INVITED REVIEW

REHABILITATION IN AMYOTROPHIC LATERAL SCLEROSIS: WHY IT MATTERS

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ABSTRACT: Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease that results in a constellation of problematic symptoms and a high patient and caregiver burden. Multidisciplinary care includes rehabilitation interventions that have the goal of assisting people to teach their fullest potential despite the presence of a disabling disease. Given the progressive nature of ALS, the clinician must be aware of the expected disease trajectory and apply appropriate interventions at each stage. This review will present rehabilitation strategies that can be utilized to maximize patient independence, function, safety, and quality of life, and to minimize disease-related symptoms. The role of bracing, exercise, assistive devices, and adaptive equipment will be discussed. At each disease stage, an experienced rehabilitation team is well positioned to make a significant impact on the life of ALS patients.

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"Can you cure ALS?"
"I am afraid I can't."
"What else can you offer then?"
"Rehabilitation for ALS? I thought there is nothing you can do for this disease."

It is this type of conversation with patients and their families that prompted this review on the importance of rehabilitation in amyotrophic lateral sclerosis (ALS). ALS is a fatal neurodegenerative disease that produces a constellation of symptoms, including muscle weakness, wasting, fatigue, spasticity, cramps, muscle twitches, dysphagia, dysarthria, respiratory failure, and, in some patients, cognitive and mood changes. The disease typically leads to death within 3–5 years after diagnosis,[1,2] with ventilatory muscle failure as the most common cause of death.[3] The only U.S. Food and Drug Administration (FDA)–approved drug for ALS, riluzole, only confers a modest survival benefit.[4,5] Thus, most of the care of people with ALS (PALS) is supportive and centered around symptom management, making ALS an incurable, yet treatable disease.

REHABILITATION AS MULTIDISCIPLINARY CARE

Rehabilitation is the process of assisting a person to maximize function and quality of life. Although there are no curative treatments for ALS, rehabilitation can assist people to continue to function independently and safely, manage their symptoms, and, most importantly, live a fulfilling life despite having a disease that is known to shorten lifespan. Therefore, rehabilitation matters to PALS, because it enables them to reach their fullest potential despite the presence of a disabling disease. Further, it is likely that rehabilitation will become even more essential when caring for PALS in the near future, as more treatments will hopefully be developed to delay disease progression and prolong lifespan.

The ALS practice parameter of the American Academy of Neurology (AAN) currently recommends early referral of PALS to a multidisciplinary clinic.[6] The multidisciplinary clinic is the ideal setting where rehabilitation needs can be assessed and coordinated.[7] In specialized ALS clinics, the team often includes a physician, physical therapist (PT), occupational therapist (OT), speech language pathologist (SLP), respiratory therapist, nurse coordinator, and social worker.[8] Additional specialists may be available, such as nutritionists, orthotists, pulmonologists, gastroenterologists, assistive technology experts, home modification/designer experts, psychologists, and palliative-care providers. This type of comprehensive approach ideally optimizes health-care delivery by consolidating diverse skill sets, providing continuity and consistency of care, and interfacing with primary-care physicians and community-based providers.[6] ALS rehabilitation specialists therefore include physicians and allied health-care professionals who work

Abbreviations: AAC, augmentative and alternative communication; AAN, American Academy of Neurology; ADL, activities of daily living; ALS, amyotrophic lateral sclerosis; ALS-FRS, Amyotrophic Lateral Sclerosis Functional Rating Scale; AFO, ankle–foot orthosis; FRO, floor reaction orthosis; FDA, Food and Drug Administration; IADL, independent activities of daily living; KAFO, knee–ankle–foot orthosis; MBS, modified barium swallow; MCP, metacarpophalangeal; NPPV, non-invasive positive pressure ventilation; NSAID, non-steroidal anti-inflammatory drug; OT, occupational therapist; PALS, people with ALS; PEG, percutaneous endoscopic gastrostomy; PLS, posterior leaf spring; PT, physical therapist; RIG, radiologically inserted gastrostomy; SLP, speech language pathologist; SOD1, superoxide dismutase 1; TENS, transcutaneous electrical nerve stimulation

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as a team to address the multifaceted needs of PALS. Given the complexity of the disease and the variability in presentation and disease course, it is best to involve professionals who have experience with ALS, can address current problems, and can provide anticipatory guidance regarding future needs. Various studies have suggested that multidisciplinary care is associated with increased survival time; higher quality of life; and increased utilization of riluzole, non-invasive positive pressure ventilation (NPPV), feeding tubes, and adaptive equipment.[9–11]

