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Communication Management in Amyotrophic Lateral Sclerosis: The Role of the Speech-Language Pathologist during Disease Progression

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ABSTRACT

During the progression of Amyotrophic Lateral Sclerosis (ALS), augmentative and alternative communication (AAC) devices will eventually become necessary to preserve communication between the affected individual with ALS and his or her family members and loved ones. Speech-language pathologists play a critical role in communication management and family education regarding communicative strengths and weaknesses and in determining the need for an alternative mode of communication during the later stages of disease progression. The aim of this paper is to explore the speech-language pathologist's changing role throughout ALS progression in relationship to the impact that the disease has on speech production and overall communication functioning.

INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) or Lou Gehrig's disease is a progressive disease that affects the motor neurons in the brain and spinal cord.[1] The incidence of ALS is approximately two individuals per every 100,000. Estimates suggest that roughly 30,000 Americans are currently living with ALS.[2] ALS is more common among men than women and symptoms often arise between the ages of 55 and 75, with prognosis being poorer at more advanced ages. ALS can occur in individuals in their 30s and 40s, though the incidence is much lower.[3] Several hallmark characteristics exist among individuals with ALS including: progressive muscle weakness and atrophy, and hyperreflexia and fasciculations, all a result of damage to both upper and lower motor neurons. [2]

The disease course in ALS is quite variable.[1] Disease progression may range from 2 to 10 years, with most individuals succumbing to the disease in approximately 5 years post onset.[4] Some of the earliest symptoms may include twitching, cramping, stiffness of muscles, muscle weakness, slurred speech, and difficulty chewing.[5] ALS symptoms typically begin in the limbs (upper or lower) thus resulting in difficulties with walking or grasping common objects. There are two forms of ALS -- bulbar and spinal. These forms are distinguished by location of pathology and clinical presentation. The term "bulbar" in ALS refers to neuronal damage in the "bulb" area of the brain stem which controls common motor functions critical to communication.[6] Patients with bulbar or brainstem forms of the disease frequently exhibit difficulty speaking, swallowing, and chewing.[4] In the spinal form of ALS, symptoms reflect a disruption of the motor signals to muscles that result in clinical signs such as muscle weakness, muscle atrophy, and hyperactive reflexes. Individuals with the spinal form of ALS frequently complain of stumbling or tripping while running or walking and difficulty with motorically simple tasks such as buttoning a shirt.[7] This review will primarily focus on the bulbar form of ALS which has a significant impact on the muscles associated with respiration, speech, and swallowing function.

The complexity and variation of this disease requires the expertise of many different team members, all working together to determine the appropriate interventions for a particular patient and his or her family members or caregivers.[8] The interdisciplinary team may include physicians, speech-language pathologists, occupational therapists, physical therapists, nutritionists, social workers, counselors, and representatives or assistants familiar with assistive technology. Another critical member of this team is the patient with ALS and his/her caregiver and/or family members.[2]

COMPLICATIONS IN ALS

Respiratory Issues

As individuals with ALS progressively lose control over muscles of inspiration and expiration, mechanical ventilation may eventually be warranted. Most deaths occur as a result of cachexia or respiratory failure and extreme weight loss.[4] There are both non-invasive and invasive options for breathing support.[6] Non-invasive options do not involve any surgical intervention. Respiratory devices such as continuous positive airway pressure (CPAP) machines are generally contraindicated for breathing management in individuals with ALS because the increase in oxygen levels can result in a significant increase in the amount of muscle effort needed for expiration.[9] CPAP devices may, however, be used to combat sleep apnea and preserve muscle strength when respiratory muscle weakness is not yet a significant issue for the individual with ALS.[10] Bilevel positive airway pressure (BiPAP) machines are also used with individuals with ALS as a mechanism to alternate the delivery of air pressures needed for inspiration and expiration.[9] BiPAP devices have been shown to improve arterial blood gas levels and improve daily respiratory functioning.[10]

