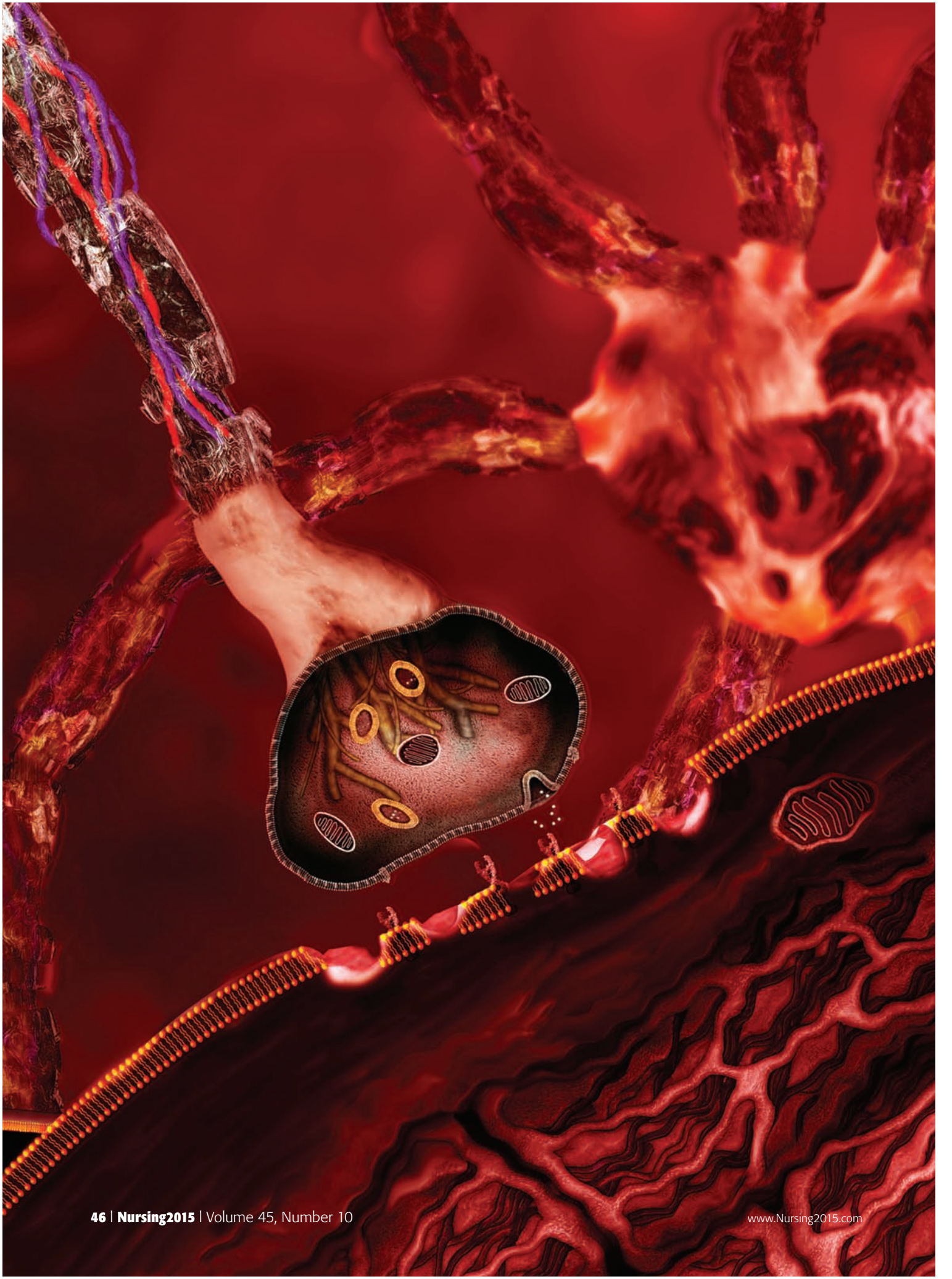

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ALS

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Amyotrophic lateral sclerosis: What nurses need to know

By Tamara L. Bellomo, MSN, RN-BC, and Lucille Cichminski, MSN, RN

A MOTHER OF THREE teenage children, Mrs. S, 49, presented to her healthcare provider with bilateral leg twitching and weakness, difficulty swallowing, and fatigue that's worsened over the past few weeks. While she was on her daily morning walk, she tripped and fell. She experienced a small laceration to her leg, prompting her visit to the healthcare facility. Her husband said that she'd had periods of slurred speech over the past few months as well. She was alert and oriented, and her vital signs were all within normal limits.

After an exam, her healthcare provider referred her to a neurologist who ordered magnetic resonance imaging (MRI), an electromyogram, and a full bloodwork panel. After multiple visits to rule out other causes, Mrs. S received the devastating diagnosis of amyotrophic lateral sclerosis (ALS).

