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Navigating ALS: An overlooked disease

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Abstract: Amyotrophic lateral sclerosis (ALS) is a rare and serious disease affecting approximately 20,000 people in the US. This article details the signs, symptoms, and diagnosis of ALS and important management considerations.

Keywords: ALS, amyotrophic lateral sclerosis, Lou Gehrig disease, motor neuron disease, rare disease

Amyotrophic lateral sclerosis (ALS) is a rare and serious disease. Reports suggest that approximately 20,000 people in the US have ALS; every year about 5,000 new cases of ALS are diagnosed. Because no records on ALS have been kept it is hard to estimate the actual number of ALS cases in the US. The CDC does not require ALS cases to be reported; however, every 90 minutes, someone is diagnosed with, and someone dies from ALS.[1,2]

Nurses are often on the front line of caring for patients with ALS. Nurses need to understand and be prepared to work with the patient and family to manage the significant challenges created by the disease. This article details the signs, symptoms, and diagnosis of ALS as well as important management considerations.

ALS was identified in 1869 by French neurologist Jean-Martin Charcot. It became well known in 1939 when baseball player Lou Gehrig was diagnosed with the

disease. It subsequently became informally known as "Lou Gehrig disease."[1]

ALS belongs to a group of disorders known as motor neuron diseases in which both upper motor neurons (UMNs) and lower motor neurons (LMNs) degenerate, but the other neurologic systems, such as bladder control and senses, are unaffected.[3] ALS can progress rapidly and is considered a fatal disorder. The National Institute of Neurological Disorders and Stroke (NINDS), a component of the National Institutes of Health, is the primary federal funder of research on the brain and nervous system, including disorders such as ALS.[3]

Epidemiology

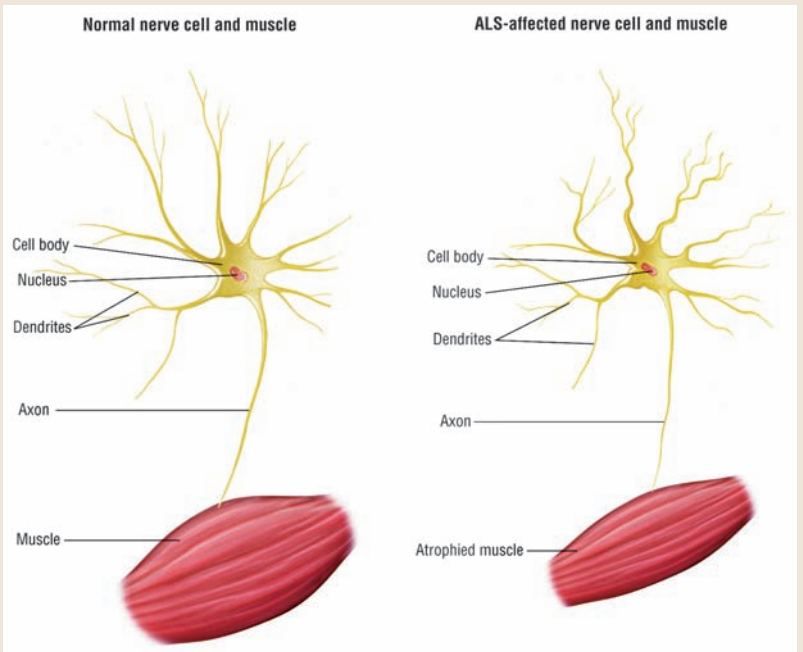
The only established risk factors for ALS are age and family history.[4]

ALS is mainly diagnosed in White males between 40 and 70 years old. Most ALS cases are sporadic; however, approximately 10% are familial and caused by an inherited mutated gene.[1] Genetic tests can determine family members' risk of developing the disease. For unknown reasons, military veterans are more likely to be diagnosed with the disease than the general public.[1]

