ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 06 | June - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PREVALENCE OF AUDITORY NEUROPATHY SPECTRUM DISORDER IN TERTIARY CARE HOSPITAL-A RETROSPECTIVE STUDY

Otorhinolaryngology

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ABSTRACT

Auditory neuropathy spectrum disorder (ANSD) is a condition in which the auditory nerve is damaged with preserved hair cell function. Individuals with ANSD exhibit difficulty to understand speech in both quiet and noisy environments irrespective of their audiogram results. A register based retrospective study done on patients who consulted at the Department of ENT, Mandya Institute of Medical Sciences and Teaching Hospital, Mandya, Karnataka, India for a specific duration of 3years.Data of basic audiological evaluations done was collected and descriptive statistics was carried out. Out of the 16,526 individuals analyzed, 11,177 individuals were diagnosed with permanent sensorineural hearing loss. Among them 77 individuals were diagnosed as auditory neuropathy spectrum disorder. Hence ANSD was seen in 0.68% (N=77) or 1 in 145 individuals from the total population of permanent sensorineural hearing loss included in the study. Hence the prevalence of ANSD in general population is a major concern in the early detection and management of the same.

KEYWORDS

Oto-acoustic emissions, cochlear microphonics, auditory brainstem response

1. Introduction

The term "Auditory neuropathy spectrum disorders" indicates the range of hearing disorders due to the dysfunction of auditory nerve or the presynaptic inner hair cells (IHCs), which contribute compromised signal processing (Siati et al,2020). Unlike in the individuals with sensory neural hearing loss, various clinical evidences are established to indicate that the cochlear outer hair cells are functioning normally in individual with ANSD (Manchaiah, Zhao, Danesh Duprey,2011). Moreover, the Individuals with Auditory neuropathy/auditory neuropathy spectrum disorder typically exhibit difficulty in understanding speech, hence poorer scores in speech recognition threshold (SRT) and speech identification scores (SIS) along with varying degrees of PTA (normal to profound) are clinically relevant in the diagnosis of ANSD (Norrix, & Velenovsky,2014).

It is a condition in which a patient's Otoacoustic emissions (OAE) and Cochlear microphonic (CM)responses are present but the auditory brainstem response (ABR) is absent or abnormal (Berlin et al, 2010).It is shown that ANSD affects individual's ability of processing the rapidly changing acoustic signals which is termed as auditory temporal processing. Advancement in accurate measurement of outer hair cell function has made it easier for diagnosing individuals with ANSD characterized by abnormal temporal coding and neural synchrony (Zeng, Oba, Grade, Sininger & Starr, 1999).

The diagnosis of ANSD is based on behavioral and electrophysiological tests like pure tone audiogram, speech identification scores, speech recognition in presence of noise, acoustic reflexes, oto acoustic emissions, auditory brainstem response. In individuals with ANSD, pure tone audiogram varies from normal hearing sensitivity to profound hearing loss(Berlin et al, 2010; Kraus et al, 2000). Speech identification is reported to be vary in individual with ANSD and recognition scores are impaired in presence of noise (Kumar&Jayaram,2006). The combination of normal OAE responses and severely impaired ABR responses reflect normal outer hair cell function with abnormal auditory nerve function in individuals with ANSD. In some individuals, oto acoustic emissions (OAE) are also absent or abnormal then ANSD is identified by the presence of Cochlear microphonic with abnormal or absent ABR (Kumar& Jayaram,2006; Berlin, Hood,Morlet, Rose& Brashear,2003). The site of lesion for ANSD is presumed to occur at the cochlear inner hair cells, synapses between inner hair cells and auditory nerve, dendrites or neural axons, afferent and efferent activity of the auditory nerve, spiral ganglion neurons, neurotransmitter biochemical anomalies (Vignesh, Jaya&Muraleedharan,2016; Starr, Sininger&Pratt,2000). Yet the exact site of lesion and pathophysiology of ANSD is not completely understood.

