Critical Review:
Which Speech Symptoms Contribute Most to Reduced Intelligibility in Individuals with Ataxic Dysarthria Secondary to Friedreich's Disease?

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This critical review examines the evidence regarding which speech dimensions effect impaired intelligibility in persons with Friedreich's ataxia (FA). Study designs include single group designs and one study comparing single group data with normal controls'. Overall, there is inconsistent evidence that word final voicing, vocal harshness, inconsistent articulatory breakdown and metered speech contribute to reduced intelligibility in this population. This evidence supports hypotheses that there are variable courses of spinocerebellar degeneration in patients with FA. This relationship between disease evolution and speech symptoms warrants further investigation. Recommendations for future studies and clinical applications are provided.

Introduction

Friedreich's ataxia (FA) is a hereditary, neurodegenerative disease that is caused by lesions in the cerebellum, cortical, bulbar and spinal pathways of the nervous system (Ackermann, Hertrich & Hehr, 1995; Folker et al. 2010; Gilman & Kluin, 1985; Hiller, 1929). Reflecting a complex etiology, Geoffrey et al. (1976) suggested there are a number of core symptoms essential to a diagnosis of FA yet presentation is heterogeneous, dependent on the stage of the disease.

Recent evidence indicates these patients share a common pathogenesis in the cerebellum but Friedreich's disease follows disparate evolutions. Mean age of onset has been shown to be as young as 10.52 years while mean age at time of death has been reported as 32 years of age. Prevalence rates are not significantly different between genders. (Geoffroy et al. 1976; Harding, 1981). Dysarthria becomes a salient clinical feature of FA with cerebellar involvement in the first two years after onset. It is universally present in this population (Blaney & Hewlett, 2007b; Geoffroy et al. 1976; Harding, 1981). Effects of this motor speech impairment on quality of life in this population are not well documented. As Friedreich's disease changes over time, individuals face innumerable challenges such as cardiomyopathy, hand wasting, ataxic gait and lower body sensory impairment, diabetes and retinopathy in addition to adjustments to speech function (Harding, 2007).

Individuals with FA may present with features of a mixed dysarthria. Flaccid and spastic elements of dysarthric speech are observed in combination with ataxic dimensions such as imprecise consonants and irregular articulatory breakdown (Brown, Darley & Aronson, 1970; Darley, Aronson & Brown, 1969). Cluster analysis shows that voicing and vowel phoneme errors contribute more to reduced intelligibility in this population than in the ataxic dysarthria population at large described by Darley, Aronson and Brown (1969) (Ackermann, Hertrich & Hehr, 1995; Gentil, 1990; Rosen et al. 2010).

A clinically useful method for assessing individuals with FA is to identify the cluster of deviant speech characteristics that is typical of the population. Given that patients with FA present with inconsistent patterns of errors across the identified speech dimensions, recent studies have focused on cluster analyses that differentiate subgroups of this population. Speech language pathologists need to be cognizant of these subgroups and how to intervene appropriately for ever-changing speech targets for improved overall communication.

Importantly, it is the role of clinicians to assess intelligibility and mitigate the deleterious effects of Friedreich's disease on speech and quality of life in this population. Measures of intelligibility hold great ecological validity, reflecting the ability of individuals with FA to communicate verbally with novel listeners. A review of prosodic, voice, respiration, resonance and articulation characteristics that contribute to impaired intelligibility is essential for effective management of individuals with FA throughout the course of the disease. (Blaney & Hewlett, 2007a).

Objectives

The primary objective of this paper is to critically evaluate existing literature with regard to the effect of specific speech error patterns on intelligibility in FA. The secondary objective is to provide evidence-based recommendations for speech language pathologists in clinical practice, e.g. charting specific speech goals for different stages of FA.

Methods

Search Strategy

Computerized databases, including PsycINFO, PubMed, CINAHL and Google Scholar, were searched using the following strategy: ((Friedreich's disease) OR (Friedreich's ataxia) OR (FRDA)) AND ((dysarthria) OR (ataxic dysarthria) OR (ataxia)) AND (speech) AND (intelligibility)). The search was limited to articles written in English.

Article references were also reviewed.

Selection Criteria

Studies selected for inclusion in this critical review were required to investigate dimensions of speech and intelligibility in patients with diagnoses of Friedreich's disease. No limits were set on the demographics of research participants or outcome measures.

Data Collection

Results of the literature search yielded the following types of articles congruent with the aforementioned selection criteria: single group tests (3) and a single group test with normal values for comparison (1).

