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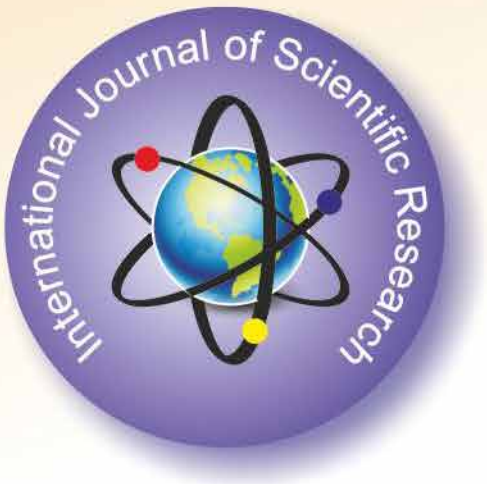

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Research Paper

Perceptual challenges in auditory neural processing in neurodegenerative conditions like Fredereich Ataxia

Medical Science

KEYWORDS : Perceptual challenges in auditory neural processing in neurodegenerative conditions like Fredereich Ataxia

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ABSTRACT

Friedreich ataxia (FA) is considered as a neurodegenerative disorder and is the most frequent type of hereditary ataxia. FA is characterized by degeneration in several spinal tracts, dorsal roots and peripheral nerves. Due to the disruption in the neural function, these individuals pose certain perceptual challenges in understanding speech in the presence of noise. Auditory nerve is considered as an important structure for the neural processing of acoustic sounds. Any break down in its function will result in auditory processing deficits. These difficulties can only be identified with detailed test battery approach which includes both behavioural and electrophysiological tests. Therefore in the present case study, we aim to highlight the auditory processing deficits in FA which resulted in the identification of auditory neuropathy spectrum disorder (ANSD) a rare neural disorder seen in individuals with FA. Further it sheds light in to the possible management options in individual with ANSD.

Friedreich ataxia (FA) was coined and described by German investigator Nicholaus Friedreich (1860).This disease is considered as a neurodegenerative disorder and is the most frequent type of hereditary ataxia. The hereditary pattern is autosomal recessive. FA is characterized by degeneration in several spinal tracts, dorsal roots and peripheral nerves, especially in the Clark column, fasciculus gracilis and cuneatus, and ascending and descending spinocerebellar tracts (Pandolfo, 2009). In some cases, other anomalies can be associated including, cardiomyopathy, pes cavus, xyfoscoliosis, hearing loss, optical atrophy, diabetes mellitus and, glucose intolerance. However, these abnormalities are not homogeneous and different patterns of abnormal auditory responses can be found.

It is reported that the breakdown in the neural process due to degenerative condition can also have its effect on the auditory signal processing. Auditory nerve is an important structure, which act as a signal carrier from peripheral auditory system to central auditory regions. The acoustic nerve consists of thousands of neural fibres which coordinate synchronously in response to an acoustic stimulation. If there is any break down in any of the neural tract, the neural firing gets affected. These individuals pose certain perceptual challenges, especially difficulty in understanding speech in the presence of various acoustic conditions. Till date to our knowledge such findings in neurodengerative conditions like FA is not reported in literature.

Therefore in the present case study, we aim to highlight the auditory processing deficits in FA which resulted in the identification of auditory neuropathy spectrum disorder (ANSD) a rare neural disorder seen in individuals with FA. Further it sheds light in to the possible management options in individual with ANSD.

Method

A 16 year old boy came to our clinic with the complaint of reduced hearing sensitivity and difficulty of understanding speech sounds. He was reported to be normal till 14 years of age and observed reduced hearing sensitivity one and half years back. Neurological evaluation was done and he was diagnosed as FA. As the primary complaint was hearing difficulty, a diagnostic audiological test battery was performed in order to identify the possible cause for hearing loss. Pure tone audiometry (PTA) and Speech in noise test (SPIN) was done by using a dully calibrated GSI-61 (Grason Staddler Inc-61) clinical audiometer. An Immittance evaluation was done to assess the middle ear status by using GSI-Tympstar. To evaluate the integrity of outer hair cell functions, Transient Evoked Otoacousitc Emission (TEOAE) was recorded by using ILO-v.6.Followed by which, a click evoked Auditory Brainstem Response (cABR) was obtained by using Intelligent Hearing System - Evoked Potential (IHS-EP). A Speech Evoked Cortical Auditory Potentials (SECAP) recording has done in order to assess the auditory cortical neuron responses for speech stimulus. After the audiological evaluation, hearing aid trail was done and subsequently, SECAP recording was done to assess the hearing aid benefit. All the testing was carried out in a sound treated room with permissible noise levels. Both subject and parents were informed about the potential importance and scientific benefit of reporting these findings and dully signed consent form were obtained.