Pathophysiology

ALS is a progressive neurodegenerative disorder primarily characterized by the degeneration of neurons (see *Nerve damage leads to muscle atrophy*).[4] UMNs send electrical signals down from the motor cortex to the spinal cord, and LMNs send electrical signals from the spinal cord to the muscles throughout the body. ALS begins as a focal process, then spreads throughout the motor system, causing neuron loss at all levels.[5,6] By the time the first signs and symptoms of ALS are noticeable, neuronal damage has already occurred. The etiology of ALS is unknown.[7] Researchers continue to study possible causes of ALS. Most hypotheses center on a complex interaction between genetic and environmental factors. Factors contributing to motor neuron deterioration include oxidative stress, defective glutamate metabolism, mitochondrial dysfunction, genetic variations, apoptosis, cytoskeletal protein defects preventing normal cell movement and division, autoimmune dysfunction, and inflammatory responses.[6]

Nerve damage leads to muscle atrophy



WERNER, R. A MASSAGE THERAPIST'S GUIDE TO PATHOLOGY: CRITICAL THINKING AND PRACTICAL APPLICATION. 7TH ED. WOLTERS KLUWER, PHILADELPHIA, 2016

Signs and symptoms

The onset of ALS can be subtle and often overlooked. However, the hallmark of ALS is the combination of UMN and LMN involvement. Weakness, atrophy, and fasciculations are direct consequences of muscle denervation; hyperreflexia and spasticity result from degeneration of the lateral corticospinal tracts.[4] Initial signs and symptoms may occur in the limbs ("limb onset") or bulbar segment, in which the patient most often presents with dysarthria or dysphagia ("bulbar onset").

Some of the first signs of limb-onset ALS usually appear in the hand or arm, and the patient notices difficulty buttoning clothes or writing. However, in other cases, the signs and symptoms start in one of the lower extremities. Not all people with ALS experience the same symptoms, sequences, or progression patterns. However, progressive muscle weakness and paralysis are universal.[1]

Initially, muscle stiffness, weakness, fasciculations, or muscle cramps is subtle. Then, muscle spasms and fasciculations will become generalized, including the tongue, causing dysphagia, dysarthria, and dyspnea. In addition, tongue fasciculations may occur first, with atrophy and swallowing or speech problems arising later.[8] Most voluntary muscles become paralyzed in the later stages of the disease, whereas involuntary muscles are unaffected. Eventually, breathing is affected, and long-term mechanical ventilation is needed.[6]

Diagnosis

Diagnosing ALS requires a detailed health history and a thorough physical assessment with diagnostic studies to rule out other diseases. Then, neuroimaging ascertains findings that may exclude more diseases.[6] Diagnostic studies include electromyography; nerve conduction studies; MRI; muscle biopsy; and lab testing of blood, urine, and sometimes cerebrospinal fluid. If the bulbar area is affected, swallow studies and pulmonary function tests are warranted.[5]

The diagnostic efficacy of the Gold Coast criteria was introduced in 2014 and it is most helpful in diagnosing ALS. It defines body regions as bulbar, cervical, thoracic, and lumbosacral. For one of these regions to be classified as involved in ALS, clinical examination or EMG must identify abnormalities in two limb muscles innervated by different roots and nerves, one bulbar muscle, or one thoracic muscle.[10] In 2019, the Gold Coast proposed simplified criteria to reflect progressive UMN and LMN signs and symptoms in one limb or body segment; progressive LMN signs and symptoms in at least two body segments; and an absence of electrophysiologic, neuroimaging, and pathologic evidence of other disease processes that might explain the signs of LMN or UMN degeneration.[10]

The ALS Functional Rating Scale (ALSFRS) is currently used to monitor changes in patients' speech, salivation, swallowing, handwriting, cutting of food and using utensils (with or without gastrostomy), dressing and hygiene, turning in bed and adjusting bed clothes, walking, climbing stairs, and breathing.[11,12] The highest score for a healthy, fully functioning patient using these 12 parameters is 40. If clinicians use the ALSFRS at regular visits, they can assess and graph the progression of ALS and prepare patients and their caregivers for possible complications and plan for the resources needed.

