
Brief Report

The Prevalence of Auditory Neuropathy in Students with Hearing Impairment in Tehran, Iran

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Auditory neuropathy is defined as a sensorineural hearing loss characterized by normal cochlear hair cell function and absent or abnormal auditory brainstem evoked potentials. These people can hear the sound but can not understand it. They have neural hearing loss and the auditory rehabilitation approach for these people may be different from those who have sensorineural hearing loss. Therefore, screening of auditory neuropathy among hearing impaired students is essential. The prevalence of auditory neuropathy among the students with hearing impairment in specific schools for them was the objective of the current research.

From 2002 through 2003, 841 hearing impaired students, aged 2 – 20 years, underwent a complete history taking, clinical examination, and audiometry. We found 13 students with auditory neuropathy who comprised 1.55% (CI95%: 0.71 – 2.38%) of the students with hearing impairment.

We suggest that a complete panel of audiological tests for detection of auditory neuropathy be performed before admission of students with hearing impairment to schools.

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Introduction

Auditory neuropathy (AN) is a term currently used to describe a condition found in some patients ranging in age from infants to adults, who display auditory characteristics consistent with normal outer hair cell function and abnormal neural function at the level of cochlear nerve.¹ These patients often complain that they can hear sounds, but cannot understand speech.² AN was first described in late 1970s as paradoxical findings because of a discrepancy between the observations of lack of auditory brainstem evoked potentials (ABR) and presence of otoacoustic emission (OAE) and hearing thresholds.³

The prevalence is presently unknown with estimates varying from 0.5 – 15% of patients with sensorineural hearing loss (SNHL).⁴ AN has been shown to occur with diffuse neonatal insults such

as anoxia and hyperbilirubinemia, infectious diseases (e.g., mumps), immune disorders (Guillain-Barre syndrome), and genetic and syndromal neuropathies.⁵ In all of these patients, SNHL is a problem and speech discrimination is very poor.

Patients with AN require a different management approach to their auditory and communication problems from approaches used for patients with usual peripheral hearing losses.⁶

In these patients, the results of conventional management strategies such as amplification⁷ or cochlear implantations may be different from those children who have sensory hearing loss.⁸ For these reasons, screening for diagnosis of AN before admission of those with hearing impairment to schools for hearing impaired children is of paramount importance. We conducted this study to determine the prevalence of AN among children with hearing impairment.

Patients and Methods

Eight hundred and forty-one students from schools for hearing impaired children in Tehran, Iran participated in this cross-sectional study. The

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children were in preschool, primary school, middle school, and high school divisions with a mean age of 13.9 years. For all children, a questionnaire was completed with the help of their parents and teachers. The questionnaire consisted of information on family and medical history. Otoscopic examination and tympanometry to detect outer or middle ear disorders were carried out for all subjects. Those who were found to have possible outer or middle ear disorders were referred for further examination.

Transient evoked OAE (TEOAE) screening was conducted in a quiet room at school. The OAE system used was the Echo- Check. Measured TEOAEs were considered true responses if showed a reproducibility index of >50%.

Children who had recordable TEOAE responses in one or both ears were given a full diagnostic audiology assessment. The assessment included pure tone audiometry, speech audiometry, immittance audiometry and acoustic reflex, repeat TEOAEs, and ABRs. In our TEOAE diagnostic assessment, the reproducibility of >70% was considered as robust response, the reproducibility between 50% and 70% was considered as poor response, and the reproducibility <50% was considered as no response.

Results

In this study, TEOAE screening was conducted for students with hearing impairment and normal tympanogram. Positive results (recordable TEOAE) were found in 13 (1.55%; CI95%: 0.71 – 2.38%) students. In six students, TEOAEs were noted bilaterally. Six students were males and seven were females.

Figure 1 shows that 69.23% of the patients with AN had robust TEOAE response in at least one ear.

