GUIDE #7

Functioning When Mobility Is Affected by ALS

ALS ASSOCIATION

LIVING WITH ALS
RESOURCE GUIDE

Functioning When Mobility Is Affected by ALS

Contributors:

Charlotte Chatto, PT, PhD

Leslie Ryan, MSPT

Nanette C. Joyce, DO

ALS ASSOCIATION

Table of Contents

A NOTE TO THE READER: The ALS Association has developed the *Living with ALS* resource guides for informational and educational purposes only. The information contained in these guides is not intended to replace personalized medical assessment and management of ALS. Your doctor and other qualified health care providers must be consulted before beginning any treatment.

Introduction

ALS is a disease of the motor neurons—the chain of nerve cells that originates in the brain, travels through the spinal cord (**Upper Motor Neurons or UMNs**) and ends with nerve cells projecting from the spinal cord to the muscles (**Lower Motor Neurons or LMNs**). The LMNs provide signals for muscle contraction and move our bodies through space, as we demand.

Defining this distinction between the types of motor neurons is important when discussing mobility and ALS because the loss of UMN or LMN will cause unique corresponding symptoms and related mobility impairments. Although various changes in mobility will occur and the challenge of everyday movements will increase over time, there are many types of equipment and devices that can be used to help maintain independence for as long as possible. For example, one person with ALS described his experience in the following way:

"ALS is a disease of daily discovery… each day I awaken to discover those things I can't do today that I could do yesterday."

Despite the ongoing changes, this individual approached each day creatively, adjusting his technique and tools so that he could continue to participate in the activities that he loved.

He loved to fish. When he lost the function of one of his hands, he fashioned a fishing pole that did not need both hands to control. When he could no longer stand, he used his power wheelchair to go to the docks. When he could no longer hold his pole, he attached it to his wheelchair. His spirit for pursuing what he loved could not be taken away from him. He spent his time focused on maintaining a life filled with meaning and joy.

What we will cover in this resource guide:

- UMN versus LMN symptoms and impairments
- Taking a proactive approach

- Team members required to address constant decline in mobility
- Energy conservation
- Activities and aids for daily living
- Exercise, flexibility, and stretching
- Durable medical equipment and assistive devices
- Home adaptions

Upper Motor Neuron (UMN) Versus Lower Motor Neuron (LMN) Predominance

People with UMN predominant ALS develop stiffness in their muscles, also known as spasticity. Spasticity is defined as an increased tension in the muscle that depends on how fast the muscle is stretched; the faster a muscle is stretched, the greater the involuntary resistance in the muscle becomes. This is due to an imbalance in the signals that influence the spinal cord and control the muscles' responses to being stretched. In healthy individuals, coordinated movements require some muscles to contract while others relax. In spasticity, this coordination is impaired. People with spasticity lose smooth, coordinated mobility of the limbs. Imagine, for instance, driving a car while applying both the gas pedal and the brakes. As you press faster and harder on the gas you also press faster and harder on the brakes. Strangely, you do not know which action, stopping or going, will prevail. Driving under these circumstances could be jerky, unpredictable and unsafe.

When you have a spastic muscle from UMN disease, your response to environmental disturbances is impaired by your inability to respond with rapid, instinctual responses. For example, a person with spastic leg muscles will walk with a stiff, jerky and dis-coordinated gait

pattern. If during an afternoon stroll this person catches their toe on a crack in the sidewalk and stumbles, they will instinctively attempt to right themselves but will instead respond with spastic stiffening of their legs and likely fall. If their upper limb muscles are also spastic, they will not be able to coordinate the protective response of reaching their arms out to slow their contact with the ground and protect their head from injury.

On the other hand, those with LMN predominant ALS have muscle weakness at the core of their mobility impairment. LMN loss causes muscles to weaken and shrink, becoming smaller and less powerful, until they are no longer able to signal muscle contraction. Progressive LMN loss in ALS eventually results in paralysis of all skeletal muscles of the body.

To meet the clinical diagnostic criteria for ALS, a person will necessarily have evidence of both UMN and LMN abnormalities. ALS, however, is quite varied from person to person. Each person presents with a unique pattern of signs and symptoms, and a combination of spasticity, muscle atrophy and weakness. Addressing your ongoing mobility needs requires frequent assessments and is best handled in collaboration with your care team, who are willing to be endlessly creative in their approach to problem solving. Table 1 lists some primary reasons why mobility might be affected.

Primary reason why mobility might be affected
Muscle weakness in limb(s)
Paralysis occurs in some muscles, while other muscles may be weak or not affected
Spasticity (involuntary spasms and stiffness in muscles)
Painful muscle cramping
Loss of range of motion and flexibility, especially in the shoulders, hands, and ankles

Being Proactive

"One of the things that is pressed upon you at ALS support group is to always stay ahead of the curve when making decisions. You don't want to make decisions after you need something; instead, make these in advance so that you are not caught off guard. This comes from folks who know and who've been there."

