Evaluation of Dizziness Handicap in Adolescents and Adults with Auditory Neuropathy Spectrum Disorder

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Abstract

Introduction Vestibular symptoms and damage to the vestibular branch of the eighth cranial nerve is reported in individuals with auditory neuropathy spectrum disorder (ANSD). However, the real life handicap caused by these vestibular problems in individuals with ANSD is not studied.

Objective The present study attempted to evaluate the dizziness-related handicap in adolescents and adults with ANSD.

Method The dizziness handicap inventory (DHI) was administered to 40 adolescents and adults diagnosed with ANSD. The study also attempted to determine if there is any gender effect on DHI scores and its correlation to the reported onset of hearing loss.

Results The results of the study showed that adolescents and adults with ANSD had a moderate degree of dizziness-related handicap. The dizziness affected their quality of life, causing emotional problems. There was no gender effect, and the level of the handicap was greater in the cases in which the onset of the hearing loss was reported soon after the diagnosis of ANSD. There could be a vestibular compensation that could have resulted in a reduction in symptoms in individuals in whom the onset of the hearing loss was reported later on.

Conclusion Thus, a detailed assessment of vestibular problems and their impact on quality of life is essential in adolescents and adults with ANSD. Appropriate management strategies should be considered to resolve their vestibular problems and improve their quality of life.

Keywords
- ► dizziness
- ► handicap
- ► quality of life
- ► emotional problems
- ► vestibular compensation
- ► onset of hearing loss

Introduction

Auditory neuropathy spectrum disorder (ANSD) can be defined as a clinical disorder in which individuals have normal otoacoustic emissions (OAEs), and the auditory brainstem response (ABR) is abnormal or absent.[1–4] The prevalence rate of ANSD in Western countries is reported to vary from 11 to 0.5%.[5–10] In the Indian population, Kumar and Jayaram[11] reported that, among individuals with sensorineural hearing loss, 1 in 183 were diagnosed with ANSD. Auditory neuropathy spectrum disorder is a retrocochlear disorder that affects the communication abilities of those diagnosed with it because of their poor speech perception.[1] Vertigo is also one of the symptoms reported by individuals with ANSD. These individuals also exhibit vestibular neuropathy along with auditory difficulties.[12–14]

Auditory neuropathy spectrum disorder is a retrocochlear disorder affecting the vestibulocochlear nerve that can lead

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to auditory and vestibular symptoms. The involvement of the vestibular branch in ANSD is extensively reported in the literature.[12–15] Sazgar et al[15] reported that isolated auditory neuropathy or vestibular neuropathy are rare and the most common pathology is "audio-vestibular neuropathy," affecting both branches of the eighth cranial nerve. Sinha, Shankar and Raja[14] reported that cervical and ocular vestibular-evoked myogenic potentials (VEMPs) were abnormal in individuals with ANSD. Sinha et al[13] showed that VEMPs were absent, and caloric tests showed hypofunctional responses in individuals with ANSD. This suggested the involvement of both the inferior and superior vestibular nerves in such individuals. However, considering all these important test findings, most of the studies have not given the proper importance to the assessment of the dizziness handicap in individuals with this disease.

The focus of the assessment and audiological rehabilitation of ANSD patients has always been on the auditory symptoms, and the vestibular symptoms are usually ignored. Hence, it is essential to understand the effect of vertigo/dizziness on the quality of life of individuals with ANSD. The dizziness handicap inventory (DHI) is a 25-item questionnaire used to determine the dizziness-related handicap.[16] It has three subscales (functional, emotional and physical) that assess the deterioration of quality of life in individuals with balance problems. Therefore, the present study attempts to evaluate the results of the DHI in adolescents and adults with ANSD. It also attempts to determine if there is any gender effect on DHI scores and any correlation between the reported onset of the hearing loss and the DHI scores.

