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]

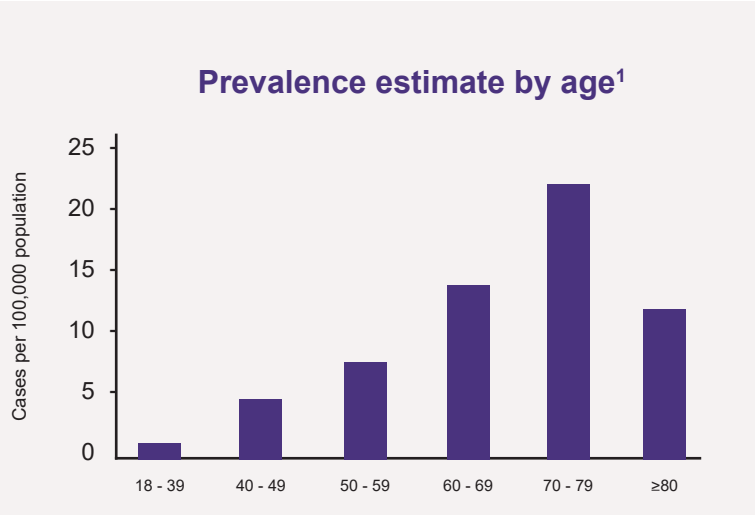

Figure 1: While ALS prevalence is highest among individuals older than 60, it can affect adults at any age[1]

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 document 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

In the middle stages of the disease, the clinical decisions are characterized by ensuring access to medication, assistive technology, physical therapy, and respiratory therapy. Documenting the disease progression is essential for payer authorization. Criteria for non-invasive ventilator coverage do not require a sleep study for ALS patients, as long as a decline in the respiratory capacity is well-documented.

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.
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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 #2: Respiratory Presentation with Bulbar and Upper Extremity Weakness

64-year-old white female

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Symptom Presentation & Initial Care Management

- Progressive dysarthria and swallowing difficulties
- The patient notes that over the period of time, she has not perceived any dramatic weakness in her hands, arms, or shoulders. There is no shortness of breath
- Riluzole prescribed — does not qualify for Radicava because of diminished FVC & low score on ALSFRS-R
- Active exerciser

ALS Clinic Visit

- Discussed and given educational handout material on PEG tube[4,5]
- Provided communication applications for iPhone & iPad
- Pulmonary consult — given PFT and overnight pulse oximeter to further assess respiratory capacity and limitations; PFT 1.09L/2.86L = 39% predicted
- Using CPAP — transition to BiPAP/cough assist/suction machine
- Referred to AT clinic appointment for evaluation for speech-generating communication device

Clinical Considerations and commentary

- Noninvasive ventilation (NIV) is proven to improve both survival and quality of life in ALS, with better response in non-bulbar patients.[1,2] Some considerations include:
  - Increasing inspiratory time helps improve lung volume recruitment and basilar atelectasis
  - All patients require a back-up rate for full respiratory support
  - Patients do not require tracheostomy to provide full ventilator support
- Practice guidelines instrumental in clinical swallowing and dysphagia management[3,4,5]
- Documentation of clinical progress note justifying need for each respiratory device

NIV (BiPAP) Device is Covered if Any of the Following Criteria are Met

A sleep study is not required to obtain coverage for an NIV (BiPAP):
Can lead to misdiagnosis for apnea and CPAP which will not help

- A Forced Vital Capacity (FVC) measured either upright or supine, with FVC <50%
- A Maximal Inspiratory Pressure (MIP) or Negative Inspiratory Force (NIF) <-60.
- Nocturnal oxygen desaturation, measured either with overnight oximetry or home sleep study, showing nocturnal desaturation (SaO2) < 88% for greater than 5 minutes (this does not need to be continuous)
- Arterial blood gas, while awake, with pCO2 > 45 mmHg

Table 2: Criteria for coverage of an NIV (BiPAP) device

1. Gruis, K. L. and N. Lechtzin. Respiratory therapies for amyotrophic lateral sclerosis: a primer. Muscle Nerve . 2012;46(3): 313-331.
2. Aboussouan LS, Mireles-Cabodevila E. Respiratory Support in Patients With Amyotrophic Lateral Sclerosis. Respir Care. 2013;58(9):1555-1558.
3. Pattee GL, Plowman EK, (Focht) Garand KL, et al. Provisional best practices guidelines for the evaluation of bulbar dysfunction in amyotrophic lateral sclerosis. Muscle and Nerve. 2019;59(5):531-536.
4. Kellogg J, Bottman L, Arra EJ, Selkirk SM, Kozlowski F. Nutrition management methods effective in increasing weight, survival time and functional status in ALS patients: a systematic review. Amyotroph Lateral Scler Front Degener. 2018;19(1-2):7-11.
5. 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.

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