Results

From the above audiological tests, PTA in both ears obtained were 71.6dBhL and diagnosed as bilateral severe rising sensori neural hearing loss (Figure-1).An audiometric SPIN test revealed a drastic reduction on speech perception scores when tested at -10dBHL signal to noise ratio (SNR).No middle ear pathology was observed, however, an absent acoustic reflex threshold was observed in both the ears. TEOAE was recorded with good amplitude, which is an indication of intact pre-neural function. No identifiable peaks obtained at 90 dBnHL in cABR, indicates poor or no synchronous firing of auditory nerve neurons (Figure-2). SECAP responses were present with a broadened peaks at a latency of 120-200ms (Figure-3a).From the results of behavioural and evoked potential tests, the client was diagnosed to have an Auditory Neuropathy Spectrum Disorders (ANSD).Hearing Aid Trial (HAT) was carried out with a 3 channel digital hearing aid and found that there is no benefit from the hearing aid. However, a SECAPs recording found that, there is a latency shift seen of about 40 ms for N1-P2 complex before and after fitting the hearing aid (Figure-3b).

Discussion

The aim of this present case report is to highlight the potential importance of indentifying the speech understanding deficits in neurodegenerative conditions like FA. The authors would like to explain how the breakdown in the neural conduction affects the auditory neural processing which is crucial for understanding the speech sounds. The possible explanation is that the neuro degenerative effect seen in these individuals could have largely influenced the spiral ganglion functions, which is prominent for the normal neural conduction (Landry, Wise & Fallon,2011). Due to the ganglion loss, the synchronous firing of auditory neuron fibres will be affected, which results in impaired neural performance in auditory brainstem response and intact outer hair cell function.

The other reason could be that the disturbances in the cerebellar control circuit which possibly send the signals towards lower extremities especially spinal cords may influence its functioning on the proximal neural regions. As the auditory nerve function is important for the temporal encoding of speech sounds, one would suggest that individuals with FA will show temporal encoding deficits. As the cochlea is responsible for frequency selectivity and frequency discrimination, these individuals would not show any effect on frequency discrimination task (Jorisa,

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Bergevinb & Kalluri.2011).

Generally in cases of ANSD, the hearing thresholds will be in the normal range or it may extent up to severe to profound hearing loss (Patricia, 2008). The obtained auditory thresholds in ANSD pose certain physiological changes in the central auditory system. In the present case, the audiogram pattern observed were a rising pattern, where the high frequency sensitivity were preserved compared to the lower frequency sensitivity, yet a considerable amount of hearing loss is seen. The possible explanation for the rising pattern seen in ANSD subjects could be explained based on the physiological changes taking place inside the auditory system. The auditory system processes the acoustic signal through place coding and through phase locking. Phase locking is defined as the aggregate neural responses for a particular acoustic stimulus in which it preserves the phase of the signal (Krishnan, 1999). In humans phase locking is up to 2 KHz and above 2 KHz place coding takes place. Therefore the affected neural synchrony will have its impact on the phase locking mechanism which results in the disruption in the signal processing especially at the lower frequencies up to 2 KHz, which results in the broader low frequency loss. However, the severity of neural dysfunction will also have an additional effect on the higher frequencies in the auditory range which results in flat hearing loss in later stages of ANSD (Gary, Colette &David, 2004). All these explanations highlight the effect of neural degeneration on the auditory system and its effects on the speech understanding deficits, which is predominantly featured in FA.

The management of ANSD is always been a challenging task for audiologists (Patricia, 2008). In the present case, the client was not appreciating the hearing aid benefit behaviourally. On SECAP recording, a considerable amount of differences in latencies has seen with and without the hearing aid (Gary et al, 2004). This has strengthens the notion that the temporal processing could possibly be affected in ANSD individuals. The changes in latency towards the better side reveals that the amplified signals through the hearing aids may process better through the auditory nerve even though the phase locking is impaired. However, it is premature to comment on such hypothesis as it requires more clinical and scientific evidence to support such findings.

