naturally make breathing efforts less effective.

A simple method to assess nighttime breathing difficulties is to measure exhaled carbon dioxide in combination with pulse oximetry, which painlessly measures blood oxygen levels through the placement of a small clip on a finger or toe. Typically, these tests may be done at home while the patient sleeps. Polysomnograms are performed in a sleep laboratory and last between seven and 12 hours.

For the general population, continuous positive airway pressure, or CPAP, is the ventilation therapy commonly prescribed for obstructive apneas. CPAP blows in a continuous flow of air at a set pressure, keeping the airway from collapsing and obstructing breathing. But CPAP often isn't appropriate for people whose problem is caused by weak respiratory muscles, because the muscles have to work harder to exhale against the constant inward flow of air.

In those cases, bilevel positive airway pressure ventilation, typically called BiPAP, is more commonly used. (BiPAP is a registered trademark of Respicare.) Air is administered at a higher-pressure level on inhalation and a lower (or zero) pressure on exhalation. In muscle diseases, the difference between the inhalation and exhalation airflow pressures is typically high to provide greater assistance to the inspiratory muscles and little or no resistance during exhalation.

Because sleep-breathing problems can be a symptom in some neuromuscular diseases, it's important to seek help at the first symptoms of underventilation. Be sure to consult an experienced medical professional or certified sleep specialist who knows which therapeutic solutions are appropriate for people with neuromuscular diseases.

Emergency respiratory distress

It's wise to plan ahead, talking with the doctor and each other about dealing with breathing problems as they arise.

A breathing crisis can arise very quickly. Call 911

immediately if your loved one is showing severe signs of underventilation such as struggling for breath or inability to breathe.

A bag valve mask (BVM or Ambu bag) is a normal part of an emergency crew's resuscitation kit or a hospital crash cart to help someone who's not breathing. Ambu bags serve essentially the same purpose as mouth-to-mouth resuscitation and are available online or through medical supply stores.

Emergency medical personnel probably will want to administer oxygen in response to signs of respiratory distress. This is fine only if combined with breathing support. A written explanation from your physician should help explain the situation to emergency personnel. Your loved one's healthcare team should be notified of the emergency, and the specialist's contact information offered to the emergency team.

Planning ahead for assisted ventilation will help you avoid having to make decisions in an atmosphere of panic.

Other than complete respiratory failure, other signs, such as continued shortness of breath, fever, malaise, or low pulmonary function test readings, require immediate medical attention. These could mean an acute infection.

Ventilatory Support Options

When Vital Capacity (VC) reaches a certain low level, usually near 50%, the respiratory team will introduce the idea of assisted ventilation. There are two primary types: noninvasive, in which air is delivered through a mask and no surgery is involved; and invasive, in which a tube is surgically inserted in the trachea and attached to a ventilator. Assisted ventilation can help your loved one feel less fatigued and safer, by improving sleep and decreasing the potential for respiratory crises. It can rest the breathing muscles and increase energy, allowing for more activities and enjoyment of life.

Discussing options together

Deciding which, if any, assisted ventilation to use is one of the most important and most personal choices you and your loved one will have to make. Discuss this well in advance of time to act on the decision, and review all the factors involved. An emergency room is a poor environment for making such a critical decision — and a respiratory crisis may decrease your options.

It's difficult to face these decisions when you're trying to focus on ways to maintain life as you know it and to hold onto hope for a long survival or research progress. An absolute decision doesn't have to be made in the first few months after diagnosis. But getting the issues out on the table and exploring the options is important. Keep the door open for a while. The options may look different as time passes.

First, do some research through sources like those suggested in this guide or through the MDA National Resource Center. Talk with your healthcare team; speak with other families of people with the same diagnosis. Check your insurance policy, and see what coverage is offered for respiratory equipment, supplies, and day-to-day care.

Don't expect a consensus of opinion from everyone you consult. Even physicians who are knowledgeable in this area have different opinions. No one can tell you what's best for your family.

It isn't unusual for adults in the early stages of muscle disease to declare they don't want to live with mechanical interventions such as a feeding tube, wheelchair, or tracheostomy. But after living with their condition for some time, and adjusting as abilities decline, each of these devices often seems more acceptable. Those with young children or other joys in living, or with certain religious beliefs, may feel that life is worth sticking around for, even if they need machines to help them move, eat, and breathe.