Multidisciplinary care should ideally start early in the disease course and should be approached in a problem-oriented fashion. The goal is to focus on what the patient needs most at any particular time in the course of the disease to maintain maximum function and quality of life. Thus, it is critical to frequently reassess rehabilitation strategies and modify them according to changes in disease status. As an example, in the early stages of ALS, therapists can educate patients on energy-conservation techniques and intermittent use of braces when performing demanding activities. Later, therapists may need to modify braces or recommend additional adaptive equipment and mobility aids to adapt to evolving weakness. Rather than writing generic therapy prescriptions, it is best to address specific problems as they arise (e.g., gait training, transfer training, wheelchair evaluation, etc.). Of note, insurance carriers limit the number of therapy sessions for which people are eligible in a given period of time. Ideally, PALS patients should develop a therapeutic alliance with a therapist or group of therapists early in the disease so that they can have a point of contact for future needs. However, in-person visits need to be spaced over time. This model is very different from the rehabilitation of other diseases, such as acute orthopedic injuries, where the focus is on concentrating therapy sessions in a relatively short period of time to obtain maximal recovery. Due to the progressive nature of ALS, a completely different rehabilitation paradigm needs to be implemented and requires involvement of therapists who are familiar with the complexities of the disease.

THE GOAL OF THIS REVIEW

Other investigators have reviewed the individual roles of each rehabilitation specialist in the care of PALS. Our goal here is to address the utility of multiple rehabilitation tools and strategies in the different phases of ALS. The presenting symptoms and disease course can vary substantially. As an example, PALS with bulbar onset may have significant difficulties with communication early on, whereas they may not have problems with limb strength until later in the course of their disease. Here we use case studies to highlight how rehabilitation interventions are critical toward maximizing patient function, safety, independence, comfort, and quality of life. One should keep in mind that the specific type and timing of these interventions must be individualized to address the patient's symptoms and rate of progression.

CASE STUDY 1: EARLY-STAGE ALS

Mary is a newly diagnosed ALS patient. She is 45 years old, otherwise healthy, and has a busy life with a full-time administrative job and 2 young children. She likes to exercise regularly, drives to work every day, and often walks with her kids to soccer practice. For the last 6 months, she has been experiencing right leg and arm weakness, as well as fatigue. She tripped and fell twice on uneven terrain while playing with her kids. She is exhausted by the end of the day and has a hard time preparing dinner for her family. Cutting food and opening jars is increasingly difficult. She must ask her husband for help with buttons and zippers. She also wonders whether she can continue to exercise, an activity she enjoys, because it keeps her connected with 2 friends with whom she has been exercising for a long time.

In the early phases of ALS, patients may develop mild, but progressive, muscle weakness, fatigue, poor endurance, and performance limitation.[12] In limb-onset ALS, weakness typically begins asymmetrically in specific upper or lower body muscles.

Fall Assessment and Lower Limb Orthoses. Lower body weakness, often unilateral or bilateral foot drop, is associated with increased work of ambulation as the patient compensates with a circumducted, steppage, or hip hiking gait to avoid tripping and falling. Proximal leg weakness may also be present, making it difficult to independently perform a sit-to-stand transfer, such as when getting out of a car or a low chair. Further, muscle weakness may be compounded by spasticity. Changes in gait pattern, difficulty with transfers, and spasticity all increase energy expenditure and contribute to fatigue. Altogether, these factors produce a substantially increased risk of falls.

Rehabilitation assessment of early lower body weakness begins with a thorough initial evaluation of gait, balance, manual motor strength, range of motion, and tone to assess fall risk and optimize remaining function. Daily functional activities are reviewed to identify areas for intervention. Evaluation and modification of the home and workplace environments (such as removing carpets and installing night lights) can enhance safety and promote independence. It is important to note that, if falls occur, a resultant injury and a subsequent period

Table 1. Ankle–foot orthoses (AFOs) used most commonly in ALS care.

Type of AFO	Description	Use
Posterior leaf spring (PLS)	Medial and lateral trim lines are placed posterior to the malleoli, somewhat flexible.	Mild-to-moderate foot drop.
Carbon-fiber lateral or posterior strut dorsiflexion assist brace	Lightweight, unobtrusive.	Moderate foot drop (also helps with knee control).
Floor reaction orthoses (FROs), such as the ToeOFF braces	Built to leverage ground reaction forces to offer a "push" at toe-off to assist with propulsion and compensate for ankle plantarflexion weakness; they also create a knee extension moment to help counteract quadriceps weakness and tendency to knee buckling.	Mild-to-moderate foot drop with quadriceps weakness; they also help compensate for ankle plantarflexion weakness.
Hinged (articulated)	Include an ankle joint; allow sit-to-stand transfers more easily than solid AFOs; anti-spasticity features (such as a plantarflexion stop) can be incorporated as needed.	Moderate foot drop with or without spasticity; sufficient knee extensor strength needed for optimal use.

of immobilization can be associated with significant functional decline, making fall prevention a high priority in ALS care. Fatigue can be addressed by recommending energy conservation techniques such as pacing, taking rest breaks, and using bracing and adaptive equipment.