–Cheryl T., caregiver

While the details of each person's story will be unique, there are more similarities than differences from one person with ALS to another. Experts who focus their practices on caring for people with ALS develop a deep understanding of how ALS symptoms spread and change. In this way, they are able to predict and anticipate the mobility and Activities of Daily Living (ADL) needs prior to major transitions, thereby avoiding crises. For instance, if your legs become weak, you will begin to have difficulty standing up from a low-seated position. Instead of waiting until you are unable to stand from the toilet (which are most often low in height) and require a visit from the emergency response crew to transfer you, the perceptive practitioner will have anticipated this problem and recommended in advance to obtain an elevated toilet seat with handrails to enable transfers on and off the toilet.

One of the goals of the ALS rehabilitation team is to help each individual remain as independent as possible for as long as possible. Just as a construction crew is more capable of completing a job when they have the right tools, you can remain more functional and have prolonged independence if you have the right tools, provided at the right time, to help with declining function.

Ultimately, you have a choice to approach your care in a proactive or reactive manner. **Taking a proactive approach provides everyone involved in your care extra time for problem-solving and can help**

you avoid the stress of being stuck without options, struggling to get by when you have suddenly lost an ability. It's best if you ask your treatment team to provide recommendations for equipment in advance of your need. Obtaining what they recommend and putting it away for the day it is needed can help you feel prepared. Getting the appropriate piece of equipment can relieve stress. People often delight in their newly found freedom when they receive a power wheelchair and realize they can resume community activities that were given up due to intolerable fatigue or difficulty walking with fear of falling. Remember that tools are used in every trade to improve the ability of the people doing the work. ALS should be no different.

Your Mobility Team: Physical And Occupational Therapists

The ALS rehabilitation team includes both physical and occupational therapists. The roles of the physical and occupational therapist are to address mobility problems resulting from muscle weakness, spasticity and contractures. These impairments affect a person's ability to walk, to perform transfers (e.g., moving from a reclined to a seated position or a seated to a standing position) and to perform daily activities such as dressing, bathing, grooming and feeding themselves. These therapists provide recommendations and treatment to improve function either by initiating corrective one-on-one therapy or by recommending and teaching how to use assistive devices. They design and teach focused range of motion exercises, ensuring that individuals with ALS and their caregivers have the skill to perform these independently in the home or a community setting.

Who to See for What

The ALS **Physical Therapist (PT) focuses on gross motor activities such as walking, sitting balance and lower limb range of motion, strength and balance.** They provide recommendations for braces, canes, walkers, standers, scooters and wheelchairs (Figure 1).

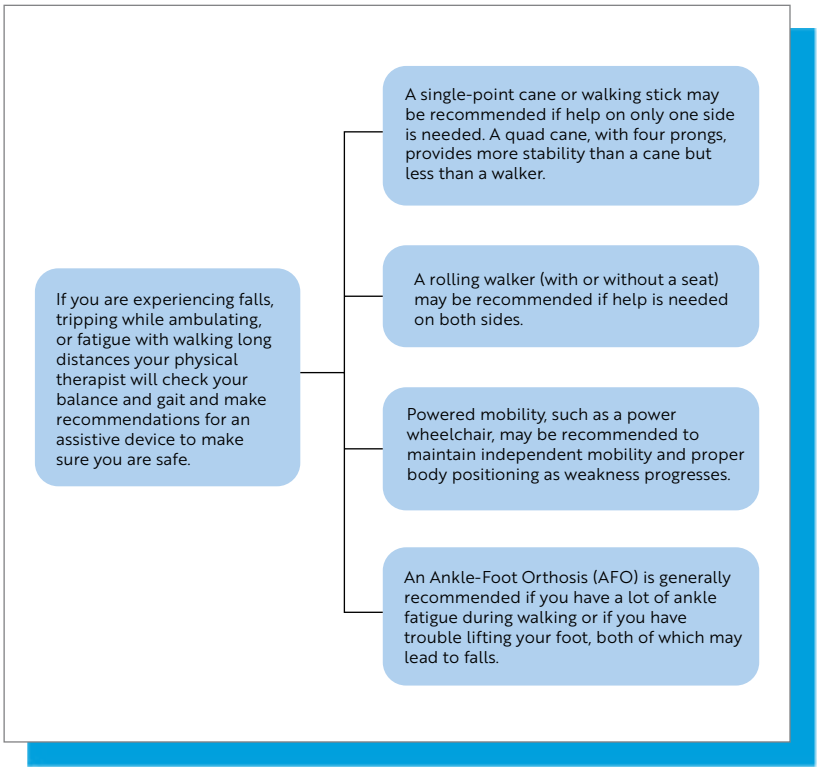

Figure 1: Physical therapist recommendations for increased mobility.

