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Subjective and Objective Estimate of Neural Fatigue in an Adult with Auditory Neuropathy Spectrum Disorder

Prashanth Prabhu*

All India Institute of Speech and Hearing, Mysore-570006, Karnataka, India

***Corresponding author:** Prashanth Prabhu, All India Institute of Speech and Hearing, Mysore-570006, Karnataka, India, Tel: +91 8904353390; E-mail: prashanth.audio@gmail.com

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Abstract

Auditory neuropathy spectrum disorder (ANSD) is a retrocochlear disorder in which cochlear functioning is normal but the transmission in the auditory neural pathway is affected. The present study reports of neural fatigue seen in subjective and objective tests of adaption in a 19 year old adult with ANSD. He had bilateral mild sensorineural hearing loss with otoacoustic emissions present and absent auditory brainstem response. Immittance evaluation showed elevated acoustic reflexes at 500 Hz and 1000 Hz in both ipsilateral and contralateral stimulation. Reflex decay test was administered at 500 Hz and 1000 Hz which showed positive reflex decay with contralateral stimulation. Olsen and Noffsinger tone decay test and supra-threshold adaptation test showed positive tone decay at 500 Hz, 1000 Hz and 2000 Hz in both ears. The results of the study showed positive reflex decay and positive tone decay which indicate abnormal neural fatigue. The abnormal neural firing seen in individuals with ANSD could have resulted in positive reflex decay and tone decay. However, further studies on larger group of population are essential for further understanding the mechanisms in detail.

Keywords: Auditory neuropathy spectrum disorder; Adaptation; Neural fatigue; Reflex decay; Tone decay

Introduction

Auditory neuropathy spectrum disorder (ANSD) is a defined as a retrocochlear disorder in which patient has normal otoacoustic emissions/cochlear microphonics and absent/abnormal auditory brainstem response (ABR) [1-5]. The middle-ear muscle reflexes are reported to be abnormal in both adults and children with ANSD [1,6,7]. Berlin et al. [1] reported that middle ear muscle reflexes were absent in 89% of the participants and it was abnormal (combination of elevated and absent) in 11% of the participants among 148 patients with ANSD. Narne et al. [8] reported that 198 patients with ANSD in their study had absent acoustic reflexes in both ears. They concluded that acoustic reflexes are absent in patients with ANSD since the auditory nerve is unable to provide sufficiently high or sufficiently synchronized rates of discharge to activate the motor neurons of the stapedius muscle [4,9]. Thus, the studies show that reflexes are rarely present and it is elevated in individuals with auditory neuropathy spectrum disorder [1,8]. There are very limited studies reported in the literature which have attempted to determine acoustic reflex decay when reflexes are present in individuals with ANSD. The pattern of acoustic reflex decay is not reported in any of the previous studies on ANSD. In addition, there is also a dearth of literature on determining neural fatigue using different subjective tests in individuals with ANSD [10]. Thus, the present study attempts to report a unique case with auditory neuropathy spectrum disorder who presents abnormal neural fatigue in subjective and objective tests.

Case Presentation and Methods

A 19 year old adult male reported to audiology clinic with a complaint of reduced hearing sensitivity in both ears since 4 years. He reported of difficulty understanding speech. He reported occasional headache and intermittent tinnitus. He also reported that the problem was more in noisy situation. The participant underwent detailed audiological evaluation which included pure tone audiometry, speech audiometry, Immittance evaluation, otoacoustic emissions (OAE), auditory brainstem response (ABR) and long latency responses (LLR) using standard protocols. Neurological evaluation included clinical neurological examination, CT and MRI scan to identify any space occupying lesion in the auditory nerve. Informed consent was taken from him to conduct the study and publish the data. The results of the audiological evaluation showed that he had bilateral mild sensorineural hearing loss. The pure tone threshold in right ear was 30 dB HL and left ear was 31.67 dB HL. The audiogram for both ears is shown in Figure 1. The speech identification scores were 96% in right ear and 56% in left ear. He had 'A' type tympanogram with acoustic reflexes present but elevated at 500 Hz and 1000 Hz in both ipsilateral and contralateral stimulation. The details of acoustic reflexes recorded in the participant are shown in Table 1. OAE were normal indicative of normal outer hair cell functioning and ABR was absent indicative of retro-cochlear pathology. Clinical neurological evaluation suggested ANSD. In addition, CT and MRI scan results showed no space occupying lesion and confirmed the diagnosis of ANSD. All the above findings correlated with the diagnosis of auditory neuropathy spectrum disorder [1,3].

