Management of Dysarthria in Amyotrophic Lateral Sclerosis

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Summary

This article describes intervention for dysarthria associated with amyotrophic lateral sclerosis (ALS). Five critical periods are presented including a stage with normal speech, detectable speech disturbance, behavioural intervention, use of augmentative communication, and loss of useful speech. Intervention strategies at each of these stages are outlined with the goal of maintaining functional communication regardless of the severity of dysarthria.

ALS is a rapidly progressive degenerative disease of unknown etiology involving the motor neurons of both the brain and spinal cord.[1] The symptoms characteristic of ALS are generally classified by site of involvement (that is, upper motor neuron versus lower motor neuron) and by whether spinal nerves (those innervating the arms and legs) or bulbar nerves (those innervating the muscles of speech and swallowing) are involved. The motor speech impairment associated with ALS is classified as a mixed dysarthria showing features associated with both spasticity (lack of fine control) and flaccidity (weakness).[2]

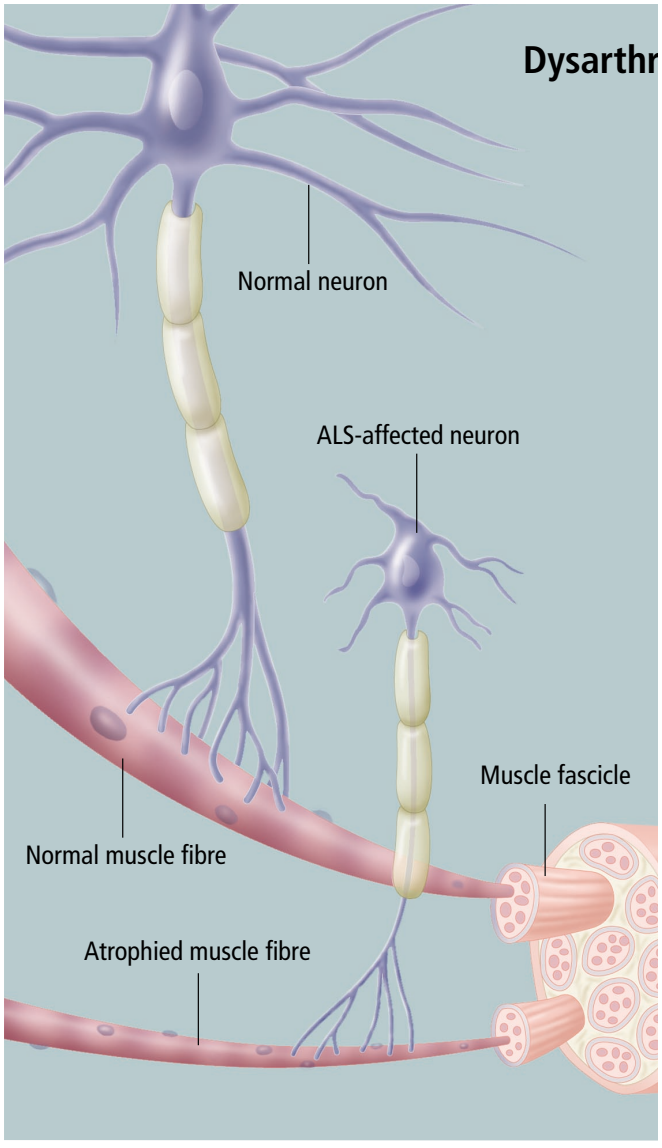
Loss, or potential for loss, of communication is one of the most distressing aspects of ALS. Approximately 80% of persons with ALS experience such severe speech impairments that they require augmentative communication strategies to meet their daily communication needs. These communication impairments are progressive. Those with bulbar signs experience relatively early loss of the speech while those with spinal signs lose their speaking effectiveness somewhat later in the course of the disease. Maintenance of functional communication skills either in the form of natural speech or augmentative communication strategies is an important intervention goals. There are few aspects of adult life that are not affected by reduced communication capabilities. This article describes principles of intervention for dysarthria associated with ALS. Symptoms and intervention for five stages of dysarthria severity are also outlined.

Principles of Intervention

Communication competence allows persons with ALS to maintain their ability to guide, direct and influence the management of both medical and personal aspects of their lives.[3]

The first intervention principle is to maintain functional communication throughout the course of the disease. This principle is most often violated when patients, their families and professionals delay their decision-making until the individual can no longer speak, before pursuing alternative communication methods. This delay is commonly the result of inadequate information and education.

