

## Nursing Considerations for Patients with Amyotrophic Lateral Sclerosis

Nawal Awad Hussain Alotaibi <sup>1</sup>, Mastura Alyan S Alzahrani <sup>2</sup>, Rehab Muteb Alruwili <sup>3</sup>,  
Maram Faleh Alhajouj <sup>4</sup>, Omamah Faleh Alnakhli <sup>5</sup>, Jamilah Sayed Attallah  
Aldahmeshi <sup>6</sup>, Ghurub Muflih Ahmed Alrashidi <sup>7</sup>, Suad Saad M Almutairi <sup>8</sup>, Sanaa  
Manzel Alrwialiy <sup>9</sup>, Walaa Qasem Alnakhli <sup>10</sup>

<sup>1</sup>- Nursing specialist, King Abdullah Medical Complex, Jeddah Second Health Cluster, Jeddah, Saudi Arabia

<sup>2</sup>- Nursing technician, King Abdullah Medical Complex, Jeddah Second Health Cluster, Jeddah, Saudi Arabia

<sup>3</sup>- Nursing technician, Turaif General Hospital, Turaif, Saudi Arabia

<sup>4</sup>- Nursing technician, Eradah Mental Health Hospital, Madinah, Saudi Arabia

<sup>5</sup>- Nursing technician, King Salman Medical City, Madinah, Saudi Arabia

<sup>6</sup>- Nursing technician, Maternity and Children's Hospital in Arar, Saudi Arabia

<sup>7</sup>- Nursing technician, Hail Health Cluster, Hail, Saudi Arabia

<sup>8</sup>- Nursing, King Abdullah Medical Complex, Jeddah, Saudi Arabia

<sup>9</sup>- Nursing, Eradah Mental Health Hospital, Madinah, Saudi Arabia

<sup>10</sup>- Nursing, Eradah Mental Health Hospital, Madinah, Saudi Arabia

### Abstract:

Caring for patients with Amyotrophic Lateral Sclerosis (ALS) requires a comprehensive approach that addresses the physical, emotional, and social needs of the individual. Due to the progressive nature of ALS, nurses should regularly assess and document the patient's neurological status, including motor function, swallowing ability, and respiratory function. It's essential to collaborate with an interdisciplinary team to create a tailored care plan that includes physical therapy, occupational therapy, and nutrition support. Nurses should also educate patients and their families about the disease's progression, advance care planning, and the potential use of assistive devices to maintain independence and quality of life for as long as possible. Moreover, emotional support is critical for both patients and their families coping with the challenges of ALS. Nurses should provide psychological support, facilitating communication and offering resources for counseling or support groups, as the disease can lead to feelings of isolation and despair. Palliative care principles should be integrated early on to manage symptoms effectively, improve comfort, and enhance overall well-being. Furthermore, attention to end-of-life discussions and advance directives is vital, ensuring that patients' wishes are respected as they approach the later stages of the disease.

**Keywords:** ALS, nursing considerations, patient care, interdisciplinary team, neurological assessment, physical therapy, emotional support, assistive devices, palliative care, advance care planning.

### Introduction:

Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig's disease, is a progressive neurodegenerative disorder that primarily affects motor neurons in the brain and spinal cord. This leads to a gradual deterioration of voluntary muscle control, ultimately resulting in paralysis and, in many cases, death. Research suggests that while the onset of ALS can vary widely among individuals, the disease typically affects middle-aged adults, with men being diagnosed slightly more often than women. Currently, there is no known cure for ALS, which underscores the essential role of nursing care in the management of symptoms, providing

palliative support, and enhancing the quality of life for those affected [1].

The nursing considerations for patients with ALS are multifaceted and complex, requiring a holistic, patient-centered approach. Nurses play an invaluable role not only in the clinical assessment and management of physical symptoms but also in addressing the psychological and emotional needs of both patients and their families. The progressive nature of the disease means that nursing care evolves over time, often transitioning from curative measures to palliative care. This shift necessitates a strong emphasis on communication, advocacy, and interprofessional collaboration to ensure that the

patient's dignity and preferences are respected throughout their disease journey [2].

One of the critical nursing considerations in the management of ALS is the need for comprehensive symptom assessment and management. Patients often experience a range of symptoms that can include muscle weakness, spasticity, dysphagia (difficulty swallowing), respiratory compromise, and significant emotional distress. Each of these symptoms requires tailored interventions, underscoring the importance of a thorough clinical understanding of the disease's progression and its impact on daily functioning. For example, nurses must monitor respiratory function closely, as compromised respiratory muscles can lead to life-threatening complications. Patients may benefit from interventions such as non-invasive ventilation support, suctioning mechanisms, and, in some cases, assistive technologies that can improve their ability to communicate or perform activities of daily living [3].

In addition to physical symptoms, psychosocial considerations play a pivotal role in the care of ALS patients. The debilitating nature of the disease can give rise to anxiety, depression, and feelings of isolation for both patients and caregivers. Therefore, nurses need to be vigilant in recognizing the signs of psychological distress and should utilize evidence-based strategies to foster emotional well-being. Interventions could include providing education about the disease process, facilitating support group participation, or involving mental health professionals when necessary. By adopting a compassionate, empathetic approach, nurses can help to diminish feelings of hopelessness and empower patients to maintain a sense of purpose and agency throughout the progression of their illness [4].

Effective communication is another cornerstone of nursing care for ALS patients. Given the communication challenges that may arise as the disease progresses, it is crucial that nurses establish a rapport early on, while patients still have the capacity to articulate their values, preferences, and goals of care. This initial dialogue can establish a foundation for advance care planning discussions, which can be emotionally charged but are necessary for respecting patient autonomy. Nurses must facilitate these conversations with sensitivity, ensuring that patients and their families feel

supported and informed in making decisions regarding their care, whether that involves aggressive treatment options or transitioning to palliative care models [5].

Moreover, the role of the nurse extends beyond direct patient care to include education and support for family members and caregivers. ALS affects not only the individual but also their loved ones, who often face significant physical and emotional burdens. Providing education on disease progression, care needs, and coping strategies can empower caregivers and improve their confidence in managing the complexities of care. Nurses should also advocate for the integration of palliative care teams early in the treatment process, even in conjunction with disease-modifying therapies, to optimize quality of life and align care objectives with the patient's wishes [6].

#### **Pathophysiology and Progression of ALS:**

Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig's disease after the famous baseball player who was diagnosed with the condition, is a progressive neurodegenerative disorder that primarily affects motor neurons in the brain and spinal cord. Despite its relatively low prevalence when compared to other neurological diseases— affecting approximately 2 to 6 individuals per 100,000 per year—it poses profound challenges for patients, families, and healthcare systems alike due to its debilitating effects and the dramatic progression of its symptoms. Understanding the pathophysiology and clinical progression of ALS is essential to understand both its profound impact and the avenues through which research and treatment might be developed [7].