In 10 (77%) of 13 students with AN, parents had consanguinity. Eleven (84%) students had severe to profound hearing loss. Overall, 73% of the students had hearing loss with a flat audiometry. Our data indicate that 87% of the students with AN had family history of hearing disorder or genetic basis for developing AN and 62% had neonatal risk factors for AN. In only one student with AN, we could not find any associated medical conditions or family history of hearing disorders.

The associated conditions included hyperbilirubinemia (n = 1), meningitis (n = 1), anoxia

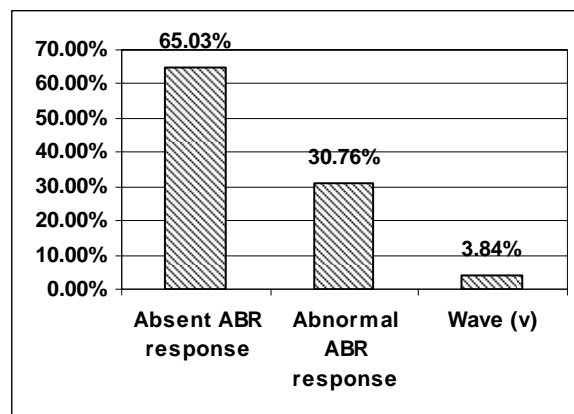


Figure 1. Distribution of transient evoked otoacoustic emission response in 13 patients (26 ears) with auditory neuropathy.

(n = 1), ototoxic drug exposure (n = 1), infection (n = 3), and family history of hearing loss (n = 11). Five patients had no associated medical conditions or other risk factors.

Eight students fitted with binaural or monaural hearing aid. Sixty-five percent of the patients had complete absence of ABR waveform regardless of the level of stimulus.

Figure 2 shows ABR response result from patient with auditory neuropathy.

Discussion

The prevalence of AN has been reported with a high variance. Rance and colleagues detected 109 infants with greater than mild SNHL after screening a population of 5,199 infants with risk factors for developing hearing loss. They found twelve infants with AN, which translates to a prevalence of 0.23% or 11% of children with SNHL.⁹ The prevalence reported in one research

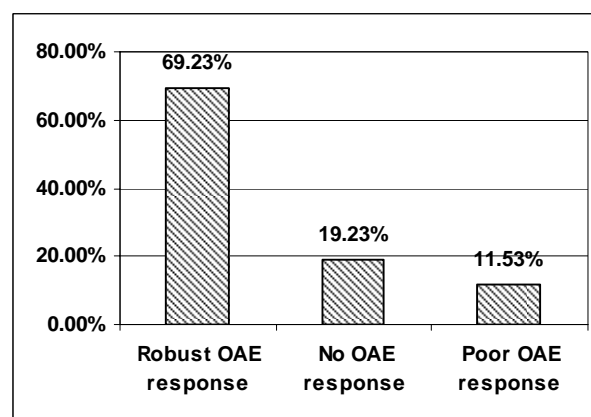


Figure 2. Distribution of ABR response in 13 patients (26 ears) with auditory neuropathy.

conducted in Hong Kong was 2.44%,¹⁰ and that from Germany was 0.94%.¹¹ We found the figure of 1.54%. In our study, seven (53%) students had unilateral AN while Kothe et al reported this condition as a rare occurrence.¹¹

Madden et al¹² investigated the clinical features of 22 students with AN in 2002 and found history of hyperbilirubinemia in 11 (50%) patients, prematurity in 10 (45%), exposure to ototoxic drugs in nine (41%), family history of hearing loss in eight (36%), neonatal ventilator dependence in eight (36%), and cerebral palsy in two (9%) patients.

The incidence of AN within a general population without any risk factors is not yet established. In our study, 13 (1.55%) children out of 841 students with SNHL with normal tympanometry had AN. Because of the special educational and rehabilitative strategies needs of such patients, it is important to identify them at an early age. So, there is an obvious need for screening programs for diagnosis of AN to be established in schools for children with hearing impairment. In addition, widespread use of screening programs will allow a more accurate evaluation of the prevalence of AN.

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