Treatment and prognosis

As of this writing, there is no cure for ALS or treatment to reverse its damage. In addition to symptomatic management, five medications have been FDA-approved for ALS: Riluzole, Nuedexta, edaravone, and Relyvrio. Riluzole, a member of the benzothiazole class, can extend survival by several months to tracheostomy. Edaravone, a member of the substituted 2-pyrazolin-5-one class, is available as an I.V. infusion and oral suspension and can slow disease progression by about 33%.[13] Evidence shows that these medications slow and reduce damage to motor neurons and nerve cells.[14]

Current research for ALS includes evaluating the causes of motor neuron death; the use of stem cells as possible treatments; comparing familial and sporadic ALS cases to

Individual and caregiver resources[15]

ALS Association	Website providing information about ALS, treatment, research, local support groups, and more
Your ALS Guide	Comprehensive guide developed by the ALS Association to help people with ALS, families, and caregivers learn what they need to know and get needed support and resources
International Alliance of ALS/MND Associations	A global network of ALS organizations
Department of Veteran Affairs	Provides services to veterans with ALS
ClinicalTrials.gov	A website with information about and locations for clinical trials
ALS Untangled	A website that provides information about alternative/off-label ALS treatment options
Patients Like Me	A website sharing information about different medical conditions, including ALS
ALS Medicare Resource Line	A website providing free individualized case management for people living with ALS, their families, and caregivers
A.C.C.E.S.S. Program	Free service for people living with ALS and other chronic conditions to help navigate Medicare and other challenges
Care Connection	A program to help people living with ALS launch, organize, and mobilize their community of support
ALS Forums	Online support group for people living with ALS
Living with ALS Resource Guides	A set of free comprehensive guides developed by the ALS Association
Team Gleason	A nonprofit organization that provides funding to people living with ALS for assistive technology, including home automation, voice and message banking, power wheelchairs, respite care for caregivers, education and webinars, and medical advocacy
ALS Pathways	A website with information about ALS assessment and management, caregiver support, and other ALS resources

identify genes involved in the development of the disease; discovering new biomarkers to improve diagnosis; and new treatments, such as drugs, stem-cell therapy, antibodies, and gene therapies.[10]

Most patients with ALS die within 3 to 5 years after diagnosis; a small percentage survive 5 to 10 years. The latter are often younger males with limb-onset ALS.[8]

Patients with ALS can maintain and improve their lives through various resources (see *Individual and caregiver resources*).[15]

Patient and caregiver support

The primary goal of the treatment plan for a patient with ALS is to promote their quality of life. Understanding the disease from the viewpoint of the patient with ALS and the caregiver helps identify factors that may contribute to a better quality of life. In addition, identifying a supportive group of family, friends, and healthcare professionals is essential.

Supportive measures to maintain the patient's health and optimum functioning as the disease progresses include assisting with nutrition, medications, physical and occupational therapy, speech and respiratory therapy, counseling, and hospice care.[16] Patients with ALS who receive multidisciplinary care have a better disease prognosis. Providing a personalized clinical approach can decrease the risk of death by 43.7% and prolong life by 7.5 months on average.[16]

Most patients with ALS are cared for at home by family. Depending upon the situation, caregiving can be stressful for the patient and the caregiver, so both should be frequently assessed for their quality of life.[16]

A person newly diagnosed with ALS can expect difficulty understanding the disease. Therefore, education for the person with ALS and their caregivers is critical. In-person and virtual ALS support groups are available.[17,18] Patients with ALS and caregivers should share their stories with these groups and learn how others have adapted to the challenges or found options for living with ALS.

Anger and fear are common in patients with ALS. Caregivers, friends, and loved ones should be encouraged to view the disease through the eyes of the patient with ALS. Doing so can promote an understanding of the patient's situation and emotions. Nonjudgmental listening can help a patient with ALS feel heard and allow them to work through emotions. As a result, the patient may develop a feeling of self-control over the disease. In addition, communication devices can help patients with ALS

A nurse's journey with ALS

My symptoms started in late 2019. Then, during the pandemic lockdowns in 2020, my symptoms worsened, specifically with my speech, swallowing, and weight loss. As a nurse, I knew something was wrong. I was officially diagnosed with ALS on December 29, 2020, the day after my 63rd birthday. This was the day my life and my family's lives changed forever.