Method

Participants

A total of 40 individuals with ANSD with complaints of vestibular problems were considered for the study. The study sample consisted of 18 males and 22 females between the ages of 14 and 35 years (mean = 21.3 years; standard deviation [SD] =5.27). All the participants had pure tone average (PTAs, at 500 Hz, 1 kHz, 2 kHz and 4 kHz) thresholds ranging from mild (26–40 dB HL), moderate (41–55 dB HL), and moderately severe (56–70 dB HL) to severe (71–90 dB HL) degrees of hearing loss.[17] They were diagnosed as having ANSD based on the presence of transient-evoked otoacoustic emissions but absent auditory brainstem responses. They had no history and/or presence of middle ear pathology with A-type tympanogram[18] and absent acoustic reflexes. The diagnosis of ANSD was confirmed by a neurologist. The time reported for the duration of the hearing loss ranged from 12 months to 180 months. The demographic details, audiological findings and the reported vestibular symptoms are shown in ►**Table 1**.

Procedure

Pure tone air conduction (AC) and bone conduction (BC) thresholds were estimated using the modified Hughson and Westlake procedure.[19] Air conduction thresholds were obtained for pure tone frequencies from 250 Hz to 8 KHz, and BC thresholds from 250 Hz to 4 KHz at octave frequencies. A two-channel diagnostic audiometer (Inventis Audiology Equipment, Padova, Italy) was used to obtain pure tone air conduction and bone conduction thresholds and speech identification scores. Speech identification scores using headphones were obtained for phonemically balanced words developed for adults in Kannada by Yathiraj and Vijayalakshmi.[20] Recorded word lists were routed from a personal computer through a two-channel diagnostic audiometer at 40 dB SL (re: speech recognition threshold – SRT). An immittance meter GSI Tympstar (Grason Stadler Inc., Eden Prairie, MN, US) was used for immittance testing. The better ear of the participant was tested to obtain tympanogram and acoustic reflexes for a probe tone frequency of 226 Hz. Acoustic reflexes were measured using 500, 1000, 2000 and 4000 Hz pure tones, presented to both ipsilateral and contralateral ears. The ILO v.6 (Otodynamics Ltd, Hatfield, UK) OAE analyzer was used to obtain transient-evoked OAEs (TEOAEs). After ensuring adequate probe fit, TEOAEs were measured for non-linear click trains presented at 80 dB pe SPL (decibel peak-equivalent sound pressure level). A waveform reproducibility of more than 50%[21], and an overall signal to noise ratio of more than 3 dB SPL[22] at least at 2 frequency bands was required to be considered as presence of TEOAEs. The Biologic Navigator Pro (Bio-logic, Mundelein, IL, US) AEP system with ER 3A (Etymotic, Elk Grove Village, USA) insert earphones was used to record the ABR. Click-evoked ABR was recorded twice and replicated for 100 µsec click stimuli delivered at a repetition rate of 11.1 clicks/second at 90 dB nHL. The recording was obtained for a total of 1,500 sweeps and a filter setting of 100 to 3,000 Hz was used. Auditory brain response was considered as absent if the peaks were not clearly identified in both recordings and lacked replication.

A detailed clinical case history was taken from all the participants of the study. Those who reported any symptoms related to vertigo/dizziness, headache, nausea, nystagmus and blurred vision were included in the study. The dizziness handicap inventory was administered to all participants of the study according to the procedure described by Jacobson and Newman.[16] The dizziness handicap inventory has 25 questions, and for each of them, the participants responded "Yes" (4 points), "Sometimes" (2 points) or "No" (0 points). The total DHI score, as well as the scores for all the three subscales (emotional, physical and functional) were analyzed. An independent *t*-test was performed to determine if there was any gender effect on the DHI scores. A correlation analysis was performed to determine the relationship between the onset of the hearing loss and the DHI scores in individuals with ANSD.

Ethical Considerations

In the present study, all the testing procedures were performed using non-invasive techniques, and adhering to the conditions of the ethical approval committee of the institute. All the test procedures were explained to the patients and their family members before testing, and informed consent was taken from all the patients or their family members for the participation in the study.

