INSTRUMENTAL EVALUATION OF VELOPHARYNGEAL DYSFUNCTION IN AMYOTROPHIC LATERAL SCLEROSIS

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ABSTRACT

The goals of this study were to (1) determine the effect of amyotrophic lateral sclerosis (ALS) on velopharyngeal function during the early and later stages of bulbar motor involvement, and to (2) identify the most sensitive measure of velopharyngeal dysfunction among several currently available options. The velopharyngeal function of 82 patients with ALS was compared to that of 20 normal controls using a nasometer and the Phonatory Aerodynamic System (PAS, PENTAX Medical). Forty-eight patients with ALS were considered as bulbar presymptomatic (BPS) and 34 patients with ALS were considered as bulbar symptomatic (BS) based on a measure of speaking rate. Findings revealed that the velopharyngeal dysfunction was manifested differently between the ALS-BPS and ALS-BS groups. In addition, between the two instrumentation approaches, the aerodynamic technique appeared to be more suitable due to the confounding effect of excessive nasal air flow on nasometer output.

Keywords: Amyotrophic lateral sclerosis, velopharyngeal dysfunction, resonance

1. INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is an adult-onset fatal neurodegenerative disease characterized by progressive deterioration of various motor functions such as ambulation, speech, swallowing, and respiration. Velopharyngeal dysfunction (VPD) is a pathophysiologic condition commonly observed in patients with ALS due to progressive paresis of velopharynx musculature. VPD, causes resonance abnormalities e.g., hyponasality, hypernasality, and mixed nasality. Severe hypernasality can lead to unintelligible speech [1, 2, 3, 4] due to acoustic disruptions such as reduced spectral energy [5, 6, 7], and collapsed vowel space area [8, 9, 10]. In addition, nasal air emission, a common symptom of VPD, can impair oral articulation by diminishing the build-up of oral air pressure, which is required to produce stops and fricatives. Nasal air emission also can alter speech respiratory function requiring more frequent inspirations to ensure an adequate air supply to support voice and speech. Increased number and duration of pauses are well-established features of abnormal speech in persons with ALS [11, 12]. Despite the impact of VPD on different speech subsystems, clinical evaluation currently relies almost exclusively on subjective ratings of abnormal resonance features, which are unlikely to be sensitive to early changes and are vulnerable to listener bias [13, 14]. Instrumentation approaches for objectively assessing VPD have been developed but not widely tested on clinical populations with dysarthria due to ALS. The two most widely used approaches, acoustic nasometry and oral-nasal aerodynamics, are most commonly used to quantify VP function in populations with anatomical abnormalities such as children with cleft lip and palate. Nasometer measures the ratio of the amplitude of the acoustic energy obtained from the nose to that of the mouth; whereas, the aerodynamic assessment provides information about the magnitude of oral pressure or nasal flow.

Various studies have suggested significant correlation between perceived nasality and nasalance score in clinical populations without ALS [15, 16, 17]. Other studies have, however, reported the potential confound of nasal air emission on nasalence scores [18, 19]. For example, in a study conducted by Dalston et al. [18] the sensitivity of nasometry for correctly identifying hyponasality increased from 0.48 to 1.0 upon the removal of data from participants with nasal air emission. Given that participants with ALS have been observed to produce stop plosives (i.e., /p/) with various degrees of nasal air emission [2, 20], we hypothesize that an aerodynamic approach will provide a more accurate assessment of VPD in this population than a nasometric approach.

This study reports on VPD in person with ALS using both nasometric and aerodynamic approaches. Our goals are to understand how ALS affects VP function and to compare the diagnostic efficacy of two commonly used approaches of measuring VPD. Because patients with ALS vary with regard to the site of symptom onset and the course of progression, the long-term goal of this research is to improve (1) the early detection of VPD in persons with ALS, (2) the monitoring of disease spread and progression, and (3) the understanding of VPD on speech loss.