Others may choose to decline invasive ventilation, and opt for pain control as their breathing further weakens and stops.

Obviously, this decision is very much an individual

one. It should have input from your loved one and be made based on complete information and a time of reflection. Ultimately the choice is made by the individual; his or her wishes can be expressed in a medical power of attorney or living will document.

Noninvasive ventilation (NIV)

Most people who need breathing help begin with NIV, and some people are able to continue with this approach for many years. At first, the machine only may be needed during sleep — restoring a good night's sleep for both of you. Later it may need to be available during some daytime hours.

The most common form of noninvasive ventilation is the bilevel positive pressure type, in which air is supplied through a facial mask (interface) or other device on a timed cycle. This involves a small, portable machine that can be placed at bedside. A Bi-level PAP machine delivers air at two pressures — a higher pressure to help the person breathe in more, and a lower pressure so they can breathe out with little resistance.

There are other NIV choices, which the pulmonologist will explain in recommending the best one for the individual situation.

NIV interfaces

The interface is the mask or other device placed over the nose, mouth, or mouth and nose, so a person can use assisted ventilation.

Ideally the individual should try several interface options to get used to the feeling and practice breathing with the device in place. The RT should ask questions about sleep habits, nasal congestion, and feelings of claustrophobia, and determine which interface might be most comfortable and supportive.

With patience, the user should get used to a comfortable interface in a few weeks or months. Begin by using it a few minutes at a time, gradually increasing duration.

A face mask is most common, but if users experience panic, claustrophobia, or general



discomfort at having something over the face, nasal pillows may be preferable. These fit inside the nostrils and put no weight on the face; some users can't even tell they have them on.

Some people use nasal pillows in the daytime and a face mask at night. Nasal pillows also make it easier to turn the head and wear eyeglasses.

Straps around the head hold the mask in place. If air escapes through the mouth at night, chin straps can keep the mouth closed.

Masks are designed to seal around the nose and/or mouth. When they leak, they can blow air into the face and eyes, and decrease the effectiveness of ventilation. Work with your respiratory therapist to find the best solution for you. Gel- and air-cushioned masks are becoming more common, are lighter on the face and seal better. Those with bulbar symptoms may have the most trouble keeping the mask sealed. Some find the best solution is a custom fitted mask or nasal plug made through the hospital's respiratory therapy department.

Some masks irritate the skin on the bridge of the nose or around the mouth. One solution is tucking moleskin, cotton or soft felt just under the nose part, being careful not to break the seal. Carefully clean

and dress any sores or wounds. Apply an antibiotic ointment such as Neosporin while the respirator isn't in use.

All parts of an NIV device require regular cleaning and sterilization, including the tubing, filters, and mask. Filters need regular replacement, and an enzyme cleaner may be required for the mask.

NIV settings

For NIV to be as comfortable and effective as possible, settings for air volume and pressure have to be correct and supportive. If they aren't, breathing distress will continue. Users have complained that their cheeks are puffed out or air bloats the stomach. These discomforts result from an air pressure or volume that's too high.

Usually, your respiratory team will set the pressure levels based on a sleep study, or begin at low levels and adjust them with frequent monitoring. They can be increased gradually to the level your pulmonologist considers most supportive and your loved one can tolerate.



Setting up the equipment, interfaces, and follow-up requires experience and careful adjustment to make it work properly for each person, although some machines come with software or controls and information that helps adjust them correctly. The respiratory team should be available to make adjustments as needed, or to perform overnight oximetry at your home. Try to find a support person with knowledge of muscular dystrophy or neuromuscular diseases.

Oximetry is a measure of oxygen saturation that tells whether the respiratory device is providing enough support. A pulse oximeter should be used regularly to check oxygen saturation while using NIV. A respiratory therapist can check the pulse oximetry on visits, or you can buy one for under \$50.

NOTE: Respiratory function is too important for guesswork. Contact your respiratory therapist who can make changes as needed.

Several studies have suggested that NIV can slow the decline in respiratory function and make a significant difference in survival time in people with more progressive forms of neuromuscular disease. This applies to those with moderate or no weakness of the mouth and throat muscles. Those with more severe bulbar (mouth and throat) muscle involvement may be less likely to tolerate NIV and may have to consider invasive ventilation sooner in the disease course. Individuals and physicians hold strong opinions about the long-term use of NIV — for and against. Whether your loved one decides to use NIV or invasive ventilation, close monitoring by respiratory experts is vital.

