National Hospice and Palliative Care Organization

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Palliative Care Resource Series

THE ROLE OF PALLIATIVE CARE IN TREATING AMYOTROPHIC LATERAL SCLEROSIS (ALS)

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INTRODUCTION

This paper will describe ALS and introduce the role of Palliative Care for patients with ALS.

DEFINITION, PROGNOSIS AND DISEASE PROGRESSION

Amyotrophic Lateral Sclerosis (ALS) is a progressive, neurodegenerative disorder, resulting from ongoing destruction of both upper and lower motor neurons in the brain and spinal cord, resulting in a combination of upper motor neuron (spasticity, clonus, and hyperreflexia) and lower motor neuron (weakness, atrophy, and fasciculation) findings. At present, there is no known cure, and the disease is ultimately fatal.

Prognosis

Mean survival with ALS is on the range of 3-5 years, although there is high variability, and prognosis is in part modifiable by interventions such as mechanical ventilation. Approximately 10% of patients with ALS survive longer than 10 years. Predictors of a poorer prognosis include older age of onset, more rapid progression at disease onset, and involvement of bulbar muscles at onset. (**2**)

Natural History

Approximately 80% of cases of ALS begin with weakness and atrophy in one limb, with symptoms eventually spreading to the other limbs and axial muscles, along with progressive impairment of the oral, pharyngeal, and respiratory muscles. Approximately 20% of cases onset with impairment of bulbar function, and tend to have a more aggressive course. (**1**)

As the disease progresses, nearly all voluntary muscles of the body are affected, with relative sparing only of the extraocular movements and muscles controlling bladder and bowel function. Ultimately, this results in near total paralysis, complete dependency for all activities of daily living, and loss of the ability to speak, swallow, and breathe, with death typically occurring from respiratory failure.

PALLIATIVE CARE IN ALS

Palliative care plays a multifaceted role in ALS, with involvement in symptom control, goals of care discussions, patient and family support, and assistance with transitions of care. Evidence has demonstrated improvement in symptom burden and quality of life, with early referral to specialist palliative care in ALS and other neurodegenerative diseases. (**3**)

Multidisciplinary ALS Care

Participation in a multidisciplinary ALS clinic, in which a person with ALS has access to practitioners from multiple disciplines, has become a standard of care in ALS. Typical disciplines participating in an ALS clinic include physicians of varying specialties (neurology, pulmonology, gastroenterology, and palliative care), nurses, social workers, respiratory therapists, dietitians, psychologists, physical and occupational therapists, and speech and language pathologists.

Evidence has demonstrated that persons with ALS benefit from participation in a multidisciplinary ALS clinic, showing improvements in both survival time and quality of life (**4, 5**). It has been theorized that these benefits may relate to increased and earlier access to specialized therapies, and improved coordination of care.

Disease Modifying Pharmacotherapy

To date, **Riluzole**, a glutamate inhibitor, is the only approved treatment for ALS. Clinical trials have demonstrated that it can prolong survival (or time to tracheostomy) by an average of 2-3 months, along with a modest slowing of decline in bulbar and limb function. (**6**) However, it does not reverse or improve existing symptoms. Common side effects are GI symptoms and fatigue. Before and during treatment with riluzole, patients may need frequent blood tests to monitor liver function.

Symptom Management

Due to the lack of disease modifying therapies, and the range of symptoms that occur with ALS, the majority of therapy in ALS is palliative in nature.

Sialorrhea is a significant concern in ALS, resulting in both embarrassment and aspiration risk. Excessive drooling can be managed by minimization of sweet and sour foods which stimulate the salivary glands, use of suction devices, and pharmacotherapy, including tricyclic antidepressants and anticholinergic medications. Botulinum toxin and radiation therapy may provide benefit in cases of severe drooling.

Thickened Secretions can result in difficulty with expectoration and mucous plugging, and may be exacerbated by anticholinergic medications used to treat sialorrhea. Thickened secretions can be managed by increasing fluid intake, air humidification, and use of mucolytics, including guaifenesin and N-acetyl cysteine. Expectoration of secretions, which becomes progressively more difficult due to increasing muscle weakness, can also be facilitated by use of a cough assist device.

