Prevalence of Auditory Neuropathy in a Population of Children with Severe to Profound Hearing Loss

Nader Saki,¹ Arash Bayat,² Soheila Nikakhlagh,*¹ Nima Rezazadeh,² Mehdi Nori¹

1. Department of Otolaryngology, Ahwaz Jundishapur University of Medical Sciences, Ahwaz, Iran
2. Department of Audiology, Tehran University of Medical Sciences, Tehran, Iran

Abstract

**Background:** The purpose of this investigation is to determine auditory neuropathy in the students with severe to profound hearing losses in Ahwaz.

**Materials and Methods:** In this cross-sectional study, 212 children of 7-11 year old with severe to profound hearing loss performed ordinary audiometric evaluations as well as ABR and OAE. The patients with normal DPOAE who had no record of acoustic reflex having normal ABR, were considered as the patients with auditory neuropathy.

**Results:** The neuropathic complication found in 14 children was appeared in 8 ones as one-sided (57.14%) and in 6 ones (42.86%) as two-sided. 68% of the patients as diagnosed had a very low Speech Discrimination Score (SDS).

**Conclusion:** we must be very vigilant in auditory neuropathy diagnosis for the purpose to be successful in appropriate treatment of severe to profound hearing losses.

Keywords: Otoacoustic Emissions (OAE), Auditory Neuropathy, Auditory Brainstem Response, Severe to Profound Hearing Loss

Introduction

Reduction in hearing sensitivity may be caused by disorders of different areas in peripheral and central auditory pathways. It may be also developed as a result of top hearing axon-neurons damages where it is called neuropathy. Auditory Neuropathy/Dys-synchrony (AD/AN) involves in a group of impairments which affect auditory nerve, cochlear inner hair cells and/or synapse between auditory nerve and inner hair cells [1, 2].

This impairment may be diagnosed by making use of physiological measurement of pre-neural hearing including cochlear microphonic (CM) and otoacoustic emission (OAE) as mostly recorded in the patient people and indicate normal functions of cochlear receptors along with the absence or abnormal results of auditory brainstem responses during recording the central auditory potentials in normal or near-normal environment [3, 4]. Among other clinical characteristics of auditory neuropathy, we may suggest to weaker speech perception than hearing thresholds in audiogram, permanent or fluctuating hearing loss with varying degrees, absence of acoustic reflex in the middle ear muscle and normal results in MRI imaging. OAE test is applied as a device to distinguish sensorineural hearing loss from auditory neuropathy by measuring indirectly the function of outer hair cells [5]. In cases where OAE is recorded and ABR is absent, auditory neuropathy could have been possibly occurred. CM as a pre-neural response resulted by cochlear hair cells activities is sensible at the beginning of ABR record in the individuals suffered from auditory neuropathy and it is continued for several milliseconds after that click stimuli is presented [1].

The auditory neuropathy is a rare impairment and the prevalence of this disorder has been reported between 5 to 13 persons in every one thousand people. The risk factors which lead to such disease include genetic, hyperbilirubinemia, asphyxia, low birth weight and suffering from a series of syndromes such as Guillain-Barré [3-5].

Few studies have been conducted on the prevalence level of auditory neuropathy in the children of school-age in Iran; thus the present investigation is carried out aiming to determine the auditory neuropathy prevalence in the students with severe to profound hearing losses in Ahwaz.

Materials and Methods

In this cross-sectional study, elementary school children of 7-11 year old suffered from severe to profound hearing loss were evaluated. All those children underwent otoscopy and tympanometry evaluations (MI 53 Device manufactured by MAICO) in order to make sure of their health in ear transmission system.

At the nest stage, the children were assessed by audiometry test of pure sounds (MA 53 manufactured by MAICO in USA) and the results of pure tone hearing thresholds in the frequencies between 8000-250 Hz and their speech tests was recorded. Then, the patients performed ABR (Octavos system manufactured by Madsen Co.) and DPOAE (Distortion product otoacoustic emissions) evaluations. In this investigation, the subjects with normal DPOAE results, absence of acoustic reflex...
record and abnormal ABR, were considered as the patients suffering from auditory neuropathy.

