

ALS: ONGOING INTERVENTION

Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disease that affects nerve cells in the brain and spinal cord. Voluntary muscle action deteriorates due to death of motor neurons. The incidence of ALS is two per 100,000 people, and it is estimated that as many as 30,000 Americans may have the disease at any given time. The life expectancy of a person with ALS averages about two to five years from the time of diagnosis, but this is widely variable. Most people who develop ALS are between the ages of 40 and 70, but younger people can also be affected. Symptoms of ALS may include muscle weakness in the upper and/or lower extremities, decreased ability to speak, swallow or breathe, and twitching and cramping of muscles. In addition, there has been recent increasing awareness of the combination of Frontal Temporal Lobe Dementia (FTLD) with ALS. Studies suggest that five to 15 percent of ALS patients may have FTLD, and as many as half may have mild-to-moderate cognitive or behavioral abnormalities. This can result in personality changes, language difficulties, and decreased judgment and insight. These deficits may not be apparent when talking to a patient with ALS, but it is important to realize that, if present, impaired judgment and lack of insight can affect wheeled mobility skills.

Wake Forest Baptist Health provides care to ALS patients utilizing a multidisciplinary team approach. The clinic is held for two days each month and is staffed by neurologists, nurses, a clinical social worker, dietitian, occupational therapist (OT), physical therapist (PT), assistive technologist, respiratory therapist, speech language pathologist, research coordinator, neuropsychologist, durable medical equipment (DME) provider, and representative of the North Carolina ALS Association. Newly diagnosed patients are encouraged to see all disciplines during their visit. Returning patients see the clinicians who are appropriate for their individual needs. Patients typically spend two to four hours at the clinic, and return to the clinic every three months. I serve as the physical therapist for the regular outpatient wheelchair seating clinic as well as for the ALS clinic, and I have found ALS patients benefit from a separate model for seating and mobility needs.

When seating a person with ALS there are unique challenges. The need for a chair may arise quickly, making longer equipment trials impossible. Many patients are walking some when they are evaluated for a wheelchair, but a fall, foot drop or increasing fatigue may lead to the suggestion that a person use a wheelchair for mobility. The patient may not have considered their options about equipment, and thus

they typically rely heavily on the recommendations of the physical therapist and the DME provider. As significant as mobility choices are, it is important to note that these patients and their families are often called upon to render many other possibly life altering decisions regarding respiratory support, feeding support, communication needs and end of life decisions during the same clinic visit. In addition, fatigue can be a significant part of the ALS disease process. This clinic serves a very specific population and draws patients from locations quite a distance away (even from neighboring states), impacting delivery. To get the equipment as quickly as possible, patients usually elect to make all choices for their equipment during their regular ALS clinic visit.

People with ALS may have frequently changing needs, thus it is helpful to think long-term when setting up the chair. As people with ALS become weaker throughout their trunk, there is a tendency to sacral sit. We plan for this eventuality by making certain the seat is long enough to accommodate sacral seating. This is coupled with a pressure relieving, positioning cushion in order to try and prevent pressure issues. Many of our patients stay in the chair most of the day as they weaken and transfers become more difficult. It is imperative their cushion

and back support are comfortable, distribute pressure adequately, and are durable. We have found multiple seat functions, including tilt, recline and power operated legrests are essential in helping patients obtain a balance between trunk support, comfort and breathing ability. A seat elevator can either help maintain independence with transfers or make assisted transfers easier, but funding limitations can be an obstacle. We work with the multidisciplinary team to coordinate funding options through insurance, loaner equipment and grants.

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to ask questions and explore possible future equipment needs. At this early stage, patients are still adapting to the diagnosis. Many do not want further information regarding wheeled mobility. At some point, the patient, physician or therapist recommends going forward with a wheelchair. The occupational or physical therapist, patient, caregivers and DME provider meet during the clinic visit. The patient sees the physician the same day, and the face-to-face visit and need for power mobility are documented. The therapist completes the written evaluation and the documentation required for insurance funding. Patients are given the option to try multiple bases both in the clinic and in their home environment, but most choose to limit trials.