The **ALS Occupational Therapist (OT) provides assessment and treatment for the skills needed to perform activities of daily living.** They focus on their client's ability to transfer, dress, bathe, groom, toilet and feed themselves. They assess upper limb range of motion and strength and provide recommendations for upper limb and neck bracing. In addition, they perform home safety evaluations and can perform wheelchair assessments. Together, the PT and OT help families negotiate the transition of their loved one from independence to dependence by providing caregiver training, teaching transfer techniques and offering recommendations for durable medical equipment and assistive devices.

Figure 2: Example of bracing option to support the head.

In addition to interacting with therapists, you may also need the services of an **orthotist, who fabricates, fits and maintains braces**; and **a durable medical equipment provider, who provides and maintains wheelchairs, transfer equipment, bath equipment and hospital beds.**

Conservation Of Energy

As the disease progresses in ALS, **individuals often struggle with fatigue.** Using your energy for those activities that matter most and applying energy conservation techniques for other tasks can be a helpful strategy for maintaining a schedule that is not too tiring. Both PTs and OTs are trained to provide recommendations and teach energy conservation techniques.

The following are a few examples of conservation strategies:

Rearrange your environment:

1. Unclutter rooms and have furniture arranged to improve access.
2. Reduce the need to go up and down stairs by moving a bedroom to the main floor used for daily living.
3. Keep regularly used items in easily accessible places.
4. Replace heavy items with lighter weight versions (e.g., plastic plates and cups as opposed to stoneware and glass).

Eliminate unnecessary activities and prioritize:

1. Eliminate activities that are not important for you to do and delegate those that are important to family and friends.
2. If possible, consider hiring help to complete chores around the house.
3. Sit, rather than stand, while performing tasks.
4. Use a rolling walker with a seat or a wheelchair with a tray to help carry objects through the house.

Plan ahead:

1. Plan enough time for activities to reduce the extra energy demand required when rushing.
2. Schedule rest breaks in the middle of planned activities.

3. Call ahead to make sure that everything you will need is available when shopping, etc.
4. Have meals made ahead and packaged into easy-to-prepare servings.

There are many other ways to conserve energy. A creative PT or OT can help you look for ways to modify your home and activities to reduce fatigue. Attending support groups or participating in online conversations with other people living with ALS is another rich resource for solving mobility challenges.

Here are some ways you can **plan and prioritize:**

- If you know that you are going to have to walk a long distance, plan for plenty of time and rest breaks.
- If you can sit to take a shower, instead of stand, make sure you have a bathtub seat.
- If you are going to prepare a meal, make sure you get all the items you need before you begin.
- Think of ways that you can slide, push or pull items rather than carry them.
- If you have more energy in the morning, do those activities that are most important to you at that time.

Activities And Aids For Daily Living

There is a long list of Durable Medical Equipment (DME) and assistive devices that are potentially helpful. The goals for using DME and assistive devices are to improve function, prolong independence and maximize safe caregiving once a person has transitioned to dependence. Activities of daily living include dressing, bathing, grooming, toileting, bed mobility, transfers and

feeding oneself. Each person's needs differ depending on the extent and location of their weakness.

For example, if your arms are weak and lifting your hand to your face becomes challenging, elevating the arm above the waist by resting the elbow on a table, tray or other elevated surface will improve arm function for eating, brushing teeth or grooming as you eliminate much of the work done against gravity.

Consulting with your OT on a regular basis and exploring online or catalog resources can provide options for commonly encountered problems.

Devices for Eating, Brushing Teeth, Bathing, Grooming, and Dressing

Figure 3: Utensils and tools to assist with eating.

EATING

When the hands become weak, it is easier to hold tools that have wide handles and are lightweight (Figure 3). Plastic or rubber utensils are lighter and easier to use than metal utensils and can extend a person's ability to eat independently. As grip strength weakens, holding objects can be made easier by building up the thickness of handles. This can be accomplished simply by using foam tubing that can be cut to size and slipped over the handle, or you can purchase ready-made utensils with widened rubber handles. Once a person loses the ability to grasp but still has the ability to bend their elbow and bring their hand to their face, a universal cuff that slips over the hand and attaches to the needed tool is useful. A universal cuff can be attached to grooming tools as well as utensils.

Brushing Teeth

Electric toothbrushes make brushing teeth easier, as the handles are generally wider than a standard toothbrush and the user does not need to exert as much energy to scrub their teeth. For persons with bulbar dysfunction or those who are dependent for their care, there are suction toothbrushes available that wick away water and saliva as the teeth are cleaned (e.g., Plak-Vac® by Trademark Medical). Electric razors are easier to use to maintain facial hair.