The second, and related principle of management is timely intervention. To accomplish this, professions need to systematically evaluate the speech performance of persons with ALS and to inform them regarding predictors of impending communication failure. With this information, persons with ALS and their families can prepare themselves to make the necessary



intervention decisions to become aware of the availability of services that they need in the future. A chief goal is to help the patient and family become informed consumers of medical and technological services.

The third principle of intervention is a system to identify "critical periods" when intervention is needed promptly and briefly. These critical points are periods when the person's capabilities have changed and communication needs are not being met, but before fatigue and weakness are overwhelming. Identification and anticipation of these critical periods requires the staging of intervention so that current needs are met and future needs anticipated.

Staging Intervention

The following section outlines intervention in five stages of dysarthria severity. These stages that define critical periods of intervention are summarized in Table 1 (page 40).

Stage 1. Normal Speech Processes

Symptoms

Individuals at this stage are functionally normal communicators. Only the patient or spouse may note some change from premorbid levels.

Intervention

The major role of a speech-language pathologist at this point is to provide information. With the encouraging developments in course-altering drugs, the neurology community is increasingly interested in early diagnosis of bulbar symptoms. The speech-language pathologist can contribute important information in this area. Subtle changes in speaking rate may signal early bulbar involvement.[4,5] In a study of 218 individuals with ALS, Ball and her colleagues[6] identified a series of early predictors of speech change including laryngeal control as reflected in voice quality, speaking rate and listener ratings of communication effectiveness. These authors report that when speaking rate decreases to 60% of normal, or approximately 100 words per minute on the Sentence Intelligibility Test[7] persons with ALS can expect a precipitous decline in intelligibility. On this task, non-disabled individuals speak, on average, at 190 words per minute.

