



Incidence of auditory neuropathy among the deaf school students

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Summary

Objective: We aimed to determine the incidence of auditory neuropathy (AN) among the deaf school students that have severe hearing loss.

Method: All students having severe hearing loss constituting a total of 75 at Deafness school in Afyon Province of Turkey were enrolled in the study. The etiological factors about the hearing loss were determined by performing an individual file survey and by interviewing the teachers and parents. First, all the children were subjected to an otolaryngologic examination. Then, auditory brainstem response (ABR) and transient evoked otoacoustic emissions (TEOAE) tests were done to all students enrolled in the study.

Results: The ages of the children were between 6 and 17 (mean age 11.9) and 32 (42.9%) of them were girls and 43 (57.1%) were boys. Three cases (4%) were diagnosed as AN in our study, however, no risk factors were determined in two of them. A history of hearing loss following a vaccination was found in only one patient.

Conclusion: Our data show that the incidence of AN in the children with severe hearing loss is not negligible. Therefore, our results suggest that automatic ABR should be also used with OAE as a routine application in the neonatal screening programmes, since the solely use of OAE in the neonatal screening programmes may result in the delay of the diagnosis of the children with AN.

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1. Introduction

Auditory neuropathy (AN) can be defined as a hearing impairment; that otoacoustic emissions (OAE) and/or cochlear microphonics (CM) are normal, despite being with no or abnormal responses at

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auditory brainstem response (ABR) at high stimulus levels [1–3]. As its name suggests, AN is described as a pathology in anywhere between the inner hairy cells of the cochlea and 8th cranial nerve, rather than a pathology in the outer hairy cells of the cochlea, as a periferic organ.

AN can be distinguished from the other hearing losses as follows: (1) the patient should have hearing loss at least in some surroundings, (2) the patient should have normal outer hairy cells function, which is detected by the normal otoacoustic emissions and normal cochlear microphonics or by both, (3) the patient should have impairment of 8th nerve function, that is detected by ABR, (4) the patient should have low speech discrimination ratio inappropriate with the obtained hearing loss by pure tone audiometry, (5) middle ear acoustic reflexes should be lost, and (6) cerebral and brainstem radiological imaging should be normal [1]. Although there are several reports on the incidence of AN in different groups in the literature, the incidence of AN among the neonates having a high risk for hearing loss was reported between 0.23 and 1.3% [4,5]. It was also reported that this ratio is between 1.8 and 14% among the children who have severe hearing loss [2,4,6].

Factors such as hyperbilirubinemia, prematurity, ototoxic drug use, artificial ventilation, cerebral palsy, perinatal asphyxia, head trauma, neonatal enfection as well as genetic factors were hold accountable for the etiology of AN [7–9]. However, the number of cases in which the etiological factors are not known exactly cannot be disregarded [10].

In this study, we aimed to determine the incidence of AN among the deaf school students having severe hearing loss and to investigate the factors that affect the etiology of AN.

2. Material and method

All deaf school students ($n = 75$) having hearing loss were included in this study. They all had severe hearing loss. Their age were between 6 and 17 (mean age 11.9) and 32 (42.9%) of them were female and 43 (57.1%) of them were male. To determine the etiological factors on the hearing loss, individual file survey and interviews with the teachers and parents were performed. All the children underwent an otolaryngologic examination. The children who have pathologies in the external ear canal, tympanic membrane and the middle ear were excluded in the study group. Important risk factors for every child were investigated. In case of having no risk factors, and also having a relative with hearing loss as well as being born as a result of an intermarriage for a child, the etiology was defined as hereditary.

ABR and transient evoked otoacoustic emissions (TEOAE) tests were carried out to all students enrolled in the study. The tests were performed by Otodynamics Ltd. EZ Screen 2 (USA), ERA Diagnostic System Evostar 2/1 + Evoselect (Germany) devices and a computer system with Intel Celeron M operator.

No sedative drug was given to the children before the test. Tests were performed in a silent room while the child was sleeping and immobile. TEOAE was repeated at least twice. While ABR test was being performed, 100 and 90 dB stimuli was applied initially, followed by no lower levels for the children who do not respond at initial levels. The hearing thresholds were measured by applying stimuli at lower levels for the children who responded at initial levels.

The following criteria were used in the evaluation of the TEOAE results:

Pass: signal/noise ratio (SNR) ≥ 3 dB at three different frequencies between 1 and 4 kHz.

Borderline: signal/noise ratio ≥ 3 dB at two frequencies between 1 and 4 kHz.

Fail: the measures except these.

ABR test was evaluated by considering the characteristics; such as wave latencies, interpeak latencies, peak amplitude, wave morphology and the repeating of the waves with stability at repeating stimuli.

The children who respond to emissions, but respond insufficiently or with no response in the ABR were diagnosed as AN. In addition, magnetic resonance imaging of the ear was performed and the acoustic reflexes were controlled for the children diagnosed as AN.

3. Results

The otolaryngologic examination for all children revealed normal results before the measurements of ABR and TEAOE. Etiological factors that are thought to be involved in the hearing loss in the study group were listed in Table 1.

The etiological factors seen most frequently were as follows: febrile convulsion in 21 (28%), heredity in 14 (18.7%) and meningitis in 4 (5.3%) of the children.

TEOAE results in accordance with the etiology of the hearing loss were shown in Table 2.

TEOAE responses were obtained from both of the ears in two out of the 31 children whose etiology of the hearing loss are not known. While no response was obtained from one of these children by ABR, 90 dB response at right ear and 100 dB response at

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