Similar to any other disorder, the prevalence of ANSD varies respective to the population in concern. For example, the prevalence of ANSD reported to be 40% in neonatal high-risk babies for hearing loss (Rea& Gibson,2003).In the age range of 6-12years, prevalence rate of ANSD was 2.46% in deaf school children (Lee, McPherson, Yuen,&Wong, 2001).The prevalence of ANSD in India, among different population has been determined by different authors. For instance, a study conducted among 217 childrendiagnosed with unilateral or bilateral sensorineural hearing loss Chennai, India, reported that 11 children are diagnosed with ANSD, hence the prevalence is determined to be 5.06% (Vignesh, Jaya &Muraleedharan,2016).Similarly,Mittal et al (2012) carried out a study on 183 children with sensorineural hearing loss in New Delhi. The prevalence of ANSD is found to be 5.3 % (26 cases). In contrary to the prevalence values of ANSD children in previous studies, another study done by Lepcha et al (2015) indicated that the prevalence of ANSD in children is found to be only 1.12%. A register-based study conducted by Kumar &Jayaram (2016) on patients diagnosed with sensory neural hearing loss in Mysuru, indicated that 1 in 183 individuals regardless of age groups, with sensory neural hearing loss are diagnosed with ANSD.While analyzing the results of these studies, it comes to the notice that there is a large difference in prevalence values of ANSD in children in different studies. Moreover, the studies conducted on the prevalence ANSD in adults and geriatric populations are limited. Hence it is important to estimate the prevalence of ANSD in general population (Including all the age groups) and understand the audiological characteristics of ANSD for formulation of proper diagnosis tools and in availing efficient management options of ANSD. Hence this study aims at estimating the prevalence and audiological characteristics of ANSD at Mandya Institute of Medical Sciences and Teaching Hospital, Mandya, Karnataka.

2. Method

A retrospective study was carried out to estimate the prevalence of

auditory neuropathy spectrum disorders (ANSD) in individuals with sensorineural hearing loss who consulted at the audiology section in the Department of Otolaryngology from June 2017 to June 2020. The study was initiated after the ethical clearance from Institutional ethical committee, Mandya institute of medical sciences (MIMS), Mandya.

2.1 Participants

The consultation details of 16526 individuals maintains as case registers at out-patient department (OPD) in MIMS, Mandya for the duration of June 2017 to June 2020 was reviewed. The OPD register was used to retrieve the information regarding the total number of cases, demographic details, medical history, otolaryngologic evaluation details, comprehensive audiological evaluation results, speech and language evaluation outcomes, neurological evaluation findings of the patients.

2.2 Participant inclusion criteria

The individuals who underwent complete otolaryngological and audiological evaluations during the period of June 2017 to June 2020 are included in the study. The individuals of all the age group were included in the study.The estimation of ANSD was carried out in patients diagnosed with either unilateral or bilateral sensorineural hearing loss, at the same time, individuals diagnosed with conductive hearing loss or mixed type hearing loss were excluded from the study. The records of infants suspecting from delayed auditory nerve maturation were meticulously analyzed and correlated with the available follow up records.

2.3 Procedure

The diagnostic data of individual with sensorineural hearing loss were retrieved and noted from the OPD register maintained in the audiology section of the department. The prevalence of the ANSD was determined based on the outcome of audiological evaluation using a standardized test battery which include behavioral and objective tests done in a sound treated room under standard conditions.

Behavioral audiological tests mainly include Puretone Audiometry, Behavior observation Audiometry (BOA), Visual Reinforcement Audiometry (VRA). The selection of the correct behavioral audiological test is based on the age of the participants. PTA is being carried out using calibrated Inventis-Harp dual channel audiometer and speech audiometry uses live voice stimulus presentation.

The objective evaluation includes Immittance evaluation (tympanometry and acoustic reflex testing), Transient evoked oto acoustic emissions (TEOAE), Auditory Brainstem Response (ABR). The tympanometry is being done using Inter-acoustics AT235 automated immittance meter with 226Hz probe tone, and the acoustic reflex is measured ipsilaterally for 500Hz,1000Hz,2000Hz &4000Hz. Likely TEOAEare being measured at 80dBPeSPL click stimulus with a reproducibility of >80% and SNR ratio of >6 at three consecutive frequencies. The integrity of the auditory neurons is measured through ABR using the Neuro audio evoked potential system.The absolute latency and interpeak latencies of I.III.IV peaks are noted. The presence of cochlear-microphonics were noted and confirmed by changing the polarity of the stimulus. The protocol for ABR testing is depicted in Table 1. The patients are further undergoing neurological evaluation by a Neurologist CT/MRI evaluation at Mandya.

Table 1: Protocol used for ABR

Stimulus parameters	Recording parameters
Click stimulus	30Hz to 3000Hz band pass filter
Starting from 90dBnHL	11.1/sec, 30.1/sec 30.1/sec repetition rates
100µs duration	Analysis window – 10ms
Alternative polarity	Montage: Cz-A1 and Cz-A2 (Ispi and contra)
2000 sweeps	50uV Artifact rejection

The Diagnosing criteria given by Starr, Sininger, Pratt (2000) was used to estimate the prevalence of ANSD in the participants. The criteria emphasize that the individual with either any of the degree of hearing loss or even absence of hearing loss in PTA along with absent or abnormal ABR with or without cochlear microphonics is a possible condition of ANSD. Along with presence of robust OAE with absent acoustic reflex clearly confirms the ANSD.

