

Guide For Caregivers



MDA[®]

Thank you to the MDA Care Center staff who contributed their time and expertise to help craft this guide into an accurate, useful resource for caregivers.



This guide for caregivers was produced solely by MDA and made possible in part by grants from Synergy Home Care and VieMed.

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FINDING CAREGIVING HELP

The information provided in this guide is designed to empower you and your family to take charge of your caregiving needs. At MDA, we are working to empower you by providing resources and support throughout your journey with neuromuscular disease. If you or someone you love is affected by muscular dystrophy, ALS or a related neuromuscular disease, please know you're not alone. Here, we offer some practical advice, recommendations, tips and resources for finding caregiving help.

Also available to provide additional support, the MDA National Resource Center is an informational hub for families living with neuromuscular disease. 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, 8 a.m. to 5:30 p.m. Central time, to answer questions and provide one-on-one support and resources from MDA's resource referral database. 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).



Emotional support and respite for the caregiver are absolutely essential forms of help.

ASK AND RECEIVE

One theme is sounded repeatedly by caregivers of loved ones with muscular dystrophy, ALS and related neuromuscular diseases: You can't do it alone. Get help!

Physical demands, emotional demands, other family responsibilities, jobs and more eventually take a toll on even the strongest, most devoted caregivers. Don't wait until stress becomes extreme. Take the advice of experienced caregivers: Get help early, get it from every possible source and accept all offers. This resource presents some of the many types of help a caregiver may find valuable.

Some of the help you'll need will be financial; some will be assistance with caregiving chores; some will be in the form of medical and other expert advice. Emotional support and respite for the caregiver are absolutely essential forms of help.

Rarely will someone provide the kind of care that the primary family caregiver can give a loved one, but many people can do some of the chores. To clarify the roles, think of yourself as the carer, the one who emotionally cares for (loves) the person who needs you, and the primary caregiver. Others who are hired or recruited as volunteers are caregivers or helpers, filling assigned tasks when needed.

HIRING CAREGIVERS

AGENCY HELP

Home health care services typically refer to those services that meet Medicare and/or private insurance guidelines. 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 registered nurse, physical therapist, occupational therapist or speech therapist who must be part of the care plan. For example, the individual may have a new medication or diagnosis in which a registered nurse will need to monitor the symptoms or side effects. A physician will guide you on what type of home care services would be recommended for you or your loved one.

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.

Assistance can include:

- Companionship
- Supervision
- Transportation for errands and appointments
- Homemaking
- Meal preparation
- Medication setup and reminders
- Assistance with personal care (bathing, dressing, grooming)
- Assistance with mobility and transferring around the home

Registries or visiting nurse associations serve as employment agencies for home care nurses and aides, matching these providers with clients and collecting finder's fees. These organizations usually aren't licensed or government regulated.

Home care providers may be found through a quick internet search under "home care," "hospice" or "nurses." The MDA Resource Center, an Area Agency on Aging or United Way chapter may also have a list.

Insurance policies and state Medicaid programs vary in the type of care they will cover, so be sure to understand the policy's provisions. Some in-home assistance for low-income people over age 60 with disabilities may be covered under the Older Americans Act, with funds administered by the state. A waiver or TEFRA waiver is a Medicaid waiver and may be available for home based Medicaid services for children and adults living with disabilities. A few states allow hiring and paying a family member as a

caregiver. Check with your counties human services office, Area Agency on Aging, or state department of social services.

WHAT YOU SHOULD KNOW ABOUT AGENCY HELP

Nurses and home care aides are trained and experienced in the various chores required to care for a person who's seriously ill. The agency handles all hiring, firing, supervising, payment, taxes and paperwork, and is responsible for finding a person the family likes.

There are costs associated with in-home care. Some agencies will accept state and local funding sources, such as waivers and Medicaid. Schedules will be individually designed to take into account the goals and needs of each client, as well as the family's care budget. In-home care often can be integrated with other community-based services to increase one's options.

A hired caregiver should be willing to learn about the family's preferences, and respectful of physical and emotional boundaries. The aide should be businesslike and competent but compassionate, willing to communicate with the person with neuromuscular disease no matter how difficult, and aware of family members' personal space and emotions. If the aide is at all uncomfortable with performing any of the duties required, especially duties related to the personal care of your loved one, don't be afraid to request another aide. Your responsibility is to make sure the aide's

behavior gives your loved one a sense that his or her needs will be taken care of competently. This helps reduce the stress of having an outside caregiver in the home.

Clarify requirements or preferences with the agency and employee at the beginning of the working relationship. Look at the first few weeks as a trial period; if someone doesn't show up on time, isn't a good fit or behaves offensively, call the agency and ask for another aide. A professional agency will comply without objection.



TIPS FOR USING AGENCY HELP



- **Be completely honest about needs.** Agencies are familiar with almost every type of assistance required; embarrassment or concern that you're asking too much shouldn't interfere.
- **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 agency will know your exact wishes.
- **Make it clear who in the family is the authority or decision-maker** — the person with neuromuscular disease or the primary caregiver or both. If everyone in the family gives orders to the employee, the result can be confusion and failure to give proper care. Discuss disagreements among family members outside the employee's presence.
- **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 employee is clear on your wishes. If that doesn't work, call the agency — it's responsible for supervision.



QUESTIONS TO ASK A HOME CARE PROVIDER

While caregivers who provide home care services may offer a similar list of tasks they can perform, it is important to understand more about the process and the caregivers themselves. Below are some questions you will want to ask before signing up to work with an agency. Working through an agency ensures that you have a partner in arranging and managing the care and the caregivers. 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.

GETTING STARTED WITH A HOME HEALTH CARE AGENCY

- What type of training, supervision and monitoring does it provide its staff?
- Is there specific training provided related to neuromuscular diseases?
- Does the agency develop a plan of care, and how often is it reviewed and by whom?
- 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 your caregivers agency employees or independent contractors (1099 tax form)?

There may be times where individuals will want to hire someone privately and not work with a home care agency team. In that case, here are some considerations to keep in mind when hiring a private home care worker.

If you are hiring a home care worker not affiliated with an agency, it is important to:

- Check references.
- Check with any state licensing boards (e.g., National Council of State Boards of Nursing).
- Complete a criminal background check (e.g., local police, sheriff, Bureau of Criminal Apprehension, driver's license division).
- Check with the IRS about tax issues.
- Check your homeowner's or renter's insurance policy to see if it covers damage to your home or any injury sustained by the caregiver.
- Determine a backup plan for when the private caregiver is sick or takes vacation.

