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Original Paper

Auditory Perception in Individuals with Friedreich's Ataxia

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Abstract

Introduction: Friedreich's ataxia (FRDA) is an inherited ataxia with a range of progressive features including axonal degeneration of sensory nerves. The aim of this study was to investigate auditory perception in affected individuals. *Methods:* Fourteen subjects with genetically defined FRDA participated. Two control groups, one consisting of healthy, normally hearing individuals and another comprised of subjects with sensorineural hearing loss, were also assessed. Auditory processing was evaluated using structured tasks designed to reveal the listeners' ability to perceive temporal and spectral cues. Findings were then correlated with open-set speech understanding. *Results:* Nine of 14 individuals with FRDA showed evidence of auditory processing disorder. Gap and amplitude modulation detection levels in these subjects were significantly elevated, indicating impaired encoding of rapid signal changes. Electrophysiologic findings (auditory brainstem response, ABR) also reflected disrupted neural activity. Speech understanding was significantly affected in these listeners and the degree of disruption was related to temporal processing ability. Speech analyses indicated that timing cues (notably consonant voice onset time and vowel duration) were most affected. *Conclusion:* The results suggest that auditory pathway abnormality is a relatively common consequence of FRDA. Regular auditory evaluation should therefore be part of the management regime for all affected individuals. This assessment should include both ABR testing, which can provide insights into the degree to which auditory neural activity is disrupted, and some functional measure of hearing capacity such as speech perception assessment, which can quantify the disorder and provide a basis for intervention.

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Auditory Perception in Individuals with Friedreich's Ataxia

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Introduction

Friedreich's ataxia (FRDA) is the most common of the inherited ataxias, affecting approximately 1 in 29000 Caucasians [Delatycki et al., 2000; Pandolfo, 2008]. The principal features of FRDA include progressive ataxia, absent lower limb reflexes, spasticity, scoliosis, impaired vibration sense and proprioception, foot deformity and cardiomyopathy [Harding, 1981; Delatycki et al., 2000]. Presentation of FRDA is usually in childhood, with an average onset age of 10 years [Delatycki et al., 1999]. To date, no therapies are proven to alter the natural history

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