Individuals with ALS are required to decide on the type of breathing support they would like implemented when they become unable to breathe without assistance. Weakened muscles of respiration have detrimental effects for the individual with ALS in several areas. First, the immune system can become compromised limiting the ability to clear secretions that may lead to infections.[9] Second, many individuals are at an increased risk for aspiration pneumonia because they are unable to expel aspirated food or liquid. Third, because the body is also working harder to exchange oxygen and carbon dioxide, affected individuals experience significantly increasing fatigue levels. As a result, when respiratory distress occurs, some individuals require a surgical procedure designed to create a new airway, or tracheostomy, which will allow the individual to receive oxygen through a ventilator. The ventilator can be modified to meet very specific respiratory needs.[9] The addition of the ventilator then presents new challenges for the individual with ALS as it changes the way they breathe. To address these issues, speech-language pathologists must play an integral role in explaining the physiological changes that occur following the tracheostomy and use of the ventilator. The speech-language pathologist must also stress the importance of oral care for patients on ventilators.

Communication and Swallowing Disturbances

Communication and swallowing abilities are common throughout the progression of this neuromuscular disease. Eventually, speech will become increasingly hard to produce and swallowing function may become unsafe.[5] An interesting hallmark characteristic of ALS is that many individuals remain cognitively intact.[11] That is, they are aware of their speech-related errors and subsequent reductions in intelligibility. Yet, research has shown that areas outside of the motor system may be affected in individuals with ALS. Dementia may be evident within a year of the onset of physical symptoms; however, fewer than 5% of all patients eventually show clinical signs of dementia.[12] Adequate deficit awareness is common among individuals with ALS and their awareness of their deficits contributes to their ability to self-monitor communication breakdowns and correct errors. Additionally, patients typically adjust to their eventual loss of speech.[7] It is in this area that the speech-language pathologist will educate the individual and caregivers in terms of communication options and the reasons behind loss of speech function. Additionally, speech-language pathologists are called upon to play a crucial role in determining appropriate communication and swallowing strategies that best suit the needs of patients and families.[5]

Individuals with ALS experience significant declines in communication abilities throughout the progression of ALS. Perceived differences arise in the areas of speech intelligibility, voice quality, resonance, volume and rate of speech, and length of utterance.[9]The main deterrent of intelligibility is dysarthria. Dysarthria, a disorder of motor speech production due to weakness, incoordination, or paralysis is quite common in ALS.[2,13] Dysarthria is often the first symptom of the bulbar form of this neuromuscular disease. Dysarthria results in either weak or spastic movements thus having detrimental effects on several systems involved with speech production (i.e. respiration and phonation). The type of dysarthria present in ALS depends upon the areas affected by the disease. For example, spastic dysarthria occurs following damage to upper motor neurons while flaccid dysarthria is caused by lower motor neuron damage.[13] As ALS progresses, many individuals are required to use augmentative and alternative communication devices to express themselves. For example, speech-language pathologists must decide which patients are candidates for speaking valves that allow them to produce voice while breathing through a trach tube or whether augmentative communication devices should be introduced as a primary means of communication.[14]

Problems with swallowing function create additional quality of life issues. The muscles of the oral cavity, pharynx and larynx deteriorate, changing the individual's ability to manipulate and transport a bolus, while laryngeal muscle weakness can leave the airway vulnerable to food and liquid entry.[14] Speech-language pathologists work closely with dieticians and the patient with ALS to determine the appropriate consistency and amount of foods and liquids the individual is to have. Many individuals with ALS require meals that are easy to swallow to limit the amount of fatigue that can occur during meal time. To accommodate the changes in swallow function, individuals with ALS are counseled to make adjustments such as moistening foods and avoiding dry, tough foods that require excessive mastication. Swallowing assessments using modified barium swallow (MBS) studies may be performed regularly to identify specific breakdowns during the swallowing process. Individuals with ALS are oftentimes at risk for aspiration of thin liquids particularly as the disease progresses. Speech language pathologists may train individuals with ALS to use compensatory strategies such as the "chin-tuck" or "head turn" to reduce the risk of these potentially dangerous problems.[4] To compensate for swallowing difficulties, many patients require the placement of a percutaneous endoscopic gastrostomy (PEG) feeding tube particularly as the disease progresses. Placement of the PEG can improve quality of life by relieving the fatigue and frustration associated with muscle weakness that many individuals experience during mealtime. Such adjustments can result in a more positive mealtime experience for the individual with ALS.[4]

