Classifying Subgroups of Individuals with ALS: Acoustic and Aerodynamic Characteristics

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Amyotrophic Lateral Sclerosis (ALS) is a progressive degenerative neuromuscular disease that involves the upper and lower motor neurons. Damage to the lower motor neurons is manifested by muscle weakness, fatigue, muscle atrophy, and fasciculations, whereas upper motor neuron damage is manifested by spasticity, muscle weakness, cramping, increased tone, and hyperactive deep-tendon reflexes. Upper and/or lower motor involvement may occur during the course of the disease, but eventually, both systems are involved.

At initial presentation, individuals are described primarily as being either bulbar or nonbulbar, as determined by presence or absence of neuromotor symptoms. Usually, bulbar signs are typified by rapid deterioration, while nonbulbar signs point to slightly slower deterioration, especially of the cranial nerves affecting oral-pharyngeal-laryngeal motor coordination and speech production (Darley, Aronson & Brown, 1975).

Progressive deterioration of the oral, velopharyngeal, and laryngeal subsystems serving speech leads to speech production difficulties and decreased speech intelligibility in individuals with ALS. Major characteristics of speech difficulties include imprecise articulation, hyposlalinguad and nasal air emission, strained-strangled, harsh, breathy, low-pitch and low intensity voice production, and slow speaking rate. These characteristics may occur in different individuals at various times throughout the course of the disease (Kent, Sufit, Rosenbek, Weismer, Martin, & Brooks, 1991).

Aerodynamic assessment of speech was accomplished with a commercial pressure-flow device (RC Electronics - Computer Scope) employing separate differential pressure transducers for oral-nasal pressure measures and a pneumotachograph-differential pressure transducer for nasal airflow measurements. Measures of peak and durational aspects of the oral-nasal aerodynamic features of speech were analyzed using the accompanying software package on the same computer used for other analyses.

Recording Procedures:
Each subject was seated comfortably in a quiet room with an electret condenser microphone held at a constant mouth-to-microphone distance of 15 cm. Speech materials were recorded on a research quality cassette tape recorder and/or directly to one channel of a computer with A-D/D-A converters and a commercial acoustical recording/analyses package (CSpeech, Milenkovic, 1987).

Acoustical information for nasalization was acquired using a Nasometer (Kay Elemetrics) with a dual oral-nasal microphone system and the resultant signal sent to a computer and accompanying software package for data display and analyses.

Results
Both inferential and descriptive statistics were employed for each of the data sets to describe differences in the subgroups of bulbar and nonbulbar individuals with ALS over two time periods (approximately 6 months apart).

Respiratory-Laryngeal Subsystem:
An analysis of variance procedure revealed significant differences (p = .008) in Maximum Phonation Time (MPT) over time for the vowel /a/ production for the ALS subjects, with the bulbar group having numerically lower MPTs than the nonbulbar at each assessment period. That is, the bulbar group decreased from 16.7 seconds to 11.3 seconds over time, while the nonbulbar decreased from 21 to 16.7 seconds over the same time period.

The bulbar subjects also demonstrated significantly (p = .003) slower vocal fold diadochokinetic rates for /ha/ production (3.46 s/s) than the nonbulbar subjects (4.88 s/s). In addition, the last segment of the repetition phase was significantly different for the Group from Time 1 to Time 2, with the bulbar subjects showing a poorer performance.

Microanalytical acoustical measures of frequency and amplitude perturbation from the sustained vowel /a/ productions showed a numerically poorer performance for the bulbar subjects for modal fundamental frequency, increased jitter and shimmer measures, and decreased signal-to-noise ratios. In addition, there were statistically significant reductions (p = .001) for vocal frequency range, intensity range, and maximum and minimum intensity range, with the bulbar individuals showing more restricted performance within and across assessment times than the nonbulbar individuals.

Velopharyngeal Subsystem:
Results of the acoustical assessment of the velopharyngeal (VP) subsystem indicated that the bulbar individuals had significantly (p = .04) greater nasalance scores of the vowel /i/, syllables /i/ and /i/ and the "Zoo Passage" than the nonbulbar subjects.