The MDA Resource Center can help provide individuals and families with home care provider resources, and be sure to consult with your health care provider for additional guidance.



HIRING AIDES DIRECTLY

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, maintaining equipment — essentially everything the primary caregiver has been doing.

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

PROS AND CONS OF HIRING DIRECTLY

- **Pro:** Caregivers can pay a lower rate to someone who doesn't come through an agency, especially a part-time high school or college student.
- **Pro:** There's no need to deal with agency guidelines, regulations and applications.
- **Con:** The person won't have been evaluated by an agency, making the primary caregiver responsible for checking out applicants' backgrounds and ensuring the person is honest and qualified.
- **Con:** Some recordkeeping is involved, primarily hours and payment history. You also may be responsible for paying federal and state employment taxes if payment to the aide is over a certain dollar threshold.
- **Con:** Health insurance may not cover the cost of an independently hired aide (although it may not cover agency help either).
- **Con:** If there's only one helper, there's no agency to send a backup person when the helper is unavailable.
- **Con:** If the helper is a family member or friend, there can be awkward consequences if things don't work out or problems arise.

FINDING AN AIDE

There are many ways to find potential assistants. Word-of-mouth is the best resource; ask your MDA Family Care Specialist, support group, faith community, friends and neighbors. Other ways to locate applicants are:

- Check with independent living centers or senior centers.
- Place classified ads in local or college 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 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 health care professionals at your MDA Care Center for other resources in your area.
- Your children's teachers or school administrators may know of parents looking for work.
- Call the community volunteer center.
- Ask at privately owned drugstores and medical clinics.

DECIDING WHAT HELP YOU NEED

Before you look for an aide, 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, or can they use yours? Do you need someone to be on call or to work set hours?

Review these guidelines with your loved one. Is a male or female assistant preferred? What other preferences does the person have? The loved one may be resistant to having "strangers" come in, but go ahead and look for helpers anyway. It may take some time to convince your loved one to allow others to provide personal care, but at a certain point the primary caregiver knows best what's needed in the house.

INTERVIEWING AND HIRING

Hiring your own caregiving assistant means you have to screen applicants for criminal records, training, job referrals, etc. You can't take a chance on allowing someone dangerous, incompetent or dishonest into your home to deal with a vulnerable person.

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 don't describe your loved one's exact physical condition or the number of people in the household. There are criminals who search out vulnerable victims from ads and notices.

You can ask a bit about experience or background over the phone; the phone call should let you weed out inappropriate applicants. If you get a good feeling from the phone call, ask the person to come for an interview. You may want to interview three to five people before choosing one.

Expect interviewees to dress appropriately (that doesn't mean suits or heels) and to arrive on time or call if they must be late. Expect someone who listens, makes good eye contact and asks relevant questions. Most applicants should bring a résumé, though a young student or a person just re-entering the job market may not have one.

You can print out a simple job application form from the internet or create one.

Ask some open-ended questions: Tell me about yourself. Why are you interested in this job? What's your school/work schedule? What do you do in your spare time? The answers will give some insight into communication skills and personality, important factors in a future relationship.

Of course, ask about experience — even volunteer or family caregiving experience could be valuable. If they've done in-home care before, ask why the job ended.

Explain your needs in some detail and find out if they have experience bathing, feeding, etc. Describe a typical day and what's expected of them, and check the reaction. Be sure to tell

them of any uncomfortable tasks you expect of them, and ask if they have the physical strength for lifting and transferring.

If you're seeking a live-in, be specific about guidelines: Can anyone move in with them? Can they bring a pet? What's included — rent, utilities, food, phone, cable, 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 OK for the partner to spend time at your home. If so, how often?

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.

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.

If at any point in the interview, you know the person is unacceptable, just conclude the meeting. There's no need to waste time talking with someone you aren't going to hire.

When you've narrowed the field to one or two leading candidates, it's time to check 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 one 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. There are several websites that will do this for a fee or will explain ways to go about searching; see lexisnexis.com; virtualchase.com; searchsystems.net. Some search programs are little better than scams. Among the good ones are ReverseRecords.org and CourtRecords.org. You can also check with your state's bureau of Criminal Apprehension, as they offer free services.

Introduce your loved one and get his or her reactions. Trust your instincts.





MANAGING CAREGIVERS AND HELPERS

- 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 neuromuscular disease and the primary caregiver, will come to accept that this helper can improve the family's quality of life.
- To make things as easy as possible for everyone, give the caregiver as much information as possible. Offer a written summary about your loved one's condition, and provide MDA materials about the disease.
- Post a daily schedule, listing times for getting up, bathing, meals, meds, etc. Be very specific about dosages of medications. A monthly or weekly calendar that lists times of therapy and doctor appointments also would help. If the individual requiring care has speech difficulties, these lists will help new caregivers understand the needs he or she may be describing. Explain any other keys to assist with communication.
- Make copies of any instructions or advice you've received from health care professionals, such as those regarding acts of daily living. Go through each new chore carefully; demonstrate several times and show the new person how to watch for 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.
- Ask your loved one for feedback and make it clear to the caregiver.
- If your loved one is reluctant to be assisted by others, explain as best you can that accepting someone else's help is a great way for your loved one to help you. Their resistance may be a form of denial of the seriousness of their condition; requiring outside help means admitting the depth of the problem.
- Let the helper take on chores gradually or assist you until the your loved one with neuromuscular disease gets used to the idea. Explain to your loved one what

you will be doing while you're gone, and make it clear you'll be back. Remind the caregiver to be patient.

- If you aren't going 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.
- For caregivers you hire directly, 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.



ASKING FOR HELP FROM FRIENDS AND FAMILY

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 chores.

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 aren't going to line up at the door. They're at home waiting for a call: People usually will help if they receive specific directions. Be aware of your own psychological, physical and mental strengths, and look for friends to help in the areas where you're less confident.

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.

List all the people who have offered to help, and all those who haven't offered but who might be willing if asked. Develop a chore list, including frequency, and start matching chores with available people.