Speech Production

Speech-language pathologists play a critical role in the maintenance of speech production during the disease progression. The speech-language pathologist is primarily responsible for monitoring changes in speech and swallowing function. Kent and Sufit (1991) suggest that measures of intelligibility, vocal or phonatory function, respiration, and articulation are be completed in patients with ALS to gauge the severity of impairment associated with the disease and disease progression.[15] For example, during the course of the disease, speech production typically deteriorates requiring the completion of a variety of speech production tasks to measure change in performance. Measurements may include range of motion of the articulators as well as the speed and strength with which the patient is able to control the articulatory muscles. It is important to note here that the speech-language pathologist should reiterate to patients and their families that speech therapy will not be aimed at exercising the muscles of speech because the use of repetitive exercises would only cause more rapid deterioration of muscle function. Individuals would also become very fatigued and oral communication would become the least viable form of communication.[9] Additionally, measures of articulatory function are completed via diadochokinetic tasks designed to measure production in various contexts (i.e. initial, final, and varying /p/ /t/ /k/). Similarly, intelligibility measures are completed that are critical aspects of speech production assessment. Speech-language pathologists may require individuals with ALS to read word lists and passages to document articulation errors and percentage of intelligibility.[2] All measures of speech production are critical aspects of assessment and short and long term intervention planning.[2]

IMPROVING COMMUNICATION

Compensatory Strategies for Communication

Speech-language pathologists and other health professionals may consider discussions about the use of compensatory strategies regardless of the patient's level of function. For example, speech-language pathologists can discuss compensatory strategies for communication to improve their understanding of how communication typically deteriorates over time and how it can be maximized in various settings. Several strategies can be initiated by the individual with ALS and their family member/caregiver to reduce the burden they may experience when they realize they are experiencing a communication breakdown. The use of a communicative partner (family member, caregiver or spouse) who repeats the words he or she is able to understand is one strategy that allows the patient to produce only the missed words, thus conserving energy and improving intelligibility. Pacing strategies may also be introduced to the patient in an effort to improve intelligibility and conserve energy. [13,16] Other strategies may include environmental adaptations such as removing outside distractions like the television when conversing with the patient with ALS.[13] Finally, if the communicative partners position themselves in a line of direct eye contact they will be more likely to pick up on nonverbal contextual cues that add to the meaning of the intended message. [2]

Augmentative and Alternative Communication (AAC)

Research has shown that those who have lost their ability to speak hold the ability to maintain social closeness with another person as the most important purpose of communication, above their desire to indicate wants and needs or to pass on information.[16] So, it is only logical for the speech-language pathologist to consider the abilities and expectations of all who will be involved in communicating with the patient with ALS in suggesting augmentative and alternative communication (AAC) devices. AAC devices play a critical role in the communicative functioning of individuals with ALS during the disease progression. AAC devices are recommended when patients are only 50% intelligible during reading tasks.[11] Thresholds for words per minute have been established on standardized assessments such as the Sentence Intelligibility Test thereby indicating it may be time to consider an alternate mode of communication.[3]

AAC Approaches: Low Tech and High Tech

There are two general approaches to AAC: low-technology and high-technology. Low-technology approaches includes writing and alphabet boards and other aides that do not require extensive training for use. High-technology approaches consist of computer-based communication devices that involve voice output and computer generated displays. High-technology approaches require some training for efficient use.[11] High-technology devices typically provide more access options, allowing for modification once the patient is unable to use his limbs. Head tracking and eye tracking capabilities are among several options available for patients.[3] Similarly, there are several types of systems for augmentative alternative communication: symbol-based, text-based, and text-to-speech machines which may all be modified to compensate for any physical or cognitive limitations the individual with ALS may experience. [9]