The pathophysiological processes underlying ALS are complex and multifactorial. At the core, ALS is characterized by the selective degeneration of upper motor neurons in the primary motor cortex and lower motor neurons in the brainstem and spinal cord. This neuronal loss disrupts the transmission of signals necessary for voluntary muscle control, leading to the progressive weakness and atrophy that define the disease [8].

Though the exact cause of motor neuron degeneration remains largely unknown, several mechanisms have been proposed. Genetic factors play a significant role in ALS, with mutations in genes such as **SOD1**, **C9orf72**, **FUS**,

and **TARDBP** being linked to inherited forms of the disease (familial ALS). The most common genetic cause, the expansion of the C9orf72 gene, accounts for a significant percentage of all ALS cases and is also associated with frontotemporal dementia. These genetic mutations often lead to toxic gain-of-function effects, where the mutated proteins misfold and aggregate, disrupting cellular homeostasis [9].

Another critical aspect of ALS pathophysiology is the role of glial cells—particularly astrocytes and microglia. These supportive cells are not only crucial for neurological health but may also contribute to neurodegeneration in ALS. Activated astrocytes may propagate neuroinflammation and release toxic factors, while microglial activation can exacerbate neuronal injury, creating a pathological cycle that accelerates motor neuron death [10].

Additionally, oxidative stress has emerged as a pivotal factor in ALS pathophysiology. Neurons exhibit high metabolic activity and are particularly vulnerable to oxidative damage. An imbalance between the production of reactive oxygen species and antioxidant defenses leads to oxidative injury, further contributing to the degeneration of motor neurons. This condition may be compounded by mitochondrial dysfunction, which is observed in many ALS cases, leading to impaired energy metabolism and increased sensitivity to stress [11].

Excitotoxicity, a process wherein neurons are damaged and killed by excessive stimulation by neurotransmitters, particularly glutamate, is also implicated in ALS. Motor neurons in ALS patients often exhibit increased levels of glutamate, leading to overstimulation of glutamate receptors. This overstimulation can initiate a cascade of events resulting in neuronal apoptosis, further contributing to motor neuron degeneration. Notably, the enzyme glutamate transporter (EAAT2) is downregulated in ALS, impairing the clearance of excess glutamate and exacerbating excitotoxic effects [12].

The clinical progression of ALS is characterized by a gradual decline in motor function and is typically divided into several stages. Although the rate of disease progression varies widely among individuals, the overall trajectory is similar. The age of onset, genetic background, and even environmental factors can influence the rate at which ALS advances.

The initial symptoms of ALS often manifest subtly and can include muscle weakness, twitching, and cramps. Patients frequently report difficulty in fine motor tasks such as buttoning shirts or writing. This phase may last for an extended period, leading to underdiagnosis or misdiagnosis as symptoms can be nonspecific and overlap with other neurological conditions. Early symptoms often affect specific muscle groups, and while both upper and lower motor neurons are involved, lower motor neuron signs often take precedence at this stage. [13]

As the disease progresses, patients will experience increased muscle weakness and atrophy. Weakness typically begins asymmetrically, affecting one limb more significantly than the other. Patients may experience dysphagia (difficulty swallowing) and dysarthria (difficulty speaking) as muscles controlling swallowing and speech become involved. Respiratory muscles can also weaken over time, leading to significant complications with breathing [14].

In later stages of ALS, individuals may become dependent on assistive devices or caregivers for mobility and daily activities. The progression can lead to total paralysis as more motor neurons degenerate. Despite the loss of motor function, cognitive function remains relatively intact for the majority of ALS patients, although a subset of individuals may experience frontotemporal dementia [15].

The quality of life for ALS patients declines considerably due to the physical limitations imposed by the disease, along with challenges surrounding communication and feeding. The emotional burden on families, mental health professionals, and caregivers becomes critical as they adapt to the progressive decline and eventual terminal nature of the illness.

With advancements in supportive care, including nutritional intervention, respiratory support, and palliative care, many patients experience improved quality of life and extended survival, although these measures do not modify the underlying disease progression [16].

#### **Clinical Assessment and Monitoring Strategies:**

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a progressive neurodegenerative disorder that primarily affects

motor neurons in the brain and spinal cord. The degeneration of these neurons leads to muscle weakness, atrophy, and ultimately paralysis, which significantly impacts the patient's quality of life and ability to perform daily activities. The complexity and variability of ALS necessitate a comprehensive and multidimensional approach for clinical evaluation and monitoring [17].

### Clinical Evaluation

Clinical evaluation of patients with ALS involves a combination of neurological examinations, functional assessments, and patient-reported outcomes. The evaluation often begins with a detailed medical history, including the onset and progression of symptoms, family history, and comorbid conditions. This history is vital, as it assists physicians in differentiating ALS from other similar conditions, such as multifocal motor neuropathy or spinal muscular atrophy [17].

1. **Neurological Examination:** A comprehensive neurological examination is crucial for identifying motor function impairment. Clinicians assess muscle strength, tone, atrophy, reflexes, and coordination. The limb function may be evaluated through dynamometry to quantitatively assess muscle strength. Additionally, cranial nerve function and bulbar involvement (dysphagia and dysarthria) are evaluated, as bulbar symptoms significantly impact communication and dietary needs.
2. **Functional Assessment:** Various scales are employed to evaluate the functional status of ALS patients quantitatively. The most commonly used is the Revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R), which assesses functions related to mobility, self-care, communication, and respiratory function, rating from 0 (worst) to 40 (best). Regular use of this scale allows for consistent tracking of disease progression over time [17].
3. **Pulmonary Function Tests:** Given the respiratory compromise associated with ALS, pulmonary function tests should be incorporated into the evaluation. Tests measuring forced vital capacity (FVC) help

determine the respiratory muscle strength and are predictive of respiratory failure. Regular monitoring of pulmonary function can guide decisions regarding interventions, such as non-invasive ventilation [18].

4. **Nutritional Assessment:** A nutrition assessment is essential, as dysphagia (difficulty swallowing) can lead to malnutrition and dehydration. Weight loss and changes in diet should be closely monitored, and dietitians may provide recommendations to ensure adequate caloric intake through texture-modified diets or supplements.
5. **Quality of Life Measures:** Assessing the psychological and social aspects of living with ALS is equally important. Instruments such as the ALS Assessment Questionnaire (ALSAQ-40) and the European Quality of Life-5 Dimensions (EQ-5D) can provide insights into the patient's quality of life, emotional well-being, and overall satisfaction with their health [18].