The literature suggests it takes 9 to 12 months from the time the symptoms begin to diagnosis; that was certainly my experience. At the end of 2020, I had multiple tests performed and acquired a second opinion confirming my diagnosis.

As my family and I gradually allowed the news to sink in after hearing the ALS diagnosis, we all tried to figure out what to do next. As a healthcare professional, I am accustomed to helping others and being the one to fix things, but not this time.

The symptoms began in the bulbar area. In the summer of 2021, after trying many things to manage the disease, I finally agreed to a gastronomy tube so my nutrition would not continue to suffer. To this day, that is the sole source now of my nutrition. Additionally, I explored various open clinical trials and was admitted into one for 9 months. In May 2022, I started on a newly FDA-approved medication.

Around this time, I began looking at options for voice banking—a technology that creates a synthetic voice using clear voice recordings—with the help of a friend who is a speech pathologist. Unfortunately, my friend thought the bulbar impact was beyond voice banking, so we began exploring alternatives. In hindsight, my biggest regret is that I did not voice bank when I noticed I was losing my voice.

Some friends recommended the Gleason Foundation, founded by the pro footballer Steve Gleason in 2011 after he was diagnosed with ALS, to help with voice banking and equipment needs. I worked with Bridging Voices through the Gleason Foundation to find voice footage from voicemails, presentations, and videos. Within a few months, they converted my "old voice" to the "new voice" and provided the equipment I needed to communicate using my "new voice." While adjusting to communicating in this new manner has been difficult, I have embraced this technology that allows me to remain a productive member of my profession and communicate with loved ones.

In October 2021, I was appointed as a Nursing Regional Dean. I have used my communication device to speak to large groups of nursing colleagues, family and friends, and nursing students. Through the communication tools provided for me through the Gleason Foundation, I can continue teaching and shaping the next generation of nurses who may one day serve patients with ALS.

I am living a full, productive, and active life. I have chosen to manage the disease and refuse to let it manage me. As my son said, "My mom isn't dying of ALS; she is living with ALS." Even though I am a nurse, I have gained a better understanding of how care coordination for any disease is of utmost importance.

achieve independence and maintain their identity and self-worth.[16]

The role of a caregiver can be physically and emotionally stressful. Caregivers must pay close attention to their health and take regular time out to avoid burnout. Support groups also exist for caregivers, where participants discuss various issues, such as stress management and obtaining resources. In addition, hearing about others' caregiving experiences lets caregivers know they are not alone.[19]

Nursing considerations

Nurses play an important role in caring for patients with ALS by conducting ongoing assessments, providing education and resources, and communicating with other healthcare team members. Nurses must develop trusting relationships with patients and caregivers and collaborate to make informed treatment and overall care decisions.

Airway

Patency of the airway must be assured at all times, and various conditions can affect the patient's breathing. Establishing a baseline respiratory status early or soon after diagnosis is important to compare breathing status later in treatment. Patients experiencing breathing difficulties may complain of shortness of breath or dyspnea, fatigue, or breathing difficulties related to changes in position. Problems swallowing food or liquids may cause aspiration and impair breathing. Increased bronchial secretions may obstruct the airway and require suctioning or medications for control. Assessment of the respiratory system should include an evaluation of the respiratory rate and quality and the presence of skin pallor or cyanosis. Nasal flaring, intercostal retraction, and accessory muscles of respiration use, such as the cervical muscles, are also signs of respiratory difficulty. The presence of cough, swallow, and gag reflexes are critical to prevent breathing difficulties. Various techniques for protecting the airway should be discussed with the patient and caregivers, including sitting up straight, using the chin-tuck maneuver (putting the chin on the chest when swallowing), and refraining from talking when eating or drinking. Frequent assessment of the patient's ability to swallow liquids and solids will help to prevent aspiration.[20]

