Amyotrophic Lateral Sclerosis (ALS): also known as Lou Gehrig’s disease, is a rapidly progressive disorder which affects voluntary muscle control in all areas of the body. Initial symptoms can be seen in the arms, legs and/or muscles used for speech, swallowing and breathing. Clients may have little control over their arms and yet walk well, or have shortness of breath without significant weakness in the extremities. Hand and ankle weakness may be more or less pronounced than shoulder and hip weakness. Because clients with ALS experience a variety of progressions, the specific timing and types of complex rehab equipment required will also vary.

One of the hallmarks of seating and wheeled mobility for the person with ALS is flexibility for long-term needs and changes. No matter how and where in the body the disease is first manifested, the seating specialist and supplier must consider how easily components can be added or changed in the future as the condition progresses. This may include seating and positioning components, alternative drive control options and an attendant control.
“REGULAR PATIENT AND CAREGIVER EDUCATION AT EVERY ALS MULTIDISCIPLINARY CLINIC VISIT MAY ASSIST WITH INCREASING POWER FEATURE USE OVER TIME, THEREFORE INCREASING COMFORT.”

The evaluation process often begins when ambulation becomes slower and non-functional at times, the client has started using a walker, or fatigue and shortness of breath are a factor. Some clients do not want to give up walking, and it is up to us to provide education on the benefits of a power wheelchair for positioning, fatigue and strength management, as well as overall function and safety. Can the client complete their MRADLs in a safe and timely manner? Has the client fallen? Can the client get where they need and want to go in and out of the home?

If the client, therapist or supplier waits too long, the client may be completely non-functional or falling with serious injuries before the wheelchair arrives due to the slow process of funding a wheelchair through insurance. If needed, the Muscular Dystrophy Association (MDA) and ALS Association (ALSA) have loan closets, which often have power wheelchairs for use while a client is waiting for permanent equipment. Being proactive means planning that the right wheelchair will be available at the right time. For example, at delivery, and for the first few months, the client may still be able to stand at times for pressure relief and may not get in the habit of using power seating for this purpose. Therapists and suppliers need to continue to provide education and reminders as each client progresses on the use of power seating for positioning, pressure relief and edema management. Ward, et. al. 2015 noted, “regular patient and caregiver education at every ALS multidisciplinary clinic visit may assist with increasing power feature use over time, therefore increasing comfort.”

For a client with ALS, a group 3 complex rehab power wheelchair should always be considered when medically appropriate for increased flexibility with long-term needs. Generally, this level of wheelchair can meet the long-term needs of a client with ALS through the lifespan. The mat evaluation is typically straightforward, since clients were often fit and healthy before diagnosis with few comorbidities. Clients typically have full range of motion, with few spinal or pelvic asymmetries. Since clients often have no prior experience with power wheelchairs, they often need to test drive various power wheelchairs, sit on multiple cushions and rely on the evaluation team for direction in product choice. At the bare minimum, the following should be included on the power wheelchair:

- Upgraded electronics that allow control of power features through the driving method and support a variety of driving methods
- Swing away joystick
- Power seating including tilt, recline, elevating leg rests
- Positioning and pressure relieving cushion
- Adjustable and removable head support
- Contoured back support
- Height adjustable armrests
- Positioning components such as lateral thigh supports, lateral trunk supports and elbow blocks, as required

Many other options can and should be considered on the power wheelchair as well such as adjustable seat height and anterior tilt; head support components such as lateral facial pads or dynamic forehead straps; alternative driving methods such as a compact joystick with light throw, mini proportional joystick, foot drive or head array; cup holder; invertor to provide power for ventilator or BiPap from the wheelchair batteries; backpack hooks; gel armrest or footplate pads; and mobile

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ARM supports for self-feeding and upper extremity support. Several manufacturers offer ALS drive control packages, which are billed once through insurance and then, as the client changes, a new driving method can be provided without additional funding approval. This may allow the client to continue driving and use other wheelchair functions over the course of their disease.