Lightweight braces (orthoses) and adaptive equipment can be used on an intermittent basis early in the disease course, when weakness is still mild, to help conserve energy and assist at times of demanding activities such as walking long distances or traversing uneven terrain. A variety of orthoses for the lower limbs are available. Ankle–foot orthoses (AFOs) assist people with ankle dorsiflexion weakness and are the most commonly prescribed braces for PALS (Table 1). Some AFOs can also enhance knee control. Knee–ankle–foot orthoses (KAFOs) may also provide knee stability in people with quadriceps weakness. However, they are usually too heavy and cumbersome to be effective in ALS. The involvement of physical therapists and orthotists with experience in ALS care is crucial to identify the brace that best suits the individual patient's needs. Importantly, the brace may need adjustments over time as the patient's functional status is likely to evolve. A few sessions of gait training with a skilled therapist is highly recommended to optimize braced gait. When using braces, patients should be instructed to perform skin checks every time the brace is used to identify any possible skin irritation early on and to allow appropriate brace modifications. Skin evaluation should be part of routine follow-up care. Because patient tolerability for braces is variable, their feedback on the comfort and fit of the orthosis is critical to enhance compliance.

Table 2. Adaptive equipment for ADLs/IADLs.

Activity	Examples of adaptive equipment
Meal preparation and self-feeding	Large-handled utensils, rocker knives, cutting boards, bendable utensils, universal cuffs for holding utensils, lightweight drinking cups, straw holders, long straws, reachers and grabbers, scoop dishes, plate guards, non-skid pads, mobile arm supports (devices to support the arm and assist with self-feeding, computer keyboard use, and other activities).
Dressing	Button hooks, zipper pulls, Velcro fasteners, sock aids, elastic shoelaces, long-handled shoe horns, dressing sticks.
Grooming and personal hygiene	Strap-fitted hairbrush, long-handled comb, cylindrical foam applied to the handle of multiple bath tools to facilitate grip, lightweight electric shavers, and toothbrushes, floss holders, long-handled sponges.
Reading and writing	Book easels, tilt-top overbed tables, manual or automatic page-turning devices, rubber thumbs, pen holders, pencil grips, foam cylinders placed around writing instruments to facilitate grip, writing splints, dry-erase writing boards.
Leisure activities	Many adaptations exist for activities such as gardening, golfing, and fishing. Many different types of adaptive sports are also available.

Adaptive Equipment for Activities of Daily Living and Hand Orthoses. Upper body weakness ranges from proximal weakness of shoulder muscles to distal weakness involving wrist and intrinsic hand muscles. Hand weakness interferes with activities that require fine motor skills such as grasping, gripping, and pinching.[12] Patients may have difficulties with doorknobs, buttons, and zippers, and with writing, cutting food, and opening cans and jars. A variety of tools are available to assist with activities of daily living, with the goal of prolonging independence for as long as possible (Table 2). Unfortunately, most insurance carriers do not cover the cost of these devices. Inevitable disease

Table 3. Hand orthoses (splints) used most commonly in ALS care.

Type of splint	Description	Use
Resting hand splint	Lightweight; may be used during the day and/or at night to maintain proper muscle length in patients with wrist and intrinsic hand muscle weakness.	Prevention of wrist and finger flexion contractures.
Anti-claw	Limits metacarpophalangeal (MCP) extension and improve grasp by keeping the joints flexed.	Reduction of "claw hand" deformity; improvement of grasp.
Volar cock-up	Supports the wrist in 20–30° of extension.	Improvement of grasp in people with wrist extensor weakness.
Short opponens	Keeps the thumb in an abducted and opposed position.	Improvement of grasp in people with thumb abduction and extension weakness.

progression needs to be considered when making recommendations for all equipment. Guidance from an OT with experience in ALS is crucial to ensure that the most appropriate device is selected and to avoid unnecessary expenditures on equipment that is not likely to benefit the patient. Hand orthoses (splints) may also be considered to assist with activities impacted by intrinsic hand muscle weakness (Table 3).

Exercise. PALS often inquire about exercise, because physical activity is often an integral part of the pre-morbid lifestyle for many. PALS often ask whether exercise is safe, whether it can help slow their disease, and what type of exercise is recommended for their particular condition. The types of relevant exercise include stretching and range-of-motion exercises, aerobic training, resistance/strengthening, and balance exercises (Table 4).