Invasive ventilation

There are several reasons an individual and their loved ones may choose invasive ventilation. Some of the reasons are outlined below:

- It can take months to find the right mask or device and get used to NIV.
- Facial features such as a crooked nose or a deviated septum can make finding a mask that doesn't leak or breathing entirely through the nose difficult.

- NIV may aggravate sinus problems or cause severe abdominal distention.
- Some find anything on the face claustrophobic.
- Facial, mouth and throat weakness can reduce necessary jaw closure and ability to use a mouthpiece with NIV.
- Bulbar muscles are weakened so that the person can't speak or swallow or keep saliva out of the airway.
- The hours the person needs to use NIV increase from overnight to most of the day. Going out becomes more difficult if the person resists wearing the mask in public.
- Because pressure-based NIV can only assist breathing, as respiratory capacity deteriorates, the user slides back into the fatigue, poor appetite, and anxiety of pre- NIV days.
- Pneumonia or a simple chest cold result in a respiratory crisis. As lungs become congested, NIV reaches its pressure limit more quickly, and less air is delivered.
- The individual has problems swallowing and often aspirates food or saliva into the lungs, creating more respiratory distress.
- Invasive ventilation can allow the longest possible survival with symptoms.



- In almost all cases, speaking valves (PassyMuir valves are one brand) now in use with trachs can redirect air flow while you're talking and allow your voice to be heard.
- While more complicated, eating may also be possible with a trach. Be sure to consult your respiratory therapist with suggestions about processes that work best for your loved one.
- Travel and portability are not necessarily hampered by having a trach. Continue to read more in this section about considerations about traveling with a trach or other invasive ventilation equipment.

The most permanent type of ventilation is the positive pressure ventilator with a surgically created tracheostomy. A ventilator is attached by a breathing hose to the tracheostomy tube, delivering air through the neck into the trachea (windpipe) on a timed cycle.

Tracheostomy surgery is considered minor and often can be done under local anesthesia with sedation. It's usually followed by several days or weeks of rehabilitation during which caregivers can learn how to clean and maintain the tracheostomy tube (trach), change supplies, and perform suctioning. Ask nurses and respiratory therapists all the questions you can think of during this phase.

Common concerns

- It's possible for a person with a trach to shower but extreme care must be taken not to get water into the opening. Use a cloth or plastic wrap to keep water out.

Problem solving

Maintenance of invasive ventilation does require greater effort by the caregiver. Problems that arise may not be easy to fix without help.

- Whenever the trach or vent tubing is moved, it sets off an aggravating but not painful coughing spell. Vent hoses may pull on the tracheal opening, causing a sore throat-type ache. These problems are usually relieved by repositioning the hoses; a caregiver may require help from an RT or medical provider to make this adjustment.
- Discomfort of the skin around the trach indicates irritation or infection and usually is easily treated with ointment.
- The most annoying discomfort is the same as with an NIV mask — air leaks. As with a

mask, adjusting the hose position may help, but persistent problems may require a medical procedure to increase the size of the trach.

- Trachs carry a risk of incision infection, and increased respiratory infections are reported. At the same time, the trach is easier than NIV for clearing lung secretions and controlling respiratory status.
- Some people are prone to granulation tissue, an excessive growth of new tissue stimulated by the trach incision and presence of the trach tube. This tissue is delicate and bleeds easily and may make trach changes difficult. Granulation tissue can be handled with cortisone, silver nitrate and, in some cases, laser removal. Seek medical advice.
- Trach users need a backup ventilator.
- The battery operating the vent has time limits and will need to be charged regularly. An extra battery and alternative power source is necessary.
- If the person needs to go to a residential care home, some won't take vent users.
- Vents often have temperature limitations, so avoid excessive cold or heat.
- When the ventilator is installed, contact the local electrical and/or gas power company and explain that this is a device required for life. Request a form to protect against the power being turned off and have it signed by a physician. A generator or other backup power strategies are advisable.

What invasive ventilation means for caregivers

Those caring for people using invasive ventilation move into a new level of caregiving. A person with a trach must have someone available at all times to suction saliva and mucus plugs and respond to emergencies, such as a detached tube or power failure. However, nurses aren't required. Anyone, even an older child, can be taught how to suction and handle vent alarms.