### Monitoring Strategies

Continual monitoring of ALS patients is essential to adapt the treatment plan based on disease progression and emerging symptoms. The frequency of evaluations can depend on the stage of the disease and the rate of progression, often recommending every few months for those in the early to middle stages and more frequently for those in advanced stages.

1. **Multidisciplinary Care Approach:** Optimal care for ALS patients often requires a multidisciplinary team, including neurologists, nurses, physical therapists, occupational therapists, speech therapists, respiratory therapists, social workers, and dietitians. Each team member plays an essential role in addressing specific aspects of the patient's care, fostering comprehensive management that enhances quality of life [19].
2. **Involvement of Palliative Care:** Incorporating palliative care early in the disease process can significantly improve the overall care experience by addressing

symptoms like pain, respiratory distress, and psychological distress. The goal is to enhance the quality of life and support patients and families in making complex decisions regarding care preferences.

3. **Assistive Devices and Rehabilitation:** Depending on the rate of functional decline, patients may benefit from physical and occupational therapy to maintain mobility and independence as long as possible. The introduction of assistive devices, such as wheelchairs, home modifications, and communication aids, should be monitored closely to ensure they meet the patient's evolving needs [19].
4. **Support and Counseling:** Regular opportunities for psychosocial support and counseling are vital for both patients and caregivers. Providing resources for stress management, caregiver support, and psychosocial interventions can help mitigate feelings of isolation and anxiety, which are prevalent in this population.
5. **Clinical Trials and Research Participation:** Participation in clinical trials can provide access to cutting-edge therapies and contribute to the growing body of knowledge regarding ALS. Patients should be informed about ongoing trials and the potential benefits and risks associated with participation, fostering a sense of involvement in advancing treatment options for future patients [20].

#### **Multidisciplinary Care Approaches in ALS Management:**

Amyotrophic Lateral Sclerosis (ALS), popularly known as Lou Gehrig's disease, is a progressive neurodegenerative condition that affects motor neurons in the brain and spinal cord. This disease leads to muscle weakness, disability, and eventually respiratory failure. While the precise cause of ALS remains unknown, the complexity of the condition necessitates a multifaceted approach to treatment and management. As such, multidisciplinary care approaches have emerged as a cornerstone in enhancing the quality of life and maximizing functionality for individuals living with ALS [21].

Multidisciplinary care involves the collaboration of healthcare professionals from various disciplines working together toward a common goal: optimal patient care. In the context of ALS, this means integrating expertise from neurology, nursing, physical therapy, occupational therapy, speech therapy, respiratory therapy, nutrition, psychology, and social work, among others. This collaborative model stands in contrast to a unidisciplinary approach, which typically emphasizes the role of a single healthcare provider and may lead to fragmented care [22].

The necessity for a multidisciplinary approach in ALS management arises from the diverse symptoms and complications associated with the disease. Patients may experience not only motor dysfunction but also cognitive and behavioral changes, respiratory issues, dysphagia (difficulty swallowing), and emotional distress. A single healthcare provider's perspective may not sufficiently address the multifarious nature of these symptoms. Hence, the amalgamation of skills and insights from various specialties is critical in delivering comprehensive care that responds adequately to the needs of the patient and their family [23].

#### **Key Components of Multidisciplinary Care**

1. **Neurological Management:** The neurologist typically serves as the leader of the multidisciplinary team. They diagnose the condition, monitor the progression of the disease, and prescribe medications that may help alleviate symptoms. Neurologists also play a pivotal role in coordinating care among other specialists [24].
2. **Physical and Occupational Therapy:** Physical therapists develop exercise regimens that focus on maintaining mobility and minimizing the impact of muscle weakness. Occupational therapists assess the patient's home environment and recommend adaptations that allow individuals to maintain independence and perform daily activities effectively, including dressing, grooming, and cooking [25].
3. **Speech and Swallowing Therapy:** As ALS progresses, patients often face difficulties with speech and swallowing.

Speech-language pathologists work with patients to develop communication methods, whether through traditional verbal techniques or augmentative and alternative communication (AAC) devices. They also provide strategies to manage dysphagia, which can significantly improve safety and quality of life [26].

4. **Respiratory Care:** Respiratory therapists are crucial for patients with ALS, particularly in the later stages of the disease when respiratory muscles weaken. They evaluate lung function and may introduce mechanical ventilation or non-invasive ventilation strategies, ensuring that patients receive adequate respiratory support.
5. **Nutritional Support:** Malnutrition and dehydration are common concerns for ALS patients, largely due to swallowing difficulties. Dietitians assess nutritional intake and provide tailored dietary plans or suggest enteral feeding options when necessary, which could involve the placement of a feeding tube [27].
6. **Psychological Support:** Psychological interventions are essential for addressing the emotional and mental health challenges that accompany the diagnosis of ALS. Psychologists or social workers can provide counseling, support groups, and resources that help patients and their families cope with the psychological toll of the disease.
7. **Palliative and End-of-Life Care:** Given that ALS is a terminal condition, the role of palliative care is integral from the time of diagnosis. This aspect of multidisciplinary care focuses on providing relief from symptoms and improving the quality of life throughout the disease's progression. It emphasizes patient-centered care that respects the individual's values and preferences [28].

#### **Benefits of Multidisciplinary Approaches**

1. **Comprehensive Care:** The integration of multiple professional perspectives enables a holistic view of patient care. By addressing all facets of the patient's well-

being, healthcare teams can tailor interventions that reflect individual patient needs [29].

2. **Improved Symptom Management:** Patients with ALS experience a range of debilitating symptoms, from weakness to respiratory complications. A coordinated approach among specialists often results in more effective symptom management, leading to improved functional ability and comfort.
3. **Enhanced Quality of Life:** Multidisciplinary care models often result in improved quality of life for patients. With access to a wider range of therapeutic interventions and support systems, patients often report enhanced psychological well-being, relief from pain or discomfort, and better management of daily activities.
4. **Family Support and Education:** A multidisciplinary approach extends beyond the patient to include family and caregivers. Healthcare providers can offer education on the disease process, facilitate communication between family members, and provide necessary resources to support them through their caregiving journey [30].
5. **Research and Best Practices:** Multidisciplinary teams often engage in collaborative research efforts that can lead to better understanding of ALS, innovations in treatment, and the establishment of best practices in management [31].