Figure 4: A sliding tub transfer bench to safely get in and out of the tub.

Showering and Bathing

Aides for showering and bathing include grab bars, long handled scrub brushes, hand-held shower heads, sliding tub and shower benches and rolling shower chairs (Figure 4). Shower chairs and sliding tub benches should be purchased with a seat belt and a backrest high enough to support the head. These features are needed in the later stages of the disease when there is no longer sufficient head and torso control when sitting.

Figure 5: A long handled shoe horn can assist with putting on shoes without having to bend forward.

Dressing

Clasps and closures can become difficult to manage. As fine motor control worsens, buttonhooks and zipper loops can be used to facilitate buttoning buttons and zipping zippers. Other dressing strategies include switching from tailored clothing to loose-fitting clothes or clothes made of stretch

fabric (e.g., Lycra®) with elastic waistbands and loose pullover shirts. Socks can be donned more easily using a sock assist tool. Long-handled shoehorns minimize the work needed to reach down and put on a pair of shoes (Figure 5). Fabric shoe closures (e.g., Velcro®), magnetic shoe closures (Zubits®) or coiled no-tie shoelaces can make fastening a pair of shoes much easier than having to tie shoelaces. For dressing, a quick internet search for "adaptive clothing" can provide multiple resources and options for improving the ease of dressing.

Figure 6: A Hoyer Lift allows a person to be lifted with minimal physical effort.

Moving Around and Getting in and out of Bed

There are three major aspects of bed mobility that will need to be addressed during the course of the disease: getting in and out of bed, moving in bed and body positioning while in bed.

Getting in and out of Bed

To improve the ability to stand from and sit down on the edge of a bed in the early stage of ALS, risers can be added to the legs of the bed to bring it to a height that is easier from which to stand. Adding impermanent adjustable bed rails or a transfer pole that extends from the floor to the ceiling next to the bed can provide a solid hold for the upper limb to assist with standing. When you are no longer able to get into and out of bed, a transfer lift system should be used for safe and easy caregiver supported transfers. Lift systems come in many varieties, powered by hydraulic or electrical means, and can be mobile units or permanent systems that are attached to the ceiling (Figure 6). These lifts use a large sling that wraps under and around the person being lifted and hooks to the machine. The system then lifts and supports the individual's entire weight, allowing them to be moved easily into a wheelchair, onto

a commode or into and out of bed safely. An OT or PT should be asked to provide your family and caregivers with training on how to use the lift system when it is ordered.

Moving in Bed

It is important to be able to move in bed, whether by your own efforts or the efforts of others. A person with normal strength can naturally move throughout the night while sleeping, repetitively repositioning themselves for comfort. As you weaken, moving in bed can become impossible. For skin health and to avoid pressure-related wounds, a person who is unable to independently adjust their position in bed requires repositioning at least every two hours. Draw sheets are helpful for repositioning by putting the caregiver at a greater mechanical advantage. One option is a two-draw sheet system where a circular piece of material that is slippery is placed on top of the bottom fitted sheet. This provides a surface that has less friction for sliding a person back and forth over the bed. This sheet is then covered with a second unfitted cotton draw sheet.

Figure 7: Draw sheet used for repositioning in bed.

The top sheet is used as the pulley that moves the person in the bed. When you use a draw sheet, the top draw sheet should be placed at the person's shoulders and extend to below their hips. The draw sheet should be rolled up close to the body of the person who is being moved, creating a handle to grab. Caregivers should always roll the person being moved toward them and use their legs instead of their back for transfers.

Positioning While in Bed

Pillows, pillows and more pillows should be available to support comfortable positions. Bolsters and wedge pillows can be very helpful in providing support as well. While lying on their side, a person will need support under their ribs so they are not lying directly on their shoulder, which can become painful, and in between their knees to keep their hips aligned. While lying on their back, support under shoulders and arms can prevent discomfort, and pillows or bolsters under knees and lower legs can prevent increased pressure on the heels.

Hospital beds provide a convenient way for positioning. There are fully electric and semi-electric beds. Medicare typically will cover a semi-electric hospital bed. These beds have electric hand controls that allow the head of the bed to be lowered and lifted and the knees of the bed lowered and elevated by a push of a button. Elevating the head of the bed to 30 degrees helps improve respiratory performance during the night. The semi-electric bed is equipped with a crank to raise and lower the height of the entire bed. The fully electric bed does this with the electric hand controls. Changing the height of the bed is helpful for both the caregiver and person in the bed, enabling better biomechanics for bedside care and safe transfers.