ALS, also known as Lou Gehrig disease after the famous baseball player who had the disease, is one of the most common neuromuscular diseases in the world. This rapidly progressive, fatal neuromuscular disease involves the degeneration and death of the upper and lower motor neurons.[1] This article discusses the diagnosis and treatment of ALS and how nurses can help their patients deal with the difficult diagnosis and find the resources they and their families need.

Who's affected?

An estimated 20,000 to 30,000 Americans are living with ALS, and about 5,000 new cases are diagnosed each year in the United States.[2] This disease affects people of all ethnic, socioeconomic, and racial backgrounds.[2,3] ALS most commonly strikes people between ages 40 and 60, but it can also develop in younger and older people. The incidence is slightly higher in men than women.[3]

The average survival after diagnosis is 3 to 5 years, but the disease progresses more slowly

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in some cases.[3] The ALS Association reports that about 30% of people with ALS live 5 years, 10% to 20% survive more than 10 years, and 5% live for 20 years.[4] However, even longer-term survival is possible when the disease manifests at a younger age. An example is the well-known theoretical physicist Stephen Hawking. Dr. Hawking was diagnosed with the disease at age 21 and has been living with it for over 50 years. Although confined to a wheelchair and now able to speak only through a computer system, he set forth his theory of relativity and quantum mechanics, worked for over 30 years as a full professor of mathematics, and established the Stephen Hawking Centre for Theoretical Cosmology (Cambridge, U.K.), all while living with ALS.[5]

Besides diagnosis at a younger age, other variables for longer survival include male gender and limb onset versus bulbar onset.[6] (See *Understanding the pathophysiology*.)

Puzzling etiology

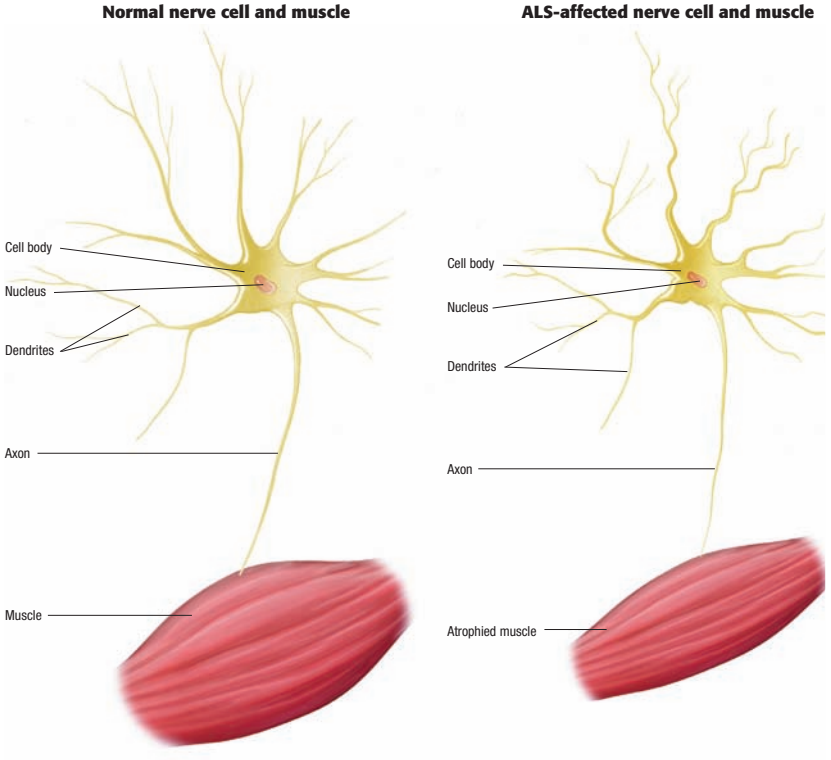
To date, no single cause of ALS has been identified. In 90% to 95% of ALS cases, the disease seems to occur spontaneously.[2] Patients with this sporadic form of the disease have no family history of ALS and their family members aren't at increased risk for the disease.[2]

Roughly 5% to 10% of all ALS cases are inherited. Familial ALS (FALS) usually results from an autosomal dominant inheritance pattern that requires only one parent to carry the gene responsible for the disease. The age of onset for FALS is about 10 years earlier than average onset of ALS and it's associated with a shorter survival.[7]

Roughly one-third of all familial cases result from a defect in a gene known as *chromosome 9 open reading frame 72*, or C9orf72, a gene whose function is still unclear. This gene mutation is also implicated in a small percentage of nonfamilial cases. Another 20% of familial cases result from mutations in the gene that encodes the enzyme copper-zinc superoxide dismutase type 1 (SOD1) gene.[2] Genetic testing is available for anyone with indications of ALS and a family history of ALS. Although genetic testing isn't a routine part of the diagnostic evaluation in ALS, it can help determine the cause of FALS in a family. Testing is the most useful when a person in the family has