2.4 Statistical Analysis

Descriptive statistical analysis was carried out to estimate the prevalence of ANSD in the participants.

3. RESULTS

Out of the 16526 individual case files analyzed, 11,177 (Male: 5,925; Female: 5,229, gender ratio:1:1) individuals were included in the study since they were diagnosed with permeant sensorineural hearing loss. Out of 11,177 individuals, 77(Male:21, Female:56, gender ratio of 1:3.) of them were identified to have ANSD, which yields a prevalence of 1 in 145 (0.68%) individuals with sensory neural hearing loss (Figure 1). The age of the individuals with ANSD is ranging from 9 to 68years with a mean age of 28years. The age of onset of symptoms was in the range of 9 to 23years with a mean of 16years.

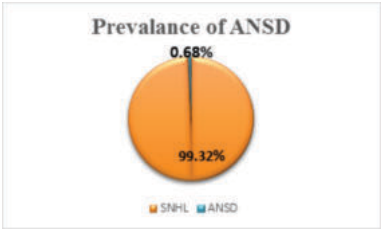

Figure 1: Prevalence of ANSD in individual with SNHL.

3.1 Audiological evaluation outcomes in individual with ANSD

The Audiological evaluation results showed that all the individual diagnosed with ANSD reported to have hearing loss unilaterally or bilaterally. Among them, 69 (90%) individuals have bilateral and symmetrical hearing loss. Only 8(10%) individuals have asymmetrical hearing loss. At the same time degree of hearing loss varied from mild to profound degrees. The details of audiogram pattern obtained from the individuals with ANSD is given below. The degree of hearing loss varied from mild to profound hearing loss. They were different configuration of audiogram pattern seen in them. Sharp peak was seen in 6, flat in 16, rising in 35, saucer shaped in 10 and sloping type in 10 individuals. Speech identification scores varied from no score to 60% score. Up to 60% of individuals did not have any quantifiable score on speech identification test. There was no correlation seen between the speech identification score and pure tone thresholds. Immittance evaluation showed normal tympanograms with absent stapedial reflex in both the ears.

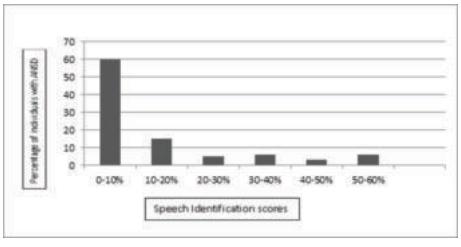

Figure 2: Speech Identification Scores Of Individuals With ANSD In Right Ear

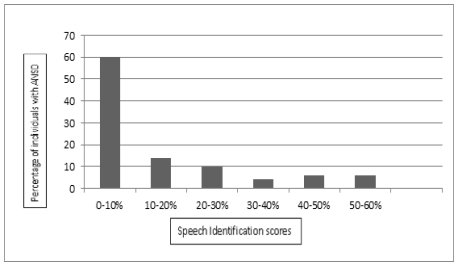

Figure 3: Speech Identification Scores Of Individuals With ANSD In Right And Left Ear

Transient evoked OAE responses were present in the 90% (70) individuals with ANSD in both the ears. The response reproducibility was >80% and SNR was >6dB in all the individuals bilaterally. The mean amplitude of TEOAE response was 13.24dBSPL in right ear

(6.30-18.90dBSPL) and 13.31dBSPL in left ear (6.60 – 18.80dBSPL). None of the individuals had ABR response for click stimulus for both the ears. Figure 2 represents ABR waveform in right and left ears.

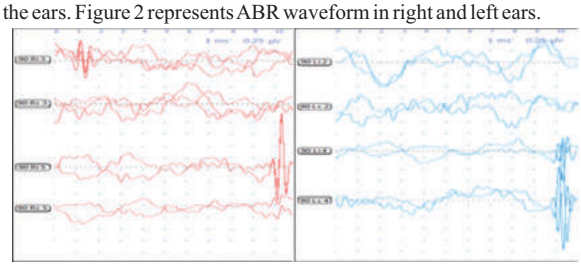

Figure 4: Representative waveform of ABR response in right and left ear

4. Discussion

The ANSD can be seen all population irrespective of the age of the individual. The current study estimates the prevalence of ANSD is0.68% (N=77) or 1 in 145; in individuals diagnosed with permanent sensory neural hearing lossduring the period from June 2017 to June 2020 at MIMS,Mandya. The low prevalence of ANSD in general public is discussed in the study (Kumar&Jayaram,2016). The authors have reported that the prevalence is only 0.54% or reported 1 in 183 persons irrespective of the age groups. Similarly, a prevalence of 1.1% was obtained in the study by Lepcha et al (2015) in a tertiary hospital, Chennai for the period of 17months.At the same time Another study by Vignesh, Jaya &Muraleedharan(2016) showed the prevalence rate of 5.06% in the age range of 6months to 12years in Chennai. Similarly, a higher prevalence rate of 14.2% among children below 12 years of age in Delhi (Mittal et al ,2012) Unlike the available literature, the present study shows a lower rate of 0.68% among individuals with sensorineural hearing loss at Mandya.

