Auditory Perception in Individuals with Friedreich’s Ataxia

Rance G. · Corben L · Barker E · Carew P · Chisari D · Rogers M · Dowell R · Jamaluddin S · Bryson R · Delatycki M

Department of Otolaryngology, The University of Melbourne, and Murdoch Children’s Research Institute, Parkville, Vic., Australia

Keywords: Friedreich’s ataxia · Auditory perception · Temporal processing · Speech perception

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.
Related Articles:

Otorhinolaryngol Nova 1999;9:105–114

Results of Pediatric Cochlear Implantation Compared with Results Obtained with Hearing Aids
Kiefer J., Pfennigdorff T., Spelsberg A., Gall V. et al.
Objectives: The performance of cochlear implanted children was evaluated in comparison with results of children wearing hearing aids. Patients and Methods: 88 cochlear implanted children were included in this study. Onset...

Go to Article

Audiology & Neurotology
Audiol Neurotol 2003;8:269–285

Acoustic and Electrical Pattern Analysis of Consonant Perceptual Cues Used by Cochlear Implant Users
Teoh S.W., Neuburger H.S., Svirsky M.A.

It is hypothesized that for postlingually deafened adult cochlear implant (CI) users, a significant source of their perceptual performance variability is attributable to differences in their ability to discriminate the basic...

Go to Article

References


External Resources

• Pubmed/Medline (NLM)
• Chemical Abstracts Service (CAS)


External Resources


   **External Resources**
   - Pubmed/Medline (NLM)
   - Crossref (DOI)
   - Chemical Abstracts Service (CAS)
   - ISI Web of Science


    **External Resources**
    - Pubmed/Medline (NLM)
    - Crossref (DOI)
    - Chemical Abstracts Service (CAS)
    - Cambridge Scientific Abstracts (CSA)
    - ISI Web of Science


    **External Resources**
    - Pubmed/Medline (NLM)
    - Crossref (DOI)
    - Chemical Abstracts Service (CAS)
    - ISI Web of Science


    **External Resources**
    - Pubmed/Medline (NLM)
    - Crossref (DOI)
    - Chemical Abstracts Service (CAS)
    - ISI Web of Science


    **External Resources**
    - Pubmed/Medline (NLM)
    - Chemical Abstracts Service (CAS)
    - ISI Web of Science


    **External Resources**
    - Pubmed/Medline (NLM)
    - Crossref (DOI)
    - Chemical Abstracts Service (CAS)
    - ISI Web of Science

**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)
- ISI Web of Science


**External Resources**
- Crossref (DOI)
- ISI Web of Science


**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)
- ISI Web of Science


**External Resources**
- Pubmed/Medline (NLM)
- Crossref (DOI)

External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- Cambridge Scientific Abstracts (CSA)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Chemical Abstracts Service (CAS)


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)

External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Cambridge Scientific Abstracts (CSA)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- Cambridge Scientific Abstracts (CSA)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- Cambridge Scientific Abstracts (CSA)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Crossref (DOI)
- ISI Web of Science


External Resources
- Pubmed/Medline (NLM)
- Chemical Abstracts Service (CAS)
- ISI Web of Science


Auditory Perception in Individuals with Friedreich’s Ataxia

Gary Rance*, Louise Corben†, Elizabeth Barker*, Peter Carew*, Donella Chisari*, Meghan Rogers*, Richard Dowell*, Saiful Jamaluddin* and Rochelle Bryson*

Martin B. Delatycki††

*Department of Otolaryngology, The University of Melbourne, and † Murdoch Children’s Research Institute, Parkville, Vic., Australia

Key Words
Friedreich’s ataxia • Auditory perception • Temporal processing • Speech perception

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.

Received: April 28, 2009
Accepted: August 21, 2009
Published online: November 05, 2009
Issue release date: May 2010

Number of Print Pages: 12
Number of Figures: 5
Number of Tables: 5

ISSN: 1420-3030 (Print)
eISSN: 1421-9700 (Online)

For additional information: https://www.karger.com/AUD