Friends and neighbors don't need to get involved in the most intimate aspects of caregiving. Household chores, errands, child care and such are more appropriate and comfortable for them, and they'll allow the primary caregiver more time for hands-on, one-to-one care. In requesting that others rake the leaves, drive the kids to soccer, pick up groceries, etc., you'll have to let go of total control over these tasks. You don't have the energy to sweat the details, and your way isn't the only way. If it gets done, you don't have to worry about it anymore.

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

- **Be clear about what you want.** Avoid vague requests: "Maybe sometime you could stop by to visit with AI for a few minutes?" 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 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 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 lists of chores by the phone or in a purse,** 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 offer but would be glad to

spend some money. If asked, suggest a book of postage stamps, an in-home massage, a takeout meal, a professional housecleaning, 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.

Be aware of your own psychological, physical and mental strengths, and look for friends to help in the areas where you’re less confident.



Children involved
in caregiving learn
compassion, cooperation,
patience and responsibility.



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 — “bring Mommy the formula for Daddy's dinner” — at first.

Children involved in caregiving learn compassion, cooperation, patience and responsibility. Sometimes, however, teens who take primary responsibility for caregiving develop depression, anxiety, insomnia, loss of interest in school, or delay becoming independent. Teen caregivers should be encouraged to get in touch with the social worker at the MDA Care Center to find support.

LONG-DISTANCE CAREGIVERS

Today, family members are spread across the country, and it isn't always possible for them to relocate to care for a parent or sibling. Long-distance caregivers have special difficulties:

- They can't see what's really happening or do hands-on care.
- It's difficult to reach doctors or social service agencies and coordinate needed care.
- They may feel guilty and sad over not being involved.
- Traveling back and forth can be exhausting, expensive and frustrating.
- Other relatives, including the individual requiring care, may not understand why the person can't just "drop everything" and come to help them.

With good planning and cooperation, even those who are miles away can contribute.

If a long-distance relative is the primary caregiver, a leave of absence from work may be possible. If not, or when young children are in the picture, frequent visits may be the only way. Try to be there for doctor visits to meet the health care team or for selection of major equipment.

The caregiving role will require a great deal of communication and coordination with those who live closer to the loved one who requires care:

- Entrust a neighbor or close friend who lives near the relative to check up and visit on a regular basis. Talk to the person frequently, and be sure they contact you if there are any serious changes.
- Go to the loved one's town to interview formal services such as visiting nurses, senior centers, adult day care or a meals program. Keep in regular contact with these agencies and make sure, through a friend or neighbor, that the person with neuromuscular disease is receiving proper care.
- Meet the physician and keep in regular contact. Call and speak to the physician directly, or ask how you can receive regular, updated notes on the visits and tests that are administered.
- Hire a private care manager through a social service agency or home health care agency. This person can coordinate services from physicians, nurses, therapists, social workers, homemakers, durable medical equipment and supply dealers, and volunteers. Private care managers can monitor daily care, assist families with implementing a long-term care plan and keep family members informed if a problem arises.
- Customize a caregiving network. Long-distance caregivers also can work with primary caregivers who see the loved one daily.
- Be informed about the individual's medical condition so you understand what's going on.
- Visit as often as possible so the family knows they're a priority in your life.
- Make the moments you are together count. Think in advance of your visits about things you can do together and ways to help. Just let your loved one enjoy your company.
- Offer financial support if possible. If family members are reluctant to take money, offer to pay for a specific need — in-home care, respiratory supplies, food, etc.
- Provide emotional support and involvement. Call or email the loved one often to keep them part of your world, and find out how the day went. Reach out to the primary caregiver as well and offer support or an ear for their frustrations.
- Maintain a website with family photos and news, or email pictures often. Encourage children to send notes and drawings.
- Respect the primary caregiver's leadership and offer cooperation; avoid disagreements whenever possible; make gentle, helpful suggestions.
- Get involved with MDA in your community; participate in fundraisers; volunteer to help at local events or even start a support group in your area for other caregivers and their families.

"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, so volunteers can offer specific help.

In care circles, a few individuals take responsibility for organizing areas such as laundry, shopping, child care, 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. If the family is so inclined, a Share the Care Circle can be pitched as a human interest story to a local newspaper or TV station or shared on social media channels. More attention equals more volunteers.



OTHER ASSISTANCE

ONLINE AND DELIVERY HELP

Many services and businesses offer home delivery and pickup, or online or mail order services: pharmacies, groceries, laundries and dry cleaners, holiday and birthday shoppers, clothing and electronics from department stores, movies and entertainment, etc. Ask a good friend to investigate these services and line them up for you. Using these efficiently can save tremendous amounts of time (and gasoline) that make it worth any extra cost.

The most efficient grocery delivery is to place a large order or a regular weekly order, rather than calling every time you need a gallon of milk. It's possible to order online from local supermarkets or from national services that ship within 24 hours.

Explain your situation to businesses such as car repair shops, etc., and they may be willing to pick up and deliver.

HOME MODIFICATIONS

Volunteers can be found to remodel a room, build a ramp, widen doorways or install a roll-in shower. Put the word out among friends and start the search for handymen or contractors who will donate labor and/or materials. Independent living centers (CILs) or home health agencies may know of someone who will help. The Homebuilders Foundation exists in several states as a philanthropic arm of the industry; they may help locate a volunteer contractor.

If it's necessary to move in order to accommodate your loved one's condition, or for financial reasons, the state's Social Services or Family Services Department may know of a program to help find housing. Some states are committed to helping people stay in their homes. CILs also have leads on low-income, accessible housing.

Disability law attorneys or legal aid organizations can help with rental disputes and agreements.

Put the word out among friends and start the search for handymen or contractors who will donate labor and/or materials.

MDA is committed to ensuring that the MDA Care Center network operates with a multidisciplinary approach. MDA Care Centers include strong medical teams, but they don't just have doctors. In addition, MDA Care Centers include a social worker, physical therapist and care center coordinator.



MEDICAL HELP

An MDA Care Center can refer your family to a range of needed assistance. Medical care is coordinated, and MDA Care Centers are directed and staffed by neuromuscular disease specialists, as well as knowledgeable therapists, social workers, medical specialists and others.

Multidisciplinary care brings together the expertise of professionals from a wide range of disciplines in one location to provide coordinated, patient-centered care to individuals with complex medical needs.

MDA is committed to ensuring that the MDA Care Center network operates with a multidisciplinary approach. MDA Care Centers include strong medical teams, but they don't just have doctors. In addition, MDA Care Centers include a social worker, physical therapist and care center coordinator.

