Original Article

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Prevalence of vestibular symptoms in individuals with auditory neuropathy spectrum disorder – A retrospective study

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Summary **The objective of the study was to retrospectively determine the prevalence of vestibular symptoms in individuals with auditory neuropathy spectrum disorder (ANSD). It was also attempted to determine the prevalence of vestibular symptoms and factors (gender and age of reported hearing loss) that could affect the prevalence in individuals with ANSD. The vestibular symptoms reported in the case history were analyzed in individuals diagnosed with ANSD. The symptoms reported by a total of 316 individuals (185 females and 131 males) with ANSD were analyzed. The result of the study showed that one in five individuals with ANSD reported at least one of the vestibular symptom. The vestibular symptoms were in more females and in individuals with earlier onset of hearing loss. The result of the study supports that there is a vestibular damage in individuals with ANSD. However, it is essential to carry out prospective studies validating these vestibular symptoms with objective vestibular tests before generalizing the results.**

Keywords: Vertigo, vestibular symptoms, gender, onset of hearing loss

1. Introduction

Auditory neuropathy spectrum disorder (ANSD) can be defined as a clinical disorder in which individuals have normal otoacoustic emissions (OAE), and auditory brainstem response (ABR) is abnormal or absent (*1-4*). In the Indian population, Kumar and Jayaram (*5*) reported that 1 in 183 were diagnosed as ANSD among individuals with sensorineural hearing loss. ANSD is a retrocochlear disorder affecting the vestibulo-cochlear nerve which can lead to auditory and vestibular symptoms. ANSD affects their communication abilities because of 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 (*6-8*).The individuals with ANSD are also reported to have vestibular neuropathy along with auditory difficulties (*6-8*).

The involvement of the vestibular branch in individuals with ANSD is extensively reported in the literature (*6-9*). Kumar *et al*.(*6*) suggested using the terminology "vestibuloacoustic neuropathy" in individuals who have both auditory and vestibular involvement. Sazgar *et al*. (*9*) reported that isolated auditory neuropathy or vestibular neuropathy is rare and the most common pathology is "audio-vestibular neuropathy" affecting both branches of the eighth cranial nerve. Sinha, Shankar and Raja (*8*) reported that cervical and ocular vestibular evoked myogenic potentials (VEMP) were abnormal in individuals with ANSD. Sinha *et al*. (*7*) showed that VEMP were absent and caloric tests showed hypofunctional responses in individuals with ANSD.This suggested involvement of both the inferior and superior vestibular nerve in individuals with ANSD. However, considering all these test findings, most of the studies have not given importance to the vestibular symptoms in individuals with ANSD. There is dearth of literature determining the prevalence of vestibular symptoms in individuals with ANSD.

The focus of assessment and rehabilitation of ANSD has always been on the auditory symptoms and the vestibular symptoms were usually ignored. Hence, it is essential to understand the prevalence and characteristics of vestibular symptoms in individuals with ANSD which would guide audiologists for appropriate assessment

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and management of these symptoms. Thus, the present study attempted to determine prevalence of vestibular symptoms in a large group of individuals with ANSD. The present study also attempted to determine the prevalence of vestibular symptoms and factors (gender and age of reported hearing loss) affecting the prevalence in individuals with ANSD retrospectively.

2. Materials and Methods

2.1. *Participants*

The individuals who were diagnosed as ANSD in the Department of Audiology between January 2001 and August 2016 were reviewed retrospectively. A total of 316 individuals diagnosed (185 females and 131 males) with ANSD was considered for the study. The selected participants were in the age range of 13 years to 42 years with mean age of 23.53 with a standard deviation of 10.29. The reported onset of hearing loss was noted and it ranged from 6 months to 180 months. The mean age of onset of hearing loss was 63.4 months (SD = 44.3). The participants were diagnosed as having ANSD in our clinic based on the criteria reported by Starr *et al*. (*10*). They had no history and presence of middle ear pathology with an A type tympanogram (*11*) and absent acoustic reflexes. The diagnosis of ANSD was confirmed by a neurologist.