Caregivers must decide who should be given the responsibility of ventilator care. For those hiring outside help, be aware that many home healthcare agencies only will provide registered nurses for

people with trachs. This introduces a cost associated with that level of care. Speak with your insurance provider, or work with your state's waiver program case worker to identify additional resources for financial assistance for hiring caregivers. You may also find more information in "Finding Caregiver Help" on [page 5](#).

In addition, new supplies are required, including dressings, tubing, filters, and batteries. Consult with your insurance provider to find out which of these are covered under your plan.

Daily care for people with invasive ventilation includes:

- Cleaning around the trach as part of bathing or washing up, using ordinary soap and clean washcloths. If there's a lot of mucus drainage around the tube, recurring infections, or redness, more cleaning or use of ointments may be required.
- Dressings where the tube connects to the body must be changed.
- The inner cannula, a tube within the trach tube, must be cleaned daily, usually with dish soap or hydrogen peroxide. Some healthcare providers suggest changing the inner cannula every day.
- Trach ties, which hold the device and dressings around the neck, should be changed when they're damp or dirty.
- The outer trach tube needs to be changed regularly — some experts advise once a day, some say once a week. People prone to granulation tissue need to have the tube changed more frequently, as do those with respiratory infections. Changing the tube is usually a simple procedure.
- Suctioning is necessary to remove mucus or saliva that the individual can't cough up. Some days people need suctioning two or three times, other days they require it a dozen or more times.

Generally suctioning can be done by a caregiver with a nonsterile disposable glove (not reused) and catheter (changed daily or several times a day). The respiratory care team will provide instructions. The caregiver passes a catheter attached to a suction

device through the trach tube and down into the airway. This must be performed carefully because suctioning itself can push bacteria from the upper airway down into the deeper and normally sterile lung passages, adding to the risk of respiratory infections, including serious pneumonias. Be sure the gloved hand doesn't touch anything but the catheter.

You also can use an insufflator-exsufflator to fit onto the trach tube and pull out secretions from both lungs at once. Use of this airway clearance device may reduce the need for suctioning.

Quality of life

People with neuromuscular diseases who are using either invasive or noninvasive ventilation go to school and work; they go out with friends and travel.

Like most electronic equipment, respiratory equipment has become smaller with time, and they can easily attach to a power wheelchair and accompany an individual almost anywhere. While it's important to have access to care if your equipment malfunctions, they're by no means confined to home. The relief that individuals experience when they're no longer fighting for breath generally seems a fair trade for any self-consciousness about their appearance. Early in the course of a neuromuscular disease,

some people feel sure they'd never want to be on a breathing machine. But by the time your loved one needs respiratory assistance, their feelings about what's necessary for a worthwhile life may have changed. Your loved one may find that being able to write poetry, paint pictures, manage their business, or watch their children grow are well worth putting up with the inconvenience of respiratory supports.

The decision to use noninvasive or invasive respiratory equipment depends on many factors, and medical issues aren't the only ones to be considered. When making the choice, it's worth keeping in mind that people (even professionals) who haven't themselves used ventilatory assistance probably don't know how your loved one will experience it.



PCA Interview Guide

This guide is intended to provide sample interview questions for an individual interviewing a personal care attendant (PCA) of any type (CNA, LPN, RN, etc.). Some of the questions may not be relevant for your needs, so feel free to add or subtract any questions that apply to your current situation. To get started, introduce yourself, explain your basic care regimen and diagnosis, and set expectations. Then, use these questions as a starting point for your interview. Happy interviewing!

Basics:

- What interests you about this position?
- What makes you a good fit for this role?
- Why are you interested in this job? What's your availability?

Background:

- Where did you go to school? What degree(s) do you have?
- What is your absolute dream job? How is this job helping you with that?

Experience:

- What experiences have you had in order to be qualified for this job?
- Have you ever worked with a patient with a neuromuscular disease before?
- How long did you hold your previous job? Why did you leave?
- Do you have any references I may contact?