Beyond this framework, the specialists included on an MDA Care Center team vary based on the conditions they manage and the age range of the individuals they work with. Likewise, not all MDA Care Centers function in exactly the same way. At some MDA Care Centers, individuals will see all their relevant specialists during every visit, while at others, the directing physician will recommend that individuals see some specialists more or less frequently.

The local MDA office or the MDA National Resource Center can point you to community services such as home health agencies, equipment providers or independent living centers.

The **social worker** at the MDA Care Center provides help in navigating the health care system, and is a great source of referrals to family counselors, in-home help agencies and assisted living facilities. The social worker is the family's advocate or case manager, who can help coordinate the various types of care needed or being received.

However, in the current health care system, families coping with neuromuscular disease need to be their own advocates. Dealing with medical professionals outside the MDA family, such as pulmonologists, skilled nurses, emergency services or therapy agencies, can require that families know what's expected and needed, and be prepared to educate the professional.

Understand what each person does in the medical setting so you can ask the right person. Get to know office staff so they'll be more receptive. When asking for information,

be firm about what you need and strive to remain calm no matter how frustrating the experience.

Remember that doctors are legally and professionally responsible to your loved one, not to family caregivers, unless a caregiver holds medical power of attorney or durable power of attorney. Your loved one must communicate to the doctor that the caregiver is to be part of exams, tests, discussions and decision making.

To become informed about providing physical care, ask occupational therapists, physical therapists and others how to do basic things: range-of-motion exercises, transfers, vent maintenance, tube feeding, basic nursing skills, etc. Manufacturers' representatives should be willing to answer questions about equipment, and demonstrations often are given at support groups. Some hospitals teach these skills, as do some home care and hospice services.

To learn about medical issues, do online research or even take a brief course.

You'll be able to ask the doctor more knowledgeable and pertinent questions and gain a higher level of information.

For example, a woman with ALS wrote up a detailed explanation of the disease, and how it did and didn't affect her. She included her needs and expectations for caregivers. Everyone — aides, physical therapists, nurses, volunteers — who dealt with her was required to read the material. Professionals who thought they knew everything about ALS learned more and got a better understanding of the woman's individual needs.

Educating staff about your loved one's medical condition and its effects is especially important when the person is in the hospital and is being treated by nurses, residents and others who may never have helped an individual with neuromuscular disease. A caregiver may have to get "politely pushy." It's better to be considered obnoxious than to let your loved one get the wrong type of care. Ideally, staying in the hospital with your loved one ensures their specific needs are made known and accommodated.

Keep medical information organized and complete so that each new person you see at the clinic, hospital or in your home easily can find answers and data.

Many hospitals have a patient advocate who speaks on behalf of patients in order to protect their rights and help them obtain needed information and services. This often is a nurse, social worker or other health care provider. If there's an ongoing problem with hospital treatment, ask to speak to the patient advocate.

A recent study funded by the National Hospice and Palliative Care Organization found that hospice care seems to prolong the lives of some terminally ill patients by days or even months.



LONG-TERM CARE

At some point, even the most devoted caregiver may need to turn over a significant portion of the loved one's care to professionals. It's emotionally very difficult to place a loved one in a long-term care facility, but his or her medical needs, or a caregiver illness, financial realities or other factors may make this essential.

Most of the time, some percentage of the costs of the facilities described here is covered by private insurance, Medicaid or Medicare, or the person's SSI or SSDI income. Once a person with neuromuscular disease goes into long-term care, Medicare may no longer provide certain equipment. Check with a medical expert and your MDA Care Center team to learn what equipment should be obtained prior to moving your loved one to a long-term care facility.

HOSPICE

Hospice services can provide a transition from care at home to full-time care away from home. Hospice care is available to people in the end stages of a terminal disease.

Hospice staff and volunteers coordinate with the patient, physicians and family members to provide day-to-day care and comfort, in the home or at other locations. Services include administering medications, including those to ease suffering; performing personal hygiene tasks and minor medical procedures; and offering compassionate companionship and support. Most hospice services provide support to the family as well. The hospice service may provide a lot of equipment for the patient at home, including bed, bedside table, alternating pressure mattress, roll-in shower, commode, medications and other supplies.

In general, hospice patients receive only comfort care and symptom management, not life-prolonging devices or therapies. However, pre-existing feeding tubes are OK and some hospice programs also accept patients who already are vented.

Hospice care often is provided in the home, but can be conducted in hospice centers, hospitals, nursing homes or long-term care facilities. The cost is covered under Medicare, Medicaid and most private insurance plans.

Call local hospice services when your loved one needs help with breathing, feeding, turning, etc. They can do a home evaluation

and explain what services may be available and when. Try to find a hospice whose staff is familiar with your loved one's diagnosis.

A recent study funded by the National Hospice and Palliative Care Organization found that hospice care seems to prolong the lives of some terminally ill patients by days or even months. Some reasons for this may include:

- More personalized attention may result in improved monitoring and treatment.
- Hospice care offers a comprehensive approach, focusing on the patient's emotional and spiritual needs in addition to physical health.
- Family caregivers are offered support and training. Their reduced stress or workload may help patients feel like less of a burden, and so increase their desire to live.

ASSISTED LIVING FACILITIES

Assisted living bridges the gap between independent living and nursing homes. Residents in assisted living centers aren't able to live by themselves but don't require constant care either.

Assisted living facilities offer help with activities of daily living such as eating, bathing, dressing, laundry, housekeeping and taking medications. The facility should create a service plan for each resident, detailing personalized services required by the resident and guaranteed by the facility, and update it regularly. People who live in newer-model assisted living facilities usually have private apartments, and there are common areas for socializing, as well as a central kitchen and dining room for preparing and eating meals.

In 2016, the national average cost of an assisted living facility was \$3,600 per month, which isn't likely to be covered by insurance. Other terms used for assisted living facilities include: residential care, personal care, adult congregate living care, supported care or enhanced care.

The caregiver and person being cared for can maintain their close relationship after placement in a long-term care facility, and the caregiver will remain vitally involved in care decisions.

NURSING HOMES

A nursing home or skilled nursing facility (SNF) is a place of residence for people who require constant nursing care and have significant deficiencies with activities of daily living. Physical, occupational and other rehabilitative therapies are offered. Nursing homes are designed to care for individuals who can't care for themselves and have numerous health care requirements.