2.2. *Procedure*

Pure tone air conduction (AC) and bone conduction (BC) thresholds were estimated using a modified Hughson and Westlake procedure (*12*). AC 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 was used to obtain air conduction and bone conduction pure tone thresholds and speech identification scores. Speech identification scores using headphones were obtained for phonemically balanced words in Kannada. Recorded word lists were routed from a personal computer through a 2 channel diagnostic audiometer at 40 dB SL (re: Speech Recognition Threshold). An immittance meter Grason Stadler Inc. Tympstar (GSI-TS) 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, 1,000, 2,000 and 4,000 Hz pure tones, presented to both ipsi-lateral and contra-lateral ears.

Otodynamics ILO v.6 OAE analyzer was used to obtain Transient Evoked Oto-acoustic Emissions (TEOAEs). Waveform reproducibility of more than 50% (*13*), and an overall signal to noise ratio of more than 3 dB SPL (*14*) at least at two frequency bands was required to be considered as presence of TEOAEs. A Biologic Navigator Pro (Bio-logic, Mundelein, IL) AEP system with Etymotic Research 3A insert earphones was used to record 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 Hz to 3,000 Hz was used. ABR was considered absent if peaks were not clearly identified in both the recordings and lacked replication.

A detailed case history was taken from all the participants of the study according to the protocol of the clinic. The case history taking included questions pertaining to presence or absence of vestibular symptoms. The details regarding the vestibular symptoms such as vertigo, imbalance, headache, nausea/vomiting and visual problems (Nystagmus/blurring of vision) were recorded from all participants of the study. Out of 316 participants with ANSD, 57 reported at least one of the reported symptoms. These symptoms were analyzed to determine the most common vestibular symptom reported by individuals with ANSD. The effect of gender and degree of onset of hearing loss on vestibular symptoms was also determined.

2.3. *Ethical considerations*

In the present study, all testing procedures were done using non-invasive technique adhering to conditions of the ethical approval committee of the institute and complied with the Declaration of Helsinki.

3. Results

The results of the study showed that 57 out of 316 (18.0%) individuals with ANSD reported vestibular symptoms. This shows that approximately 1 out of 5 individuals with ANSD report at least one vestibular symptom. The results of the study showed that 29 out of 57 (50.8%) reported vertigo, 9 out of 57 (15.8%) reported balance problems, 7 out of 57 (12.3%) reported headache, 4 out of 57 (7.0%) reported nausea/vomiting and 8 out of 57 (14.0%) reported problems in vision as shown in Figure 1.

Interestingly, the results of the study showed that

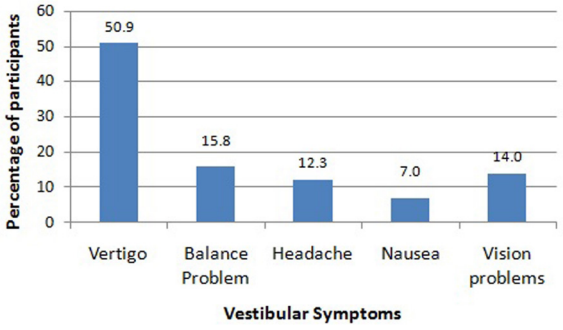

Figure 1. The percentage of participants with different vestibular symptoms.

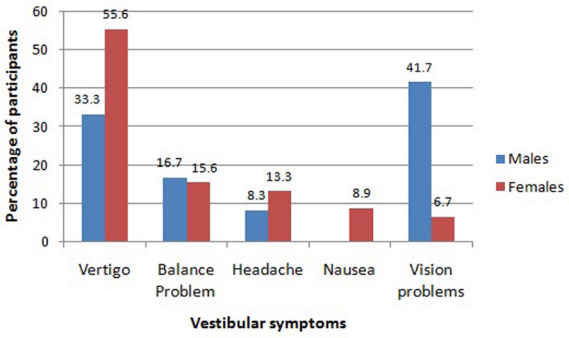

Figure 2. Percentage of participants with different vestibular symptoms across gender.