Decreased breath sounds and/or the presence of adventitious breath sounds, such as crackles or wheezes, require immediate intervention. Suction equipment should be readily available at home to remove secretions and maintain a patent airway. If the patient has a tracheostomy, suctioning and tracheostomy care will be part of the regular ongoing treatment for preventing airway obstruction and respiratory infection. Deep breathing, coughing, and an incentive spirometer or a cough assist machine can be used to encourage improved expansion of the lungs. The head of the bed should be elevated 45 degrees to improve shortness of breath. Various medications, such as anticholinergics, may be prescribed to provide comfort, reduce secretions, and promote gas exchange.[21]

After the patient has had time to adjust to the diagnosis, options for mechanical ventilation and cardiopulmonary resuscitation should be discussed with the patient and family. Although it can be difficult, the patient should consider these decisions before they are needed. Advanced care planning must also be discussed.[16]

Mobility

The patient's ability to move is another important area of assessment, especially since it contributes to the patient's ability to reposition for better breathing or elimination of secretions. Assessment should include the patient's motor strength, spasticity or flaccidity, and the development of contractures over time. Throughout the patient's disease, the nurse should encourage the continuation of daily routines and activity, exercise, and active range of motion (ROM). If the patient has limited mobility, passive ROM exercises can help prevent contracture and joint pain. If extremity weakness compromises the patient's mobility, safety, or independence in activities of daily living (ADL), a referral should be made to a physical or occupational therapist. As the disease progresses, patients may lose their ability to move or perform ADL. When this occurs, frequent repositioning, at least every 2 hours, will be essential to promote circulation and prevent complications of immobility, such as pressure injuries. When repositioning the patient, always assess the skin for discoloration or temperature change from surrounding tissues. People with light skin may demonstrate erythema, blanching, or even bruising, whereas those who are darkly pigmented may not have visible blanching or evidence of color changes.[22] Pressure-reducing support surfaces will also be helpful when the patient loses the ability to self-position in bed. In chairs or wheelchairs, using a full-seat wheelchair cushion will help prevent skin breakdown.[23]

Nutrition

Periodic dietitian consults are important for assessing the patient's ongoing and changing nutritional needs. Research has demonstrated the benefits of proper nutrition, particularly antioxidants and carotenoids, in treating people with ALS.[24] Specific nutrients are believed to trigger the disease; therefore, a proper diet is also critical for

those at risk for ALS.[25] A diet rich in nutrients, with enough calories, protein, fluid, vitamins, and minerals to meet nutritional requirements, has been shown to reduce the severity and progression of the disease, and it is associated with improved outcomes.[26]

Foods that are difficult for people with ALS to swallow should be avoided, including items of mixed consistencies; highly textured foods, such as red meats and raw vegetables; dry foods; sticky foods; small hard-textured foods, such as peanuts; and dairy products. Commercial thickeners may be used to thicken fluids and help prevent choking and aspiration. Nutritional supplements may be used to promote higher calorie intake. As the patient's swallow, cough, and gag reflexes become reduced or ineffective, feeding tube insertion must be considered. Studies show that early feeding tube placement may allow the patient to supplement oral intake and eat for pleasure.[27] Feeding tube insertion is a very personal decision that must be discussed at length with the patient, family, and physician or advanced practice clinician (APC).[15]

Hydration and elimination

Dehydration, a common problem for adults as they age, can be an even more significant problem for people with ALS. Dehydration can contribute to several other problems, such as skin breakdown and difficulty with elimination. Thus, it is better to prevent it than to try to correct it.

Elimination is also an important area for assessment, whose management should be focused on preventing complications. The average adult needs about 35 mL of water/kg/day for normal renal function. This number can be used to calculate a person's fluid requirements.[28] Fluid intake and output must be monitored and recorded daily, with discrepancies reported to the physician or APC as soon as possible. The nurse should also assess for signs and symptoms of urinary tract infection, such as frequency, urgency, dysuria, or fever.[15,16]