Inquire about assisted living facilities and nursing homes in the community and be sure they can provide all the care your loved one requires. Be sure they permit ventilators, feeding tubes, etc.

After years of caring for a loved one with neuromuscular disease, having him or her live outside the home is a drastic transition. It's another loss to grieve, and usually isn't a change that anyone welcomes. The caregiver faces the shock of a suddenly changed role, just as the individual with neuromuscular disease may feel abandoned or adrift in unfamiliar surroundings. There's also a sense of relief for the caregiver as duties are reduced, but it comes mixed with sadness, loss and emptiness.

The caregiver and person being cared for can maintain their close relationship after placement in a long-term care facility, and the caregiver will remain vitally involved in care decisions. The loved one still needs a caring person to be sure that things are going well and the right care is being provided. Get to know the staff at the facility to understand who's responsible for what and who is your best ally if there are problems.

One positive aspect is that now time together can be focused on each other, rather than on chores to be performed.

REWARDS OF CAREGIVING

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

One recent study noted that “individuals who reported providing instrumental support to friends, relatives and neighbors, and individuals who reported providing emotional support to their spouse, had a 30 to 60 percent decreased chance of dying over the course of the study,” compared to their non-helping counterparts.

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!



Schedule respite regularly — you’ve earned it...

RESPITE HELP

One way to experience the rewards of caregiving is to get support through respite care. Call on your network of paid or volunteer helpers to stay with your loved one so you can get a haircut, go to a ball game, shop, enjoy a hobby, or just sit in the park and watch birds. Ask for babysitting or for someone to take the kids overnight occasionally.

Every community has respite facilities such as adult day care centers that will take the loved one for a few hours or 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 — you’ve earned it, and 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 this is necessary for you to do the best job possible over the long haul.

...no professional counseling or family support is quite as meaningful as peer support.



SUPPORT GROUPS

Caregivers credit support groups with new friendships, important advice on equipment and more. Individuals and families who are farther along on the journey help new members come to the realization that they can get through this; no professional counseling or family support is quite as meaningful as peer support. People don't have to explain their condition or any of the needed care or devices. They don't have to explain the exhaustion of caregiving or the other emotions and stresses they experience. Everyone who's there already knows.

Support groups aren't just places to share emotions and pick each other's brains. Groups often 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

DAILY CARE FOR INDIVIDUALS WITH NEUROMUSCULAR DISEASE

This section covers offers advice and resources for handling some topics that might arise when caring for your loved one. 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.

Proactively planning for future accessibility needs will allow for changes to be made in a timely manner and not during a care crisis.

ACCESSIBILITY AT HOME

Home modifications can make it easier on everyone. Plan ahead and make modifications before they're needed. 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. Modifications 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, building a new bedroom/bathroom suite, etc. Modifications may be tax deductible to some degree. If your home is not easily modified for increased accessibility consider a move to a home that can meet everyone's needs today as well as into the future, as additional needs may arise. Proactively planning for future accessibility needs will allow for changes to be made in a timely manner and not during a care crisis.

ACCESSIBILITY OUTSIDE THE HOME

While the Americans with Disabilities Act (ADA) requires that places open to the public be accessible to people with disabilities, this is not always the case. Whenever possible, check accessibility ahead of time and be ready to be flexible. Notify inaccessible businesses about your problems with access. Get a permit for using handicapped parking spots.

ALTERNATIVE THERAPIES

Nontraditional treatments — such as vitamins, supplements, acupuncture, electrical stimulation and heavy metal chelation — currently lack solid scientific proof that they work. Different people react differently, so an alternative therapy may be worth a try, but proceed with caution.

The important thing is to integrate alternative therapies into conventional medical care. Consult your primary care or MDA Care Center doctor before starting anything new. If a treatment won't cause harm, many physicians are willing to work with you in giving it a try. Alternative therapies usually aren't covered by insurance and can be pricey. Anything that's touted as a miracle cure or that requires a lot of money upfront should be viewed with a great deal of skepticism or ignored.

BATHING

As a neuromuscular condition progresses, bathing techniques change. Early on, handrails, shower chairs and handheld showerheads make it possible for people to continue bathing themselves. In middle stages of disease, more help may be needed. Later 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. Often, a person with late-stage neuromuscular disease needs a sponge bath, not a full bath, daily or every other day. A bathing sling is used on a standard lift and has a cutout for easier transfer into the tub.

Children with muscle disease who have difficulty sitting independently may need extra support in the bathtub early on. Selections range from simple positioning support chairs to fully adjustable reclining sling chairs, we encourage you to work with your occupational therapist to help find a solution best for your loved one.

A few other tips: Too-hot water and too-vigorous toweling off can cause dry, itchy skin. Similarly, a hot tub or spa may be inadvisable because the heat can overtax the breathing system, and jets can damage weak muscles. For sponge baths, a height-adjustable bed, such as a hospital bed, eases the strain on caregivers' backs. Look for a no-rinse shampoo or a shampoo cap that can simplify the task.

Assistance with bathing and daily hygiene can be found through home care aides, Medicare home health care and hospice staff.

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; or 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.

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. 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 (every day or every three days or whatever) 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. Don't mistake this involuntary release as diarrhea and treat your loved one with an anti-diarrheal product.

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 health care provider. Suppositories may be given between manual removal attempts to help clear the bowel. If you have any doubts, see your health care provider for a diagnosis.



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.

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.

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.

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's no guarantee that these will remain useful if the condition continues to progress. 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.



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.

In some individuals, drooling (sialorrhea) can be controlled or at least brought to a tolerable level. In some cases, an occupational therapist 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.

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.

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.

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. 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 pillow case.

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. 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 health care 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, power outages, terrorist attacks — 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. Order an adapter kit for your ventilator so it can be plugged into your car or a marine battery.
12. 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.
13. Have a list of the style and serial numbers of medical devices.
14. 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?

(continued)

...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.

EMERGENCY PREPARATION *(continued)*

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. 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?

ENERGY/FATIGUE

Some forms of neuromuscular disease lead to muscle fatigue which may manifest as general fatigue. Mental exertion also may be fatiguing because of overall effects of the disease. 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 assist with 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 your loved one's physician about any concerns about sleepiness and alertness of your loved one.

Assistive equipment enables greater independence and safety for your loved one, while making it easier for you to be an effective caregiver.

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 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.