#### **Challenges in Implementing Multidisciplinary Care**

Despite its numerous benefits, the implementation of multidisciplinary care in ALS management is not without challenges. Healthcare systems often operate in silos, where communication among departments can be inadequate. This lack of collaboration may lead to delays in care and fragmentation of services. Furthermore, limited access to specialists in some regions may hinder the establishment of effective multidisciplinary teams. Financial constraints also pose barriers, as patients may face high out-of-pocket costs for therapies and treatments not fully covered by insurance [32].

### **Physical and Occupational Therapy Interventions:**

Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disorder that primarily affects motor neurons responsible for voluntary muscle control. The gradual degeneration and death of these neurons lead to muscle weakness, atrophy, and eventually paralysis, significantly impacting an individual's quality of life. While there is currently no cure for ALS, various therapeutic interventions can improve patients' functional abilities and overall well-being. Among these interventions, physical therapy (PT) and occupational therapy (OT) play a critical role. Nurses, in collaboration with physical and occupational therapists, provide comprehensive care that addresses the multifaceted challenges faced by ALS patients [32].

ALS is characterized by the selective degeneration of motor neurons in the brain and spinal cord, leading to the progressive loss of muscle control. Patients often experience a wide range of symptoms, including muscle cramps, weakness, difficulty in coordination, speech and swallowing difficulties, and respiratory dysfunction. As the disease advances, patients may require assistance with mobility, self-care, and other daily living activities. The decline in physical functioning, coupled with the emotional and psychological impact of the disease, presents unique challenges that require a holistic care approach [32].

Physical therapy focuses on maintaining and improving the physical function of ALS patients through specialized exercises, mobility training, and supportive interventions. Physical therapists work to develop individualized exercise programs that are tailored to the patient's abilities, goals, and needs. Nurturing the patient's physical function and addressing mobility limitations can lead to improved quality of life [33].

### **Nursing Interventions Related to Physical Therapy**

1. **Assessment and Monitoring:** Nurses must conduct comprehensive assessments to gauge the patient's initial physical condition, including strength, range of motion, coordination, and balance. Regular monitoring helps in tracking disease

progression and adjusting therapeutic interventions as necessary [34].

2. **Collaboration with Physical Therapists:** Nurses should maintain open communication with physical therapists to understand the specific goals of the therapy and contribute to the patient's care plan. Input from nurses can be invaluable in devising strategies to incorporate therapy into the patient's daily routine effectively [34].
3. **Encouraging Participation in Physical Therapy:** Nurses play a vital role in encouraging patients to adhere to prescribed physical therapy regimens. This can involve motivating patients during sessions and providing education about the benefits of maintaining physical activity, despite the challenges presented by ALS [34].
4. **Preventing Complications:** ALS patients are at increased risk for complications such as muscle atrophy, contractures, and pressure ulcers. Nurses can help implement preventative measures, including conducting skin assessments, encouraging changes in position, and recommending appropriate supportive devices.
5. **Assistive Devices Education:** Nurses can educate patients and caregivers about the use of assistive devices, such as walkers, wheelchairs, and braces, which can enhance mobility and safety during physical activity. Educating families about these devices can empower them to support the patient's independence [35].
6. **Pain Management:** Chronic pain can come from muscle tension, spasticity, and joint pain in ALS patients. Nurses can assess pain levels and recommend appropriate pain management interventions to support the effectiveness of physical therapy.

### **The Role of Occupational Therapy**

Occupational therapy focuses on helping patients improve their ability to perform activities of daily living (ADLs), such as dressing, bathing, cooking, and self-care. Occupational therapists work to

enhance a patient's independence and adapt the living environment to minimize functional limitations [36].

### **Nursing Interventions Related to Occupational Therapy**

- 1. Identifying Daily Living Challenges:** Nurses must assess the patient's ability to perform ADLs and identify specific challenges. This assessment can help occupational therapists develop targeted interventions that address the patient's individual needs.
- 2. Fostering Independence:** Encouraging patients to engage in adaptive techniques for daily tasks is crucial. Nurses can offer guidance on simpler methods to complete tasks, suggest adaptive tools (e.g., button hooks, dressing aids), and encourage patients to practice these methods during care routines [37].
- 3. Creating a Safe Environment:** Environmental modifications can significantly enhance the safety and efficacy of occupational therapy. Nurses should assess the home environment and recommend necessary changes to reduce fall risks and promote ease of access. This may include the installation of grab bars, use of non-slip mats, and ensuring adequate lighting [37].
- 4. Education for Family Members:** Nurses can provide education and training for the family and caregivers about how to assist ALS patients with ADLs while respecting the patient's dignity and independence. This includes techniques for safe transfer, bathing, and feeding that minimize physical strain on caregivers and ensure patient safety.
- 5. Emotional and Psychological Support:** Occupational therapy often encompasses addressing psychosocial aspects. Nurses can facilitate discussions about emotional support, coping strategies, and referrals to mental health professionals, helping patients and families navigate the psychological impact of ALS.

- 6. Cognitive and Sensory Engagement:** As ALS may affect cognitive functions and sensory inputs, nurses can encourage participation in activities that stimulate cognitive engagement and sensory exploration, thus enhancing the patient's overall mental well-being [37].

### **Integrating Nursing Strategies**

The most effective care for ALS patients often arises from an interdisciplinary collaboration that incorporates the skills of nurses, occupational therapists, physical therapists, and other healthcare professionals. Proper communication is pivotal, as regular meetings can help develop comprehensive care plans and offer insights into the changing needs of the patient.

Additionally, education is vital. Nurses can serve as resource persons, providing both patients and families with important information about ALS, therapeutic interventions, and what to expect during the disease's progression. This knowledge empowers patients and caregivers, enabling them to make informed decisions regarding care strategies [38].

### **Nutritional Considerations and Swallowing Management:**

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a progressive neurodegenerative condition primarily characterized by the degeneration of motor neurons in the brain and spinal cord. This decline leads to widespread muscle weakness, paralysis, and ultimately, the inability to perform voluntary movements, including swallowing and speech. Given that ALS affects not only motor function but also the muscles involved in the ingestion of food and liquids, nutritional management becomes paramount in maintaining the quality of life for affected individuals [39].

The primary impact of ALS on nutrition arises from dysphagia, commonly experienced in patients as the disease progresses. Dysphagia refers to difficulty in swallowing, which can lead to various complications including inadequate nutritional intake, dehydration, aspiration pneumonia, and social isolation due to fear of eating in public. Studies indicate that dysphagia can occur in up to 80% of individuals with ALS, often resulting in

unintentional weight loss, malnutrition, and increased morbidity. Additionally, the hypermetabolic state often seen in ALS patients can increase caloric requirements, further exacerbating the need for careful nutritional planning [40].