In addition, patients are educated that they are free to choose any DME provider they wish. In the past we have had multiple providers at the clinic, but because of the number of choices each patient must make in regard to the chair, as well as other aspects of care, we now have one DME provider at the clinic who takes care of the majority of our ALS patient's equipment needs. This provider is also present at future clinics to provide adjustments, maintenance and recommendations as needs change. In addition, this provider is contracted through the state ALS Association to deliver and pick up loaner equipment. This coordination has worked best in our clinic. During the clinic visit, patients try seats and bases to determine best fit and function. From measurements and trial with the recommended equipment, an order form is completed with collaboration between the DME provider and therapist, along with the patient and family. The DME provider obtains a copy of the PT or OT equipment evaluation with embedded letter of medical necessity, the physician notes from the visit, and a completed detailed prescription within two to three days of the clinic. Unless prior authorization is required from the insurance company, the equipment is ordered at this time. In addition, a loaner power wheelchair as close to the patient's prescribed equipment is provided if appropriate and requested. The DME provider typically delivers the equipment within three weeks from the clinic date. The therapist and provider then see the patient during their next clinic visit for adjustments and changes. This allows the patient to receive the equipment as soon as possible thus allowing them to take full advantage of the equipment and hopefully prevent falls. The DME provider is also available to make adjustments in the patients' homes on an ongoing basis, especially if the patient is no longer able to travel to clinic appointments.

Mr. S. is a 62-year-old male, who lives in a one level home with his wife and 19-year-old developmentally disabled son. He had symptoms of weakness in his right hand and voice difficulties beginning in 2009, and he was diagnosed with ALS in June 2010. He has been seen approximately every three months in ALS clinic since that time. I saw him mainly to check in during clinic visits and he reported hiking up to four miles, playing racquetball with his left hand, and exercising at the YMCA. In June of 2012, he developed mild right foot drop and was prescribed an ankle foot orthosis. In September 2012, he reported a fall and was prescribed a rollator walker. At this time he had very limited grip in his right hand, but was able to place his hand on the walker and maintain contact. OT noted increasing weakness in his left hand and upper extremity.

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He returned to the clinic in November 2012 with an increased number of falls, and his right hand tied onto the walker. In addition, the patient had very limited communication. He had received an iPad for communication, but used it very little and did not wish to explore other options. He and his family requested an evaluation for a power wheelchair.

The wheelchair evaluation during clinic was fairly straightforward (*Figure 1*). The patient, his wife and several out of town family members were present, as well as the DME provider. We offered Mr. S. the opportunity to try different bases at his home, but after trying the Permobil M300 in the clinic he elected to forgo further trials. The team did not anticipate any issues with accessibility in his single level home. Mr. S was able to position his left hand on a standard joystick and demonstrated safe driving (*Figure 2*). We considered starting with alternative controls, but decided to stay with the simpler joystick set-up, knowing changes may be made if necessary.

Expandable electronics were ordered to allow alternative access in the future as well as control of power seating through the joystick. Mr. S. tried several cushions and was most comfortable with the Comfort Company Maxx. We included power tilt, recline, power elevating legrests, retractable joystick, arm trough for the right side, and tray. Wheelchair specifications were made based on a combination of measurements and fitting in a demonstration Permobil M300. In addition, a seat elevator was provided through special funding. Mr. S. performed stand pivot transfers, which were somewhat difficult due to the lack of

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strength in both his arms and legs. The seat elevator made transfers independent.

Patient's wife was concerned about funding for a wheelchair lift for the back of her car. She was given information on local companies to contact. I completed the letter of medical necessity, and the clinic physician signed it that day. This paperwork and the face-to-face visit from the clinic physician allowed the DME provider to order the chair immediately after clinic.



During our post clinic meeting, Mr. S's condition was reviewed, and there was some discussion about his need for immediate hospice care due to his declining respiratory function. However, the general feeling during the clinic visit was that there were several items, including the wheelchair, that were needed for the patient, so he was offered palliative care. The family declined these services. Shortly after clinic the DME provider was called by the hospice physician. The patient had declined further, and the patient and wife wanted him enrolled in hospice care as soon as the wheelchair was delivered. The power wheelchair was delivered to the patient's home three weeks after he was seen in ALS clinic. A Jay Union cushion was provided instead of the Comfort Company Maxx, most likely due to last minute confusion in order to get the wheelchair delivered as soon as possible. The provider made adjustments and emailed me pictures (*Figure 3*).

Mr. S. was next seen in ALS clinic in March 2013. I had hoped to observe the patient in his wheelchair, but he was unable to transport it to clinic. Mr. S. and his wife reported that he was able to drive his wheelchair throughout his home and operate the seat functions independently. He reported he was comfortable in the chair and used it for rest as well as for mobility. He did not request any changes to the chair or the seating system. I will continue to follow the patient in future ALS clinic appointments, and if his chair needs to be brought to the appointment, the DME provider will transport the chair. ➤



FIGURE 1.
DISCUSSING
SHOULDER
HEIGHT

FIGURE 2.
TRIAL OF
STANDARD
JOYSTICK

FIGURE 3.
FITTING
OF THE
WHEELCHAIR
AT HOME

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HELPFUL RESOURCES

The ALS association webpage - <http://www.alsa.org>