Clinical Considerations for Treating the Dental Patient with ALS

A Peer-Reviewed Publication
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Abstract
Amyotrophic lateral sclerosis (ALS) is a progressive degenerative disorder that affects the nervous system, particularly the motor neurons. It may affect either spinal or bulbar neurons, which classifies how an individual's symptoms present. The prognosis for individuals with ALS is not promising, usually resulting in death within three to five years of diagnosis. Most management of amyotrophic lateral sclerosis focuses on palliative care, determined by the affected individual's ongoing symptoms. An essential component of palliative care is management of the individual's oral conditions. Manifestations of ALS may affect the dental practitioner's care of patients with this disease. These modifications will be dependent upon the individual's current symptoms and needs.

Educational Objectives
At the conclusion of this educational activity participants will be able to:
1. Describe amyotrophic lateral sclerosis (ALS) and the manner(s) in which it may present
2. Recognize common types of palliative care offered to individuals with ALS
3. Describe the symptoms associated with spinal and bulbar degeneration
4. Integrate appropriate modifications into the dental treatment of patients with ALS

Author Profile
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Amyotrophic lateral sclerosis (ALS) is a progressive degenerative disorder that affects the nervous system, particularly the motor neurons. It may affect either spinal or bulbar neurons, which classifies how an individual’s symptoms present. The prognosis for individuals with ALS is not promising, usually resulting in death within three to five years of diagnosis. Most management of amyotrophic lateral sclerosis focuses on palliative care, determined by the affected individual’s ongoing symptoms. An essential component of palliative care is management of the individual's oral conditions. Manifestations of ALS may affect the dental practitioner’s care of patients with this disease. These modifications will be dependent upon the individual's current symptoms and needs.

**Introduction**
Amyotrophic lateral sclerosis (ALS), commonly referred to as Lou Gehrig’s disease, is a progressive degenerative disorder that affects the nervous system, particularly the motor neurons.\(^1\)\(^,\)\(^2\) It may affect either spinal or bulbar neurons, which classify how symptoms present in the individual.\(^1\) Currently, the cause of ALS is unknown, although research indicates the possibility of genetic and environmental factors affecting its incidence.\(^1\)\(^,\)\(^2\) It is important for health care practitioners to understand amyotrophic lateral sclerosis to enable them to make modifications to patient care as needed.

**Epidemiology and Prognosis**
The sporadic form of amyotrophic lateral sclerosis, characterized by motor neuron degeneration and premature death, comprises the majority of all ALS cases.\(^6\) Juvenile-onset ALS, where disease onset occurs before age 30, comprises only five percent of all ALS cases. Familial ALS occurs only five to ten percent of the time. The more common sporadic form comprises the vast majority of cases. The average onset of sporadic ALS occurs between 55 and 65 years, with a median age of 64 years.\(^6\) There is a slightly higher incidence of sporadic ALS in men than women, with a ratio of 1.5:1.\(^5\) The age of onset of familial ALS occurs approximately ten years earlier than sporadic ALS, with males and females equally affected.

The prognosis for individuals with amyotrophic lateral sclerosis is not promising. Although there are possible treatments on the horizon, they are still in early developmental stages. This rapidly progressive disease usually results in death within three to five years of diagnosis, primarily from respiratory complications.\(^6\) Slower disease progression can occur in younger individuals and those whose disease first presents in the extremities. Only ten percent of individuals with ALS live longer than eight years.\(^5\)

**Management of the Disease**
Currently, management of ALS patients primarily focuses on palliative care.\(^6\)\(^,\)\(^7\)\(^,\)\(^8\)\(^,\)\(^9\) Palliative care is a philosophy of care that addresses the prevention and relief of suffering.\(^6\) Symptoms of ALS greatly impact an individual's quality of life and, therefore, must be continually managed.\(^8\) A multidisciplinary approach to care provides the individual with ALS with the most optimal care choices, thus enhancing their quality of life.\(^6\) The National Consensus Project Clinical Practice Guidelines for Quality Palliative Care (NCPQPC) outline eight domains of palliative care.\(^6\) These include: (1) physical care, such as pain management and physical symptoms; (2) psychological care that addresses depression, loss, grief, and other symptoms experienced by both the patient and his/her loved ones; (3) social systems available for the patient; (4) faith-based beliefs; (5) cultural norms that are integrated into patient care; (6) ethical and legal choices for the patient and family to define; (7) designation of property and other legal aspects of care for the imminently dying patient; and (8) structure and processes associated with the outcomes of ALS.\(^6\)\(^,\)\(^8\)