The Job Itself:

- From what I described for my needs, are you comfortable with those requirements?
- How comfortable are you with doing a one-person transfer? Two-person transfer? Using a Hoyer lift?
- Have you ever had an emergency situation with a client?
 - o If yes, how did you react?
 - o If no, how would you expect you'd react?
- We will work closely alone. Are you comfortable with that set-up?
- How comfortable are you in changing a trach, G-tube, colostomy bag?
- How comfortable are you with driving me alone?
- Are you comfortable with doing light housework?
- Are you comfortable with change?
- Are you willing to be a live-in?
- What does your ideal relationship with a client look like?

Getting to Know the PCA:

- What are your hobbies?
- What is your favorite movie?
- Do you have a fun fact to share?

Next Steps:

- Do you have any questions/comments/concerns?
- How do you feel after this conversation? Would you still like to be considered for this position?

Resources

MDA resources

Caregiving-related articles and blog posts can be found on MDA's website (mda.org) and on MDA's Quest Media platform (MDAQuest.org) using the search feature.

Be sure to contact the MDA Resource Center at **833-ASK-MDA1 (275-6321)** or **ResourceCenter@mdausa.org** for additional support and assistance. MDA's trained resource specialists are available Monday through Friday, 9 am to 5 pm Central time, to answer questions and provide one-on-one support and resources. Questions typically are answered within 24 hours of a request (or on the next business day). Go to mda.org to send a message to the Resource Center (scroll to the bottom of the home page to fill out an easy-to-use online form). You can visit the Resource Center website for more information about resources for Caregivers.

Quest Media Articles

- Working Together: How to establish and maintain good relationships with personal care attendants (February 2022)
MDAQuest.org/working-together
- 10 Tips for Finding Personal Care Attendants (November 2020)
MDAQuest.org/10-tips-for-finding-personal-care-attendants
- Hiring Good PCAs (June 2022)
MDAQuest.org/hiring-good-pcas
- Positive Relationships: How adults with neuromuscular disease can form and nurture healthy romantic relationships (February 2022)
MDAQuest.org/positive-relationships



- How to Help Your Child Make Friends in School (August 2023)
MDAQuest.org/how-to-help-your-child-make-friends-in-school
- Parent Pressure: How parents of children with neuromuscular diseases combat stress and develop coping skills (May 2021)
MDAQuest.org/parent-pressure
- Transition to Adulthood: How parents can help kids prepare for adult life (November 2021)
MDAQuest.org/how-parents-can-help-kids-prepare-for-adult-life
- Smart Steps to Take in a Medical Emergency (August 2023)
MDAQuest.org/smart-steps-to-take-in-a-medical-emergency
- How Palliative Care Can Support Your Health Journey (August 2023)
MDAQuest.org/how-palliative-care-can-support-your-health-journey

- 7 Budget-Friendly Accessible Home Mod Strategies (May 2023)
[MDAQuest.org/7-budget-friendly-accessible-home-mod-strategies](https://mdaquest.org/7-budget-friendly-accessible-home-mod-strategies)

Local community services

Check county and state government listings for:

- Health and Human Services Department
- Public Health Department
- Social Security Administration
- Mental Health Department

Contact the social service departments of hospitals and clinics. Locate adult day care centers and faith-based agencies, Visiting Nurse Association, and hospice programs. For other local supports, check with your [MDA Care Center](#) and the MDA Resource Center at [833-ASK-MDA1 \(275-6321\)](tel:833-ASK-MDA1) or ResourceCenter@mdausa.org.

National and state organizations

Check with the Area Agency on Aging or the state department of social services to learn if your state will provide funds for hiring and paying a family member as a caregiver.

Caregiver Action Network
[800-896-3650](tel:800-896-3650)
caregiveraction.org

Family Caregiver Alliance
Provides a listing of state-funded services for family caregivers
[800-445-8106](tel:800-445-8106)
caregiver.org/connecting-caregivers/services-by-state

Personalized web pages

CaringBridge
[651-452-7940](tel:651-452-7940)
caringbridge.org

CareCircle
carecircle.org
Caring for children

Raising a Child with a Neuromuscular Disorder: A Guide for Parents, Grandparents, Friends and Professionals, by Charlotte Thompson, MD Oxford University Press, 1999

Muscular Dystrophy in Children: A Guide for Families, by Irwin M. Siegle, MD Demos Health, 1999

A Guide to Duchenne Muscular Dystrophy: Information for Teachers and Parents, Jessica Kingsley Publishers, 2017

Caring for elders

The Eldercare 911 Question and Answer Book, by Susan Beerman and Judith RappaportMusson, Prometheus Books, 2005

American Association of Retired Persons
[888-687-2277](tel:888-687-2277)
aarp.org

Aging Life Care Association
aginglifecare.org

US Administration on Aging Eldercare Locator
[800-677-1116](tel:800-677-1116)
eldercare.acl.gov

US Department of Veterans Affairs Office of Geriatrics and Extended Care
[877-222-8387](tel:877-222-8387)
va.gov/geriatrics

Training for family caregivers

Community-based resources may offer training and classes for family caregivers. Check with local hospitals, home care agencies, Area Agency on Aging, voluntary health agencies, and county and state departments of health.