Many clients with ALS are seen at an ALSA or MDA/ALS clinic every three months, which provides a chance to check in on needs and progression in relation to the power wheelchair. Almost every client in a power wheelchair requires changes at every clinic visit. Changes might be as simple as moving an armrest pad forward or adjusting the throw on the joystick, or as complex as changing from driving with the hand to driving with the head. Often parts are used from the loan closet, if available, to meet the immediate need while equipment is ordered through insurance. If the supplier is working with a client who is not going to an ALS clinic for care, frequent follow-up with that client for medically necessary changes and upgrades will be needed.

Although each client is unique, certain equipment changes are commonly required with this population. The first is the cushion. Many clients who are still transferring and standing at the evaluation require a firmer cushion for transfers and often chose a foam or hybrid cushion. As the client progresses and becomes dependent for transfers, comfort and pain management become more of a priority. At some point, nearly all clients switch to a pressure relieving cushion, such as air or other off-loading cushions.

Another common change is the amount of postural support the client requires. Many clients have fair to good trunk strength early on and may not require lateral trunk or pelvic support or a positioning backrest at evaluation. Many times, as the client weakens, these items must be added for balance and support. A positioning backrest will be required with more contour, lumbar support and/or ability to capture the pelvis. Most power wheelchairs at this level will support numerous backrest options.

A third common change is the driving method and control of power features. Many clients begin by using a standard hand controlled joystick or a foot controlled joystick (due to significant arm/hand weakness). Most clients are able to use buttons on a standard joystick to control power seating, but will eventually need switches for power and mode. Programming provides adjustments to sensitivity and throw of the joystick to accommodate increasing weakness. Positioning changes of the joystick itself may provide a mechanical advantage. As the client loses the ability to drive with these methods, the next option will depend on the where the client has volitional movement. Mini-proportional joysticks, which require less activation force and distance, can be placed by fingers or the chin. If the client is using a larger movement, for example from the shoulder, a compact light throw joystick may work better. Switches can be placed at the fingers, head, lips, chin, medial and lateral knees or even heels. Some clients will require single switch scanning, if only one switch site can be identified.

Sip and puff control often is not appropriate for this population as a drive control, as it relies on control of air pressure within the mouth, rather than breath control. To manage air pressure within the mouth, the client must have good lip closure and a competent soft palate. Many clients with ALS have difficulty with lip closure due to muscle weakness and lose competency of the soft palate due to bulbar involvement. Some clients will be able to use sip and puff for a mode switch rather than for driving (See picture 1). Head arrays can also become challenging as the client loses trunk and head control and may rely more on recline. The neck may weaken more on one side than the other and the head array must have options to compensate for this. For example, one head array has both force and proximity options built into the pad, allowing the head to rest on the pads for support. Another option for drive control is a sensor worn on a headband or ball cap, which moves the chair as the client moves their head. Because the sensor moves with the client, the problems with slouching and moving away from the head array mounted to the chair are not issues with this style (See picture 2). Successful control has also been found with four proximity sensor switches mounted to each side of both knees, as many clients with ALS retain the ability to move their legs slightly side to side until late in the disease. Eventually, driving with any method becomes difficult, and the client may use single or dual switches for control of power seating while the caregiver uses the attendant control to drive the chair.
The client may also need to use other assistive technology, such as a communication device or computer. As muscle weakness progresses, the client may have limited switch sites for control of multiple assistive technologies. The driving method may need to also be used to control other technologies through interfacing.

For a client with ALS, the power wheelchair becomes forefront in importance for comfort and functionality. The client will typically be in the wheelchair all day, including naps and often at night when the bed becomes too restrictive or uncomfortable. The ability to continually change position through power seating may assist with pain management, circulation, edema, range of motion and positioning. Ward, et.al. 2015 sent out surveys to respondents at one month and six months after wheelchair delivery – “at the one- and six-month follow-ups, when asked a question about how the power wheelchair affected their quality of life, 89 percent and 90 percent respectively noted an improvement in their quality of life because of the power wheelchair.” It is amazing that the power wheelchair assists in improving quality of life even as the disease takes away strength, and means that our job is all the more important.

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REFERENCES:

RESOURCES
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