Speech Perceptual Characteristics of Individuals with Dysarthria Secondary to Huntington's Disease

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Background

Huntington's disease (HD) is an inherited neurodegenerative disorder characterized by chorea, cognitive decline, and personality change (1). Onset occurs around 35-45 years of age with death approximately 10 to 20 years after diagnosis (2). Although progression primarily involves basal ganglia degeneration, cerebellar atrophy and cerebral white matter may be involved (3-4). Existing research of speech perceptual characteristics of HD is limited but describes dysarthria as hyperkinetic with variable rate, prolonged intervals and inappropriate silences, reduced pitch variability, irregular/imprecise articulation, phonatory deviations, and sudden forced inspiration or expiration (5). It is possible that the speech perceptual characteristics of HD are much more variable and perhaps resemble a variety of dysarthria types.

Purpose

The purpose of this research study was to determine speech perceptual characteristics and identify potential distinct clusters, or subgroups of speakers, with HD within a mild speech severity range.

Research Questions

1. What are the speech perceptual characteristics consistent with diagnosis of HD and how do they compare to previous literature on hyperkinetic dysarthria?
2. Are there distinct clusters of speech perceptual characteristics within speakers with mild dysarthria due to HD?

Participants

Speakers: 49 individuals with genetically confirmed Huntington’s disease. All participants:
- Were native English speakers
- Had genetically confirmed HD
- Were at least 70% intelligible
- Seen for routine follow-up in VUMC interdisciplinary HD clinic

Materials & Procedures

Patient Screening Materials:
- Sentence Intelligibility Test (SIT) - to determine speech impairment severity.
- The Rainbow Passage - to determine speech perceptual characteristics

Speech Rating Materials:
- Speech perceptual characteristics checklist (Darley et al., 1969a, 1969b)

Recording equipment:
- Tascam digital recorder (DR-100MKII) and lapel microphone (Audio-Technica AT899) with a microphone-to-mouth distance of approximately six inches

Procedures

Collection of Speech Samples:
All participants completed a brief speech screening during visits to the VUMC HD clinic between 5/2016 & 7/2017 consisting of:
- Patient history and interview
- Sentence Intelligibility Test (SIT)
- Reading of “The Rainbow Passage”

Speech Perceptual Ratings:
- Following training, 4 graduate students completed ratings using a checklist of 38 speech perceptual characteristics (Darley et al., 1969a, 1969b) for all 49 participants
- Ratings scale ranged from 1 (normal) to 7 (severe)
- 3 directional items: pitch, loudness, & rate (indicated by +/-)

Data Analysis

Analysis for Research Question 1:
- For each speaker ratings were averaged across 4 listeners for each speech characteristic
- Across speakers, rating of speech characteristics were averaged to determine the group’s speech perceptual characteristics
- For directional items, absolute value of average items were used
- Final group averages for the 49 speakers were ranked in descending order (most salient to least salient)

Analysis for Research Question 2:
- Unsupervised k-means clustering analysis
- Clustering determination made by AIC method
- Submitted all speakers’ average ratings to cluster analysis

Results

As a group, the participants with HD in the current study show some similar speech characteristics to those found in previous studies (Darley et al., 1969a, 1969b). However, there are differences in speech characteristics across all speakers as highlighted by the outcome of multiple clusters.

Conclusion

Among the 4 identified clusters (subgroups), preliminary findings suggest that:
- No speech characteristics were found to be unique for subgroup 1; however, some were only shared with subgroup 2. These were an abnormally slowed rate, phonic prolongations, excess and equal stress, and a harsh voice.
- Strained-strangled voice, vowel distortions, and variable rate were unique to subgroup 2. Further, intelligibility and bizarreness were rated as most deviant compared to the average rating of these items in the other subgroups.
- Subgroup 3 was the only cluster to have the feature of reduced stress and an abnormally fast rate.
- Subgroup 4 did not present with any speech characteristics that were rated on average as clinically relevant (2 or above); yet, their speech was still rated as mildly bizarre or unnatural (rating close to 2).

Finally, all 4 clusters (subgroups) shared some salient features that were in congruence with the speech characteristics described by Darley, Brown, and Mefferd. However, the rating of deviation from normal differed between subgroups.

Implications & Future Research

The 4 identified clusters (subgroups) with their distinct combinations of speech perceptual characteristics suggest that different treatment approaches may be required to improve speech in these individuals with HD.

Further research is warranted to better determine the factors underlying the variable speech rate and vocal quality across subgroups.

The findings in this study will be expanded in the future to incorporate a larger number of raters. In addition, other clinical parameters of HD with speech will be examined in relation to the discovered subgroup (e.g., disease duration, CAG repeat length, medications).

Selected References