Table 1 Demographic details, audiological findings and vestibular symptoms of the 40 participants considered for the study

Participants	Age	Gender	PTA	SIS	Vestibular symptom
P1	14	Male	41.25	44%	Vertigo
P2	35	Female	70	32%	Vertigo
P3	15	Female	57.5	72%	Vertigo, headache
P4	16	Male	50	84%	Vertigo
P5	17	Male	43.75	80%	Vertigo, blurred vision
P6	18	Male	50	84%	Vertigo, nausea
P7	19	Female	38.75	68%	Vertigo
P8	27	Female	53.25	60%	Vertigo
P9	32	Male	32.5	68%	Vertigo
P10	19	Female	40	72%	Vertigo
P11	18	Female	47.5	12%	Vertigo
P12	16	Female	56.25	8%	Vertigo, blurred vision
P13	33	Male	55	8%	Vertigo, nystagmus
P14	22	Male	30	20%	Vertigo
P15	21	Female	50	16%	Vertigo
P16	19	Female	55	24%	Vertigo
P17	27	Male	25	68%	Vertigo, headache
P18	26	Male	41.25	32%	Vertigo
P19	26	Female	43.75	0%	Vertigo
P20	21	Female	42.5	0%	Vertigo
P21	19	Female	68.75	60%	Vertigo, headache
P22	18	Male	66.25	52%	Vertigo
P23	20	Male	50	12%	Vertigo
P24	21	Male	68.25	8%	Vertigo
P25	16	Male	50	8%	Vertigo
P26	18	Female	40	20%	Vertigo
P27	28	Female	28.75	16%	Vertigo
P28	19	Female	22.5	24%	Vertigo
P29	19	Male	42.5	68%	Vertigo, headache
P30	26	Male	52.5	32%	Vertigo
P31	29	Female	30	0%	Vertigo
P32	16	Female	23.75	0%	Vertigo, headache
P33	21	Female	33.75	60%	Vertigo
P34	16	Female	30	52%	Vertigo
P35	24	Male	83.75	28%	Vertigo
P36	27	Male	82.5	32%	Vertigo
P37	17	Female	45	0%	Vertigo, Headache
P38	18	Female	43.75	0%	Vertigo
P39	18	Male	40	48%	Vertigo
P40	21	Female	63.75	80%	Vertigo

Abbreviations: PTA pure tone average; SIS, speech identification scores.

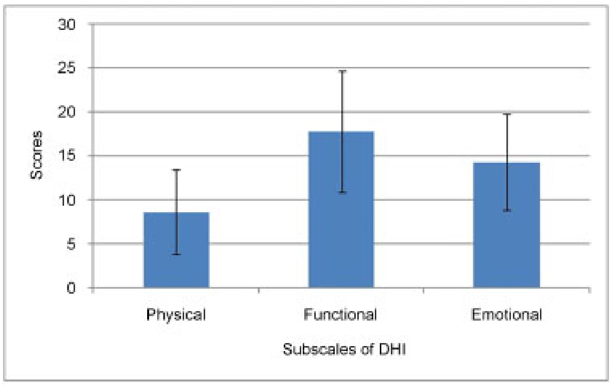

Fig. 1 Mean and standard deviation of the scores obtained for three sub-scales of the DHI.

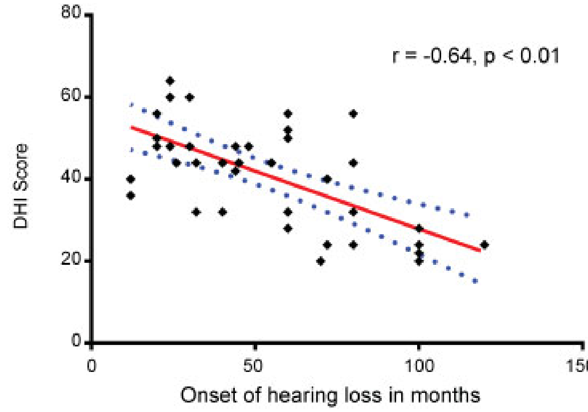

Fig. 2 Scatter plot representing the correlation between the DHI score and the reported onset of the hearing loss.