Hospital beds come with standard hospital mattresses, which are not very comfortable for most people. Alternating **air pressure relief overlays or pressure relief mattresses** are usually more comfortable. They reduce the frequency with which a person with ALS needs to be repositioned. These mattresses are continually shifting the pressure applied to the body, thereby reducing the likelihood of developing pressure-related wounds. It can take time to find a mattress that is soft enough for comfort and firm enough to enable bed mobility.

Personal Hygiene Challenges

Do not underestimate the importance of developing a plan to address needs related to personal hygiene and toileting. This can be one of the most distressing problems for you. It is a fact of life that what goes in must come out. Between the ages of two to three years we take control of this part of life, and it remains a private personal issue until we are challenged by our mobility and can no longer meet our own toileting needs. While it is a good strategy to perform timed toileting, scheduling bathroom breaks every four or so hours during the day to prevent accidents, there will always be some randomness to our urges that make our needs unpredictable. **It is not a good strategy to avoid drinking fluids to avoid needing to use the bathroom**. This behavior leads to dehydration and constipation, both of which cause discomfort and are problems of their own.

This transition will occur when you have sufficient weakness limiting your ability to get on and off the toilet, or you do not have the upper extremity mobility and coordination to wipe yourself clean (medical term: **perform perineal care**). Accidents may happen if you cannot make it to the bathroom in time. While there are adult diapers for protection when accidents occur, sitting in a wet or soiled diaper for an extended period will lead to painful rashes and increase the risk of developing pressure-related sores, if habitual.

So, what can you do? **Bedside commodes** improve the convenience of toileting by lessening the distance to the toilet. In addition, they remove the transfers required for toileting out of a small bathroom into a wide-open space. Their height can be adjusted, and they provide handrails which improve your ability to stand from a seated position, prolonging the ability to get on and off the toilet without help. Another option is a **rolling shower chair/commode combination** that can be rolled into the bathroom and backed in over the toilet. Again, this equipment removes the dependent transfer out of a small space. A **bidet** attachment for the toilet is another option that eliminates the need for wiping and can aid in independence.

Actual examples of choices made by two persons with ALS:
Example 1. A woman with ALS declared that she would move into a skilled nursing facility as soon as she could no longer clean herself after toileting. She did not want to burden her children with her perineal care. Her weakness began in her upper limbs. Early into her disease she lost the ability to wipe herself clean; her legs, however, were still very strong and she was able to walk and transfer with ease. She was still considering moving away from her family, the people she loved the most and who brought her the most joy. The OT recommended she consider getting a bidet, a tool to help with toileting, which she did. Her distress was relieved, and she remained with her family.

Example 2. Another woman with ALS had psychosocial distress when out in public related to toileting challenges. Maintaining an active social life was very important to her; however, she felt deterred by fears of needing to use the restroom in public. Her husband having to accompany her into a public women's restroom and transfer her to the toilet in a stall with limited space and of questionable cleanliness was overwhelming. She searched for a solution and decided she wanted to have a suprapubic catheter. A suprapubic catheter is a permanent tube inserted through the skin of the lower abdomen into the urinary bladder. The catheter tube connects to a bag that is attached to the leg and collects urine as it drains from the bladder. After a lengthy discussion with her physician to address all the considerations, she moved forward with the procedure. She claimed, without question, that it was the best decision she made.

Men have another option that is less permanent: a condom catheter. There are many different styles of catheters, all of which need to be sized to fit well. A physician will need to provide a prescription for monthly supplies that include the condom, tubing and a urine collection bag. In addition, men can use a urinal to avoid having to transfer out of a wheelchair or bed during the night to urinate if they have adequate hand function or a caregiver who provides assistance.

For younger women with ALS, menses can be troublesome. If there is no desire for pregnancy, extended and continuous cycle oral contraceptives that reduce the number of menstrual cycles or an intrauterine device (IUD) may be the best solution. This strategy should be discussed with an obstetrician/gynecologist and the benefits weighed against any risks.

Exercise, Flexibility And Stretching

You may be interested in the role exercise plays in the disease. Can you continue your pre-diagnosis lifestyle? Does exercise have a reparative role with positive effects on endurance and strength?

There is limited scientific evidence regarding the risks and benefits of aerobic and strength training exercise in ALS. Animal studies in mice with ALS have shown that moderate-intensity aerobic-type exercise delayed disease onset and increased survival times in the exercised mice compared to mice that were not exercised (Carreras et al., 2010; Mahoney et al., 2004). On the other hand, high-intensity endurance exercise has been detrimental (Carreras et al., 2010; Kirkinezos et al., 2003; Veldink et al., 2003).