The affected individual’s ongoing symptoms determine the type of palliative care provided.\(^7\) Some common types of palliative care offered to individuals with amyotrophic lateral sclerosis may include medication for the following symptoms: depression or anxiety, drooling, bronchial secretions, pain, pathological laughing, constipation, gastro-esophageal reflux, sleep disturbances, and venous thrombosis or pulmonary embolism. In addition, dietary changes, increased hydration, physiotherapy, and/or stretching may be utilized to address other symptoms.\(^7\)

Management of oral conditions may be required as part of the palliative treatment of an individual with ALS. Oral conditions may arise due to the effects of medication, sialorrhea caused by ineffective clearance of saliva, thickened mucus, and/or xerostomia that arises from mouth breathing.\(^10\) Management of these oral conditions is essential for patient comfort. Adequate hydration and the use of a room humidifier can be effective in decreasing xerostomia from mouth breathing during sleep.\(^10\) In addition, an expectorant or beta-adrenergic blocking agents, such as propranolol or metoprolol may be effective in 75% of patients experiencing thickened secretions.\(^10\) To address sialorrhea, an anticholinergic, such as glycopyrrolate can be 90% effective.\(^10\) Another option for sialorrhea is the injection of botulinum toxin into the parotid and submandibular glands. However, this type of care may result in temporary side effects, such as bulbar and masseter muscle weakness.\(^10\)
Table 1

<table>
<thead>
<tr>
<th>ALS Symptom</th>
<th>Treatment Modification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Advanced disease</td>
<td>Care provider to learn oral home care</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>Semi-supine treatment position</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>Nutritional counseling</td>
</tr>
<tr>
<td>Excessive saliva</td>
<td>Additional practitioner to assist with HVE</td>
</tr>
<tr>
<td>Muscle weakness/spasticity</td>
<td>Shorter appointment time or frequent breaks during appointment</td>
</tr>
<tr>
<td>Muscle weakness/spasticity</td>
<td>Bite block/assistance with holding mouth open</td>
</tr>
</tbody>
</table>

Currently, the only medication approved to slow the course of the amyotrophic lateral sclerosis disease process is riluzole.³, ¹¹ Riluzole, originally developed as an anti-epileptic drug, inhibits the release of glutamate. However, this drug’s mechanism of action in ALS is currently unknown.³ Presently, the use of riluzole prolongs the life of an individual with ALS by approximately four months. Its safety and efficacy have been established for individuals with ALS, including those with advanced disease and in elderly patients.³ More than half of ALS patients in the United States utilize it.³

Symptoms Associated with Spinal Degeneration

Individuals that experience degeneration of the spinal neurons usually exhibit weakness or spasticity of the lower limb muscles. As a result, they may also experience disturbances in gait.¹ Initial presentation of symptoms in an individual with spinal neuron involvement includes changes in the functional strength and coordination of lower extremity muscles. Eventually this progresses to muscles of the upper extremities, and finally to the bulbar muscles.¹

Symptoms Associated With Bulbar Degeneration

Individuals with involvement of the bulbar neurons present initially with speech problems resulting in dysarthria (motor speech disorder), deterioration of respiratory function, and weakness of the muscles of deglution, which results in dysphagia.¹ Dysphagia occurs in each ALS patient with bulbar involvement and eventually occurs in all ALS patients, regardless of the time of disease onset. Dysphagia in ALS patients is characterized by impairment of the oral and pharyngeal phases of swallowing. Patients with amyotrophic lateral sclerosis present with abnormal lingual movement. This abnormality makes the act of swallowing effortful, inefficient, and fatiguing.¹, ¹² Decreased tongue strength may also result in decreased frequency of swallowing. This leads to accumulation of saliva in the oral cavity, which can result in drooling, aspiration, and coughing.⁹ Salivary control, along with other secretions, is a common challenge for patients with ALS.¹

