Optimizing Access to Care in ALS

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Introduction

ALS is a is a progressive, currently incurable neurodegenerative disorder that causes muscle weakness, disability, and eventually death. ALS is a rare disease, affecting nearly 3 to 7 people in 100,000 per year in Europe and North America.[1] ALS mainly affects the elderly, though it can affect adults at any age (Figure 1). Most ALS patients die within three to five years of diagnosis; 30% of ALS patients are alive five years after diagnosis, and 10%-20% survive for more than 10 years.[2]

While there is currently no cure, a well-orchestrated, multidisciplinary treatment plan can improve quality of life and increase survival. The integrated multidisciplinary team will include a neurologist, pulmonologist, speech therapist, assistive technology specialist, physical and occupational therapists, and psychosocial support.[3]

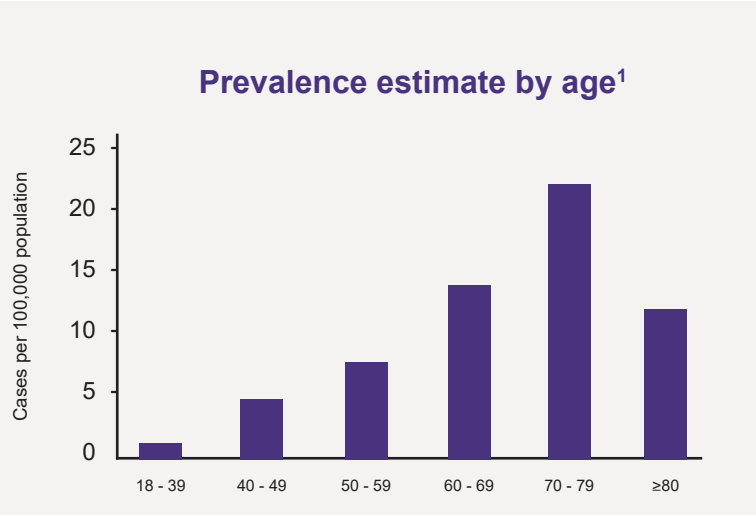

Figure 1: While ALS prevalence is highest among individuals older than 60, it can affect adults at any age[1]

Optimal care for ALS can be hindered by structural, financial, and institutional barriers. Many clinicians do not have direct experience treating patients with ALS and may not be aware of available resources or evidence-based supportive therapy. Experienced ALS centers have developed strategies to overcome barriers to ensure access to treatments and equipment. This includes philanthropic resources, strategic utilization of insurance benefits, implementation of patient-centered evidence-based care, and appropriate clinical documentation that ensures insurance reimbursement.

The cases presented in this case series illustrate three key phases of the ALS patient journey and the clinical decisions at each phase.

- Case 1: Newly Diagnosed
- Case 2: Respiratory Presentation with Bulbar and Upper Extremity Weakness
- **Case 3: Respiratory Disease with End-of-Life Care**

The clinical decisions at the end stages of disease are characterized by ensuring access to home modifications and specialized equipment that can improve quality of life in the last months. An important consideration is the timing of referral to hospice, which can have implication on billing and access to equipment.

1. Mehta P, Kaye W, Raymond J, et al. Prevalence of Amyotrophic Lateral Sclerosis - United States, 2014. MMWR Morb Mortal Wkly Rep. 2018;67(7):216-218.
2. Kiernan MC, Vucic S, Cheah BC, et al. Amyotrophic lateral sclerosis. Lancet. 2011;377(9769):942-955.
3. Traynor BJ, Alexander M, Corr B, Frost E, Hardiman O. Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: A population based study, 1996-2000. J Neurol Neurosurg Psychiatry. 2003;74(9):1258-1261.

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Patient #3: Respiratory Disease With End-of-Life Care

56-year-old white male

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Symptom Presentation & Initial Care Management

- Presenting complaint: Weakness of right index finger and thumb
- Diagnosed: Nov. 2015
- Riluzole: Dec. 2015
- FVC 5.19L/5.33L = 97% predicted: Dec. 2015
- Voice/message banking: Dec. 2016
- Home safety evaluation & physical therapy: June 17
  - Right AFO & manual wheelchair
  - Home construction & job telecommute
- Radicava/PICC: Sept. 2017
- FVC 1.60L/5.33L = 30% predicted: June 2018
  - Terminated Radicava because of disease progression

ALS Clinic Visit

- Discussed and recommended PEG tube placement[2]
- Discussed and recommended BiPAP, cough assist machine, and oral suction machine[3]
- Discussed and recommended Gleason Foundation request for seat elevator funding for power wheelchair
- Referral to local hospice program: Oct. 2018
  - Ongoing ALS clinic provider support to patient/wife/hospice staff
- Patient died Dec. 2018

Clinical Considerations and commentary

- Home renovations & equipment, power wheelchair, hospital bed, Hoyer lift, respiratory equipment, Gleason Foundation assistance for power wheelchair seat elevation & Tobii Dynavox
- All durable medical equipment, respiratory equipment, PEG tube placement should be addressed clinically and decisions regarding these interventions decided upon prior to hospice referral
- Difference between palliative care and hospice (Table 3)
  - Hospice referrals can be made by patient/family or clinic, however most follow Medicare guidelines and admission criteria, such as dysphagia, dyspnea (FVC <50% predicted), significantly progressive muscle weakness
  - An educational hospice consult is often requested to alleviate patient/family concerns, especially if no previous experience with hospice has occurred

Palliative Care	Hospice
Comparable to a house call practice	Medicare benefit program
Visits provided by MD or NP for symptom management	Services provided by interdisciplinary team
Visits occur based on clinical needs.	Visits occur usually twice per week
No requirement for prognosis of less than 6 months	Required to have a prognosis of 6 months or less
Services are consultation based. Curative measures can be maintained	Services are comprehensive and include DME, medications, 24 hour support
Patient can access home health or skilled nursing days	Patients forego skilled rehab, home health and curative treatments
Billed through Medicare Part B	Billed through Medicare Part A
Palliative specialist receives Medicare, Medicaid and private insurance	Hospice receives a per diem rate from Medicare depending on the level of care
Patient is responsible for copay based on consultation rate	Insurance reimburses case-by-case, carve out for certain treatments, such as TPN

Table 3: Differences between palliative care and hospice

1. Karam CY, Paganoni S, Joyce N, Carter GT, Bedlack R. Palliative Care Issues in Amyotrophic Lateral Sclerosis. Am J Hosp Palliat Med. 2016;33(1):84-92.
2. Kasarskis EJ, Scarlata D, Hill R, Fuller C, Stambler N, Cedarbaum JM. A retrospective study of percutaneous endoscopic gastrostomy in ALS patients during the BDNF and CNTF trials. J Neurol Sci. 1999;169(1-2):118-125.
3. Lechtzin N, Rothstein J, Clawson L, Diette GB, Wiener CM. Amyotrophic lateral sclerosis: Evaluation and treatment of respiratory impairment. Amyotroph Lateral Scler Other Mot Neuron Disord. 2002;3(1):5-13.

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