Results

Blaney and Hewlett (2007a) conducted a single group study examining the relationship between listener intelligibility ratings and perceived speech error profiles of 11 adult males diagnosed with Friedreich's disease. This group included only volunteers solicited through the Ataxia Association of Ireland and who had experienced early onset of the disease. Standardized tasks on the Frenchay Dysarthria Assessment (FDA) and single word stimuli from the Phonetic Intelligibility Test (PIT) were used to create error profiles for each participant. Total error scores on the PIT were calculated and classified by severity groups. Visual inspection revealed final plosive voicing status was the single largest source of error. Approximately 65% of the errors were made across a cluster of seven speech error categories: final plosive voicing; glottal vs. null; stop and nasal place; final consonant vs. null; stop vs. fricative; high vs. low vowel; and initial plosive voicing. The mean error rate for these categories was significantly correlated with the speakers' overall intelligibility scores as determined by the PIT which were, in turn, correlated with severity on the FDA.

Altogether the authors of this study used valid measures to attain clinically useful results and the study was somewhat well-formulated. Participants' ages were wide-ranging but all were male. Categorical speech dimension data from the PIT were appropriately analyzed using both visual inspection and correlations. Overall, this study provides compelling evidence that word final plosive voicing and six additional speech error patterns are significantly associated with intelligibility in patients with FA.

Blaney and Hewlett (2007b) also conducted a follow-up investigation to determine which acoustic parameters of word final voicing are associated with perceptions of speech errors and impaired intelligibility. Ten male subjects diagnosed with Friedreich's disease and mild to mild-moderate dysarthria completed a single word intelligibility assessment based on the PIT. Participants spoke four sets of minimal pairs that contrasted by final plosive voicing. Speech and language students scored responses off-line using a standard 4-choice recognition paradigm. Perceived voicing status scores were tabulated and used in regression analyses with acoustic data. Acoustic features of each response were measured by oscilloscope and spectrograph. Results of this study showed there are significant associations between properties of the acoustic signal produced by individuals with FA and listeners' perceptions of word final plosive voicing. Perceived voiceless quality was significantly correlated with vowel duration, voicing duration into closure, first formant (F1) at 20 ms before offset, F1 at 10 ms before offset and drop in F1 at vowel termination.

This study was somewhat well-formulated. Tests of association between acoustic variables and perceived production errors are clinically valid methods for determining treatment targets. As discussed, the PIT is a valid measure of assessment. It constrains listener choices and allows clinicians to measure the frequency and, indirectly, source of each speech error. In spite of these strengths, there were methodological shortcomings however. The sample in this study is similar to the group previously discussed. The stated sample size is large enough for adequate power, yet participants were not representative of the greater FA population. As well, authors of this study did not use a standard protocol

for acoustic measurements of speaker samples. Only 10% of acoustic measurements were re-measured by the same judge. Intra-rater reliability was calculated, but no data were reported. Statistical methods used in this article were appropriate. Multiple regression and linear regression determined straightforward and expected relationships between vowel length, onset and closure with perceived word final plosive voicing. Overall, the results of this study provide a compelling level of evidence that vowel productions critically influence listeners' perceptions of word final plosives and overall intelligibility in this population.

Joanette and Dudley (1980) investigated associations between speech variable ratings, outlined by Darley, Aronson and Brown (1969), and intelligibility in a single group study of participants diagnosed with FA. Twenty-two French-speaking individuals between the ages of 19 and 49 years volunteered as subjects. They were not grouped by severity. Each participant provided conversational speech samples, which were subsequently rated using 7-point equal interval scales for 16 dimensions along with the variable intelligibility. Analysis revealed a "general dysarthric" factor and a "phonatory stenosis" factor that distinguished three participant groups. Intelligibility was found to be highly correlated with the first factor and, specifically, with the dimensions 'imprecise consonants,' 'excess and equal stress' and 'prolonged phonemes.'

Joanette and Dudley (1980) employed a simple and mostly effective design for this population. A large group of participants provided samples of spontaneous connected speech for rating. While ecologically valid, these samples were neither phonetically balanced nor standardized for direct comparison with data from normal controls. Both authors and one graduate student then rated samples unblinded using a seven point equal interval scale on the described speech dimensions (Darley, Aronson & Brown, 1969). Significant intra- and inter-rater reliability data were reported.

Appropriate statistical manipulations including an intercorrelation matrix, principle components analysis and stepwise discriminant analysis together showed several speech dimensions are associated with judgements of intelligibility. Results provided compelling evidence that individuals with FA can be grouped by factors, which are distinguished by voice dimensions 'harshness' and 'pitch breaks' along with intelligibility.

Folker et al. (2010) used a single group test design with normal values for comparison in their study of individuals with Friedreich's ataxia (FRDA). A total of 21 female and 17 male participants were compared with 20 unimpaired individuals matched for age with the FRDA participants further subdivided by severity (mild/moderate). Participants completed the Friedreich's Ataxia Rating Scale (FARS) for a score of overall disease severity, read the 'grandfather passage' and completed the Assessment of Intelligibility of Dysarthric Speakers (ASSIDS). Samples were rated independently by two speech language pathologists in 30 speech dimensions identified by Darley, Aronson and Brown (1969).