Stretching and range-of-motion exercises are part of the standard of care for PALS. Neuromuscular specialists should encourage simple daily stretching exercises that target the major joints starting early in the course of the disease. Experience shows that a regular, proactive stretching program helps prevent development of painful and function-limiting contractures, especially at the shoulder and ankle joints. When ambulation is no longer possible, daily standing and weight-bearing with assistance provides effective stretching of the ankles. The use of night-time splints to keep the hands and ankles in a resting, neutral position can supplement daily stretching and range-of-motion exercises.

With respect to aerobic and strengthening exercise, disease-specific guidelines are not available, as the evidence regarding benefits or harm of

Table 4. Types of exercise relevant to patients with ALS.

Exercise	Description	Benefits	Practical considerations
Flexibility	Stretching; range of motion.	Part of the standard of care for prevention and management of contractures; might also help reduce pain and spasticity.	Encourage regular stretching and range-of-motion exercises early in the course of disease. Caregiver participation is needed when muscle weakness prevents the patient from performing program independently.
Strengthening	Repeated muscle actions against resistance.	Potential role in maintaining muscle strength and delaying onset of functional impairment.	Avoid high-resistance exercise. A practical approach is to find a weight that the patient can lift comfortably 20 times. Then ask the patient to perform 2 or 3 sets of 10 repetitions each with that weight. Progression to heavier loads depends on the stage of disease. Do not exercise muscles that do not have antigravity strength. Avoid eccentric exercise.
Aerobic	Dynamic activity using large muscle groups.	Potential role in reducing deconditioning and improving functional independence, mood, sleep, spasticity, and quality of life.	Select a mode of exercise with minimal risk of injury from falling (e.g., recumbent stationary bike as opposed to treadmill). Aerobic exercise should be performed at a moderate, submaximal level. A practical approach is to begin with bouts of 10 minutes of exercise 2 or 3 times a week and progress as tolerated. If the patient cannot talk comfortably during exercise, the program is too vigorous.
Balance	Balance training using different modalities.	Potential role in fall risk reduction.	Perform under supervision of a physical therapist.

exercise in ALS is limited.[13] However, a few general exercise recommendations can be made based on preclinical data, small human studies, and research on exercise in other neuromuscular diseases. Preclinical evidence gathered in the transgenic mutant superoxide dismutase 1 (SOD1) mouse model of ALS has suggested that moderate endurance exercise can delay disease onset and increase survival.[14–16] High-intensity endurance training, on the other hand, was counterproductive in these mice.[16,17] In a study in humans, Drory and colleagues suggested that a daily program of 30 minutes of moderate-intensity aerobic exercise improves function.[18] This study included 25 PALS who were randomized to perform a moderate daily program of activities, such as walking, stationary bicycling, or swimming, as opposed to avoiding any physical activity beyond their usual daily requirements. At 3 months after study initiation, PALS who performed regular exercise showed less deterioration on the ALS Functional Rating Scale (ALS-FRS) and the Ashworth Spasticity Scale.[18] At 6 months, there was no significant difference between groups, although a trend toward less deterioration was observed in the exercising group.[18] Another 6-month, randomized, controlled trial tested moderate resistance exercise in 27 PALS. The exercise group had significantly better function, as measured by total ALS-FRS scores and quality of life, without adverse effects, as compared with subjects who received usual care.[19]

These studies suggest that moderate exercise may be safe for PALS. General practical exercise considerations are summarized in Table 4. When PALS engage in an exercise program, it is important to monitor for signs of overexertion. Excessive or prolonged post-exercise fatigue, muscle pain, or soreness are indicators that the patient is overworking. Post-exercise fatigue should not interfere with daily activities. If a patient has fatigue or pain that lasts longer than 30 minutes after exercise, the exercise program needs to be modified.

In summary, in the case of Mary, a woman with early-stage ALS described in case study 1, early referral to therapists with experience with ALS is essential to maximize function and improve quality of life. Therapy may include education on energy conservation techniques, safety, and exercise. Lower limb braces can be considered to improve gait efficiency and safety. Adaptive tools for hand weakness can promote independence with household chores and work-related tasks. Education on home and workplace modifications, community resources, and expected progression is important to allow the patient and family to plan accordingly. Unfortunately, reimbursement for therapy is often limited to a set number of sessions in a given time period. It is therefore important to carefully plan services while keeping in mind the likelihood of disease progression and future needs.

CASE STUDY 2: MIDDLE-STAGE ALS

John is a 65-year-old man who was diagnosed with ALS 3 years ago. He has managed his gait difficulties by using bilateral AFOs, a cane, and, more recently, a walker. He has trouble with transfers and is now looking into wheelchair options. Since his last visit, he has developed worsening leg stiffness and back pain. He is having a hard time swallowing and has dropped a few pounds of weight. His speech has become slower and more effortful.

In the case of John, the disease has progressed considerably. He presents with a multitude of rehabilitation needs. The combined expertise of physicians, nurses, and therapists from different disciplines is now essential to allow him to maintain mobility; address secondary complications, such as pain and discomfort; and manage progressive bulbar symptoms.

