

ORIGINAL ARTICLE

## Prevalence and characteristics of tinnitus in individuals with auditory neuropathy spectrum disorder

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### Abstract

**Objective:** Tinnitus is one of the reported complaints in individuals with Auditory Neuropathy Spectrum Disorder (ANSD). The study highlights the prevalence of tinnitus and its characteristics in individuals with ANSD. **Study design:** The study consisted of 95 patients with tinnitus out of 149 individuals with ANSD. The overall prevalence, laterality and gender effect was studied in these 95 patients. The study also reports on the subjective nature of tinnitus in 23 subjects and its relationship with the configuration of the audiogram. **Results:** The prevalence of tinnitus was found to be around 67%, mostly bilateral (89.5%) and seen more often in females (70.52%). The subjective pitch was low-pitched in individuals with low frequency hearing loss and the perceived pitch was high with a flat configuration of loss. **Conclusion:** Our results show prevalence of tinnitus is more than 50% in individuals with ANSD and the tinnitus matched with the individual's frequency of maximal hearing loss.

**Key words:** *auditory neuropathy spectrum disorder, tinnitus, prevalence, low pitch, configuration of audiogram, neural synchrony*

### Introduction

Auditory neuropathy spectrum disorder (ANSD) is a condition in which a patient's outer hair cells are normal (represented by a normal otoacoustic emission response) and an absent/abnormal auditory brainstem response (ABR) (1). In some instances, when otoacoustic emissions (OAEs) are abnormal, ANSD is identified on the basis of the presence of cochlear microphonics (CM) and abnormal or absent ABRs (2–5). ANSD is presumed to occur at the junction of the spiral ganglion cells and the auditory nerve (6,7). The diagnosis of ANSD is based on multiple behavioural and electrophysiological tests (such as ABR, OAE, acoustic reflexes, word recognition scores, speech perception in noise) (8). The patient's pure tone audiogram, if it can be acquired reliably, may range from essentially normal hearing sensitivity to a profound hearing loss (1). Speech recognition ability is generally poor, particularly in noise, although it may be good in quiet in some patients (9).

The overall incidence rate of ANSD as reported varies from 11% to 0.54% of the hearing-impaired

population (5,10,11). Picton (8) reported that 90–95% of all cases of ANSD are bilateral. ANSD may be present in some 10–15% of all children with hearing loss and ANSD may be present in higher percentages (15–20%) among children with severe to profound hearing loss (12). Prabhu, Avilala and Manjula (13) reported that 83.3% (25 out of 30) of the participants of the study reported tinnitus. Kumar and Jayaram (11) also reported 50% of the auditory dys-synchrony population had bilateral tinnitus. Thus, literature reviews suggest that most of the individuals with auditory dys-synchrony reported difficulty in understanding speech, and tinnitus was found to be one of the complaints (11,13).

Tinnitus is the conscious expression of a sound that originates in an involuntary manner in the head of its owner, or may appear to do so (14). The prevalence of tinnitus is significantly higher among hearing impaired persons than in the normal hearing population. In the general population, it is estimated at around 10–15% (15), while for patients attending

otology clinics it is estimated that 70–90% experience tinnitus either as the main or associated symptom (15). Seidman, Stranding, and Dornhoffer (16) reported that 10–15% of the adult population as a whole suffers from tinnitus; in the hearing impaired population the prevalence is 70–85% (17). Tinnitus can be a symptom of a variety of auditory disorders, but exact causal mechanisms are not well understood. In more than 95% of cases, the perceived tinnitus is purely subjective in nature.

The review of the literature was conducted from October 2012 to December 2012 in academic databases such as ERIC (Education Resources Information Centre), Medline, Ingentaconnect, Springerlink, Google Scholar, Scirus and PubMed using key words pertaining to auditory neuropathy, ANSD, auditory dys-synchrony, tinnitus and hearing loss. The review shows that there are a limited number of studies regarding the prevalence, nature and characteristics of tinnitus in individuals with ANSD. Kumar and Jayaram (11) found that 50% of the individuals with ANSD in their study had bilateral tinnitus and Prabhu, Avilala and Manjula (13) reported that 83.3% of those with ANSD had tinnitus. Thus, there are only few reports regarding the prevalence of tinnitus in ANSD and there are no studies that describe the characteristics of tinnitus in patients with ANSD. Hence, it is necessary to determine the prevalence and characteristics of tinnitus in individuals with ANSD and this was the aim of the present study.