Understanding the pathophysiology[19]

ALS leads to the progressive degeneration of the motor neurons that supply voluntary muscles. It's characterized by the deterioration of pyramidal neurons in the motor cortex (upper motor neurons) and motor neurons in the brainstem and central spinal cord (lower motor neurons). This progressive degeneration of the motor neurons eventually leads to their death. When the motor neurons die, the ability of the brain to initiate and control muscle movement is lost and muscles begin to waste. This is called *amyotrophy*. The loss of nerve fibers in the lateral columns of the spinal cord causes a rigidity, or *sclerosis*, to the central nervous system tissue; thus the term *lateral sclerosis*. However, the total sensory system and the regulatory methods of control and coordination of movements aren't affected. The neurons for ocular movement, the parasympathetic neurons in the sacral spinal cord, and the intellect all remain intact.



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already been diagnosed with the disease. About 50% of those with FALS will have an identified mutation. If a mutation has been identified, asymptomatic biological family members can be tested to determine if they inherited the genetic mutation. This is called predictive testing.[8]

Other possible causes under investigation include smoking, viral infection, autoimmune disease, and environmental exposure to toxins.[4] Studies are being performed of military personnel who were deployed during the 1991 Gulf war. A recent study demonstrated that those veterans were more likely to develop ALS compared with military personnel who weren't in the region.[2,4]

Signs and symptoms

The initial signs and symptoms of ALS involve a slow, chronic progression that varies among those affected.[9] Eighty percent of patients present with asymmetric limb weakness while 20% have bulbar onset, presenting with dysarthria or dysphagia.[10]

The first manifestations of the disease depend on the location of the affected neurons and may be so slight that the person doesn't recognize them as a problem.[11] The person may not seek treatment until muscle atrophy and weight loss are apparent.[9]

The clinical hallmark of ALS is the combination of upper motor neuron and lower motor neuron signs and symptoms.[10] Upper motor neuron findings of weakness, hyperreflexia, and spasticity result from degeneration of frontal motor neurons. The lower motor neuron findings of weakness, atrophy or amyotrophy, and fasciculations are a direct consequence of degeneration of lower motor neurons in the brainstem and spinal cord.[10]

Bulbar signs and symptoms include dysphagia, which increases the risk of aspiration, and dysarthria.[9,11]

New directions for ALS research

Current ALS research topics include the following:

- Understanding the mechanisms that selectively trigger motor neurons to degenerate in ALS and to develop effective strategies to halt the processes leading to cell death.
- Developing biomarkers for ALS that could aid in diagnosis, serve as markers for disease progression, or correlate with therapeutic targets.
- Testing druglike compounds, gene therapy approaches, antibodies, and cell-based therapies.[14]
- Exploring the impact of cell transplantation to manage signs and symptoms.[20,21]
- Studying the potential association between cigarette smoking and ALS.[22]

Bladder and bowel function is usually spared because the spinal nerves that control these muscles aren't affected.[11,12] Although about 15% of patients with ALS may develop frontotemporal dementia, cognitive function is usually spared in most of those affected by ALS.[12]

Nearly all patients develop signs and symptoms of respiratory insufficiency that may require mechanical ventilation. Nurses must ensure that the risks and benefits of invasive ventilation are discussed with patients and their families long before the time when the decision must be made. Approximately 5% to 10% of patients select the option of a tracheostomy with permanent mechanical ventilation when respiratory compromise becomes severe. The most common cause of death in ALS is progressive neuromuscular respiratory failure. Other causes of death include aspiration and infection.[10]

Diagnosing ALS

Establishing an ALS diagnosis is a difficult task that can take a long time because no single test can confirm it.[2,12] Instead, diagnosis is usually based on the presenting signs and symptoms and the exclusion of other possible causes.[12] Neuroimaging including brain CT and MRI is used to rule out disorders that can mimic ALS, such as multiple sclerosis or a brain tumor.[9,12] Electrodiagnostic studies are helpful in evaluating weakness, muscle wasting, and sensory symptoms. They typically reveal combined features of acute and chronic denervation in ALS.[13] Sensory and nerve conduction studies can determine the ability of the nerves to send electrical signals even though the results of these studies are most often normal in ALS patients.[13] However, in cases of severe atrophic and denervated muscles, the conduction studies results may exhibit reduced amplitude.[2,9]

Lab testing usually includes serum and urine protein electrophoresis, thyroid studies, and 24-hour urine heavy metal panel.[4,9] Other diagnostic studies include a complete blood cell count with differential, erythrocyte sedimentation rate, C-reactive protein, vitamin B_{12}, and folate levels.[9]