Pain has a significant impact on quality of life in ALS (**7**). Sources of musculoskeletal pain include immobility and increased pressure on bones and joints resulting from muscle atrophy. Such pain can be improved with careful attention to positioning, gentle stretching and range of motion exercises, and analgesics, including acetaminophen, NSAIDS, and opioids.

Muscle Cramps occur in up to 95% of cases of ALS, and may be improved with stretching, repositioning and massage. Although medications including antiepileptics (carbamazepine, levetiracetam, and phenytoin), benzodiazepines, and baclofen have been used for management of cramps, evidence for their benefit is mainly anecdotal. Recently, mexiletine, a voltage gated sodium channel blocker, has shown promise in a phase II randomized controlled trial for reduction of frequency and intensity of muscle cramps in ALS (**8, 9**).

Spasticity, another source of pain in ALS, can also result in falls, worsen contractures, and interfere with care. Non pharmacologic interventions for spasticity include stretching, repositioning, and massage. Baclofen and tizanidine, while they may improve spasticity, should be used with caution, as they can also worsen muscular weakness.

Pseudobulbar Affect, characterized by sudden outburst of crying or laughing out of proportion to the situation or the person's underlying emotional state, results in ALS from degeneration of the corticobulbar tracts. Commonly mistaken for depression, pseudobulbar affect leads to embarrassment, social isolation, and is associated with a reduced quality of life (**10**). In the presence of pseudobulbar affect, patients and families should be provided with education and reassurance regarding the existence of this symptom. Pharmacotherapy involves use of combination dextromethorphan-quinidine (20 mg/10 mg) (**11**). Tricyclic antidepressants and SSRI's can also be used as alternatives.

Cognitive Impairment occurs in approximately 50% of people with ALS, and may range from subtle findings to overt dementia. Deficits typically present as executive dysfunction (planning, organization, and multitasking, mental flexibility) with relative sparing of memory. Approximately 15% of people with ALS develop frontotemporal dementia, which in addition to deficits in executive functioning, can result in significant personality and behavioral disturbances. The risk of developing cognitive impairment and dementia in ALS necessitates ongoing assessment of capacity for medical decision making in the context of the ongoing goals of care discussions which ALS typically requires.

Communication Deficits in ALS

Due to progressive impairment of the oropharyngeal muscles, persons with ALS develop gradually worsening dysarthria, eventually progressing to a total inability to produce speech. Early compensatory strategies include overarticulating each sound produced, speaking more slowly, and having listeners repeat the understood words so the person with ALS can focus on repeating only the unclear words.

As the disease progresses, **augmentative** (devices and strategies used to support or enhance impaired speech) **and alternative** (devices and strategies used in the absence of speech) **communication** plays a growing role. A range of augmentative and alternative communication techniques and devices exist, ranging from low tech letter and picture boards to high tech eye gaze activated speech generating devices. While communication needs change throughout the disease, early implementation of augmentative/alternative communication strategies improve mood and quality of life in patients and caregivers. (**12**)

ALS AND NUTRITION

Multiple barriers to feeding in ALS (progressive dysphagia, physical barriers to feeding such as arm weakness, fear of choking) result in significant time and energy spent, and great difficulty maintaining adequate nutritional status. This is frequently a significant source of concern for patients and families. BMI < 18.5 is associated with shorter survival. (**13**)

Interventions to maximize nutritional status in the earlier stages of disease include addition of calorie rich foods and oral supplements, alteration of food consistency, and involvement of a swallowing therapist who can instruct in various techniques to facilitate swallowing.

Feeding tubes: Placement of a feeding tube is a highly personal decision which should be discussed early in the course of the disease and then periodically revisited. Placement, if consistent with goals of care, should be considered in the setting of weight loss, insufficient oral intake, dehydration, or choking episodes.