## Material and methods

### *Participants*

The number of patients with tinnitus, its nature and characteristics in individuals with ANSD was determined using a register-based study design. The individuals diagnosed as having ANSD in the Department of Audiology, All India Institute of Speech and Hearing between September 2000 and June 2012 were reviewed retrospectively. One hundred and forty-nine individuals were diagnosed as having ANSD during that period; 95 were females and 54 were males. The mean age of the patients was 16.73 years (SD = 6.74) for males, and 14.78 (SD = 7.89) for females. The age range was 13–22 years for males and 10–21 years for females. The onset of the problem was variable among the patients in the study and most of the patients reported an approximate age of onset; the range of onset of the disorder was from 10 to 16 years.

The diagnosis of ANSD was based on the criteria recommended by Starr, Sininger, and Pratt (18). They are preserved cochlear amplification, reflected

by the presence of transient evoked otoacoustic emissions and/or cochlear microphonics; altered auditory nerve responses as indicated by absent or severely abnormal auditory brainstem responses; and normal otological and tympanometric findings with absent acoustic reflexes. A detailed neurological examination was carried out on all participants by a neurologist to rule out any space occupying lesion. Radiological investigations were performed using CT/MRI. The detailed audiological evaluation carried out included pure tone audiometry, speech audiometry, immittance evaluation, otoacoustic emissions and/or cochlear microphonics and auditory brainstem response testing.

### *Testing procedure and instruments*

It was ascertained from case records that all these subjects had been tested under standard conditions. All subjects were tested with calibrated (ISO 983) audiometers in sound treated rooms. Pure tone testing at octave frequencies 250 Hz–8 kHz had been performed using a Modified Hughson and Westlake procedure (19). Speech identification testing had been carried out with monitored live voice presentation of phonemically balanced monosyllables or phonemically balanced word lists in Kannada (20) at 40dB SL (re: Speech Recognition Threshold).

Immittance evaluation (tympanometry and acoustic reflex threshold testing) for a 226-Hz probe tone was carried out with a calibrated middle ear analyser (GSI 33 or Tymptstar V 2.0). Transient evoked otoacoustic emissions were measured from a calibrated OAE analyser (ILO 292 or ILO V6 DP Echoport) and auditory brainstem response testing was performed using a Biologic Navigator or IHS Smart EP (Version: 3140) evoked potential system. An identical protocol was used to test all the patients. The records confirmed that auditory brainstem testing was carried out twice to ensure reproducibility of waveforms. A group of 30 normal hearing adults had been tested to establish the nHL values.

### *Tinnitus evaluation*

Information about the pitch of tinnitus subjectively was obtained from 23 participants. Based on the subjective description of the tinnitus by the patients it was classified as low-pitched or high-pitched tinnitus. The tinnitus was considered to be low-pitched if the description of the tinnitus was humming, murmuring, rumbling or deep droning. It was considered as high-pitched if the description was hissing, ringing, whistling or buzzing. The relationship between the type of audiogram configuration

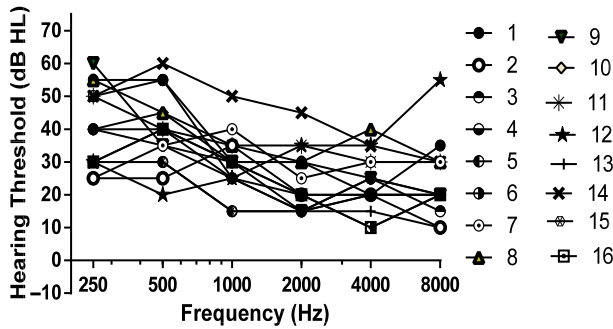


Figure 1. Audiograms of 16 patients with ANSD having low-pitch tinnitus.

and the type of tinnitus was considered in the study.

#### Ethics considerations

In the present study, all the testing procedures performed non-invasive techniques adhering to the conditions of the Ethics Approval Committee of the All India Institute of Speech and Hearing, Mysore. All the test procedures were explained to the patients and their family members before testing and informed consent was obtained from all the patients or family members for participation in the study.