Transfers. Patients with progressive leg weakness have difficulty with transfers (e.g., from sitting to standing or from bed to chair). The inability to transfer safely and efficiently needs to be addressed promptly, as it can prevent patients from getting out of bed or leaving their homes, thus hampering their ability to participate in social events and leading to decreased quality of life. Commonly available transfer aids are described in Table 5. Patient and caregiver education and training in proper transfer techniques is essential to maximize safety and prevent potential soft tissue injury and pain for the patient. Caregiver burden during transfers is also an important consideration. Poorly performed transfers can predispose the caregiver to musculoskeletal strain and injury, which in turn has a negative effect on patient care.

Assistive Devices for Ambulation and Wheelchairs. As leg weakness progresses, PALS eventually require the use of assistive devices for ambulation, ultimately transitioning to a wheelchair. Assistive devices for ambulation include canes, crutches, and walkers (Table 6). Canes provide the least stability and are used when there is only mild lower extremity weakness or imbalance. Walkers provide the most support (Table 6). The decision as to which walking aid to prescribe, however, depends not only on the degree of weakness in the lower limbs but also on upper body and grip strength. Canes are available in a variety of styles and sizes of hand grips. Offset canes have a flat handle that can be built up to allow for better grip. Crutches have limited use in ALS, as their use requires a high degree of upper body strength,

Table 5. Transfer aids.

Equipment for transfers	Use
Firm cushions (2–3 inches thick)	Allow the hips to be higher than knees when sitting, thereby facilitating sit-to-stand transfers.
Swivel cushions	Lightweight seats that swivel in both directions (allow car transfers).
Self-powered or electric lifting cushions	Cushions that assist in rising to standing; their use requires good trunk control and balance.
Powered recliner lift chairs	Assist in rising to standing by using an electric control, can also be installed in cars and vans.
Sliding boards	Assist in low-level transfers; can be used alone if the patient has good upper extremity strength and sitting balance or with assistance from a caregiver.
Transfer belts (safety belts)	Allow caregivers to provide safe transfer and ambulation assistance; belts are positioned around the patient's waist and hips and prevent traction on the shoulders, which may be painful; they also ease caregiver burden and potential musculoskeletal strain when assisting the patient. Belts with handles are often preferred.
Stairway lifts and chair glides	For multilevel homes when patients cannot negotiate stars safely; they are costly and require an extra wheelchair at each level of the staircase; patient may require assistance to transfer.
Mechanical lifts	Allow for safe lifting of people who do not have the preserved strength to transfer independently; they may be electric or hydraulic; they are attached to a sling system with head support.
Tools for bed mobility	Facilitate repositioning while in bed and getting in and out of bed (e.g., step stools, bed canes, hospital beds, and overhead trapezes). Electric hospital beds allow adjustment of total bed height in addition to head and knee adjustments.

coordination, and energy. Wheeled walkers, as opposed to standard walkers, do not need to be lifted and are preferred in ALS. However, one must ensure that the patient can maneuver them safely. If a 4-wheeled walker is used, the patient needs to be able to activate the brake system. Most 4-wheeled walkers are equipped with squeeze brakes, which may be hard to use if the patient has a weak grip. Alternatively, one can choose a 4-wheeled walker with push-down brakes (secured when the patient loads his or her weight on the walker) or a 2-wheeled walker with glides in the back.

Transitioning to a wheelchair is ultimately needed in ALS and may be challenging from both a psychological and logistical point of view (Table 7). Manual wheelchairs may be used in the early stages to help conserve energy when traveling long distances. However, one should keep in mind that, as the disease progresses, the patient will probably develop difficulty propelling the wheelchair in a relatively short period of time. Of note, at present, Medicare and most private insurers limit reimbursement to only 1 wheelchair every few years. Therefore, one may wish to rent or borrow a manual wheelchair instead of purchasing one. Power scooters are sometimes considered as an alternative to a manual wheelchair, especially for outdoor use and energy conservation when one needs to traverse longer distances. However, scooters should be recommended with caution, because reimbursement for a scooter will preclude reimbursement for a power wheelchair. This is an important financial consideration, as most PALS will ultimately need a power wheelchair. Power scooters may be an option for those who can borrow or afford to purchase one (most scooters are available for $700–$2000 depending on the model). Some patient organizations maintain loaner closets that allow patients to borrow pieces of equipment. However, other important limitations regarding the use of scooters should be discussed with ALS patients. Good upper limb and trunk strength is needed to drive them. In addition, they cannot be modified for disease progression or to accommodate other equipment such as mounted trays and electronic equipment. Finally, because of their long wheelbase and wide turning radius, there may be difficulty using them indoors.