2. Methods

2.1. Participants

Eighty-two patients with ALS and 20 normal controls (NCs) participated in this study. The participants with ALS varied in both the total score and bulbar subscore of the ALS Functional Rating Scale-Revised (ALSFRS-R) [21]. The ALSFRS-R consists of 12 survey questions to assess the degree of functional impairment with the score of each question ranging from 4 (least impaired) to 0 (most impaired). The ALSFRS-R scores ranged from 21 to 46 (M = 37.25, SD = 5.83). The bulbar subscore, based on the first three questions of the scale, assessed the bulbar function with a maximum score of 12 ranged between 5 and 12 (M = 10.05, SD = 2.08). Patients with ALS were stratified into two groups: (1) bulbar presymptomatic (ALS-BPS), and (2) bulbar symptomatic (ALS-BS) based on the cut-off speaking rate of 150 words per minute (w/m) following Rong et al. [22]. Speaking rate was used as a criterion to stratify patients with ALS because previous research demonstrated that the decline of speaking rate precedes intelligibility and occurs at a faster rate than intelligibility during the early stage of ALS [20, 23]. Forty-eight patients with ALS exhibited speaking rate above 150 (w/m) and were classified as ALS-BPS; whereas, 34 patients showed speaking rate below (150) w/m and were classified as ALS-BS.

Table 1: Demographic and medical information of participants

	NC (N= 20)	ALS-BPS (N= 48)	ALS-BS (N= 34)
Sex	9 M 11 F	29 M 19 F	16 M 18 F
Age (yr)	M= 62 SD= 8.36	M= 61 SD= 9.74	M= 66 SD= 10.52
ALSFRS-R Total	N/A	M= 36.91 SD= 6.18	M= 37.64 SD= 5.47
ALSFRS-R Bulbar	N/A	M= 10.76 SD= 1.69	M= 9.18 SD= 2.21
Site of onset	N/A	2 bulbar 41 spinal 5 unknown	11 bulbar 20 spinal 3 unknown
Disease Duration (mth)	N/A	M= 9 SD= 7	M= 18 SD= 33
Speaking rate (w/m)	M= 181 SD= 28.22	M= 186 SD= 21.65	M= 109 SD= 27.70
Intelligibility	M= 99 SD= 1.17	M= 98 SD= 2.76	M= 89 SD= 15.84

All participants of this study were a subset of participants recruited for a larger multi-site longitudinal study to investigate speech deterioration in ALS. All participants were native speakers of English with no history of speech-language impairments. Participants with ALS had no history of other cognitive and/or neurological disorders.

2.2. Data Collection

2.2.1. Speech Performance

Speaking rate and intelligibility of each participant were measured following procedures described in the Sentence Intelligibility Test (SIT) manual [24]. Speaking rate was calculated as the number of words produced per minute from the SIT stimuli.

2.2.2. Assessment of velopharyngeal function

VP function was assessed using both aerodynamic and nasometer techniques. Simultaneous recording of intra-oral air pressure and nasal air flow was acquired using the Phonatory Aerodynamic System (PAS) (Model 6600, PENTAX Medical). During the aerodynamic evaluation of the VP status, participants were asked to produce /pi/ and /mi/ syllables seven times. To detect the intra-oral air pressure, a polyethylene oral catheter was placed in the participant's mouth and the end of the catheter was connected to a calibrated pressure transducer. Nasal air flow was detected using a nasal mask snugly fitted around the participant's nose and was connected to a heated pneumotachograph, the end of which was ported to a calibrated airflow transducer. In addition, participants were asked to repeat the word "hamper" five times at their normal speaking rate and loudness. Because the word "hamper" consists of the /m-p/ sequence, the interval from peak nasal flow of /m/ to peak oral pressure of /p/ represents the adequacy of VP function and velum control. The PAS software was used to measure the aerodynamic indices of (1) the peak oral pressure, and associated nasal airflow, of /p/ in the syllable /pi/; (2) the peak nasal flow and associated oral pressure of /m/ in the syllable /mi/; as well as (3) the time lag from the peak nasal flow to peak oral pressure of /m-p/ sequence in the word "hamper." All measures were then averaged across several repetitions for each participant.

In addition to aerodynamic measures, participants were asked to read two sentences "buy Bobby a puppy" (BBP) – loaded with oral vowels and oral stop consonants – and "momma made lemon jam" (MMJ) – loaded with nasal consonants – three times each while maintaining a normal, self-determined speaking rate and loudness. A nasometer (Model 6400, PENTAX Medical) was used to measure the mean nasalance score (i.e., the ratio of the nasal energy to the sum of nasal and oral acoustic energy) of each sentence. Mean nasalance scores were subsequently averaged across the three repetitions for each participant. Kruskal-Wallis tests with

Mann-Whitney U tests as post hoc comparisons were used to determine group differences in variables under investigation.