Treatment choices

ALS is progressive and incurable, and patients require multidisciplinary care. To date, the only treatment that's been shown to improve survival time is riluzole.[14] A member of the benzothiazole class, riluzole is thought to reduce glutamate-induced excitotoxicity.[15,16] Riluzole can lengthen survival time by several months for some patients and can also extend the time before the patient requires mechanical ventilation. However, this medication can't reverse any damage already incurred. The most commonly observed adverse reactions include asthenia,

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nausea, vomiting, diarrhea, abdominal pain, anorexia, and dizziness. Because about 10% of those using this medication experience liver damage, liver function should be monitored frequently.[16]

Most other treatments for ALS are used to improve patients' quality of life and relieve signs and symptoms. The goal of this supportive care is to help the patients be as mobile, independent, and comfortable as possible. Pharmacotherapy may be indicated to treat dyspnea, muscle spasm, spasticity, sialorrhea (excessive flow of saliva), fatigue, pain, depression, dysphagia, constipation, and sleeping problems.

For patients with ALS, physical and occupational therapy play an important role in delaying loss of strength, maintaining endurance, preventing complications, reducing or controlling pain, and promoting functional independence. Patients can benefit from low-impact aerobic exercises such as swimming, walking, and riding a stationary bicycle, which may help improve cardiovascular health as they fight fatigue and depression. The goal is to provide strategies and equipment that help patients retain independence and ensure their safety while they perform activities of daily living. Speech-language pathologists can help patients experiencing dysphagia and dysarthria.[17]

To maintain nutrition, encourage patients to eat small, frequent meals. Meals should provide fiber, fluid, and calories with foods that are easy to swallow. Because choking due to excess saliva is a concern, always keep suction devices nearby.[18]

Patients may require percutaneous gastrostomy tube placement for nutritional support.[6] Pulmonary support may include noninvasive positive pressure ventilation, tracheostomy, and mechanical ventilation.[6]

Palliative care should be provided from the time of diagnosis all the way through the bereavement process to provide support to the patient with ALS, the family, and the caregiving team. Palliative care isn't restricted to end-of-life care but rather focuses on relieving suffering throughout each stage of the disease. Other objectives of palliative care include establishing goals of care that are consistent with the patient's physical, psychosocial, emotional, and spiritual values. When the patient's expected prognosis reaches 6 months or less, or when goals shift exclusively toward comfort, hospice should be introduced.[6]

Current research

The National Institute of Neurological Disorders and Stroke, part of the National Institutes of Health, is the leading advocate of biomedical research on ALS. The goals of this research are to find the cause of ALS, understand the mechanisms involved in disease progression, and develop effective treatments. Investigators are optimistic that these clinical research studies will eventually lead to more effective treatments for ALS. (See *New directions for ALS research.*)

Back to the patient...

Mrs. S's healthcare provider started her on oral riluzole and referred her to a nutritionist and physical therapist. She's scheduled for a follow-up visit in 2 weeks. Mrs. S and her family will require extensive education about her disease. Besides providing emotional and physical support, her nurse will inform the patient and family about available resources in the community and provide educational materials.

Patient education

Each patient with ALS is affected in a different way, so tailor patient education and subsequent interventions to meet the patient's specific needs. Nurses need to teach patients and their caregivers the importance of adhering to prescribed treatment regimens and to notify the healthcare provider should changes occur. Topics to teach include:

- monitoring for changes in respiratory, swallowing, or speech status
- providing skin care and preventing pressure ulcers
- making sure the patient's diet is well balanced
- adhering to physical and occupational therapy regimens
- monitoring for alterations in bowel and bladder patterns
- implementing fall prevention strategies such as removing potential trip hazards, installing handrails and a shower chair in the shower, and utilizing assistive devices such as a walker or wheelchair when needed.

Resources for patients and families

The following organizations provide assistance and support to patients with ALS and their families:

- **ALS Association**
  www.alsa.org
- **ALS Therapy Development Institute**
  www.als.net
- **fALS Connect**
  https://fals.patientcrossroads.org
- **Family Caregiver Alliance**
  www.caregiver.org
- **Les Turner ALS Foundation**
  www.lesturnerals.org
- **MDA (Muscular Dystrophy Association)**
  www.mda.org
- **National Alliance for Caregiving**
  www.caregiving.org
- **National Family Caregiver Action Network**
  www.caregiveraction.org
- **Prize4Life**
  www.prize4life.org
- **Project A.L.S.**
  www.projectals.org

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Emotional support and resources

Providing emotional support and education to patients and their families throughout the course of the disease are key nursing interventions. Communication and active listening are encouraged. Encourage patients and their families to use resources that will help support them as they deal with the challenges of ALS. (See *Resources for patients and families.*) ■

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