Normal DPOAE response was considered as a response with amplitude above the background noise level of 3dB in the frequencies under evaluation. ABR normal response was considered a response where absolute latencies of waves, latencies between wave peaks as well as the waves’ amplitude were in normal range. All evaluations were conducted in acoustic conditions and before starting any evaluation, we made sure of the machines to be calibrated.

Results

The patients under study include 114 boys and 98 girls with an average age of respectively 8.2±2.3 years and 8.6±1.1 years. The results of tympanometry for all the patients were normal, but no response was achieved in recording acoustic reflex in ipsilateral and contralateral approaches. In Table 1, frequency distribution of auditory neuropathy has been shown in details. In this investigation, 14 children (9 boys and 5 girls) have been diagnosed as suffering from auditory neuropathy. Among such patients, the complication was appeared as one-sided in 8 persons (57.14%) and as two-sided in 6 persons (42.86%). In case of one-sided complication, both ears got almost equally involved (4 cases of left ear vs. 5 cases of right ear). The prevalence of auditory neuropathy in this study was calculated as 6.6 percent.

About 68 percent of diagnosed patients had a very low SDS. The hearing loss pattern was recorded in 11 children (8 boys and 3 girls) as descending and in 3 children as flat.

Table 1. Frequency distribution of auditory neuropathy according to gender

<table>
<thead>
<tr>
<th>Gender</th>
<th>Rt ear</th>
<th>Lt ear</th>
<th>Both ears</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boys</td>
<td>3</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Girls</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>4</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>

Discussion

Considering the results achieved in this study and the low SDS, 68% of auditory neuropathy diagnosis cases has become very important. The auditory neuropathy is an impairment, during which the function of outer hair cells remains normal, but the responses related to eighth nerve fibers and brainstem is damaged.

In this investigation, prevalence of auditory neuropathy was measured as nearly 6.6 percent which is less than the values reported by Rance [5] as 10%, Kim [6] as 11% and Tallat [7] as 13.4%. Furthermore, the statistics indicated in our study is more than the prevalence level stated by Lee [8] as 2.5% and Lotfi [9] as 1.6%.

One of the factors of difference between such figures may be attributed to differences in the ages of the subjects evaluated. Reduced incidence of auditory prevalence may be found in older populations. As the patients’ age increases, the damage to outer hair cells will more likely increase and even may lead to full destruction of such cells; that is why we could not effectively record specified OAE or CM responses. Meanwhile, in a group of children whose auditory problems is diagnosed in younger ages, making use of powerful hearing aids (with high efficiency) will damage outer hair cells and consequently remove cochlear originated response. Though a particular hearing loss pattern was not suggested in the patients suffered from auditory neuropathy, such pattern was found descending in 78.5% of cases (11 children).

In this investigation, auditory neuropathy appeared in 8 patients (57.24%) as one-sided and in 6 patients (42.86%) as two-sided; this level is less than what was reported by Tallat (two-sided, 86.7%). To justify one-sidedness of this phenomenon, Cheng [10] argued that the risk factor causing such phenomenon will not involve all the outer hair cells of both ears, but it might simply involve one side and finally, this impairment will appear as one-sided neuropathy.

Duman [4] et al. evaluated 76 Turkish school-age children of 6-17 years old with severe sensorineural hearing loss by OAE and ABR tests and concluded that 3 children (4%) were suffering from auditory neuropathy. These researchers found such statistics as remarkable and finally suggest that it is necessity to implement auditory neuropathy screening program in all deaf schools.

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Authors’ Contributions

All authors had equal role in design, work, statistical analysis and manuscript writing.

Conflict of Interest

No conflict.

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Ahwaz Jundishapur University of Medical Sciences.

References