3. RESULTS

Among the seven variables (5 aerodynamic indices and 2 nasometric measures), peak oral pressure for /p/, oral pressure on peak nasal flow for /m/, and the interval from the peak nasal flow of /m/ to peak oral pressure of /p/ in "hamper" did not show statistically significant group differences. Descriptive statistics of these variables are summarized in Table 2.

Table 2: Means and standard deviations (in parentheses) of peak oral pressure (Po) of /p/ in the syllable /pi/, Po at peak nasal flow (NF/) of /m/ in the syllable /mi/, and the time interval from the peak NF of /m/ to peak Po of /p/ in "hamper".
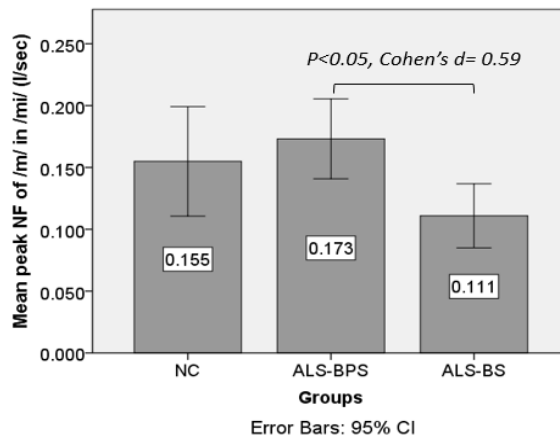
	NC	ALS-BPS	ALS-BS
Peak Po of /p/ (cm H_2O)	6.54 (2.38)	6.73 (1.97)	6.22 (2.60)
Po at peak NF of /m/ (cm H_2O)	0.96 (0.66)	0.96 (0.65)	0.92 (0.74)
NF-Po in hamper (ms)	70.27 (62.20)	66.58 (46.99)	82.03 (49.46)

In contrast, group differences were observed for mean nasal flow at peak oral pressure of the /p/ sound, mean peak nasal flow of the /m/ sound, and mean nasalance scores for BBP and MMJ sentences (as represented in Figures 1 to 4, respectively). Participants with ALS-BS showed significantly larger nasal flow for /p/ and larger mean nasalance score for BBP compared to both NC and ALS-BPS groups (p<0.05). Although the ALS-BS group exhibited a larger mean nasalance score for the sentence MMJ compared to participants in the ALS-BPS group, they were observed to produce /m/ with significantly reduced nasal flow (p<0.05).
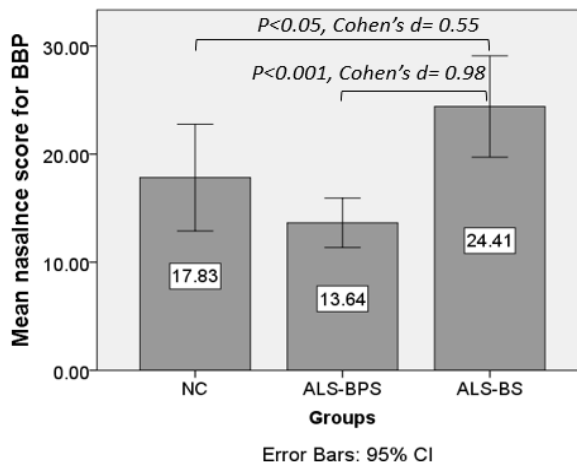
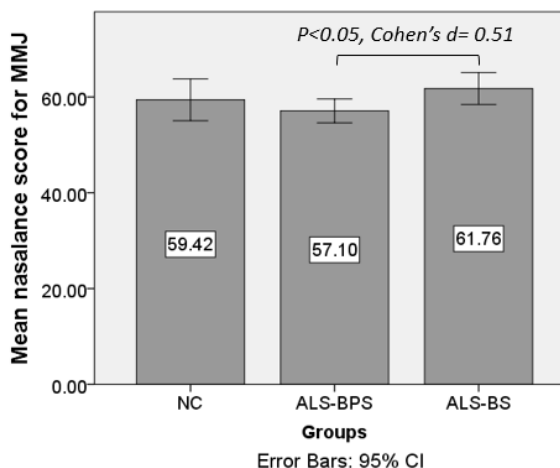**Figure 1**: Mean NF at peak oral pressure (l/sec) of /p/ sound in NC, ALS-BPS, and ALS-BS groups.
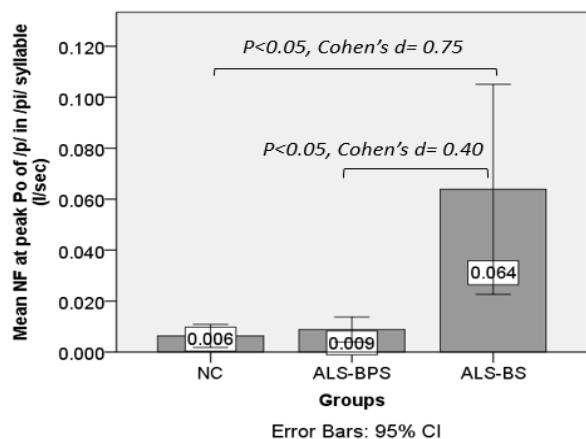
Figure 2: Mean peak NF (l/sec) of /m/ sound in NC, ALS-BPS, and ALS-BS groups.