People with ALS also typically have slowed bowel motility and difficulties with elimination. A referral to an occupational and physical therapist is often necessary to determine the patient's ability to transfer to the toilet. Ongoing assessments of mobility and the use of trunk muscles required to support normal posture needed for defecation must be done if there is a decline in status. The patient or caregiver should be encouraged to keep a daily record of bowel movements, including amount and consistency. Dietary fiber enhancers and stool softeners may be useful in helping the patient maintain normal bowel function. Chronic use of laxatives should be avoided; mineral oil should never be used due to the risk of aspiration.[27]

Communication

Changes in muscle strength and function will also affect speech, eventually causing total speech loss. A speech therapist should be contacted early after diagnosis to evaluate the patient's speech, ability to communicate, and begin assisting in voice banking.[29]

As soon as possible, voice banking should be considered when the patient's voice begins to deteriorate. There are a variety of alternative modes of communication that can be discussed with the therapist to determine the most appropriate one for the patient's condition. A "magic slate" may be used if the patient can use their hands; word boards or letterboards may be helpful when the patient cannot use arms.[29]

Emotional

Receiving an ALS diagnosis may cause a range of patient and caregiver emotions. Maintaining a good support network can help a patient and others manage the numerous and difficult life changes resulting from ALS. Numerous organizations and resources are available to help the patient and caregiver learn to manage ALS and the resulting health challenges. Learning to have the necessary but difficult conversations with family, friends, coworkers, and others can help adjust to the new reality. Patients with ALS may experience depression and anxiety; however, research is unclear whether this is related to the progression of the disease or its emotional impact.[30] Family, friends, and caregivers must learn to recognize the signs of a change in emotional status and be ready to help. Ongoing changes in independence may lead the patient and family to experience grief, a normal response to loss. Evaluation of the support systems and coping skills can help team members provide education and counseling relative to the patient's situation. Encouraging the patient and family to share their concerns and fears will enable the family to cope more effectively and promote appropriate decision-making. Finally, a referral to a counselor, medical provider, or psychologist may be needed if coping is ineffective. Referral to a support group can help patients and caregivers realize that they are not alone in managing the disease and their lives.[18,19] ■

REFERENCES

1. The ALS Association. Understanding ALS. 2022. www.als.org/understanding-als. Accessed 2022.

2. Sibat HF, Valdes L. Introduction to novel aspects of motor neuron disease. In: Sibat HF, ed. *Novel Aspects of Motor Neuron Disease*. London, UK: IntechOpen; 2021:1-21. doi:10.5772/intechopen.92610.

3. National Institute of Neurological Disorders and Stroke. www.ninds.nih.gov/. Accessed 2022.

4. www.uptodate.com/contents/disease-modifying-treatment-of-amyotrophic-lateral-sclerosis?search=ALS&source=search_result&selectedTitle=5~150&usage_type=default&display_rank=5

5. Amyotrophic Lateral Sclerosis (ALS) Fact Sheet | National Institute of Neurological Disorders and Stroke. Ninds.nih.gov. 2022. www.ninds.

nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Amyotrophic-Lateral-Sclerosis-ALS-Fact-Sheet.

6. Martin S, Al Khleifat A, Al-Chalabi A. What causes amyotrophic lateral sclerosis? *F1000Res.* 2017;6:371. doi:10.12688/f1000research.10476.1.

7. Warner T. Motor neuron diseases. *Practice Guide to Neurogenetics.* 2009. www.sciencedirect.com/topics/medicine-and-dentistry/pathology-of-amyotrophic-lateral-clerosis#:~:text=Pathophysiology,thought%20to%20contain%20disarrayed%20neurofilaments.

8. Lockhart E. ALS: 8 ice cold facts about the fatal neurological disease. ActiveBeat. 2014. www.activebeat.com/your-health/als-8-ice-cold-facts-about-the-fatal-neurological-disease.

9. Paganoni S, Macklin EA, Lee A, et al. Diagnostic timelines and delays in diagnosing amyotrophic lateral sclerosis (ALS). *Amyotroph Lateral Scler Frontotemporal Degener.* 2014;15(5-6):453-456. doi:10.3109/21678421.2014.903974.