MDA offers a durable medical equipment program that connects individuals in need with new or gently used medical equipment in good condition, including wheelchairs, shower chairs, hospital beds, walkers and canes, communication devices and similar items, to the extent feasible and when available. A prescription from your physician is required.

Forget the motto “no pain, no gain.” This doesn’t apply to people with muscle disease. Focus on maintaining functional strength, endurance and independence.

EXERCISE

It’s not known how much exercise — if any — 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.



EXERCISES 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 always should 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 15-30 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.

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 always should be observed.

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.

Assistive equipment actually allows more independence and fights the disease by preserving precious strength and energy.

FALLING

This symptom is of great concern to caregivers. 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 balk at using more “medical” devices like a walker or wheelchair out of embarrassment or resistance to “giving in to the disease.” Assistive equipment actually allows more independence and fights the disease by preserving precious strength and energy. Caregivers often try to convince their loved ones to use the appropriate equipment, with varying degrees of success.

Some tips:

- 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 or leaving the door unlocked.
- Protective equipment like knee and elbow pads and a helmet can make falls less injurious. Lighter-weight shoes are easier to negotiate.

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, 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 and more. Consult an occupational therapist 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 in order to be high enough to support weakened shoulder muscles. This is especially true if a pressure relief cushion is put on the seat.



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.

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 NMD needs help with something, it can be very frustrating to have a helper take over the task and do it their way. The end 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. Other 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 unattended a loved one with a history of choking or falling. Families have found several ways to monitor loved ones, such as cell phones, baby monitors, emergency response buttons, friendly neighbors, etc.

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.

Practice on other family members to get a feel for it and to allow your loved one to see it in action.

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.

LIFTS

As neuromuscular diseases progress and muscles become weaker, 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.

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.

To go upstairs and downstairs, a chair lift can be installed on the home's major staircase.



MASSAGE

Regular massage by a professional or a caregiver is physically and emotionally therapeutic for people with NMD, and also 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.

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 MDA Care Center physician when necessary. Regular medical checkups, flu shots and pneumonia vaccinations are essential to preventing respiratory complications. Conditions such as diabetes, cancer, hypertension, Alzheimer's, heart disease, etc., may affect what medications or treatments a person can take. 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 of 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, there are a handful of medications have been approved by the U.S. Food and Drug Administration to treat neuromuscular diseases, and others that may be used "off-label." 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.

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.

NECK

Neck muscles may be affected in some forms of neuromuscular disease, which can cause them to weaken and make it hard to turn or hold up the head. This can lead to decreased mobility and — if untreated — 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 flops 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.

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.

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



NIGHTMARES

Bizarre dreams or nightmares, coupled with morning headaches and mental fuzziness, may indicate respiratory problems in neuromuscular disease. We encourage you to speak with your pulmonologist for further testing that may be available.

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 disease 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," page 56, 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.

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.

...many women with muscle disease have successfully given birth. A child may give both parents joy and hope that will strengthen them in the battle against neuromuscular diseases.

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.
- 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.
- 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 disorder.

However, many women with muscle disease have successfully given birth. A child may give both parents joy and hope that will strengthen them in the battle against neuromuscular diseases. Consulting many experts may help couples make this very personal decision.

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.

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





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 medic alert bracelet or medallion will alert strangers to call the doctor.

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

For skin infections:

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 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.

- Herbal/natural remedies include applying apple cider vinegar or vitamin E; eating six cloves of fresh garlic or six to nine garlic capsules a day; eating yogurt with live cultures; and taking more B-complex vitamins.

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.
- Skin damaged by pressure sores requires extra protein, zinc and vitamins A, C and K.

Taking extra time to ensure comfort when a loved one goes to bed can cut down on call-backs for repositioning later in the night.



SLEEP

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 call-backs for 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 p.m. and 3 p.m., 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.

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.

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.

SOCIALIZING

Isolation is a risk for those with neuromuscular diseases. Symptoms such as drooling, immobility, 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 neuromuscular diseases, remind them he or she is still the same person, and encourage them to talk about things other than neuromuscular 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 spell caregivers at times so they 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.

SWALLOWING CONCERNS

Swallowing can be a concern in more advanced stages of some forms of neuromuscular disease. This often requires a consult with a dietitian to determine best food choices and some exercises to assist with eating and medication management. Many medications cannot be swallowed due to the size but cannot be crushed so have to work with the physician to understand the medication needs.

Because individuals may be concerned about swallowing issues and choking, they may or be encouraged to eat slowly. Caregivers can ensure that they are not alone during meal times and provide some companionship during these extended meal times.

Caregivers can ensure that food is prepared in a way to limit swallowing or choking concerns and have plenty of options available for the individual to choose from.

While the person is in bed, elevating the swollen parts above the level of the heart can reduce edema; prop the hands, knees and feet on pillows.

SWOLLEN EXTREMITIES

In some people with neuromuscular disease, fingers, hands, arms, ankles, feet or legs may appear swollen, sometimes extremely so. This is edema, the retention of fluid, and it has several causes. Often medications used to control drooling cause the body to retain fluids, leading to edema.

Most commonly in neuromuscular diseases, edema is due to being immobile. Muscle activity helps push blood through the veins to the heart. Without it, 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, 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. Adjust the length 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.

Any exercise possible also will help, whether it's being assisted to walk a little, tightening and pushing with the calf muscles, or just range-of-motion exercises.

Wearing compression hose can help, and shoes that lace are far better than slippers at controlling edema. 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 prevent discomfort. 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 figure out a balance between controlling edema and excess saliva.

Edema also can be a sign of congestive heart condition, cirrhosis or kidney problems, so be sure medical personnel check for these, especially if the eyes are puffy. Edema just in a leg can be a sign of a blood clot.

TEETH

Dental care for a person with a neuromuscular disease may eventually 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 also 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 diseases specialist can help ensure the coverage, though it's not a guarantee.

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.

In hot temperatures, breathing can be affected. Extreme hot or cold environments can affect the functioning of ventilators.

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 lack of embarrassment.

- **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.

People with neuromuscular diseases have bladder and bowel control for the most part — they know when they need to go — and always should go when they need to.

- **Raise the seat:** 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. Get the highest one that works because your loved one will need more height as neuromuscular diseases progresses. 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.

- **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.
- **Catheters:** 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 suprapubic catheter may be used when the Foley is too uncomfortable or causes continued infections. It's inserted directly into the bladder through the abdomen wall.
- **Toilet Training:** Toilet training a toddler or young child with a neuromuscular

(continued)

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.