When comparing with western studiessuch as Rance et al (1999) &Maris, Venstermans & Boudewyns(2011) reports the prevalence 11% to 1.8% of prevalence rate, the current study reports much lower prevalence rate (.68%). The prevalence rate is lower in current study which might be due to the population being studied which differs in age range as that of other studies. But there is similar lower rate reported by (1 in 200) (Davis &Hirsh,1979).In the present study the prevalence rate might be better than reported as not every patient in 11,177 underwent complete audiological test battery like ABR, OAE and neurological evaluation. Starr et al (2000) reported of 16% of population in whom OAE's are absent with progress of the disease so there might be individuals in whom OAE is absent and not identified as ANSD. Individuals with asymptomatic ANSD might have been omitted when the desired symptoms were not reported.

The audiological characteristics of the individuals with ANSD showed varied degree of hearing loss from mild to profound hearing loss. It is correlating with the findings in study BY Starr et al (1996); Vignesh, Jaya&Muraleedharan (2016), where the major complaints reported in them is hearing loss. About 90% of individuals with ANSD were bilateral in the present study which correlates with the study given by Sininger&Oba (2001).The most common audiometric configuration seen was raising type in this study. The occurrence of such an audiogram may be due to anatomical makeup of the auditory nerve. The longest fibersare most susceptible to damage and the longest fibers are the low frequency fibers from the apex of the basilar membrane. Hence the low frequency is the ones which get affected than the higher frequency) Starr, Sininger&Pratt,2011) and thus the raising type audiogram is most prevalent in our study.

Speech identification scores varied from 0 to 60% in current study. This might be due to damage to the temporal processing that is reported as a key feature in individuals with ANSD. The temporal desynchrony of the auditory nerve might be responsible for the poor identification scores in majority of them. Few individuals had better speech identification scores and, in such cases, it depends on the amount of damage to the cochlear fibers and the amount of cochlear dead regions. Majority of them had raising type of audiogram which indicates low frequency fibers that are damaged and low frequency are the ones which are coded by phase locking of the type 1 auditory nerve fibers. Due to inability to use phase locking cue, the extent of temporal desynchrony is more which in turn reduces speech perception abilities (Zeng, Kong.Michaleski, & Starr A,2005).

The current study shows the presence of OAE wasin 90% of individuals. Similar findings were reported in the studies given by Deltenre et al (1999),Rance et al (1999)&Sininger, Oba (2001)Some individuals with ANSD lose their OAE over the period of time but their hair cell function will be evident from cochlear microphonics testing. Numerous studies have found that OAEs are sensitive to even minor cochlear insults and when we look into current data, absent OAE is seen majorly in >50years of age individuals with ANSD. Hence with the progress of the disease/ over a period individual with ANSD lose their OAEs (Starr, Sininger, &Pratt,2000); The All of them had absent acoustic reflexes which correlate with Berlin et al who reports of 89.19% of absent reflexes. ABR was absent in 100% of the individuals and it correlates with the findings given by Vignesh. Jaya&, Muraleedharan (2016), Sininger&Oba (2001) which reports of absent ABR in majority of the population. Audiological characteristics reported in the present study correlates with much previous study in the literature. From these findings we suggest a battery of test for assessment in individuals with ANSD. Also, irrespective of the prevalence rate, early identification is necessary and use of conventional/routine management techniques is not advisable as it will not benefit the individual and sometimes it might be dangerous too.

5. Conclusion

As the understanding of ANSD deepens, it has become evident that perceptual consequences of ANSD are quite different from those with permanent sensorineural hearing loss. The individuals with cochlear pathology the spectral processing is affected and temporal coding is intact whereas in ANSD the temporal processing is affected with normal spectral resolution. The amplification devices are not designed to alleviate the temporal processing problems instead alleviate the speech spectrum. Hence in an individual with ANSD, the degree of temporal distortion determines if he/she can make use of the amplified signal for understanding acoustic signal through a hearing aid. Therefore, it is not the number of individuals who is identified as ANSD but it is how early they get diagnosed as ANSD which will help to prevent them from ill effects of amplification devices in future. Hence early identification with a group of team members is most essential for further management and counseling.

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