A key characteristic of ALS is the progressive weakness of not only swallowing muscles but also the muscles responsible for chewing. This renders a normal diet increasingly difficult to consume and can lead to a range of symptoms including prolonged meal times, choking, and a reduced enjoyment of food. Consequently, a comprehensive understanding of the nutritional needs of ALS patients is crucial for optimizing their health outcomes [41].

### Nutritional Considerations

1. **Caloric Needs:** Due to a hypermetabolic state, individuals with ALS often require higher caloric intake than the average population. It's essential for healthcare professionals to conduct thorough assessments of the patient's body weight, dietary intake, and energy expenditure. Once caloric needs are determined, meals should be planned to ensure adequate energy support that can help maintain weight and muscle function [42].
2. **Macronutrients:** Emphasizing a balanced diet that includes carbohydrates, proteins, and fats is vital. Protein is particularly important for muscle health, but patients must balance their intake to prevent challenges in digesting solid foods. Healthy fats can provide concentrated calories without increasing the volume of food, making them beneficial for those struggling with intake. Carbohydrates should primarily come from whole grains, fruits, and vegetables that also provide vital micronutrients [42].
3. **Hydration:** Maintaining hydration is essential for all patients, but for those with swallowing difficulties, it becomes a complicated task. Dehydration can lead to further complications and can impact medication effectiveness. Monitoring fluid intake through thicker liquids or pureed foods becomes necessary, and the use of

hydration supplements may be recommended in some instances [43].

4. **Supplements:** Nutritional supplements may be beneficial for ALS patients experiencing challenges in maintaining adequate dietary intake. Various products that provide concentrated proteins, vitamins, and minerals can help fill nutritional gaps. These can be in the form of shakes, powders, and fortified foods designed for individuals experiencing dysphagia [44].

### Swallowing Management Strategies

1. **Swallowing Assessments:** Early and comprehensive evaluations by a speech-language pathologist (SLP) can establish a baseline for swallowing capabilities and identify specific challenges faced by the patient. Different assessment methods—including clinical swallowing evaluations and instrumental assessments like videofluoroscopic swallow studies—are critical for understanding the swallowing difficulties and planning appropriate interventions [45].
2. **Diet Modification:** To address dysphagia, dietary modifications are essential. The texture of food should be adjusted based on the level of swallowing difficulty. This can range from pureed diets, which require minimal chewing, to mechanically altered soft foods that ease the swallowing process while being appealing and nutritious. Thicker liquids may also be recommended to help prevent aspiration [45].
3. **Adaptive Equipment:** Utilizing specialized eating utensils and swallowing aids can contribute to a better dining experience. Products such as cups with lids to control liquid flow or weighted utensils designed for patients with limited hand strength can enhance independence at mealtime [46].
4. **Postural Techniques:** Training patients in proper posture during eating can help promote safer swallowing. These techniques often include adjustments such as sitting upright and positioning the head

in a specific manner, which has been shown to reduce the risk of aspiration [47].

5. **Feeding Assistance:** Patients may require additional support during meals. Encouraging family members or caregivers to assist with feeding can enhance meal enjoyment and ensure adequate intake. Educational programs for caregivers can improve their understanding of the challenges faced by ALS patients and provide guidance on effective feeding techniques [47].

### **The Multidisciplinary Approach**

Effective nutritional and swallowing management in ALS requires a collaborative, multidisciplinary approach. This involves a team of healthcare professionals including primary care physicians, neurologists, dietitians, speech-language pathologists, occupational therapists, and nursing staff. Each member plays a vital role in assessing, planning, and implementing interventions tailored to the individual needs of the patient. Regular communication between team members is vital to ensure continuity of care and adapt strategies as the disease progresses.

Moreover, engaging the patient and their family in decision-making is crucial. Patients should be informed of their nutritional and feeding options, empowering them to participate actively in their care. This collaborative approach not only improves nutritional outcomes but also enhances the patient's quality of life and provides necessary emotional support [48].

### **Emotional and Psychological Support for Patients and Families:**

Amyotrophic lateral sclerosis (ALS), commonly referred to as Lou Gehrig's disease, is a progressive neurodegenerative disorder that affects motor neurons in the brain and spinal cord. It leads to muscle weakness, disability, and ultimately, respiratory failure. As patients navigate the complexities of living with ALS, the need for emotional and psychological support becomes vital. This support is equally important for families, who often face overwhelming challenges when caring for a loved one with this debilitating disease. In nursing homes specially designed for ALS patients, these supportive measures play a crucial role in enhancing

the quality of life for both patients and their families [48].

Before delving into the emotional and psychological support required, it is essential to understand the condition itself. ALS typically manifests in adulthood, although it can occur at any age. The exact cause of the disease remains unknown, and there is no known cure. Patients generally experience a gradual but relentless decline in their muscular abilities, leading to various challenges such as difficulty speaking, swallowing, and breathing. Consequently, many families experience increased stress and emotional turmoil as they witness the physical and cognitive decline of their loved ones [49].

Emotional and psychological support is crucial for ALS patients and their families. For patients, the diagnosis can be devastating, leading to feelings of despair, anxiety, and isolation. The knowledge that the condition is progressive can place patients in a state of chronic grief, as they must continuously confront their declining abilities and the loss of independence. Furthermore, patients experience a unique psychological burden, as they must contend with the implications of this irreversible disease, including thoughts about mortality and existential questions about life's meaning [49].

For the patient's family members, the ramifications of living with an ALS patient can be equally profound. Family caregivers often experience caregiver burnout, characterized by physical, emotional, and mental exhaustion from the demands of providing care. The emotional toll of witnessing a loved one's decline can lead to anxiety, depression, and feelings of helplessness.

When families place their loved ones in nursing homes that specialize in ALS care, they often look for more than just physical assistance. They seek an environment that fosters mental well-being and emotional connections. These specialized nursing homes recognize the unique needs of ALS patients and their families, implementing a multi-faceted approach to support [50].

Many nursing homes offer access to licensed mental health professionals who specialize in working with patients with chronic illnesses. These counselors can provide one-on-one therapy for both the patient and family members. Individual counseling allows for processing grief, managing anxiety, and developing

coping strategies. Group therapy sessions can also offer a sense of community and belonging, as patients and families connect with others who understand their struggles [50].

In addition to addressing physical disabilities, palliative care teams consist of interdisciplinary professionals who focus on improving the quality of life for ALS patients. These teams include doctors, nurses, social workers, and chaplains who collaboratively address various aspects of a patient's and family's emotional and psychological plight. Palliative care emphasizes the importance of pain relief and symptom management while providing support for emotional and spiritual concerns [50].