TOILETING *(continued)*

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.

- **Cleansing:** 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, hang 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.
- **Public restrooms:** In public places, there's usually a toilet stall which is 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 diseases, 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.

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.

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’re flying to a distant destination in the U.S., you may be able to get a loaner wheelchair through the MDA equipment program in the city you’re visiting. Talk with your local MDA office about this before the trip.

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

Society for Accessible Travel & Hospitality
www.sath.org

Accessible Journeys
www.disabilitytravel.com

Wheelchair Travel.org
wheelchairtravel.org

URINARY URGENCY

Incontinence isn’t a feature of neuromuscular disease because in general the smooth muscle of the bladder and bowel aren’t affected by the disease. But 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.



RESOURCES

MDA RESOURCES

Caregiving-related articles and blog posts can be found on MDA's website (mda.org) and on MDA's Strongly blog (strongly.mda.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, 8 a.m. to 5:30 p.m. 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: <https://www.mda.org/services/caregiver-resources>

MDA QUEST ARTICLES:

October 2017

Choosing with Care: Deciding whether family members or professionals serve as primary caregivers comes down to a nuanced calculation of means, availability and personal preference

<https://www.mda.org/quest/article/caregiving-choosing-care>

October 2016

The Heart of Care: When one partner is the primary caregiver for the other, a little creativity and a lot of communication help keep the fire burning

<https://www.mda.org/quest/article/heart-care>

January 2014

Preventing and Dealing with Theft by Hired Caregivers:

<https://www.mda.org/quest/article/preventing-and-dealing-theft-hired-caregivers>

October 2012

Getting Care: There's No Place Like Home

Professional home care providers can reduce family caregiver burnout while providing individualized service and greater independence for loved ones

<https://www.mda.org/quest/article/getting-care-theres-no-place-home>

ADDITIONAL RESOURCES

CareCircle

914-741-6742

carecircle.com

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.

Check with your local MDA office and MDA Care Center for other local supports.

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

caregiveraction.org

Family Caregiver Alliance

800-445-8106

caregiver.org

Provides a listing of state-funded services for family caregivers.

PERSONALIZED WEB PAGES

CarePages

888-852-5521

carepages.com

CaringBridge

651-452-7940

caringbridge.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 Rappaport-Musson, Prometheus Books, 2005
American Association of Retired Persons
888-687-2277

aarp.org

Indian Health Service Elder Care Initiative
ihs.gov/MedicalPrograms/ElderCare

Aging Life Care Association
aginglifecare.org

U.S. Administration on Aging (AoA)
800-677-1116
eldercare.gov

U.S. Department of Veterans Affairs Office of Geriatrics and Extended Care
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.

Medifecta Training Program and Resources

medifecta.com

Offers video-based training resources for caregivers and senior care professionals.

LONG-DISTANCE CAREGIVING

Handbook for Long-Distance Caregivers

Family Caregiver Alliance

800-445-8106

caregiver.org

‘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 Jahnna 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)
nahhh.org

NAHHH represents organizations that provide lodging and service for families receiving medical care away from home.

National Patient Travel Center
800-296-1217
PatientTravel.org

Helps locate air transportation for patients who need distant specialized medical care.

Patient Advocate Foundation
800-532-5274
patientadvocate.org

Visiting Nurse Association of America
888-866-8773
vnaa.org

LONG-TERM CARE

National Clearinghouse for Long-Term Care Information
U.S. Department of Health and Human Services
longtermcare.gov

HOSPICE

American Hospice Foundation
americanhospice.org

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

Hospice Net
hospicenet.org

Hospice Patients Alliance
hospicepatients.org

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

ASSISTED LIVING FACILITIES

Consumer Consortium on Assisted Living
ccal.org

National Center for Assisted Living
ncal.org

New LifeStyles
800-869-9549
NewLifeStyles.com

Nursing homes

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

RESPIRE HELP

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

Easterseals
800-221-6827
easterseals.com

Easter Seals provides services, including adult day care.

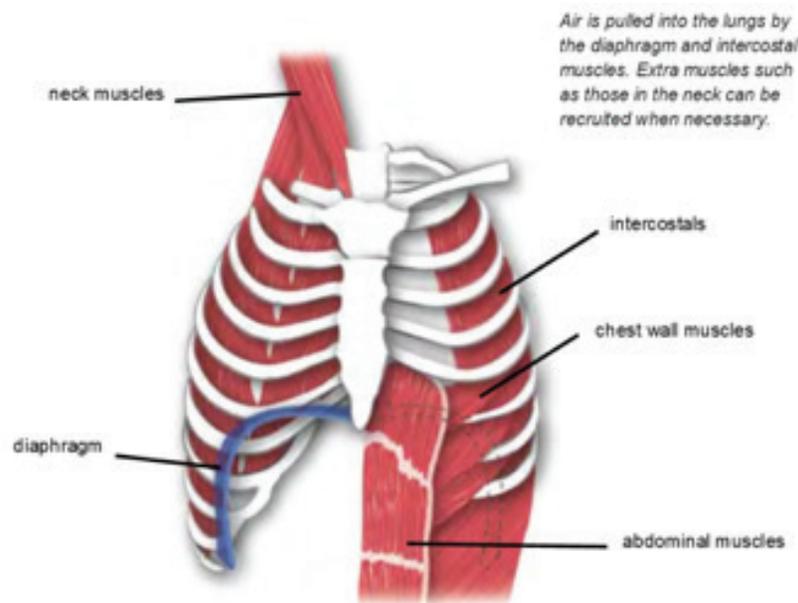
National Adult Day Services Association
877-745-1440 nadsa.org

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

U.S. Department of Veterans Affairs Office of Geriatrics and Extended Care
877-222-8387
va.gov/geriatrics

Veterans eligible for outpatient medical services can receive in-home respite care.

RESPIRATORY CARE IN NEUROMUSCULAR DISEASE



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

Fortunately, expert physicians such as those at MDA Care Centers know a great deal about treating the respiratory effects of neuromuscular diseases. This guide will help you understand how to manage your loved one's respiratory health and intervene before it reaches a crisis level, under the direction of your health care team.

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 of these muscles. 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 in your loved one, and risks that may be associated with a neuromuscular disease.