Long-distance caregiving

Resources for Long-Distance Caregivers (Family Caregiver Alliance)

800-445-8106

caregiveraction.org/i-dont-live-my-family-im-caregiver

‘Share the care’ circles

AARP Meditations for Caregivers: Practical, Emotional and Spiritual Support for You and Your Family, by Barry J. Jacobs and Julia L. Mayer, 2016

A Guide for Caregiving: What’s Next? Planning for Safety, Quality, and Compassionate Care for Your Loved One and Yourself, by Tina Marrelli, 2017

Caregiver Defined: Words that honor the work of the caregiver, by Michael Fortuna, with illustrations by Peg Lee, 2017

The Conscious Caregiver: A Mindful Approach to Caring for Your Loved One Without Losing Yourself, by Linda Abbit, 2017

Living with Dying: A Complete Guide for Caregivers, by Katie Ortlip, RN, LCSW and Jahna Beecham, 2017

Share the Care: How to Organize a Group to Care for Someone Who is Seriously Ill, by Cappy Capossela and Sheila Warnock, Simon and Schuster Adult Publishing Group, 2004

Lotsa Helping Hands
lotsahelpinghands.com

Share the Care
212-991-9688
sharethecare.org

Finding or adapting a home

A contractor’s organization may donate labor. Also look for a local Home Builders Foundation.

Medical help

Get Palliative Care
getpalliativecare.org

National Association of Hospital Hospitality Houses (NAHHH)

Represents organizations that provide lodging and service for families receiving medical care away from home
nahhh.org

Mercy Medical Angels

Provides free transportation to medical care
mercymedical.org

Patient Advocate Foundation
800-532-5274
patientadvocate.org

Visiting Nurse Association
vnatoday.org

Long-term care

National Clearinghouse for Long-Term Care Information

US Department of Health and Human Services
longtermcare.gov

Hospice American Hospice Foundation
americanhospice.org

Hospice Foundation of America
800-854-3402
hospicefoundation.org

National Hospice and Palliative Care Organization
800-646-6460
nhpco.org

Assisted living facilities Consumer Consortium on Assisted Living
ccal.org

American Healthcare Association National Center for Assisted Living
ahcancal.org

New LifeStyles
800-869-9549
NewLifeStyles.com

Long Term Care: Everything You Need to Know About Long Term Care Nursing and How to Plan and Pay for Long-Term Care Insurance, by Betty Wilson, 2014
Nursing Homes and Assisted Living: The Family's Guide to Making Decisions and Getting Good Care, by Peter S. Silin, 2009

Respite help

Arch National Respite Network and Resource Center
archrespite.org
respitelocator.org

Easterseals
Provides services including adult day care
800-221-6827
easterseals.com

National Adult Day Services Association
877-745-1440
nadsa.org

Shepherd's Centers of America
816-960-2022
shepherdcenters.org

US Department of Veterans Affairs Office of Geriatrics and Extended Care
Veterans eligible for outpatient medical services can receive in-home respite care.
877-222-8387
va.gov/geriatrics

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With more than a decade of research in amyotrophic lateral sclerosis (ALS), MTPA strives to make a difference in the lives of those with this debilitating disease. We are focused on supporting the advancement of new treatment options as we continue to deepen our understanding of ALS.



To learn more about ALS, scan the QR code or visit ALSPathways.com



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 Instagram: @mdaorg

 Facebook: MDAorg

 LinkedIn: Muscular Dystrophy Association

 Twitter: @MDAorg

 Advocacy Twitter: @MDA_Advocacy

 YouTube: YouTube.com/MDA

 TikTok: @mdaorg

 Twitch: MDA_LetsPlay

 Discord: MDA Let's Play



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