ASSIDS results and perceptual ratings for the FRDA group and controls were compared using Mann-Whitney U. A "modified" Bonferroni procedure controlled for Type I error in this comparison. Dysarthria severity was significantly related to disease duration, identification of the genetic marker GAA2 and FARS score. Ten speech dimensions were found to differentiate controls from participants with Friedreich's ataxia and these dimensions were used to define subgroups using cluster analysis. Agglomerative hierarchical cluster analysis showed a main subgroup and two smaller subgroups. Groups differed significantly in disease duration. On visual inspection, the first group was characterized by mild impairment in dimensions of consonant imprecision, reduced pitch variation, loudness maintenance, reduced phrase length, reduced breath support for speech and hypernasality. Intelligiblity in running speech for this group was mildly reduced. The second and third subgroups' speech was more severely impaired. Subgroup 2's pattern of speech dimension errors reflected velopharyngeal incompetence. A factor related to laryngeal functioning differentiated subgroup 3. This group was characterized as having strained-strangled voice quality and low hypernasality ratings.

Folker et al. (2010) collected a large and representative sample of participants. The study was well-designed. Variables such as genetic confirmation, disease duration and FARS' scores of overall disease severity were included in factor analysis. Appropriate hierarchical cluster analysis was used to define and compare subgroups' speech profiles. This study was well-designed, methodologically and statistically valid. Therefore, it provides compelling evidence that both disease duration and voice-related errors have some association with impaired understanding of individuals with FA.

Discussion

Overall, the reviewed research provides suggestive evidence that word final voicing status,

imprecise articulation, metered speech and vocal harshness are associated with reduced speech intelligibility in speakers with Friedreich's ataxia. Related evidence suggests that there are measurable acoustic correlates of misperceived speech dimensions (Blaney & Hewlett, 2007b). There is clinically relevant data that suggests subgroups of individuals with FA can be distinguished by unique clusters of speech dimensions and by variable ratings of intelligibility (Folker et al. 2010; Joanette & Dudley, 1980). Results of these studies should be interpreted with caution. Sample groups were limited with regard to disease duration and overall severity.

No conclusive statement about speech dimensions' effect on intelligibility can be made for this population. Methodologies and statistical analyses were easily compared across the examined literature. For example, studies widely incorporated gold standard clinical assessments of intelligibility, used perceiver ratings for the speech dimensions described by Darley, Aronson and Brown (1969) and reported results of factor analyses (Blaney & Hewlett, 2007a; Folker et al. 2010; Joanette & Dudley, 1980). Despite these commonalities, findings were inconsistent, owing to variable presentation of dysarthric symptoms across all participants with FA. Factors such as overall disease severity and duration since diagnosis were related to dysarthria severity ratings.

Subgroups were differentiated by clusters of symptoms such as "phonatory stenosis" which suggests there may be various evolutions of FA (Joanette & Dudley, 1980). Future research could endeavor to focus on pathological differences in dysarthric speakers with FA. Cross-sectional and longitudinal designs should be employed to determine speech pattern differences across all stages of this disease. The relationship between neurological changes and motor speech physiology needs further investigation.

Joanette and Dudley's (1980) results support the hypothesis that Friedreich's ataxia is a general descriptor that encompasses many syndromic disorders marked by spinocerebellar degeneration. Specific disorders follow separate evolutions and effect "heterogeneous speech dysfunctions." This theory is further supported by Folker et al. (2010) who suggest there is common involvement of the cerebellum with selective involvement of cranial nuclei.

Blaney and Hewlett (2007a) identify the highest single source of speech errors as word final voicing contrast. Given that dysarthria is one of the earliest symptoms of the onset of FA, clinicians can use this error to plan speech goals and develop comprehensive intervention strategies in therapy. Additional important findings show spectrographic analysis has clinical utility in treatment for this population (Blaney & Hewlett, 2007b). Segment duration reduction and reduced voice onset time are symptomatic of impaired cerebellar mediation in speech tasks. From a treatment perspective, Blaney and Hewlett (2007b) found that preceding vowel length strongly influences perceived final plosive voicing status and would be a strong candidate as a therapeutic goal for improving overall intelligibility.

Conclusion and Clinical Implications

In the present review, word final voicing, imprecise articulation and harshness were all associated with intelligibility ratings. Identification of these speech errors may inform treatment for clinicians working with this population.

Therapeutic and compensatory interventions strategies need to be designed and implemented for subgroups of patients with FA. Finally, clinical outcomes need to be studied to support communication function across the life span of individuals with FA.

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