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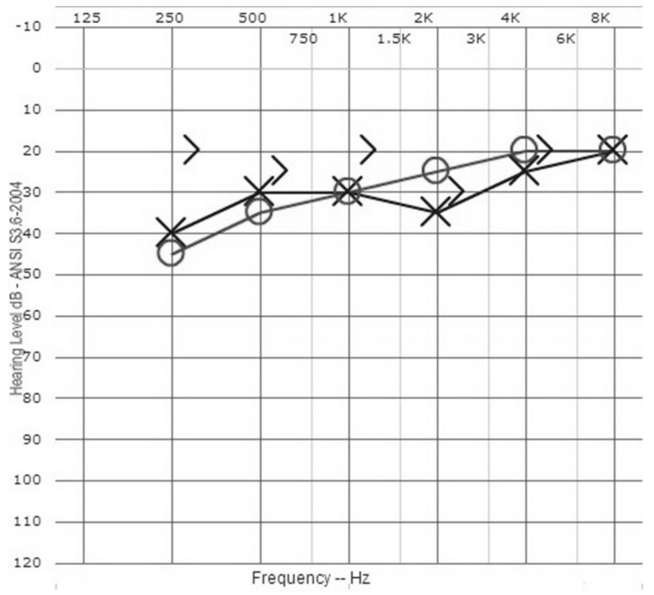

Figure 1: Audiogram of right and left ear air conduction thresholds and left bone conduction thresholds.

Ear		500 Hz	1000 Hz	2000 Hz	4000 Hz
Right Ear	Ipsilateral	105	100	No Response	No Response
	Contralateral	105	110	No Response	No Response
Left Ear	Ipsilateral	105	105	No Response	No Response
	Contralateral	110	110	No Response	No Response

Table 1: Ipsilateral and contralateral acoustic reflexes elicited at different frequencies in both ears.

Immitance evaluation (tympanometry and acoustic reflex testing) was carried out using 226 Hz probe tone in a calibrated Middle ear analyzer (GSI-Tympstar V 2.0). Acoustic reflexes were checked at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz tone for both ipsilateral and contralateral ear. Acoustic reflex decay test was administered based on the standard protocol by Anderson, Barr and Wedenberg [11]. Acoustic reflex decay was administered on him at 500 Hz and 1000 Hz in both ears. The results of the study showed positive reflex decay with a decay time of 2 seconds at 500 Hz and 1000 Hz in left ear. In the right ear, positive reflex decay time of 2 seconds and 1 second was noticed at 500 Hz and 1000 Hz respectively. The reflex decay test graphs are with positive decay in right ear and left ear are shown in Figure 2 and Figure 3 respectively.

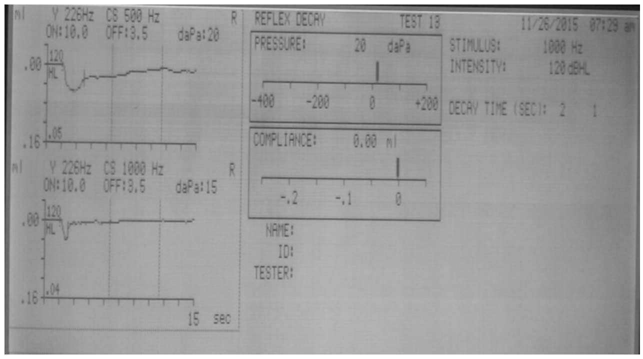

Figure 2: Result of acoustic reflex decay seen at 500 Hz and 1000 Hz in the right ear.