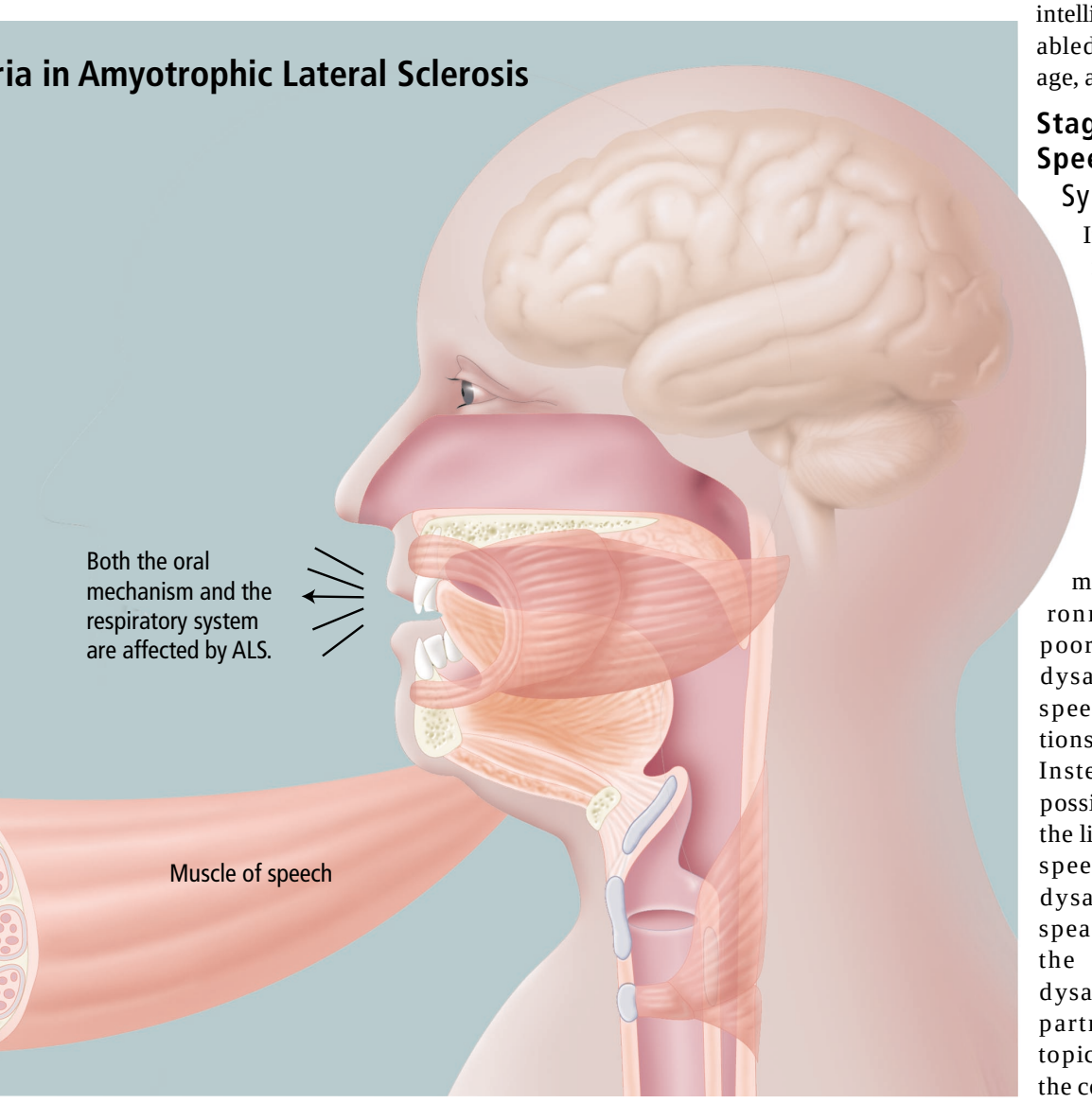
Stage 2. Detectable Speech Disturbance

Symptoms

Individuals at this stage report noticeable changes in speech which are made worse by fatigue or stress. Despite obvious changes, speech remains intelligible.

Intervention

Management of speech symptoms at this stage includes techniques such as minimizing noise in the environment. For individuals with poor respiratory support and dysarthria, efforts to increase speech loudness in noisy situations can be extremely fatiguing. Instead, we suggest that, when possible, noise and distance from the listener be reduced. When the speech is distorted, as it is in dysarthria, it is useful for the speaker to establish the topic of the message. Speakers with dysarthria should confirm their partners' understanding of the topic before proceeding. Setting the context of the message should



be done whenever a topic is changed abruptly within a conversation. Because ALS typically occurs in an older population, hearing loss in spouses of people with ALS is not uncommon. Therefore, it is beneficial to maximize the hearing of frequent communication partners. This may take the form of referrals to an audiologist who can fit them with hearing aids if necessary. People with ALS at this level of severity frequently complain that communication in a group is difficult. Because many aspects of group conversation are dependent on rapid pacing of exchanges, the speech-language pathologist can help to develop strategies for coping with groups.

Stage 3. Behavioural Modifications

Symptoms

Individuals at this stage are experiencing some reduction in speech intelligibility, especially in adverse listening conditions. Frequently they will need to repeat portions of a message in order to resolve communication breakdowns.

Intervention

In addition to the environmental management techniques described earlier, individuals are encourage to slow their speaking rate. They are encouraged to do this by exaggerating speech, especially important words that they wish to stress or make prominent. Because fatigue may have a substantial impact on the quality of their speech, training in energy conversation techniques is also warranted. This includes counseling the speaker not to exercise speech muscles to the point of fatigue and to avoid noisy communication situations where the energy demands of speech are increased. Palatal lift fitting may also be considered for some individuals at this stage.[8] A more complete description of candidacy issues can be found elsewhere.[9] Although natural speech remains a functional mode of communication for speakers at this stage of ALS, intelligibility is frequently compromised, and speakers need to learn to resolve communication breakdowns. Although speakers with ALS do not typically need extensive training, speech treatment may involve developing techniques to increase articulatory precision by (1) slowing articulatory rate, (2) exaggerating oral articulatory movements, (3) paying special attention to inclusion of final consonants, and (4) increasing the overall forcefulness with which speech is produced. Strengthening exercises of the tongue and jaw are to be avoided, as they have not been shown to be effective with this population and may be counter productive.[10]

Table 1
Stages of Severity of Dysarthria in ALS

STAGE 1: Normal Speech Processes
Normal Speech: Patient denies any difficulty speaking. Examination demonstrates no abnormality.
Nominal Speech Abnormality: Only the patient or spouse notices that speech has changed. Maintains normal rate and volume.

STAGE 2: Detectable Speech Disturbance
Perceived Speech Changes: Speech changes are noted by others, especially during fatigue or stress. Rate of speech remains essentially normal.
Obvious Speech Abnormalities:Speech is consistently impaired. Affected are rate, articulation and resonance. Remains easily understood.

STAGE 3: Behavioural Modifications
Repeats Messages on Occasion: Rate is much slower. Repeats specific words in adverse listening situations. Does not limit complexity or length of message.
Frequent Repeating Required: Speech is slow and laboured. Extensive repetition or a "translator" is commonly needed. Patient probably limits the complexity or length of messages.