Factors that Influence AAC Selection and Use

Speech-language pathologists are likely to play an instrumental role in the identification of the most appropriate AAC devices for individuals with ALS. It is estimated that "95% of people with ALS will become unable to speak prior to death," thus requiring the use of AAC devices.[3] To preserve contact between the individual and his environment, AAC devices can provide the primary means of communication. Speech-language pathologists should consider the multitude of factors that have the potential to influence the family's decision to proceed with using the devices offered to them. Among those factors are: the patient's physical limitations, their experience with technology prior to disease onset, environment, and the expectations and needs of the patient with ALS and his caregivers.[11]

There are a range of options for the use of AAC devices. Patients who have experience with technology have more options and are typically more willing to consider technology to aid their communication. Research suggests that the use of AAC devices changes depending on the environment.[11] For example, individuals with ALS may use high-technology devices when they are communicating with unfamiliar partners and low-technology devices when they are communicating with familiar partners.[11] This may be a result of comfort levels and the familiar partner's ability to detect and even predict messages in typical contexts. They also noted that men preferred high-technology devices while women felt more comfortable using low-technology devices.[11] The ALS disease stage plays a critical role in the type of augmentative and alternative communication device available for use. During the later stages of ALS, there may be more concern with communicating basic wants and needs in the form of yes and no responses or the use of low-technology approaches that require little effort. Therefore, speech-language pathologists must be patient and flexible as the needs of the individual change, particularly since the emotional and psychosocial aspects of the disease can result in families delaying their acceptance of AAC devices. Additionally, patients may see acceptance of AAC devices as an indication that they are no longer the independent and verbal individuals of their past.[11]

PATIENT FAMILY ISSUES

The educational needs of the individual with ALS and their family members should be considered early in the disease course. Issues such as the use of AAC devices should be discussed early in the disease process. Patient/family education provided early in the disease can allow patients with ALS and their family members to make informed decisions at which time it is necessary to implement these strategies.[16] Early after the initial diagnosis, and possibly during the first meeting with the patient with ALS and his family members or caregivers, the speech-language pathologist should describe changes in speech and swallowing that may be expected as the disease progresses. It may be helpful to provide all information in writing since the amount of information presented to these individuals may be daunting and overwhelming. It is critical that a wide range of topics related to communication and swallowing are included in early educational sessions. For example, emotional lability or extreme outbursts of inappropriate laughter or crying may be an appropriate topic as this is a poorly understood aspect of the disease. These early discussions could provide added insights into the disease for both the individual with ALS and his or her family.[16] According to Murphy, 2004, education about this topic may lessen their degree of embarrassment or frustration.[16]

END OF LIFE ISSUES

A primary goal of patient management of individuals with ALS is to enable individuals to remain active in the decision making process throughout the progression of their disease. Thus, maintaining their ability to express their wants and needs is particularly critical. Speech language pathologists can assist with communication of important decisions. Early in the disease process, individuals with ALS and their family members may be asked to make decisions about what types of treatments would be desired and what issues are critical to their maintenance of sufficient quality of life. The development of an advance directive which outlines what kinds of medical intervention or care the individual would like to receive in the event that they are unable to communicate these decisions at the time they are needed are recommended in the early stages of the disease. For example, an individual with ALS may not wish to use a ventilator for breathing support. Another type of advance directive is the medical directive which is placed in medical records and details wishes such as a Do Not Resuscitate (DNR) order.[17] All aspects of future

patient management should be considered during the development of advance directives to ensure that the individual's quality of life is preserved and medical decisions are made based on their wishes and desires.