RISKS OF HYPOVENTILATION

Decisions about ventilation are some of the most important that an individual with neuromuscular disease will need to make. Information can make these decisions a little easier.

The effects of underventilation can be wide-ranging and in some cases, serious. Difficulties related to respiration include pneumonia, choking, sleep-disordered breathing, shortage of oxygen to the brain and cessation of breathing, which often contribute to death. Careful monitoring and a choice of interventions can manage respiratory health to avoid a dangerous crisis and prolong life, sometimes by many years.

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's 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.

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.

Be sure to consult an experienced medical professional or certified sleep specialist who knows which therapeutic solutions are appropriate for people with neuromuscular diseases.

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 or see if one is available from the MDA equipment loan program.
- 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 to 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.

A more sophisticated assessment tool is a sleep study or polysomnogram (PSG), which pinpoints the causes of disrupted sleep through a combination of measurements, including encephalographic (brain) activity, eye movement, muscle activity, heart rhythm, respiratory effort and others. Polysomnograms are performed in a sleep laboratory and last between seven and 12 hours.

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SLEEP APNEA AND MUSCLE DISEASE *(continued)*

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 Respironics.) 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.

RESPIRATORY CARE IN DUCHENNE MUSCULAR DYSTROPHY

By the time a boy with Duchenne muscular dystrophy (DMD) is in his teens — and sometimes earlier — he'll likely experience some respiratory distress, although this may not be recognized for what it is. Interrupted nighttime sleep, excessive daytime fatigue, headaches, and trouble concentrating and thinking may be due to poor air exchange.

In DMD, and some other muscular dystrophies, it's important to realize that cardiac function is closely tied to respiratory function. A deteriorating heart muscle, also common in DMD and other conditions, can have a negative effect on breathing.

The relationship between heart function and breathing also works the other way: Respiratory abnormalities can contribute to cardiovascular problems. Doctors who study cardiomyopathy in muscular dystrophy say that using noninvasive assisted ventilation, particularly at night, helps heart function in boys with DMD.

Poorly functioning cardiac and respiratory systems can be life-threatening. Early diagnosis, regular follow-up and early therapy for the heart or breathing problems can extend a young person's quality of life and length of life.

Experts strongly suggest that boys with DMD begin using noninvasive ventilatory support at night when breathing becomes insufficient during sleep or overnight blood oxygen

measurements are below normal. When daytime breathing becomes inadequate, they should add noninvasive ventilation during the day.

When these methods begin to lose effectiveness, the young man and the family should consider the option of a tracheostomy. The guidelines warn that supplemental oxygen shouldn't be used to treat inadequate breathing during sleep unless ventilatory assistance is also being used. And they caution against starting mechanical ventilation before it's required. Doing so can interfere with adequate respiratory monitoring and lead to a false sense of respiratory security.

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.

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

ASSISTIVE COUGHING (QUAD COUGH)

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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



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 health care 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 percent, 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 health care 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.

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.

NONINVASIVE VENTILATION

Most people who need breathing help begin with noninvasive assisted ventilation (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.





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.

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

Several studies have suggested that **Non-Invasive Ventilation can slow the decline in respiratory function and make a significant difference in survival time in people with more progressive forms of neuromuscular disease.**

Respiratory Therapist who can make changes as needed.

Several studies have suggested that Non-Invasive Ventilation 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
can allow the longest
possible survival with
symptoms.**

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 preNIV 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.

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, 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.
- 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.



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 health care 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 the "Finding Caregiver Help" section of this guide.

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

health care 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 non-invasive 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.

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



Partners to maintain Independence

Going through life with different abilities is not always easy. The world we live in expects us all to be strong and capable of doing every task all the time. Door handles, shower stalls and bathtubs, ATM Machines, steering wheels and even pots and pans assume we have full function. Some of these can be adjusted to help us get through the day, but others cannot. SYNERGY HomeCare helps clients living with Muscular Dystrophy and neuromuscular disease maintain independence, by helping them accomplish the day's tasks, and more importantly allows them to enhance the quality of their lives by assisting them with the activities that deplete their energy and can become barriers to pursuing their interests, passions, and hobbies.

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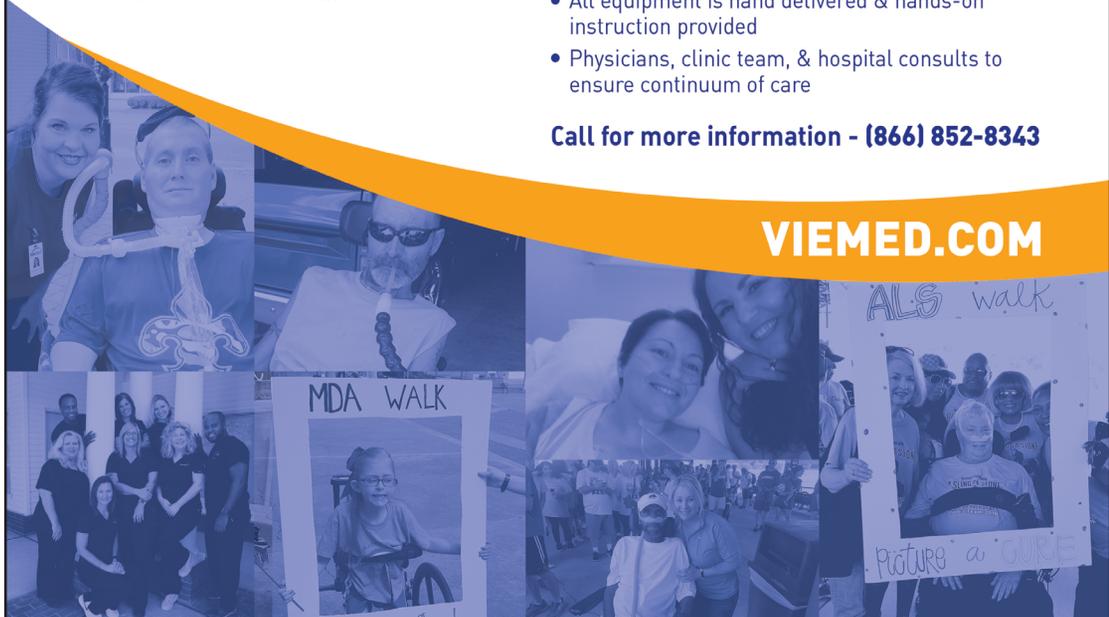
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