Small clinical studies in people with ALS also support the safety of moderate-intensity exercise. The most recently published study on this subject was a higher quality randomized controlled trial assessing the effects of exercise in ALS (Lunetta et al., 2015). The study findings were consistent with results of two prior studies of exercise in ALS (Drory, et al., 2001 and Bello-Haas et al., 2007). **All three trials showed less functional decline as measured by the ALS Functional Rating Scale (ALSFRS) in those who participated in a moderate exercise program**. The most recent study showed the greatest benefit in people living with ALS who performed a moderate-intensity strength training and cycling program.

Ongoing benefit was observed up to 12 months after the initiation of the program. However, none of the three clinical trials demonstrated a prolonged survival benefit.

Based on the available evidence it appears safe for people living with ALS to participate in moderate-intensity exercise. Overexertion, as demonstrated by prolonged fatigue after exercise, muscle pain or soreness, however, should be avoided (Petrof, 1998). Restorative gentle exercise can be used as a tool to prevent deconditioning to improve sleep and mood. Aerobic exercise practiced in a community setting (e.g., accessible pool, adaptive golf, chair yoga, tai chi) helps provide opportunities for socializing. Stretching and range-of-motion exercises should be started soon after diagnosis as part of a gentle, wellness-oriented, daily routine. Performing simple stretching exercises to target the major joints helps prevent painful and function-limiting contractures, especially at the shoulders. Training by a physical therapist, who will develop an appropriate exercise routine for you, is recommended. The program can be performed independently in the early stages of disease and transitioned to caregiver-assisted, range-of-motion exercise as needed. Linking the range-of-motion program to a dressing schedule, when limbs need to be maneuvered into clothing, is convenient and encourages performance twice daily during morning and night dressing.

Durable Medical Equipment And Assistive Devices

Medicare covers Durable Medical Equipment (DME) meeting the following criteria: "The equipment must be long-lasting, used for medical reasons, and would not be considered useful to someone who is not sick or injured." DME is for use in your home and has an expected lifetime of at least three years. To be covered by Medicare, the equipment needs to be prescribed by a physician. All people

with Medicare Part B have DME benefits with coverage at 80%. Medicare will only replace an item after it is so worn out it can no longer be fixed, and never less than five years from original purchase.

If you have an insurance provider other than Medicare, you will need to review your policy to determine what is covered, the timing of coverage and how to optimize benefits. Your ALS clinic team can help you understand your benefits regardless of your provider.

DME that Medicare covers includes, but isn't limited to:

- Air-fluidized beds and other support surfaces (these supplies are only rented)
- Canes (however, white canes for the blind aren't covered)
- Commode chairs
- Hospital beds
- Manual wheelchairs and power mobility devices
- Oxygen equipment and accessories
- Patient lifts (hydraulic, not electric)

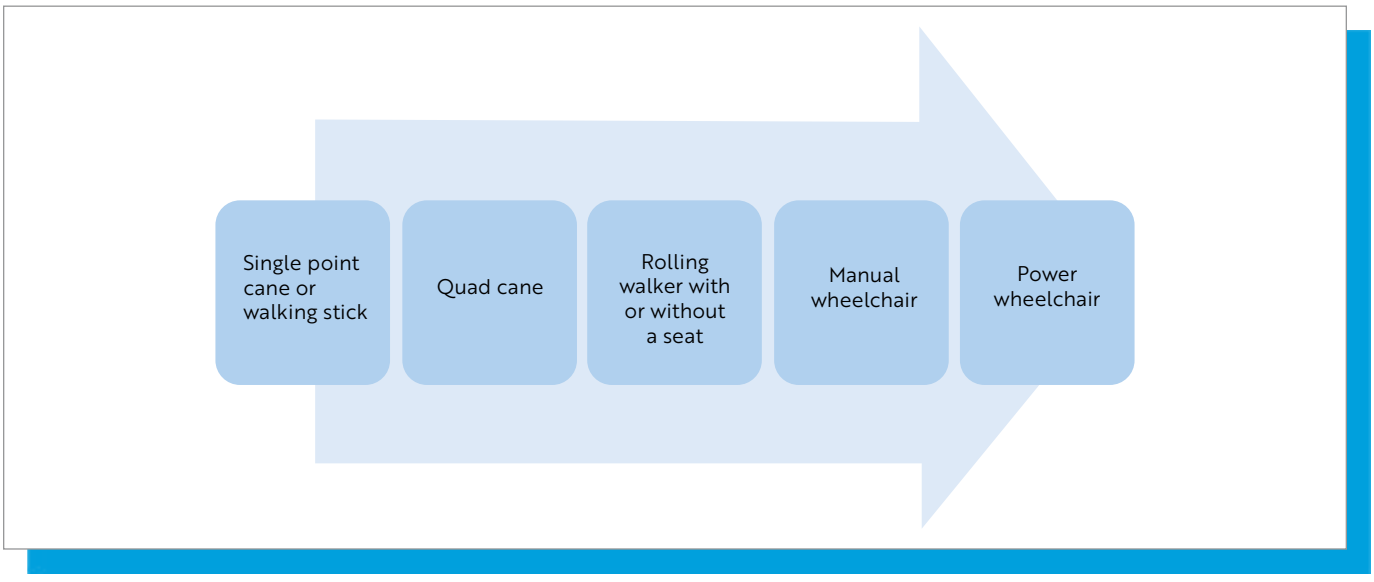

Figure 8: Transitions in assisted devices.