Dental Treatment Modifications

Manifestations of amyotrophic lateral sclerosis affect the dental practitioner’s care of patients with ALS (Table 1). Due to the high occurrence of dysphagia in individuals with ALS,¹, ¹² modification of patient treatment in the dental chair is often necessary. First, the dental practitioner may need to modify patient positioning. Since pooling of saliva can occur in the oral cavity, the patient may be more comfortable in a semi-supine position to help control aspiration of excess saliva. In addition, the dentist and dental hygienist may need another practitioner present to assist him/her with high volume evacuation to help keep saliva from pooling and interfering with dental work, and to make the patient more comfortable. Third, shorter appointments, or appointments with built in breaks, may be necessary for the patient with ALS. Since individuals with ALS experience muscle weakness and spasticity,¹ it can be difficult for them to keep their mouths open for the time necessary to complete treatment during a regular appointment. Treatment may need to be broken down into smaller divisions to allow for shorter appointment times. Frequent breaks may also need to occur during the appointment. In addition, a bite block may be necessary to hold the mouth open for the duration of the dental procedure.

Many ALS patients have difficulty swallowing,³ which can affect an individual’s intake of food. As a result, nutritional counseling by the dental practitioner may be warranted. Oral health education regarding nutrition and its effects on the oral cavity is important in emphasizing how food intake can affect the health of an individual’s intraoral tissues. The health of the intraoral tissues can also influence the individual’s food choices and nutrient intake. This important cycle should be addressed with ALS patients so that the dental professional and patient can work together to devise a nutritional plan that will complement the patient’s current physiological needs.

Lastly, oral health education may need to address modification of home care due to loss of muscular function.¹ These alterations will include modification of the daily homecare regimen, such as modified toothbrushes or brushing techniques. In addition, the introduction of auxiliary aides may enable the patient to enhance biofilm removal. Individuals who have lost substantial motor function or who experience severe spasticity may need to bring a care provider, such as a family member, to their dental appointment to assist in the nutritional and homecare planning.

Conclusion

Amyotrophic lateral sclerosis, a degenerative neural disorder, ultimately affects multiple body systems. Its poor prognosis and rapid progression lead to quick, visible symptoms.³ The initial symptoms vary according to the type of neural involvement in the individual. All patients afflicted with ALS show signs of dysphagia and other oral complications. The dental implications of ALS span both home and professional clinical care. As such, it is important for the dental practitioner to have an understanding of the disease and the modifications needed to treat patients with ALS.
Questions

1. What is the name for the progressive degenerative disorder that affects the nervous system, particularly the motor neurons?
   a. Amyotrophic latent sclerosis
   b. Amyotrophic lateral spasticity
   c. Amyotrophic lateral sclerosis
   d. Alveolar lateral sclerosis

2. What form of ALS comprises the majority of ALS cases?
   a. Juvenile onset
   b. Sporadic
   c. Familial
   d. Geriatric onset

3. Which of the following groups is most often inflicted with sporadic ALS?
   a. Men
   b. Women
   c. Children
   d. Men and women are equally inflicted

4. Death in patients with ALS occurs primarily from which of the following complications?
   a. Cardiac
   b. Skeletal
   c. Nervous
   d. Respiratory

5. What determines the type of palliative care provided to individuals with ALS?
   a. Length of time afflicted with ALS
   b. Individual’s current financial situation
   c. Individual’s ongoing symptoms
   d. Individual’s prognosis

6. Which of the following oral conditions may arise as a result of medications used for ALS patients?
   a. Thickened mucus
   b. Xerostomia
   c. Sialorrhea
   d. All of the above

7. Management of sialorrhea may be achieved through which of the following?
   a. Use of an anticholinergic
   b. Chewing medicinal gum
   c. Use of a room humidifier
   d. Adequate hydration

8. A medication currently approved to slow the course of ALS is:
   a. Metoprolol
   b. Warfarin
   c. Riluzole
   d. There is no approved medication

9. Individuals with degeneration of the spinal neurons initially exhibit:
   a. Difficulty maintaining control of the head and neck
   b. Weakness or spasticity of the lower limb muscles
   c. Problems with speech
   d. Tingling or numbness of the hands

10. Individuals with degeneration of the bulbar neurons initially exhibit:
    a. Difficulty maintaining control of the head and neck
    b. Weakness or spasticity of the lower limb muscles
    c. Problems with speech
    d. Tingling or numbness of the hands

References

Clinical Considerations for Treating the Dental Patient with ALS

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Objective #3: Yes No  Objective #4: Yes No

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