10. www.uptodate.com/contents/diagnosis-of-amyotrophic-lateral-sclerosis-and-other-forms-of-motor-neuron- disease?search=als&source=search_result&selectedTitle=2~150&usage_type=default&display_rank=2#H16

11. Georgetown University. The Amyotrophic Lateral Sclerosis Functional Rating Scale. www.sralab.org/sites/default/files/2017-07/PMandR_ALSRatingScale033111.pdf.

12. ALS Pathways. ALS information and resources. 2021. https://www.alspathways.com/als-overview/. Accessed 2022.

13. Mitsubishi Tanabe Pharma America. FDA approval of RADICAVA ORS® (edaravone) for the treatment of ALS. 2022. www.prnewswire.com/.

14. U.S. Food & Drug Administration. FDA approves drug to treat ALS. 2017. www.fda.gov/news-events/press-announcements/fda-approves-drug-treat-als.

15. Your ALS Guide. www.youralsguide.com/communication.html.

16. Galvin M, Gavin T, Mays I, Heverin M, Hardiman O. Individual quality of life in spousal ALS patient-caregiver dyads. *Health Qual Life Outcomes.* 2020;18(1):371. doi:10.1186/s12955-020-01551-5.

17. The ALS Association. Find local support for ALS. www.als.org/local-support. Accessed 2022.

18. ALS of Michigan. Helping people with ALS, their families, and caregivers. 2022. www.alsofmi.org/. Accessed 2022.

19. Centers for Disease Control and Prevention. Organizations that support ALS patients and caregivers. 2017. www.cdc.gov/als/OrganizationsthatSupportPALS.html. Accessed 2022.

20. Onesti E, Schettino I, Gori MC, et al. Dysphagia in amyotrophic lateral sclerosis: impact on patient behavior, diet adaptation, and Riluzole management. *Front Neurol.* 2017;8:94. doi:10.3389/fneur.2017.00094.

21. Garuti G, Rao F, Ribuffo V, Sansone VA. Sialorrhea in patients with ALS: current treatment options. *Degener Neurol Neuromuscul Dis.* 2019;9:19-26. doi:10.2147/DNND.S168353.

22. Pan Pacific Pressure Injury Alliance. 2019 International Pressure Injury Guideline. 2019. https://pppia.org/resources/.

23. Roy KM. Amotrophic lateral sclerosis and the trajectory of care. 2022. www.myamericannurse.com/als-amyotrophic-lateral-sclerosis-and-the-trajectory-of-care/.

24. D'Amico E, Grosso G, Nieves JW, Zanghì A, Factor-Litvak P, Mitsumoto H. Metabolic abnormalities, dietary risk factors and nutritional management in amyotrophic lateral sclerosis. *Nutrients.* 2021;13(7):2273-2293. doi:10.3390/nu13072273.

25. Tanenbaum B, Taft J. Maintaining good nutrition with ALS. ALS Association. www.als.org/sites/default/files/2020-04/Maintaining%20good%20nutrition%20with%20ALS.pdf.

26. Dorst J, Doenz J, Kandler K, et al. Fat-rich versus carbohydrate-rich nutrition in ALS: a randomised controlled study. *J Neurol Neurosurg Psychiatry.* 2022;93(3):298-302. doi:10.1136/jnnp-2021-328331.

27. Your ALS Guide: Feeding Tubes for ALS. 2022. www.youralsguide.com/feeding-tubes.html.

28. Scagnelli C, Waheed W, Tandan R. Hydration in amyotrophic lateral sclerosis. In: Preedy V, Patel V, eds. *Handbook of Famine, Starvation, and Nutrient Deprivation.* Springer; 2019:1-19. doi:10.1007/978-3-319-40007-5_108-1.

29. ALS Association. A Guide to Voice Banking Services. 2020. www.als.org/navigating-als/resources/fyi-guide-voice-banking-services.

30. Heidari ME, Nadali J, Parouhan A, et al. Prevalence of depression among amyotrophic lateral sclerosis (ALS) patients: a systematic review and meta-analysis. *J Affect Disord.* 2021;287:182-190. doi:10.1016/j.jad.2021.03.015.

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