STAGE 4: Use of Augmentative Communication
Speech Plus Augmentative Communication: Speech is used in response to questions. Intelligibility problems need to be resolved by writing or a spokesman.
Limits Speech to One word Response: Vocalizes one word response beyond yes/no: otherwise writes or uses a spokesman. Initiates communication nonvocally.

STAGE 5: Loss of Useful Speech
Vocalizes for Emotional Expression: Uses vocal inflection to express emotion, affirmation and negation.
Non-Vocal: Vocalization is effortful, limited in duration, and rarely attempted. May vocalize for crying or pain.

Source: Yorkston KM, et al. Speech deterioration in amyotrophic lateral sclerosis: Implications for the timing of intervention. Journal of Medical Speech/Language Pathology, 1993. 1(1):35-46. Reproduced with permission.

Stage 4. Use of Augmentative Communication

Symptoms

Individuals at this stage must rely on augmentative systems, either as their primary means of communication or to supplement natural speech when it is not understood.

Intervention

The transition to reliance on augmentative communication approaches is one of the major critical periods in the management of people with ALS.[11] Techniques may range from "light-tech" to "high-tech." For example, alphabet supplementation is a transitional technique that may bridge the gap between total reliance on natural speech and dependence on augmentative communication technology. Speakers with severe dysarthria can be encouraged to

point to the first letter of each word as they speak. This technique is useful for individuals whose natural speech is difficult to understand but may still be functional for some types of communication, with some communication partners, or in particular communication settings. Augmentative communication systems may also include devices alerting systems (such as simple buzzers or devices such as baby monitors that transmit signals to distant locations within the house) or systems that handle telephone communication.[12] Many people with ALS use portable writing systems to augment natural speech. These systems are typically introduced as back-up systems to resolve communication breakdowns when natural speech has not been understood. They can also be used to introduce communicative topics when natural speech is difficult to understand. As symptoms progress, computer-based multipurpose augmentative communication systems are currently commercially available. We will not review these systems in detail because technology is changing so rapidly that today's systems will no doubt be replaced by more efficient and effective ones in the near future. Rather, readers are referred to a comprehensive web site that provides links to vendors of AAC system, device tutorials and other current information (University of Nebraska, AAC Center: http://aac.unl.edu/).

Stage 5. Loss of Useful Speech

Symptoms

Individuals at this stage have lost natural speech as a functional means of communication. Therefore, they must rely entirely on alternative communication technology and techniques.

Intervention

In addition to the augmentative communication strategies just described for individuals in Stage 4, a non-fatiguing and reliable means of indicating yes or no is a mandatory element of communication management plans. Because eye gaze is usually preserved in individuals with ALS, eye pointing or eye gaze can frequently be used as a selection technique when head and hand movement are no longer functional. Eye gaze systems can be quite complex and incorporate encoding strategies, so partner training is critical. More complete descriptions of selection techniques and encoding strategies are available.[12] The majority of individuals on ventilators also experience significant cranial nerve involvement. Because many of these individuals have poor oral movement, use of natural speech via a modified tracheostomy tube or electrolarynx is not possible. For these people, the eye-gaze augmentative communication system described earlier may be appropriate. ◆

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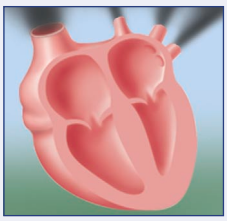
Is Poor Health Hardening Your Heart?



New evidence from Germany suggests that infections may contribute to the pathogenesis of atherosclerosis. Researchers studied a group of 572 patients and measured their IgG or IgA antibodies to eight different pathogens, including herpes simplex virus 1 and 2, cytomegalovirus, Epstein-Barr virus, *Hemophilus influenzae, Chlamydia pneumoniae, Mycoplasma pneumoniae* and *Helicobacter pylori*. Patients underwent coronary angiography, carotid duplex sonography and evaluation of the ankle-arm index, to determine the extent of atherosclerosis. What the researchers found was a correlation between infectious burden and the presence of advanced atherosclerosis. After a follow-up of three years, the mortality rate in patients with advanced atherosclerosis who were seropositive for up to three pathogens was only 7%, compared to a rate of 20% in those patients with between six and eight pathogens.

Further study will be required to determine whether the infections are actually causal for atherosclerosis, or whether they merely serve as a marker for poor health. ◆

Source

1. Espinola-Klein C, Rupprecht HJ, Blankenberg S, et al. Impact of infectious burden on extent and long-term prognosis of atherosclerosis. Circulation. 2002;105:15-21.