- Bilevel Positive Airway Pressure (BiPAP) or Average Volume Assured Ventilation (AVAP) devices and accessories
- Suction pumps
- Traction equipment
- Walkers

Beds, assisted breathing devices and other equipment are rented to own. The cost of the equipment is divided over 13 months and once paid, becomes the property of the person living with ALS.

When the loss of independent ambulation occurs, braces, canes, walkers and wheelchairs should be considered. The transition to and from each of these assistive devices should be monitored in the clinic (Figure 8). Braces to stabilize the ankle, called ankle-foot orthoses, are helpful if you have a foot drop. They prevent catching your toe and tripping.

Figure 9: Four-wheeled walker.

Functioning When Mobility is Affected by ALS

Canes and Walkers. A cane or walker can help prevent falls if prescribed appropriately or add to the risk of falls if prescribed inappropriately. For example, when you have significant spasticity, you may be at greater risk of falls and injuring yourself while using a cane because it does not have adequate stability to provide reliable support. A walker, however, may provide the support needed to continue walking safely. People living with ALS most often do well with a four-wheeled walker with a seat. Hand strength is required to be able to lock the

"This past week I had to give up using my walker and now fully rely on my "Go Chair." This causes some mixed feelings since it means giving up a major source of independence, walking. However, using the scooter does provide some freedom that the walker did not. Since I don't get as tired as I did with the walker, I am not as hesitant to get up and move, I can travel with ease around my house or most important, all over the mall; shoppers, watch out here I come. Either with the walker or the scooter, I have to count the cost, preserve my energy, and realize each step in life is precious."

–Susan C.

brakes, and it should be the type of walker that has enough clearance behind the seat to allow standing within the arms of the wheelchair. Falls are very common in ALS and every attempt should be made to try to prevent them. In one study, nearly 2% of people with ALS died prematurely from a fall.

Wheelchairs. As strength and coordination worsen, hand-held devices will eventually be inadequate to support safe ambulation and a wheelchair will be required. While a slingback manual wheelchair conveniently folds and is easily transported in a car, these wheelchairs do not have good cushion support and are not appropriate for prolonged sitting. It is important to work with a specialist who knows about ALS and can predict what will be needed in a wheelchair for long-term comfort (Figure 10).

Figure 10: Manual wheelchair.

The most appropriate wheelchairs when living with ALS have complex features such as tilt-in-space and recline that enable repositioning for maintaining skin integrity, comfort, communication, safe eating and respiratory health. Many people with ALS spend most of their time in their wheelchairs, so the chairs must be comfortable and facilitate good health. There are tilt-in-space

manual wheelchairs and power wheelchairs (Figure 11). The benefit of a power wheelchair is that you can continue to be independent and navigate around the home and community, as well as reposition yourself. However, if there is dementia, a manual wheelchair with tilt-in-space and recline may be the best option, as it will require caregiver participation for use. Medicare typically only covers the purchase of one wheelchair. It is in your best interest to have Medicare help purchase the complex wheelchair while you borrow or purchase your own lightweight foldable transport chair or manual wheelchair. Also, a good quality supportive seat cushion is a must to avoid pain and pressure wounds caused by prolonged sitting.

Custom Wheelchairs. A custom wheelchair will be provided by a Medicare-approved DME vendor. Your physician's office or ALS clinic team can refer you to a provider with experience in ALS to obtain the equipment. You will require a face-to-face visit with a physician. They will write the prescription for the wheelchair and place a referral to an OT or PT who will provide the specifics for the wheelchair to the DME provider. The process of receiving a custom wheelchair can take months, so it is best to plan ahead. If ALS progresses faster than anticipated and a wheelchair is needed before the custom chair is available, contact your local ALS Association chapter to inquire about equipment loan programs that may be able to provide short term DME solutions.

Figure 11: Power wheelchair for easier navigation around the home and community.

Transporting a power wheelchair is difficult unless you have access to an adapted van. The adapted van will need a raised top, side or rear entry, and either a wheelchair lift or ramp for entering and exiting. The van also requires tie downs that lock the wheelchair safely in place and match those on the wheelchair needing to be transported. While this is a costly solution,

transferring someone in and out of a car becomes increasingly difficult as the disease progresses. Having the ability to roll the wheelchair into the vehicle and eliminate a challenging transfer increases safety for all involved. Many public and private transportation systems have accessible vans available.