Figure 3: Mean nasalance score for the sentence BBP in NC, ALS-BPS, and ALS-BS groups.

Figure 4: Mean nasalance score for the sentence MMJ in NC, ALS-BPS, and ALS-BP groups.

4. DISCUSSION

During the production of high-pressure sounds (i.e., stops, fricatives, and affricates), the VP port is tightly sealed to allow for the build-up of intraoral air pressure and to prevent nasal air emission. In the presence of VPD, however, the coupling of oral and nasal cavities can result in increased resonance disturbances.

Our findings on persons with ALS are consistent with the presence of "mixed nasality" with hypernasality during vowels and voiced consonants and hyponasality for nasal sounds. Specifically, in comparison to the ALS-BPS group, the ALS-BS group produced the /p/ with relatively increased nasal flow and the /m/ with reduced nasal flow. These paradoxical findings of too-much and too-little nasal flow might be explained by the effects of spasticity on velar function. A diagnosis of ALS requires the presence of both spastic and flaccid symptoms. Weakness of the VP muscles due to flaccidity would be expected to result in increased nasal flow for all speech contexts; whereas abnormal nasal emission due spasticity might be expected to be more variable and context dependent.

Despite having excessive nasal emission during production of /p/, peak oral pressure was surprisingly unaffected in the ALS-BS group. This finding was unexpected because (1) we anticipated that peak oral pressures would be reduced due to nasal air escape and (2) we expect persons with speech involvement to have reduced respiratory drive. The observation of normal oral pressures in patients with bulbar motor weakness, however, raises the possibility that despite the weakness, affected patients compensate for nasal air escape, perhaps by overdriving their respiratory system. Possible strategies include initiating /pi/ syllables at higher lung volumes or using increased expiratory muscle effort to maintain adequate oral pressure.

Nasometer findings, however, were not as responsive as the aerodynamic measure of nasal flow in identifying bulbar motor involvement. Although nasometer findings for the BBP sentence are consistent with nasal flow data for /p/ sound in both ALS-BP and ALS-BPS groups, the mean nasalance scores for the MMJ sentence do not support the corresponding aerodynamic data (i.e., the peak nasal flow for /m/) in both ALS-BS and ALS-BPS groups. This inconsistency can be attributed to the fact that the energy of nasal emission may fall outside the range of the bandpass filtering defined for the nasometer (center at 500 Hz; -3dB bandwidth of 300 Hz). Therefore, the nasometer may fail to adequately capture VP status when the input nasal signal contains excessive nasal air emission. In support of this explanation, in a study conducted by [18], the sensitivity of nasometry in correctly identifying hyponasality tremendously increased upon the removal of patients with nasal air emission.

These findings suggest that aerodynamic measures should be included in both research and clinical assessment protocols for a comprehensive evaluation of VP function and resonance profile of individuals with ALS, specifically those at an early stage of the disease progression.

5. CONCLUSION

Findings of this study improved our understanding about the speech aerodynamic system as well as the resonance profile in persons with bulbar motor involvement due to ALS. While aerodynamic indices, more specifically measures of nasal flow, were able to demonstrate subtle differences between ALS subgroups, nasometer findings failed to show such distinctions. Nasometric measures alone should not be used to make specific judgement about the status of VP mechanism in patients with ALS.

6. ACKNOWLEDGEMENT

7. REFERENCES

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