#### Results

Of the 149 patients, 67 out of 95 females had tinnitus and 28 out of 54 males. Hearing loss of the individuals ranged from minimal hearing loss (pure tone average between 16dB and 25dB HL) to severe hearing loss (pure tone average between 71dB and 90dB HL). A detailed history was obtained from the 149 patients regarding the presence of tinnitus. Of 149 patients, 63.75% (95 patients) reported tinnitus. Of the 95 patients with tinnitus, 29.48% ( $n=28$ ) were males, 70.52% ( $n=67$ ) females. The study showed that the majority of patients with ANSD (89.5% ( $n=85$ )) had bilateral tinnitus.

The subjective frequency of tinnitus was also analysed in 23 patients as low-pitched or high-pitched. Out of these, 16 experienced low-pitched tinnitus and seven experienced high-pitched tinnitus. The relationship between configuration of audiogram and the type of tinnitus perceived was also determined. The audiogram of 16 patients with low-pitched tinnitus is shown in Figure 1. The configuration of audiograms shows that 14 had a rising type and two had a saucer shaped audiogram configuration. The audiogram of seven patients with ANSD with high-pitch tinnitus is shown in Figure 2. It shows that five had flat configurations with two having a rising audiogram shape.

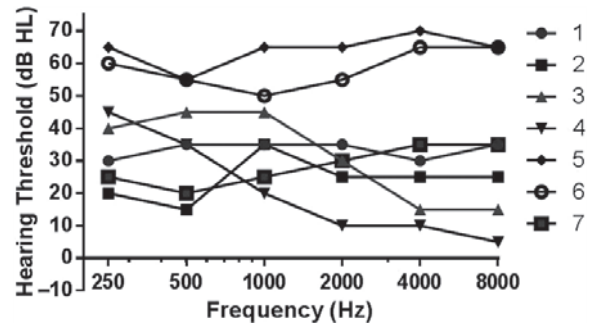


Figure 2. Audiograms of seven patients with ANSD having high-pitch tinnitus.

#### Discussion

The results show that more than 60% of patients report with symptoms of tinnitus and that this is one of the significant complaints in individuals with ANSD. Similar results were also obtained by Kumar and Jayaram (11), who reported tinnitus in 50% of the patients in their study, while Prabhu, Avilala and Manjula (13) found that 83% of patients with ANSD had tinnitus. The results show also that reports of tinnitus are more frequent in females compared to males. However, the male: female ratio in ANSD is reported to be 2:1 (11) and hence the larger incidence of ANSD in females could have resulted in the higher incidence of tinnitus in females.

The results of the study show that tinnitus perceived subjectively correlates with the degree of maximal loss, especially when the loss is at low frequencies in the majority of the patients with ANSD. In most of the patients, when the loss was flat, the perceived tinnitus was high-pitched. The common configuration seen in patients with ANSD is 'peaked' type with greater loss at low frequencies and a peak at around 2000 Hz (11). This could be because low frequency information is usually coded by phase locked responses in type 1 auditory nerve fibres (21) and individuals with ANSD cannot use phase locking cues to the same extent as normal hearing listeners due to dys-synchronous discharges of auditory nerve fibres (22). There are other models of tinnitus which also predict that changes in the processing of neuronal activity occur predominantly in the frequency range of reduced sensory input, which finally results in ongoing increased neuronal activity and/or synchrony in the respective central auditory pathways (23,24). Thus, according to these theories and explanations, the frequency of tinnitus perception should correspond to the frequency of hearing loss.

#### Conclusion

The study considered the prevalence and characteristics of tinnitus in individuals with ANSD.

The results suggest that the majority of the patients with ANSD report tinnitus as one of the complaints, and it is usually bilateral and seen most frequently in females. The subjective estimation of tinnitus showed that tinnitus pitch was low-pitched in most of the individuals with ANSD. The individuals reporting low-pitched tinnitus also had low-frequency hearing loss. Thus, tinnitus must be evaluated and treated in individuals with ANSD.

In future, studies relating to the severity of tinnitus in individuals with ANSD should be carried out in order to understand the extent of the impact of tinnitus on the life of persons with ANSD.

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