As mobility continues to decline, most patients will need a power wheelchair. The design and prescription of a power wheelchair should be coordinated with a physical therapist who is experienced in ALS to ensure that the appropriate components are selected to accommodate current needs and anticipate future modifications (Table 7). Proper seating and positioning are essential to ensure a comfortable and functional sitting posture, allow for maximal breathing capacity, and prevent secondary injuries, such as skin breakdown and back pain. Transitioning to power mobility may require a number of corresponding environmental modifications, because power wheelchairs do not fit into narrow doorways and standard cars. Ramps are needed to enter the home if there are steps, and a modified van is needed for transportation. Many individuals do not have the financial means to purchase a modified van and thus utilize a back-up manual wheelchair for travel or use accessible public transportation, if available in their community.

Table 6. Assistive devices for ambulation (mobility aids).

Device	Type	Practical considerations
Canes	• Wooden (standard, affordable). • Aluminum (light, adjustable length). • Offset (flat handle, easier to grip). • Quad (provide greater stability, but heavy to lift and therefore rarely used in ALS).	Patients must have adequate upper body and arm strength to use a cane safely. Canes are typically used on the side opposite to the affected leg. They can be used on stairs. Patients may need to negotiate stairs on an angle and 1 step at a time. One should lead with the stronger limb on flat ground and when ascending stairs, and with the more affected limb when descending stairs ("up with the good and down with the bad").
Crutches	• Axillary (rarely used in ALS). • Forearm (Canadian, Lofstrand; limited use in those with mainly leg weakness).	Limited use in ALS because they are heavy, and their use requires preserved arm and trunk strength along with proper balance.
Walkers	• Standard. • Wheeled (2 or 4 wheels).	Standard walkers are rarely used in ALS because they lack wheels and must be picked up and lowered to advance forward, causing fatigue. Wheeled walkers do not need to be lifted and are preferred in ALS as long as the patient can maneuver them safely. Four-wheeled walkers should be equipped with brakes for safety. They may have an additional attached seat, which the patient can use when fatigued. If the patient cannot use the grip brakes due to hand weakness, 2-wheeled walkers with glides in the back can be used.

Having a back-up manual wheelchair is also important in case the power chair needs repairs or upgrades, which is a common occurrence.

Musculoskeletal Symptoms. Although progressive muscle weakness in the absence of pain or sensory disturbance is a typical symptom of ALS, over time patients can develop secondary musculoskeletal pain and discomfort due to poor mobility, suboptimal transfer techniques, spasticity, loss of range of motion, difficulty with positioning in bed or in the wheelchair, and contractures. A careful history combined with detailed neurologic and musculoskeletal exams can help identify the pain generator(s) and direct treatment. It should be noted, however, that recommendations for treatment of pain and spasticity in ALS are based mostly on expert opinion, as very few studies have been conducted to analyze the efficacy of specific medications or other interventions.[20–22]

Table 7. Wheelchairs.

Type	Practical considerations
Manual	Manual wheelchairs should be lightweight or ultra-lightweight. With disease progression, patients have a hard time propelling the wheelchair. Consider renting or borrowing manual wheelchairs instead of purchasing one. Removable wheels make it easier to transport wheelchair in a small vehicle.
Transport (companion)	Transport wheelchairs are light and inexpensive, but must be pushed by a caregiver. They fold and fit into a car's trunk. Many families borrow or buy one as a back-up chair for travel.
Power	Power wheelchairs do not fit into standard cars (modified van is needed for transportation). Ramps are required to enter homes. Wheelchairs can be equipped with ventilators and augmentative and alternative communication devices. Tilt-in-space mechanism is used for pressure and pain relief. Multiple drive controls are available to allow users with different degrees of weakness to control the power wheelchair (e.g., joystick, head array, eye gaze).

For all wheelchairs, positioning and modifications with disease progression are essential to ensure proper seating and avoid secondary complications such as skin breakdown and back pain.

Pain is reported by some PALS even in the early stages of their disease.[23] Common sites of pain include the low back, the neck, and the shoulder area.[24] Loss of range of motion, subluxation, and contractures in the shoulders are particularly frequent complications of ALS, and yet they are probably preventable with the institution of a regular stretching and range-of-motion program early in the course of the disease. The use of shoulder approximation sleeves can help minimize shoulder subluxation and pain. Simple modifications, such as adjusting the seating system or arm rests of the wheelchair, provide additional comfort and pain relief. In the upper limbs, patients may complain of hand pain due to finger contractures or finger flexor tenosynovitis (the latter being responsive to local steroid injections). Resting splints may be used to allow the hands to rest in a neutral position and can be used throughout the day or at night.