Recreational therapy is another essential component of emotional support within nursing homes for ALS patients. Trained therapists design programs that cater to the individual interests and abilities of patients, encouraging participation in activities that stimulate the mind and promote feelings of joy and accomplishment. Engaging in art, music, or games can serve as effective outlets for expression and emotional release. Social interaction with peers also helps combat the feelings of isolation that often accompany ALS [51].

Education plays a crucial role in emotional support for families. Nursing homes often provide educational resources for family members regarding the progression of ALS, caregiving techniques, and available resources. Knowledge empowers families to make informed decisions and reduces feelings of helplessness. Furthermore, family support groups can create a space where caregivers share their experiences, concerns, and coping strategies, fostering a sense of camaraderie and understanding [51].

For many individuals, spirituality becomes a crucial aspect of coping with illness. Nursing homes may have chaplains or spiritual counselors available to provide emotional support and comfort. These professionals can offer a safe space for patients and families to explore their feelings, beliefs, and fears. Spiritual care can be particularly beneficial in addressing existential questions and facilitating conversations about life, death, and the concepts of hope and acceptance [51].

## **End-of-Life Care and Advance Care Planning in ALS:**

Amyotrophic Lateral Sclerosis (ALS), commonly known as Lou Gehrig's disease, is a progressive neurodegenerative condition that affects motor neurons in the brain and spinal cord. This relentless disease leads to muscle weakness, paralysis, and eventually, respiratory failure. The prognosis for individuals diagnosed with ALS is often grim, with a median survival time of 2 to 5 years from the onset of symptoms. Given the nature of the disease and its trajectory, end-of-life care, and advance care planning become critical components of managing the complexity of ALS, both for patients and their families.

Before diving into end-of-life care and advance care planning, it's important to understand the implications of ALS. The condition results in the degeneration of motor neurons, which are responsible for voluntary muscle movements. Initially, symptoms may appear as muscle twitches, cramps, or weakness in limbs. Over time, the disease progresses, leading to loss of mobility, difficulty swallowing, and impaired respiratory function. While cognitive function often remains intact until the later stages, emotional and psychological challenges can dramatically affect the quality of life of both patients and caregivers [52].

### **The Importance of End-of-Life Care**

End-of-life care encompasses a range of medical, emotional, and spiritual services designed to provide comfort and support to individuals facing a terminal illness. For patients with ALS, the emphasis is on managing symptoms, ensuring dignity, and enhancing quality of life. This care is usually provided through a multidisciplinary team that may include physicians, nurses, social workers, and hospice providers [52].

1. **Symptom Management:** Effective symptom management is a cornerstone of end-of-life care in ALS. As the disease progresses, patients may experience a range of symptoms, including pain, respiratory distress, and muscle spasms. Palliative care approaches aim to alleviate these symptoms, often using opioids or additional medication to manage pain, coupled with interventions to ease

respiratory distress such as non-invasive ventilation. [53]

2. **Emotional Support:** The emotional toll of ALS is profound. For many patients, the realization of impending death can lead to anxiety, depression, and existential distress. Psychological support through counseling services or support groups can offer necessary emotional outlets and coping strategies. It is equally important for caregivers to access support services, as they often face their own challenges of stress, grief, and decision fatigue.
3. **Spiritual Care:** Many patients and families find solace in spiritual support during the end stages of life. Healthcare teams can facilitate conversations with chaplains or spiritual advisors to help patients and their families explore their beliefs, fears, and hopes. This aspect of care can help in addressing existential questions that often arise in the context of terminal illness [53].

### Advance Care Planning

Advance care planning is the process of discussing and documenting future health care preferences. It is essential for individuals with ALS, as the disease course is predictable, allowing patients and families to engage in discussions about goals of care and end-of-life wishes before acute medical crises arise.

1. **Communication of Wishes:** An integral component of advance care planning is openly communicating wishes regarding future treatment options. Patients should discuss their values and preferences regarding interventions like feeding tubes, intubation, and resuscitation efforts. Concentrated discussions with healthcare providers can aid in establishing a care plan that aligns with the patient's goals [54].
2. **Advance Directives:** Advance directives are legal documents that outline a person's wishes regarding medical treatment in the event they are unable to communicate these preferences. Two primary types of documents are durable power of attorney for health care and living wills. Individuals with ALS are encouraged to complete these

documents to ensure their desires are honored, particularly as cognitive function may decline at later stages [54].

3. **Involvement of Family:** Advance care planning is not solely an individual process; it also incorporates family input and involvement. Patients should discuss their plans with family members to ensure everyone understands the patient's wishes, which can prevent conflicts and provide clarity when making decisions.
4. **Revisiting the Plan:** Advance care planning is not a one-time conversation; it should be revisited periodically, especially as the disease progresses and circumstances change. Regular discussions with healthcare providers can help ensure that the advance care plan remains relevant and reflective of the patient's current values and medical situation [55].

### Legal and Ethical Considerations

End-of-life care and advance care planning in ALS do intersect with various legal and ethical considerations. Medical decision-making often involves navigating complex ethical dilemmas, particularly concerning the extent and appropriateness of life-sustaining treatments. Healthcare providers have an ethical obligation to respect patient autonomy, which includes honoring advance directives and discussing the risks and benefits of proposed treatments openly [56].

Additionally, legal guardianship can pose challenges. As ALS progresses, patients may lose the capacity to make decisions for themselves; therefore, having a designated health care proxy is vital. Understanding local laws surrounding advance directives is crucial for ensuring that patient wishes are legally protected and upheld [57].

### Conclusion:

In conclusion, nursing considerations for patients with Amyotrophic Lateral Sclerosis (ALS) are vital to improving patient outcomes and enhancing quality of life throughout the disease's progression. Given the complex and multifaceted nature of ALS, nurses play a critical role in assessing physiological changes, providing effective symptom management, and facilitating communication among interdisciplinary care teams. By incorporating

individualized care plans that address the unique needs of each patient, including physical therapy, nutritional support, and emotional well-being, nurses can help patients maintain as much independence as possible.

Moreover, the importance of emotional and psychological support cannot be overstated, as both patients and their families navigate the significant challenges posed by this debilitating disease. Early integration of palliative care principles and proactive discussions around advance care planning can empower patients and ensure their wishes are respected as they approach end-of-life stages. Ultimately, a holistic and compassionate nursing approach is essential for fostering resilience and comfort in ALS patients, enabling them to face their journey with dignity and support.