To improve the subject's communication abilities in school and other noisy environments, we advocated the importance of total communication. But it is yet to be proven, how effective are these strategies, when the client is forced to communicate under adverse noisy conditions. If not trained structurally, ANSD individuals cope up with the communication environment with certain tailor-made methods or strategies by themselves.

Conclusion

To conclude, auditory processing of speech sounds are important for communication aspects .Individuals with ANSD pose certain perceptual challenges in adverse noisy conditions. Such individuals can only be identified with appropriate application of behavioural and evoked potential tests, especially in neurodegenerative conditions like FA. Auditory Management of these individuals with ANSD is also challenging task for audiologists. However, with amplification and other communication strategies can help them to improve their communication skills.

Figure-1: Bilateral rising audiogram pattern in ANSD

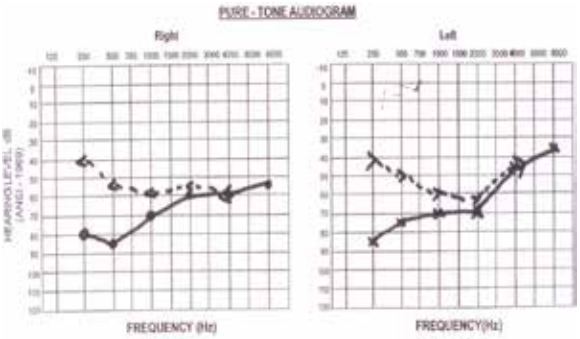


Figure-2: No identifiable cABR obtained at 90dbnHL in both ears

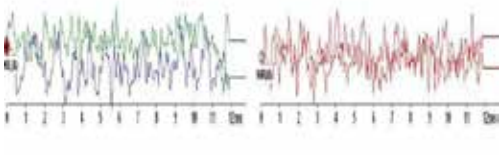


Figure-3a- Unaided SECAP responses in ANSD

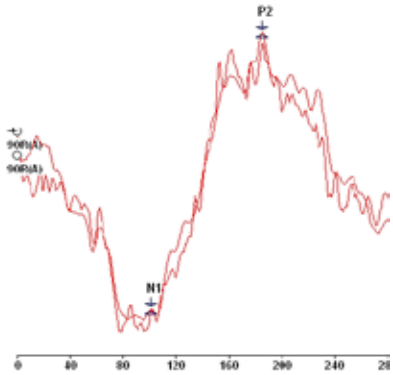
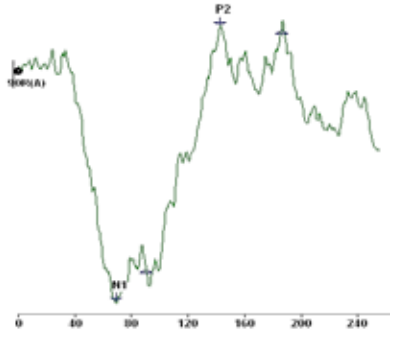


Figure 3b – Aided SECAP responses in ANSD



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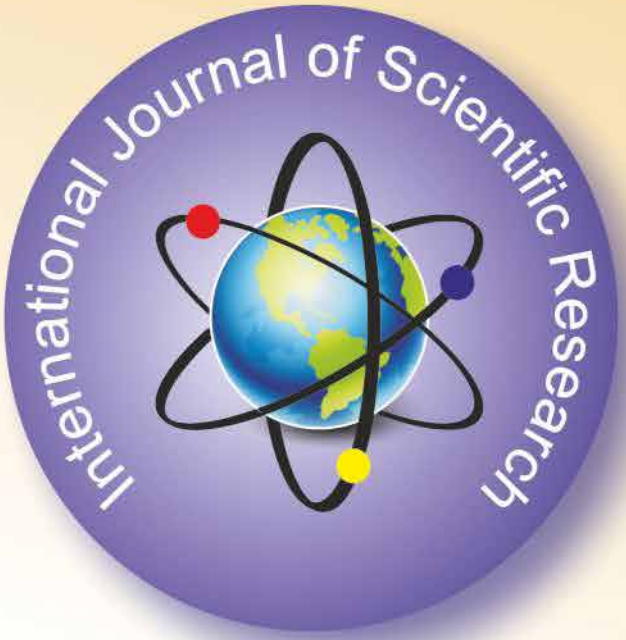

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