In the legs, discomfort can be associated with spasticity, dependent edema, loss of range of

motion, and contractures. If dependent edema is a problem, leg elevation, massage, and compression stockings may provide some relief. Gentle sustained muscle stretches, weight-bearing with assistance, and night-time neutral position splinting may be used for spasticity at the ankles. In addition, one can use dynamic splints, which provide a low-load, prolonged-duration stretch to prevent ankle contracture development. In wheelchair users, back pain can often be relieved by providing adequate lumbar support and good cushioning. Some features of power wheelchairs can help with pain management. Power-activated leg rests can help maintain hamstring length and ease back pain. Tilting the wheelchair relieves pain from gluteal pressure. For those with advanced weakness, the bed should be fitted with pressure relief over bony prominences to avoid pain and pressure ulcers. Foam wedges can be used to facilitate proper positioning.

Pharmacologic treatment of pain and spasticity is indicated in some patients, although evidence supporting one regimen versus another is lacking.[21] Depending on the pain generator, one can use medications for neuropathic pain (such as gabapentin, pregabalin, tricyclic antidepressants, duloxetine, and topical lidocaine or capsaicin) or non-steroidal anti-inflammatory medications (NSAIDs) if there is evidence of an inflammatory process such as arthritis or bursitis. As-needed or standing doses of acetaminophen may augment or, if there are contraindications, replace the use of NSAIDs. Oral medications for spasticity include baclofen, tizanidine, benzodiazepines, and cannabinoids. As an adjunct to medications, strategies to alleviate pain and spasticity include stretching, bracing, massage, topical heat and ice (given alone or sequentially, as in contrast therapy), transcutaneous electrical nerve stimulation (TENS), acupuncture, ultrasound, and iontophoresis.[25–28]

Dysphagia. Oropharyngeal and tongue weakness produce chewing and swallowing difficulties (dysphagia), along with sialorrhea, due to reduced ability to swallow saliva. Symptoms of dysphagia include coughing during or after meals, episodes of choking, frequent throat clearing, wet vocal quality (suggesting pooling of secretions), and feeling that food is "stuck" in the throat. Importantly, dysphagia can lead to aspiration and malnutrition, a negative prognostic factor in ALS.[29,30] Assessment and management of dysphagia are generally conducted in collaboration with speech and language pathologists (SLPs). The development of dysphagia is expected in ALS, and bedside assessment of swallowing is generally sufficient. In some cases, a modified barium swallow (MBS) can be offered to document penetration and/or aspiration and to help educate patients on swallowing strategies.

From a rehabilitation perspective, there is no evidence that exercise improves swallowing function. Compensatory techniques, however, can be used to reduce aspiration risks, including prolonged chewing time, head tilt, and chin tuck. Behavioral strategies include taking small bites and sips, alternating bites of solid food with sips of liquid, sitting upright, avoiding talking while eating, and paying increased attention to each individual swallow. These recommendations, however, need to be strictly individualized, as there is no prescribed regimen that is appropriate for all patients with dysphagia. Education about types of food and liquids that are easier to chew and swallow is essential. As an example, soft, moist foods are easier to swallow than dry, crumbly, or chewy foods. Thicker liquids (fruit nectar, smoothies) are easier to handle than water. A modified diet might be recommended, ranging from soft (tender foods) to puréed (blenderized foods).

Dieticians can help monitor a patient's caloric intake and recommend additional supplements to maintain body weight. Strategies to help avoid weight loss include eating smaller meals with high-calorie snacks at scheduled times, choosing calorie-dense foods, and adding high-calorie supplements. When oral intake becomes inadequate, too effortful or fatiguing, and/or compromises safety, alternative routes for nutrition may be considered, including percutaneous endoscopic gastrostomy (PEG) or radiologically inserted gastrostomy (RIG) tubes. The risk of gastrostomy tube placement increases when functional vital capacity falls to $<50\%$, and PEG/RIG use is probably effective in prolonging survival.[31] Therefore, early intervention is suggested, although there is no evidence to support specific timing of gastrostomy tube insertion in ALS.[31] Importantly, placement of a gastrostomy tube does not necessarily imply elimination of all oral feeding, but it offers a convenient method for administering medications, fluids, and extra calories with the goal of stabilizing body weight.[31]

Dysarthria. Dysarthria can manifest either as a presenting symptom of ALS or as an associated feature when disease progresses to involve bulbar muscles. ALS patients often exhibit a reduced speaking rate before the loss of intelligibility.[32,33] Dysarthria can be spastic, flaccid, or mixed, depending on the degree of upper vs. lower motor neuron involvement.[34] Patients often ask whether oral motor exercises can improve their ability to communicate. Unfortunately, these exercises are not likely to help. However, speech therapy can

assist patients in developing compensatory speech techniques to improve communication. The first steps to make communication as easy and effortless as possible include reducing ambient noise, minimizing the distance between the patient and listeners, speaking face-to-face in a well-lit room, slowing the speaking rate, overarticulating, and repeating or spelling-out words. Taking naps or rest breaks before anticipating having a long conversation can allow a patient to maximize speaking endurance. It is also important to develop personalized communication strategies between patient and caregivers, such as a system for confirming understanding.