Results

The results of the study showed that the total score of the DHI ranged from 20 to 64, indicating a mild to moderate perception of the handicap. The mean DHI was 41.2 (SD =12.6), suggesting a moderate perception of the handicap in adolescents and adults with ANSD. The mean scores of the subscales were as follows: physical = 8.6 (SD = 4.8); functional = 17.8 (SD = 6.9); and emotional = 14.3 (SD = 5.5). The means and standard deviations of the scores on the subscales are shown in ►**Fig. 1**. This suggests that all subscales were affected, but relatively higher scores were obtained for the functional and emotional subscales.

An independent *t*-test was performed to determine if there was any gender effect on the DHI scores. The results showed that there was no significant difference ($t\ [38] = 0.76$, $p > 0.05$) in the DHI scores regarding gender. The result of the independent *t*-test to assess gender effect on the physical ($t\ [38] = 0.83$, $p > 0.05$), the functional ($t\ [38] = 0.9$, $p > 0.05$) and the emotional sub-scales ($t\ [38] = 1.1$, $p > 0.05$) also showed no significant difference. Pearson's correlation analysis showed that there was a moderate negative correlation ($r = -0.64$, $p < 0.01$), which was significant between the reported onset of the hearing loss and the DHI scores. The result of the correlation analysis is depicted in ►**Fig. 2**.

Discussion

Previous studies have reported substantial vestibular nerve damage in individuals with ANSD.[12,13,15] The abnormal results on the cervical VEMP suggests a sacculo-collic pathway dysfunction affecting the inferior vestibular nerve.[12] In addition, the ocular VEMP is also affected, which is suggestive of abnormal superior vestibular nerve functioning.[14] The neuropathy may not be restricted only to the auditory branch or the vestibular branch of the eighth cranial nerve. The most common variant is the "audio-vestibular neuropathy" involving both the auditory and vestibular branches.[15] However, the previous studies did not assess the real life dizziness-related handicap experienced by individuals with ANSD. The result of the present study shows that adolescents and adults with ANSD experience a moderate dizziness-related handicap. It also shows that dizziness affects their quality of life by limiting their functional abilities and causing emotional difficulties.

The results also showed that the dizziness handicap score did not vary across gender in individuals with ANSD. In addition, the dizziness handicap was greater in the cases in which the onset of the hearing loss was reported soon after the diagnosis of ANSD. Thereby, we can hypothesize that there could be some form of vestibular compensation that could have resulted in a reduction in symptoms in individuals who reported the hearing loss as happening later on. The previous studies have reported vestibular nerve involvement in individuals with ANSD.[12–15] Despite this, audiological rehabilitation has always been the primary focus in individuals with this disease. Thus, the present study recommends the assessment of the real life handicap experienced because of vestibular problems in individuals with ANSD. It is also warrants the appropriate management of vestibular problems to improve the quality of life in these individuals. The study also highlights the need for the assessment of the vestibular handicap in these individuals during the routine audiological evaluation.

Limitations of the Study and Future Directions

This study was performed using an English version of the DHI that is not standardized for the Indian population. There is a need for replicating the study using standardized dizziness handicap questionnaires in the local language of individuals with ANSD. The changes in dizziness handicap scores with vestibular rehabilitation should also be addressed. It is also essential to carry out qualitative studies using interviews or focus group discussions to better understand their dizziness-related handicap.

Conclusions

The present study attempted to evaluate the dizziness-related handicap in adolescents and adults with ANSD. The results of the study showed that a moderate degree of dizziness-related handicap was observed in adolescents and adults with this disease. The dizziness affected their quality of life, causing emotional problems. There was no gender effect, and the handicap was greater in cases in which the onset of the hearing loss was reported soon after the diagnosis of ANSD. Thus, a detailed assessment of vestibular problems and their impact on quality of life is essential in adolescents and adults with ANSD. Appropriate management strategies should be considered to resolve their vestibular problems and improve their quality of life.

Conflicts of Interest Statement
The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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