#### References:

1. Ozanne, A. O., Graneheim, U. H., & Strang, S. (2013). Finding meaning despite anxiety over life and death in amyotrophic lateral sclerosis patients. *Journal of Clinical Nursing*, 22(15–16), 2141–2149.
2. Burke, T., Elamin, M., Galvin, M., Hardiman, O., & Pender, N. (2015). Caregiver burden in amyotrophic lateral sclerosis: A cross-sectional investigation of predictors. *Journal of Neurology*, 262(6), 1526–1532.
3. Graneheim, U. H., Lindgren, B.-M., & Lundman, B. (2017). Methodological challenges in qualitative content analysis: A discussion paper. *Nurse Education Today*, 56, 29–34.
4. Anderson, N. H., Gluyas, C., Mathers, S., Hudson, P., & Ugalde, A. (2019). “A monster that lives in our lives”: Experiences of caregivers of people with motor neuron disease and identifying avenues for support. *Bmj Supportive & Palliative Care*, 9(4), e27–e27.
5. Josefsson, K., Bomberg, M., & Krans, M. (2018). Creating a sense of security in palliative home care: Interviews with public health nurses. *Nursing and Palliative Care*, 3(1), 1–6.
6. Kläppe, U., Longinetti, E., Larsson, H., Ingre, C., & Fang, F. (2021). Mortality among family members of patients with amyotrophic lateral sclerosis—a Swedish register-based study. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 23(3–4), 1–10.
7. Pusa, S., Hägglund, K., Nilsson, M., & Sundin, K. (2015). District nurses’ lived experiences of meeting significant others in advanced home care. *Scandinavian Journal of Caring Sciences*, 29(1), 93–100.
8. Soundy, A., & Condon, N. (2015). Patients experiences of maintaining mental well-being and hope within motor neuron disease: A thematic synthesis. *Frontiers in Psychology*, 6, 606.
9. Danielsen, B. V., Sand, A. M., Rosland, J. H., & Førland, O. (2018). Experiences and challenges of home care nurses and general practitioners in home-based palliative care—a qualitative study. *BMC Palliative Care*, 17(1), 1–13.
10. de Wit, J., Schröder, C. D., El Mecky, J., Beelen, A., van den Berg, L. H., & Visser-Meily, J. M. (2019). Support needs of caregivers of patients with amyotrophic lateral sclerosis: A qualitative study. *Palliative & Supportive Care*, 17(2), 195–201.
11. Nyholm, L., Salmela, S., Nyström, L., & Koskinen, C. (2018). Sustainability in care through an ethical practice model. *Nursing Ethics*, 25(2), 264–272.
12. Oh, H., & Schepp, K. G. (2013). Wives becoming caregivers: The lived experiences of wives caring for amyotrophic lateral sclerosis patients. *Journal of Hospice and Palliative Nursing*, 15(4), 244–250.
13. Galvin, M., Gavin, T., Mays, I., Heverin, M., & Hardiman, O. (2020). Individual quality of life in spousal ALS patient-caregiver dyads. *Health and Quality of Life Outcomes*, 18(1), 1–13.
14. Norris, S. P., Likanje, M.-F. N., & Andrews, J. A. (2020). Amyotrophic lateral

- sclerosis: Update on clinical management. *Current Opinion in Neurology*, 33(5), 641–648.
15. Lerum, S. V., Solbrække, K. N., & Frich, J. C. (2017). Healthcare professionals' accounts of challenges in managing motor neurone disease in primary healthcare: A qualitative study. *Health & Social Care in the Community*, 25(4), 1355–1363.
  16. Kläppe, U., Longinetti, E., Larsson, H., Ingre, C., & Fang, F. (2021). Mortality among family members of patients with amyotrophic lateral sclerosis—a Swedish register-based study. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 23(3–4), 1–10.
  17. International Association for Hospice and Palliative Care. (2018). *Palliative Care Definition*.
  18. The Swedish Privacy Protection Authority (DPA). (2016). *GDPR, 2016/679 General data protection regulation*.
  19. Oyeboode, J. R., Smith, H.-J., & Morrison, K. (2013). The personal experience of partners of individuals with motor neuron disease. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 14(1), 39–43.
  20. Ozanne, A., & Graneheim, U. H. (2018). Understanding the incomprehensible—patients' and spouses' experiences of comprehensibility before, at and after diagnosis of amyotrophic lateral sclerosis. *Scandinavian Journal of Caring Sciences*, 32(2), 663–671.
  21. Alvariza, A., Mjörnberg, M., & Goliath, I. (2020). Palliative care nurses' strategies when working in private homes—A photo-elicitation study. *Journal of Clinical Nursing*, 29(1–2), 139–12.
  22. Wolfe LF, Joyce NC, McDonald CM, Benditt JO, Finder J. Management of pulmonary complications in neuromuscular disease. *Phys Med Rehabil Clin N Am*. 2012;23:829–853. doi: 10.1016/j.pmr.2012.08.010.
  23. Carreras I, Yuruker S, Aytan N, Hossain L, Choi JK, Jenkins BG, et al. Moderate exercise delays the motor performance decline in a transgenic model of ALS. *Brain Res*. 2010;1313:192–201. doi: 10.1016/j.brainres.2009.11.051.
  24. Drory VE, Goltsman E, Reznik JG, Mosek A, Korczyn AD. The value of muscle exercise in patients with amyotrophic lateral sclerosis. *J Neurol Sci*. 2001;191:133–137. doi: 10.1016/s0022-510x(01)00610-4.
  25. Chiò A, Logroscino G, Hardiman O, Swingler R, Mitchell D, Beghi E, et al. Prognostic factors in ALS: a critical review. *Amyotroph Lateral Scler*. 2009;10:310–323. doi: 10.3109/17482960802566824.
  26. 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;74:1258–1261. doi: 10.1136/jnnp.74.9.1258.
  27. Ashworth NL, Satkunam LE, Deforge D. Treatment for spasticity in amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev*. 2012;2:CD004156.
  28. Lacomblez L, Bensimon G, Leigh PN, Guillet P, Meininger V. Dose-ranging study of riluzole in amyotrophic lateral sclerosis. *Lancet*. 1996;347:1425–1431. doi: 10.1016/s0140-6736(96)91680-3.
  29. Kirkinizos IG, Hernandez D, Bradley WG, Moraes CT. Regular exercise is beneficial to a mouse model of amyotrophic lateral sclerosis. *Ann Neurol*. 2003;53:804–807. doi: 10.1002/ana.10597.
  30. Bello-Haas VD, Florence JM, Kloos AD, Scheirbecker J, Lopate G, Hayes SM, et al. A randomized controlled trial of resistance exercise in individuals with ALS. *Neurology*. 2007;68:2003–2007. doi: 10.1212/01.wnl.0000264418.92308.a4.

