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The frequency of auditory neuropathy detected by universal newborn hearing screening program

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Summary

Objective: Auditory neuropathy/auditory dyssynchrony (AN/AD) has become a well-accepted clinical entity. The combined use of oto-acoustic emissions (OAEs) and auditory brainstem response (ABR) testing in the universal newborn hearing screening (UNHS) has led to the easy recognition of this disorder. Although, we are now able to diagnose AN/AD reliably, little is known about its epidemiology, etiology, and especially the frequency of its occurrence. The primary goal of this study was to determine the frequency of AN/AD in the Western Anatolian region of Turkey. The secondary goal was to compare the detection rate of AN/AD before and after the implementation of the UNHS in the audiology department of Dokuz Eylul University Hospital.
Method: Between 2005 and 2007, among the 23,786 newborns who were screened by automated click evoked oto-acoustic emissions (a-CEOAE) and automated auditory brainstem responses (a-ABRs), 2236 were referred to our department. All necessary audiological tests were performed for all the referred newborns. Among them, babies with deficient or abnormal ABR in combination with normal OAEs were considered as having AN/AD. These babies were evaluated with additional diagnostic audiological tests. Furthermore, comparison of the incidence of children diagnosed with AN/AD before and after the implementation of UNHS in our audiology department was also performed.
Results: Among the referred newborns, 65 had abnormal or deficient ABR test results. Ten of these 65 newborn babies (mean diagnostic age: 5.7 months) with hearing impairment showed electrophysiological test results that were consistent with AN/AD. The frequency of AN/AD in these 65 children with hearing loss was 15.38%. Moreover, the frequency of AN/AD within UNHS was found to be 0.044%. Seven of the 10 babies with AN/AD had hyperbilirubinemia as a risk factor, which is a high rate to be emphasized. On the other hand, the retrospective investigation of children diagnosed

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with AN/AD in the same audiology department between 1999 and 2005 (i.e. before the implementation of UNHS) revealed only 7 children, with an average diagnostic age of 34 months.
Conclusion: After implementing the UNHS, the incidence of AN/AD in the audiology department increased from 1.16 to 4.13. Furthermore, the age of diagnosis of AN/AD decreased from 34 months to 5.7 months. This study shows that AN/AD, when screened, is a comparatively common disorder in the population of hearing-impaired infants. While newborn hearing screening provides early detection of babies with hearing loss, it also helps to differentiate AN/AD cases when the screening is performed with both a-ABR and automated oto-acoustic emission (a-OAE) tests. Thus, the routine combined use of a-ABR and a-OAE tests in UNHS programs, especially for the high-risk infants, can provide better detection of newborns with AN/AD. Furthermore, hyperbilirubinemia is merely an association and maybe etiologically linked.

1. Introduction

Auditory neuropathy/auditory dyssynchrony (AN/AD) is a hearing disorder showing normal oto-acoustic emissions (OAEs) in contrast to the lack of auditory brainstem response (ABR). While the hearing loss could be variable with respect to the severity and pattern of this disorder, the word recognition ability is usually poorer than expected from the pure tone thresholds, and the acoustic reflexes to both ipsilateral and contralateral stimulations are usually absent or elevated [1—3]. The primary defects in these children could be delayed speech acquisition and suspected verbal learning disability. These patients have been reported to show normal OAEs as well as preserved cochlear microphonics (CMs) [1], but no obvious central nervous system abnormalities could be observed to account for these findings on the brain magnetic resonance imaging (MRI) [4]. Berlin et al. [5] reported the absence of efferent suppression of evoked OAEs in contrast to the normal OAEs.

Studies by Berlin et al. [6] and Hood et al. [7] recommended the term auditory dyssynchrony (AD) as a more accurate definition of the disorder for some of their patients. AN/AD has been recently described in the field of audiology and has been the focus of many studies in recent years. However, the specific risk factors for AN/AD have not been fully revealed yet. While a significant amount of patients have no risk factors, a number of infants diagnosed with AN/AD have a history of major neonatal illnesses, including prematurity, low birth weight, anoxia, and hyperbilirubinemia [7—10]. Other associated disorders mentioned in the history of children with AN/AD include Friedreich's ataxia, hydrocephalus, ischemic—hypoxic neuropathy, spinocerebellar degeneration, Charcot—Marie—Tooth neuropathy syndrome, and other hereditary sensory motor neuropathies [11]. It has been found that the AN/AD may be associated with other peripheral neuropathies and can be the result of generalized neuropathology, but isolated forms of AN/AD have also been reported [4,12]. Genetics also plays an important role in the etiology of AN/AD [8].

Cone-Wesson and Rance [13] reported that 1 in every 200 hearing-impaired children (0.5%) could have audiological findings consistent with a contemporary diagnosis of AN/AD. Berlin et al. [5] estimated that AN/AD is present in at least 4% of the children having permanent hearing loss. Rance et al. [14] found that the likely prevalence of AN/AD in a group of 5199 infants screened with neonatal and hereditary risk factors for hearing loss was 0.23%. Among the 109 children with permanent, moderate, or severe hearing loss within the defined group, the AN/AD prevalence rate was 11%. It is suggested that the occurrence of AN/AD is likely to increase in future, as reductions in the mortality rates for low birth weight and premature infants may lead to an increase in the adverse neurological consequences in the surviving neonates [14]. Ngo et al. [15] detected 9 infants with AN/AD out of the 14,807 newborns screened. In their study, the percentage of AN/AD cases within the newborn hearing screening program was found to be 0.06%.

The early diagnosis of AN/AD has been possible with the implementation of universal newborn hearing screening (UNHS) programs, based not only on OAEs but also on ABR testing in many countries. The protocol of OAE and ABR is more reliable, when compared with using ABR or OAE alone [16]. The UNHS program, on a national scale, officially began on 15 January 2005 in Turkey. Four university hospitals, including Dokuz Eylul University, School of Medicine, were constituted as referral centers for this screening program. While children who were born in our hospital were screened, those from other hospitals located in the Western Anatolian region of Turkey who could not pass the screening tests were