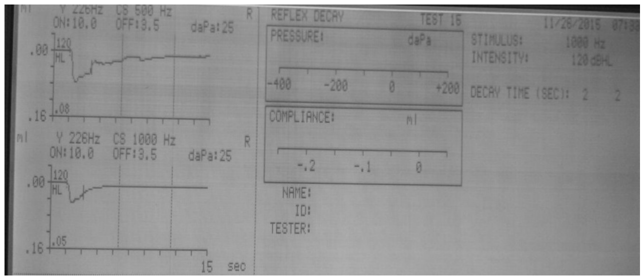

Figure 3: Result of acoustic reflex decay seen at 500 Hz and 1000 Hz in the left ear.

Olsen and Noffsinger tone decay test was tested in both ears by presenting tones of 500 Hz, 1000 Hz and 2000Hz at 20 dB SL. He was instructed to keep pressing the patient response switch till he hears the sound. If the participant was unable to hear the sound for one minute intensity of the sound was increased till 35 dB SL [12]. Olsen and Noffsinger tone decay test showed positive tone decay as he was not able to hear the tone for one minute even at 35 dB SL at all the frequencies. Supra threshold adaptation test (STAT) was also administered on him to assess neural fatigue at higher intensity level. Tones at 500 Hz, 1000 Hz and 2000 Hz were presented at 100 dB HL, 105 dB HL and 110 dB HL, respectively to both ears separately for one minute. Broadband noise at 90 dB SPL was presented to the contralateral ear while testing [13]. He was instructed to indicate the tester till he hears the sound. The test was considered as positive if he failed to hear for one minute. STAT results were also positive and he was not able to hear the tone at all frequencies for one minute. To confirm the positive STAT results, pulsed tones were presented and he was able to hear the tones for one minute.

Discussion

There are very few studies which have used subjective and objective tests of abnormal adaptation in individuals with ANSD. The result of the study shows that he exhibited abnormal adaptation in both subjective and objective tests of neural fatigue. The results are similar to previous studies which report that loudness adaptation is seen in individuals with auditory nerve disorders [10]. In the present study, objective acoustic reflex decay test was administered in a participant with ANSD. The positive reflex decay seen in him provides an objective evidence of abnormal neural fatigue. In addition, subjective tests of adaptation also showed positive tone decay suggestive of neural fatigue. The previous studies on ANSD have focused mainly on spectral and temporal processing deficits [14]. The present study shows that loudness adaptation can also been seen in individuals with ANSD which needs to be explored in further detail. In individuals with ANSD, abnormal loudness adaptation may be associated with reduction in neurotransmitter and energy resources in the auditory nerve [15]. The abnormal adaptation may further exacerbate abnormal speech perception seen in individuals with ANSD [16-18]. Ciccone et al. [19] reported that endothelial functioning is affected in individuals with idiopathic sensorineural hearing loss. However, it was not assessed in him because of lack of facility to perform the test which needs to be addressed in future studies. Thus, the present study suggests that further studies are essential on other individuals with ANSD with acoustic reflexes to understand the mechanisms of neural fatigue.

Conclusions

The present study reported results of subjective and objective tests of adaptation in an adult with ANSD. There are very limited studies which have explored reflex decay test in individuals with ANSD as majority of them don't have acoustic reflexes. In addition, subjective tests of adaptation are rarely reported in the literature. Thus, the present study is a unique attempt which tries to report abnormal neural fatigue in a male with ANSD. The results of the study showed positive reflex decay and positive tone decay suggestive of abnormal adaptation. The abnormal neural firing seen in individuals with ANSD could have resulted in positive reflex decay and tone decay. However, further studies on larger group of population are essential for further understanding the mechanisms in detail.

Conflict of interest statement

The author reports no conflicts of interest. The author alone is responsible for the content and writing of the paper.

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