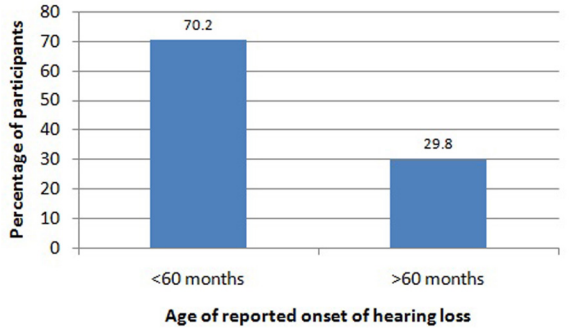

Figure 3. Percentage of participants with vestibular problems across age of reported onset of hearing loss.

out of 57 participants considered, 45 were females and 12 were males. Among females, vertigo was reported in 25 out of 45 (55.5%), balance problems in 7 out of 45 (15.5%), headache in 6 out of 45 (13.3%), nausea/vomiting in 4 out of 45 (8.8%) and problems in vision in 3 out of 45 (6.6%). Among males, vertigo was reported in 4 out of 12 (33.3%), balance problems in 2 out of 12 (16.6%), headache in 1 out of 12 (8.3%), problems in vision in 5 out of 12 (41.6%) and none of them reported nausea/vomiting. The differences in vestibular symptoms across gender are shown in Figure 2.

In addition, age of reported onset of hearing loss was less than 60 months in 40 out of 57 (70.2%) participants and reported onset of hearing loss greater than 60 months in 17 out of 57 (29.8%) participants as shown in Figure 3. This result suggests that the vestibular symptoms are lower in individuals with longer onset of hearing loss. A similar trend was observed in individuals from all age groups.

4. Discussion

The results of the study showed that vertigo was a common symptom and these vestibular symptoms are seen in 18% of individuals with ANSD. The previous studies on ANSD have reported substantial vestibular nerve damage in individuals with ANSD (*6,7,9*). The abnormal results on cervical vestibular evoked myogenic potential suggests a sacculo-collicpathway dys-function affecting the inferior vestibular nerve (*6*). In addition, ocular vestibular evoked myogenic potential are also affected and is suggestive of abnormal superior vestibular nerve functioning (*8*). 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 "audio-vestibular neuropathy" involving both the auditory and vestibular branch (*9*). Thus, the study highlights that vestibular symptoms are seen in many individuals with ANSD. The study warrants appropriate assessment and management of these vestibular symptoms in such individuals.

The results of the study also showed that the vestibular symptoms were in more females compared to males. ANSD is more common in females compared to males (*5*). In general, vestibular symptoms are reported to be more in females than males (*15,16*). Neuhauser *et al*. (*15*) studied the prevalence and incidence of vestibular problems in a large population group. They reported that females had more vestibular problems than males and the female gender was found to be a good predictor of vestibular symptoms (*15*). Yardley (*16*) also reported that the demand for vestibular rehabilitation was found more in the female population. The increased vestibular symptoms in females are usually associated with changes in levels of sex hormones (estrogen and progesterone) in females (*17*). ANSD is also reported to be more in females than males because of abnormal changes in hormones (*5*). In addition, the vestibular symptoms are also linked with increased risk of migraine in females (*18*). However, the exact reason for female preponderance is not well understood.

In addition, the results of the study showed that the symptoms were more common in individuals with earlier reported onset of hearing loss. Thus, we can hypothesize that there could be some form of vestibular compensation which could have resulted in a reduction of symptoms in individuals with a longer onset of hearing loss. The audiological assessment and rehabilitation has always been the primary focus in individuals with ANSD. The present study highlights that there could be substantial vestibular involvement, which requires attention from an audiologist in individuals with ANSD.

In conclusion, the result of the study showed that one in five individuals with ANSD reported at least one vestibular symptom. Vestibular symptoms reported in a few individuals with ANSD should be appropriately assessed and appropriate management should be suggested. It was also found that the vestibular symptoms were greater in females and in individuals with earlier onset of hearing loss. However, it is essential to carry out prospective studies validating these vestibular symptoms with objective vestibular tests before generalizing the results.

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