CONCLUSION

There are a wide range of options for the management of communication deficits in patients with ALS. Speech-language pathologists are key members of the interdisciplinary team involved in the management of ALS during the disease progression. Speech-language pathologists are called upon to assess the communicative and swallowing functioning of individuals with ALS. They may also serve as educators for patient and family issues such as communication, swallowing, and ensuring the individual and his or her caregivers are aware of how end of life issues will be addressed. AAC devices offer hope for patients who suffer from the debilitating effects of ALS on communication functioning. Patients with ALS can benefit from the services of speech-language pathologists and in particular those who have experience with management of communication deficits with the use of AAC devices. Speech-language pathologists can play a critical role in the improvement of quality of life by identifying the correct devices for patients during disease progression.

REFERENCES

1. Amyotrophic Lateral Sclerosis (ALS) Association. (2008). What is ALS. Retrieved July 22, 2008 from: http://www.alsa.org/als/what.cfm?CFID=256111&CFTOKEN=6ad62d6ad1a9c9d2-4DAD5EC6-188B-2E62-80835A7FD53A64C2
2. 2. Brownlee A, Palovcak M. The role of augmentative communication devices in the medical management of ALS. [Review]. *NeuroRehabilitation, 2007;22*(6): 445-450.
3. Beukelman D, Fager S, Ball L, Dietz A. AAC for Adults with Acquired Neurological Conditions: A Review. *Augmentative and Alternative Communication. 2007;*23(3): 230-242.
4. Walling AD. Amyotrophic Lateral Sclerosis: Lou Gehrig's Disease. *American Family Physician.1999;59* (6): 1489-96.
5. National Institute of Neurological Disorders and Stroke. (2008). Amyotrophic Lateral Sclerosis Fact Sheet. Retrieved July 22, 2008 from: http://www.ninds.nih.gov/disorders/amyotrophiclateralsclerosis/detail_amyotrophiclateralsclerosis.htm#109814842.
6. McCarthy J. ALS Society of Canada.(2005). A Manual for People Living with ALS. 3rd edition.
7. Falvo D.(2005). *Medical and Psychosocial Aspects of Chronic Illness and Disability.* Jones and Bartlett Publishers.
8. Mitsumoto H, Munsat T. (2000). Amyotrophic Lateral Sclerosis: *A Guide for Patients and Families*. Demos Medical Publishing, LLC.
9. Muscular Dystrophy Association ALS Division. (2005). Everyday Life with ALS: A Practical Guide. Retrieved November 7 2008 from http://www.als-mda.org/publications/everydalifeals/.
10. Oppenheimer EA, Guth D, Fischer J. (2004). Treating Respiratory Problems in ALS Patients Can Improve Quality of Life. Retrieved January 1 2009 from: http://www.alsindependence.com/Treating_Respiratory_Problems_in_ALS.htm.
11. Doyle M, Phillips B. Trends in Augmentative and Alternative Communication Use by Individuals with Amyotrophic Lateral Sclerosis. *Augmentative and Alternative Communication.* 2001;17(3): 167-178.
12. Eisen A, Krieger C. (1998). *Amyotrophic Lateral Sclerosis: A Synthesis of Research and Clinical Practice*. Cambrige University Press.
13. Duffy JR. (2005). *Motor Speech Disorders: Substrates, Differential Diagnosis and Management*. St. Louis: Mosby.
14. Dikeman K, Kazandjian M. (2003). Communication and Swallowing Management of Tracheostomized and Ventilator-Dependent Adults, Second Edition. Delmar Learning.
15. Kent R, Sufit R, et al. Speech Deterioration in Amyotrophic Lateral Sclerosis: A Case Study. *Journal of Speech and Hearing Research.*1991;34:1269-1275.
16. Murphy J. Communication strategies of people with ALS and their partners. *ALS and other motor neuron disorders.* 2004;5:121-126.
17. International Alliance of ALS/MND Associations.(2006) "Advanced Directives" retrieved November 10 2008 from http://www.alsmndalliance.org/uploads/pdfs/Advanced_directives.doc.

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