Home Adaptations

The OT and DME provider can perform home safety evaluations. During the evaluation, they will assess the home layout and provide recommendations for making the home more accessible and safer. These recommendations may include advice for minor adaptations or major home remodeling. Common recommendations include placing grab bars in the shower or bathtub or by the toilet, and ramps for entering and exiting the home.

Building a ramp requires adequate outside space to meet building code requirements of no more than a 1-inch downslope for every 12 inches of ramp. If there is no bedroom space on the main level of the home, a stair lift may provide a temporary method for moving up and down stairs as long as there is adequate upper body strength. Keep in mind this is a significant investment for only a temporary solution before upper body weakness occurs. Home elevators are a longer-term solution. Older homes often have narrow doorways that are not wide enough for wheelchairs; these can be widened structurally or with swing away hinges. If you have the resources, expanding a small bathroom to allow room enough for a roll-in shower and roll-over toilet is an investment that will make bathing, grooming and toileting safer and more convenient. Planning home adaptations is best done early in the disease so that any changes may be completed prior to when they are absolutely needed.

Lastly, technology options are available that enable those who are lacking the independent mobility to control their environment.

Through a smart phone, computer or tablet, one can turn on and off the lights, control the television, see who is at the door, adjust the thermostat and more. Thankfully, there are more options available now than ever before to improve independence.

Summary Statement

ALS is a progressive disease that causes growing mobility impairment and dependence on caregivers. The good news is there is an ever-expanding number of assistive devices and DME to extend independence and mobility. Partnering with a PT and OT who specialize in ALS will make for a good team as they can perform serial assessments, make recommendations and educate you and your caregivers on the use of assistive equipment and mobility devices. The goals of treatment are to improve health, safety, independence and community access. Moderate-intensity exercise and energy conservation techniques can reduce fatigue and improve the energy needed for the activities that are the most important to you and provide you with the most meaning and joy. It is critical to discuss and plan ahead with your ALS clinic team to allow the goals of treatment and optimal quality of life to be achieved.

Bibliography

Bello-Haas V.D., Florence J.M., Kloos A.D., Scheirbecker J., Lopate G., Hayes S.M., Pioro E.P., Mitsumoto H. (2007). A randomized controlled trial of resistance exercise in individuals with ALS. Neurology, 68(23): 2003-2007.

Carreras I., Yuruker S., Aytan N., Hossain L., Choi J.K., Jenkins B.G., Kowall N.W., Dedeoglu A. (2010). Moderate exercise delays the motor performance decline in a transgenic model of ALS. Brain Research, 1313:192-201. doi:10.1016/j.brainres.2009.11.051. Epub 2009 Dec 5.

Drory V.E., Goltsman E., Reznik J.G., Mosek A., Korczyn A.D. (2001). The value of muscle exercise in patients with amyotrophic lateral sclerosis. Journal of the Neurological Sciences, 191(1-2): 133-137.

Lunetta C., Lizio A.,Sansone V.A., Cellotto N.M., Maestri E., Bettinelli M., Gatti V., Melazzini M.G., Meola G., Corbo

M. (2015). Strictly monitored exercise programs reduce motor deterioration in ALS: preliminary results of a randomized controlled trial. Journal of Neurology, Oct 17, 2015. [Epub ahead of print]

Kirkinezos I.G., Hernandez D., Bradley W.G., Moraes C.T. (2003). Regular exercise is beneficial to a mouse model of amyotrophic lateral sclerosis. Annals of Neurology, 53(6): 804-807.

Mahoney D.J., Rodriguez C., Devries M., Yasuda N., Tarnopolsky M.A. (2004). Effects of high-intensity endurance exercise training in the G93A mouse model of amyotrophic lateral sclerosis. Muscle & Nerve, 29(5): 656-662.

Petrof B.J. (1998). The molecular basis of activity- induced muscle injury in Duchenne muscular dystrophy. Molecular and Cellular Biochemistry, 179 (1-2): 111-123.

Veldink J.H., Bär P.R., Joosten E.A., Otten M., Wokke J.H., van den Berg L.H. (2003). Sexual differences in onset of disease and response to exercise in a transgenic model of ALS. Neuromuscular Disorders, 13(9): 737-743.

Notes

Notes

The ALS Association
1300 Wilson Boulevard
Suite 600
Arlington, VA 22209
www.ALS.org

About The ALS Association

The ALS Association is the largest philanthropic funder of ALS research in the world. The Association funds global research collaborations, assists people with ALS and their families through its nationwide network of care and certified clinical care centers, and advocates for better public policies for people with ALS. The ALS Association is working to make ALS a livable disease while urgently searching for new treatments and a cure.