Method
Subjects:
The subjects selected for presentation included 95 men and women with diagnosed ALS. Subgrouping led to classifications with 44 persons with bulbar symptoms and 51 with nonbulbar symptoms upon initial classification by two neurologists (AH, MS). The subjects ranged from 40 years to 78 years of age. The length of illness ranged from 4 months to 12 years, with a mean duration of illness of approximately 2 years. The subjects were assessed twice, approximately 6 months apart.

Stimulus Material:
The stimuli were chosen to investigate subtle changes in the laryngeal, velopharyngeal and oral articulatory subsystems underlying speech production. Respiratory-laryngeal function was assessed using a maximum phonation time technique employing production of the isolated vowel /a/. Vocal fold diadochokinetic function of vocal fold activity was sampled using the syllable glottal fricative /h/ + vowel /a/ ("haa"). Velopharyngeal function was sampled employing the fricative blend in the word "hamper", sustained vowel /i/, syllables /ip/ and /is/, a nasal sentence, and an all "oral" element passage ("Zoo Passage"). Oral articulatory function was sampled employing 12 CVC words consisting of 6 word initial plosives (p,t,k,b,d,g) combined with the vowel /i/.

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Aerodynamic assessment of the VP port area demonstrated a significant difference ($p = 0.003$) in nasal airflow duration for the /mp/ blend at both Time 1 and Time 2 between the bulbar and nonbulbar individuals. In addition, significant differences ($p = 0.02$) in timing of closure between the /m/ and the /p/ elements was determined, with the bulbar group demonstrating the shortest time (i.e., most overlap between the nasal and non-nasal elements) when compared with the nonbulbar subjects.

**Oral-Articulatory Subsystem:**

Analyses of the acoustical voice-onset-time (VOT) data for the two groups over time indicated a significant difference ($p = 0.001$) for voicing contrasts (with voiceless phonemes being significantly longer in VOT than voiced consonants). Further, at assessment Time 2, the bulbar individuals showed a significant ($p = 0.05$) difference in VOT when compared to the nonbulbar subjects. A significant ($p = 0.05$) Voicing by Position (i.e., anterior versus posterior articulator position) was determined showing the bulbar group having more deviant articulator activity at Time 1 and 2. In addition, the bulbar individuals were more variable within and across time than the nonbulbar subjects.

**Discussion:**

The series of investigations of the respiro-laryngeal, velopharyngeal, and oral-articulatory subsystems described in this presentation provides acoustical and physiological information concerning the course of "short term" deterioration of the subsystems serving speech in individuals with ALS. These data support the approach of differentiating ALS subjects into bulbar and nonbulbar groupings based upon speed and variability of deterioration within the various subsystems (Leeper, Millard, Bandur, Hudson, 1996; Renout, Leeper, Bandur, Hudson, 1995; Delorey, Leeper, Bandur, Hudson, 1993; Herbert, Leeper, Bandur, Hudson, 1996). The present data are also generally consistent with acoustical and perceptual information from previous investigations of ALS subjects (Kent, et al., 1991; Langmore and Lehman, 1994; Caruso and Burton, 1987).

Given the differences in respiratory-laryngeal support, the loss of aerodynamic integrity at the VP port, and the differential reduction in labial, tongue tip, and tongue blade control between the two subgroups of ALS subjects studied, it would seem important to continue such descriptive categories for management purposes. There is, however, as Strong (1995) has suggested, no current medical therapies that can stop or even slow down the progress of ALS. However, management related to "quality of life" issues such as breathing and communication are important to the well being of the individual and his/her supporting family. A variety of augmentative and assistive communication devices may be used to improve quality of life within the suggested life span. Monitoring of changes in communication skills via non-invasive acoustical and aerodynamic assessment methods will allow for more precise allocation of these techniques for individuals with one of the subgroupings of ALS.

**References:**