31. Mayadev AS, Weiss MD, Distad BJ, Krivickas LS, Carter GT. The amyotrophic lateral sclerosis center: a model of multidisciplinary management. *Phys Med Rehabil Clin N Am*. 2008;19:619–631. xi. doi: 10.1016/j.pmr.2008.04.004.
32. Veldink JH, Bar PR, Joosten EA, Otten M, Wokke JH, van den Berg LH. Sexual differences in onset of disease and response to exercise in a transgenic model of ALS. *Neuromuscul Disord*. 2003;13:737–743. doi: 10.1016/s0960-8966(03)00104-4.
33. Miller RG, Mitchell JD, Moore DH. Riluzole for amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) *Cochrane Database Syst Rev*. 2012;3:CD001447. doi: 10.1002/14651858.CD001447.pub3.
34. Mahoney DJ, Rodriguez C, Devries M, Yasuda N, Tarnopolsky MA. Effects of high-intensity endurance exercise training in the G93A mouse model of amyotrophic lateral sclerosis. *Muscle Nerve*. 2004;29:656–662. doi: 10.1002/mus.20004.
35. Dal Bello-Haas V, Kloos AD, Mitsumoto H. Physical therapy for a patient through six stages of amyotrophic lateral sclerosis. *Phys Ther*. 1998;78:1312–1324. doi: 10.1093/ptj/78.12.1312.
36. Brettschneider J, Kurent J, Ludolph A. Drug therapy for pain in amyotrophic lateral sclerosis or motor neuron disease. *Cochrane Database Syst Rev*. 2013;6:CD005226. doi: 10.1002/14651858.CD005226.pub3.
37. Chio A, Bottacchi E, Buffa C, Mutani R, Mora G. Positive effects of tertiary centres for amyotrophic lateral sclerosis on outcome and use of hospital facilities. *J Neurol Neurosurg Psychiatry*. 2006;77:948–950. doi: 10.1136/jnnp.2005.083402.
38. Wu, Y.-H., Hsieh, H.-Y., Kuo, Y.-L., & Wu, C.-Y. (2022). The experiences and needs of nurses providing home-based palliative care: A qualitative meta-synthesis. *Journal of Palliative Care*, 08258597221105167. doi: 10.1177/08258597221105167.
39. Winther, D., Kirkegaard Lorenzen, C., & Dreyer, P. (2020). Everyday life experiences of close family of people with amyotrophic lateral sclerosis receiving home mechanical ventilation—a qualitative study. *Journal of Clinical Nursing*, 29(13–14), 2306–2316. doi: 10.1111/jocn.15239.
40. Watson, J. (2008). *Nursing: The philosophy and science of caring*. The University Press of Colorado.
41. World Medical Association. (2014). *World Medical Association Declaration of Helsinki: Ethical Principles for Medical Research Involving Human Objects*.
42. Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshe D, Johnston W, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology*. 2009;73:1218–1226. doi: 10.1212/WNL.0b013e3181bc0141.
43. Blackhall LJ. Amyotrophic lateral sclerosis and palliative care: where we are, and the road ahead. *Muscle Nerve*. 2012;45:311–318. doi: 10.1002/mus.22305.
44. Yunusova Y, Green JR, Lindstrom MJ, Ball LJ, Pattee GL, Zinman L. Kinematics of disease progression in bulbar ALS. *J Commun Disord*. 2010;43:6–20. doi: 10.1016/j.jcomdis.2009.07.003.
45. Gaujoux-Viala C, Dougados M, Gossec L. Efficacy and safety of steroid injections for shoulder and elbow tendonitis: a meta-analysis of randomised controlled trials. *Ann Rheum Dis*. 2009;68:1843–1849. doi: 10.1136/ard.2008.099572.
46. Tomik B, Guilloff RJ. Dysarthria in amyotrophic lateral sclerosis: a review. *Amyotroph Lateral Scler*. 2010;11:4–15. doi: 10.3109/17482960802379004.

47. Paganoni S, Deng J, Jaffa M, Cudkowicz ME, Wills AM. Body mass index, not dyslipidemia, is an independent predictor of survival in amyotrophic lateral sclerosis. *Muscle Nerve*. 2011;44:20–24. doi: 10.1002/mus.22114.
48. Pagnini F, Rossi G, Lunetta C, Banfi P, Castelnuovo G, Corbo M, et al. Burden, depression, and anxiety in caregivers of people with amyotrophic lateral sclerosis. *Psychol Health Med*. 2010;15:685–693. doi: 10.1080/13548506.2010.507773.
49. Rivera I, Ajroud-Driss S, Casey P, Heller S, Allen J, Siddique T, et al. Prevalence and characteristics of pain in early and late stages of ALS. *Amyotroph Lateral Scler Frontotemporal Degener*. 2013;14:369–372. doi: 10.3109/21678421.2012.751614.
50. Vitacca M, Comini L, Tentorio M, Assoni G, Trainini D, Fiorenza D, et al. A pilot trial of telemedicine-assisted, integrated care for patients with advanced amyotrophic lateral sclerosis and their care-givers. *J Telemed Telecare*. 2010;16:83–88. doi: 10.1258/jtt.2009.090604.
51. Gracies JM. Physical modalities other than stretch in spastic hypertonia. *Phys Med Rehabil Clin N Am*. 2001;12:769–792.
52. Ho DT, Ruthazer R, Russell JA. Shoulder pain in amyotrophic lateral sclerosis. *J Clin Neuromuscul Dis*. 2011;13:53–55. doi: 10.1097/CND.0b013e31821c5623.
53. Lillo P, Mioshi E, Hodges JR. Caregiver burden in amyotrophic lateral sclerosis is more dependent on patients' behavioral changes than physical disability: a comparative study. *BMC Neurol*. 2012;12:156. doi: 10.1186/1471-2377-12-156.
54. Baldinger R, Katzberg HD, Weber M. Treatment for cramps in amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev*. 2012;4:CD004157. doi: 10.1002/14651858.CD004157.pub2.
55. Green S, Buchbinder R, Hetrick S. Physiotherapy interventions for shoulder pain. *Cochrane Database Syst Rev*. 2003:CD004258. doi: 10.1002/14651858.CD004258.
56. Green S, Buchbinder R, Hetrick S. Acupuncture for shoulder pain. *Cochrane Database Syst Rev*. 2005:CD005319. doi: 10.1002/14651858.CD005319.
57. Bede P, Oliver D, Stodart J, van de Berg L, Simmons Z, O'Brannágain D, et al. Palliative care in amyotrophic lateral sclerosis: a review of current international guidelines and initiatives. *J Neurol Neurosurg Psychiatry*. 2011;82:413–418. doi: 10.1136/jnnp.2010.232637.