Benefits of feeding tube placement include stabilization of weight loss, improvement in overall survival time, possible improvement in quality of life, and ability to focus on eating for pleasure without the pressure to maintain adequate nutrition through the oral route only. Risks include infection, blockage and ongoing risk of aspiration. (**14**)

Timing of placement should also be considered within the context of the ongoing respiratory decline associated with ALS. Feeding tubes should be placed before respiratory vital capacity falls below 50% of predicted, as there is an increased procedure risk with declining respiratory status. (**14**)

RESPIRATORY STATUS IN ALS

As ALS progresses, weakening muscles of respiration result in worsening hypoventilation. Early signs of respiratory involvement include headaches, daytime fatigue, orthopnea, and nighttime dyspnea. Respiratory decline is best assessed by serial vital capacities (<50% of predicted is frequently associated with respiratory symptoms, and <30% of predicted is associated with respiratory failure or death) (**15**).

Use of **non-invasive positive pressure ventilation** can result in reduced symptom burden and improvements in quality of life and survival time. It tends to be less well tolerated in bulbar ALS and dementia.

Dyspnea: In addition to the use of a non-invasive positive pressure ventilator, dyspnea should be managed by addressing other potential contributing factors such as respiratory infections, pulmonary edema or bronchospasm. Cautious use of opioids and benzodiazepines can also provide symptomatic benefit. Supplemental oxygen is not recommended in the absence of overt hypoxia (**16**).

Mechanical Ventilation: Potential use of mechanical ventilation, when initiated in the setting of ALS associated respiratory failure, should ideally be discussed early in disease course and then periodically revisited. Mechanical ventilation maximizes the potential for long term survival, increases airway protection, and for some, allows for continued meaning and acceptable quality of life. However, initiation of mechanical ventilation also brings with it loss of speaking ability, increasing financial concerns and caregiver burden, risk of nursing home placement, and for some, a loss in quality of life. Ultimately, <10% of persons with ALS opt for tracheostomy/ventilator use. When discussing initiation of mechanical ventilation, one should also attempt to clarify any circumstances under which the person would wish for the mechanical ventilator to be withdrawn.

SUPPORTIVE RESOURCES FOR ALS

Identification of available support services for both patients and families is central to the management of ALS.

Benefits for Veterans: Because studies have demonstrated an increased frequency of ALS with military services (independent of location, timing, or military branch), ALS is considered a service connected diagnosis (**17**).

This entitles persons who developed ALS either during or after a period of military services (defined as 90 days or move of active duty) to a range of benefits including:

- Coverage of medical care, medications, and medical/adaptive equipment
- Disability compensation
- Special monthly compensation
- Grants of adaptive housing and automobiles
- Aid and attendant allowance
- Dependency and indemnity compensations.

The ALS Association, a national organization with locally based chapters, in addition to funding ALS research, also sponsors ALS multidisciplinary clinics, provides extensive supportive educational materials for patients and caregivers, and supplies necessary medical equipment and communication devices through equipment lending libraries.

It is helpful for the palliative care practitioner to become familiar with other locally based organizations that may provide support to persons with ALS and their families/caregivers. Patients and families may also benefit from in person and online support groups and communities.

END OF LIFE CARE

As end-of-life approaches, common concerns, such as fear of choking and suffocation, should be addressed, and reassurance provided regarding the ability to manage symptoms such as dyspnea, and secretions.

Both persons with ALS as well as their caregivers and family members can benefit from the interdisciplinary services that hospice provides during the later stages of the disease. **Hospice referral** should be considered in the following circumstances:

1) Crucial impairment in breath capacity (evidenced by vital capacity <40% of predicted, dyspnea or other signs of respiratory decline) and mechanical ventilation is declined), or
2) Rapid progression of ALS or critical nutritional impairment (evidenced by dysphagia, progressive weight loss, bed bound status, and dependency for activities of daily living).

SUMMARY

- Patients with ALS benefit from a multidisciplinary approach to care.
- Goals of care should be discussed with patients and their families early on and throughout the course of the disease.
- Despite the lack of a cure for ALS, interventions do exist that may prolong survival and improve quality of life.

CASE STUDIES

Case studies can be found in the accompanying PowerPoint presentation.

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