As dysarthria progresses, augmentative and alternative communication (AAC) devices may be needed. Low-technology options include communication boards with manual writing and letter/word/picture boards. Portable voice amplifiers allow the amplification of diminished voice volume, which is relevant to ALS patients, as concurrent weakness of the breathing muscles impairs the ability to develop sufficient respiratory support to speak loudly. Message banking is a commonly utilized technology in ALS. Patients record words and phrases while still intelligible and play them back when they are no longer able to speak. These messages can be added to a high-technology system that has voice output capability. These systems include computerized voice synthesizers on personal computers and tablets. Selection of information on personal computers/high-technology AAC devices can be manual, by eye gaze, or by head-movement-tracking technology. More recently, tablets such as the iPad have provided easy-to-use and affordable applications for speech production. These applications utilize text-to-speech technology that allows patients to type or select words on the screen that are then spoken aloud in a high-quality synthesized voice. Typing or word selection on iPads, however, cannot be accomplished by eye gaze or head-movement-tracking technology at the present time, requiring the use of other AAC devices when patients can no longer use their hands.

CASE STUDY 3: ADVANCED ALS

Peter is a 55-year-old man with end-stage ALS. He is tetraplegic and anarthric, but can communicate by using a computer that he controls by eye gaze. He receives all his nutrition and hydration through a gastrostomy tube. He is using non-invasive positive pressure ventilation. He has decided against placement of tracheostomy and long-term mechanical ventilation. He is cared for at home by his family under the guidance of an expert hospice team.

The natural history of ALS leads to profound generalized muscle weakness and ultimately death, which most commonly occurs due to ventilatory muscle failure. Patients with advanced disease face heavy physical and emotional demands. Continued engagement with the multidisciplinary team is critical to assist patients and their caregivers in coping with worsening functional status and for making informed decisions about the goals of care. Home visits are often needed to decrease the burden of travel to ALS clinics, and telemedicine is now being explored as another means of providing patient monitoring.[35] Early involvement of palliative-care experts as part of a multidisciplinary team approach helps integrate care across the continuum of the illness and can help facilitate appropriate and timely utilization of hospice services.[36,37] Active management of symptoms (especially pain and dyspnea) and a proactive effort to avoid complications of immobility (such as skin breakdown and contractures) are crucial to optimize quality of life in the late stages of the disease. Finally, caregiver burden and education must be addressed by the multidisciplinary team, particularly as the patient experiences progressive physical and behavioral decline.[38,39]

FUTURE DIRECTIONS

Given the complex range of symptoms related to ALS that benefit from multidisciplinary care, it is important to conduct research to define the most effective rehabilitation modalities for this patient population. A promising example of research in this direction is the ongoing randomized clinical trial of exercise in ALS (NCT01521728). In this trial, 60 participants with ALS are being randomized to 6 months of endurance vs. resistance vs. stretching/range-of-motion exercise. The goals of the study are to establish the safety and tolerability of these exercise programs and to obtain a preliminary assessment of their effect on function. It would also be important to conduct controlled studies to define the impact of other commonly utilized rehabilitation interventions, such as bracing, on patient safety and independence.

Finally, there is an urgent need to collect patient-centered outcome data to document the impact of rehabilitation on the ability to perform desired activities despite the expected progressive decline in muscle strength. The traditional reimbursement model for rehabilitation services is based on the principle of "improvement." In this model, rehabilitation services are reimbursed only if there is an expectation for the person to "improve" as a result of the intervention. However, improvement is generally measured by the amount of change in impairments such as strength. This paradigm does not fit with a progressive disease such as ALS, as improvement in strength is not expected. Thus, reimbursement for ALS rehabilitation services is limited. However, it is common experience that

rehabilitation interventions can have a tremendous effect at the level of performance of certain activities despite declining strength. As an example, prescription of assistive devices and training on proper transfer techniques with appropriate modifications as the disease progresses may allow patients to safely and efficiently get out of bed, leave their home, work, and be active in their communities. Collecting prospective patient-centered outcome data may help document the positive effect of rehabilitation on activity limitations and participation restrictions, thus providing evidence on the importance of rehabilitation in ALS.

CONCLUSION

ALS is a progressive, fatal disease, and PALS have multiple complex, evolving needs. At each stage of the disease, an experienced rehabilitation team is well positioned to make a significant impact on the life of PALS by maximizing safe mobility and independence, minimizing disease-related symptoms, facilitating communication and informed decision making, and enhancing quality of life.

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