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ORIGINAL ARTICLE

Management of children with auditory neuropathy spectrum disorder (ANSD)☆

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KEYWORDS
Auditory neuropathy spectrum disorders;
Sensorineural hearing loss;
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Abstract
Introduction: ANSD is a challenging problem.
Objective: To present our experience on management of the children with ANSD with respect to clinical data.
Methods: This retrospective study included all children younger than 16 years of age who applied to the department between 2005 and 2013 (with the exception of newborn hearing screening NHS referrals). The data were derived from pure tone, OAEs and ABR tests, and further medical risk factors of the subjects were evaluated.
Results: ANSD was recognized in 74 ears of 40 children (B/U: 34/6) among 1952 children with SNHL (2.04%) detected among 9520 applicants to the department (0.42%). The clinical tests revealed that hearing loss greater than 15 dB was present in both ears of 38 cases. The degree of hearing loss was profound in 48% children, severe in 12% children, moderate in 28% children, mild in 10% children and normal in 5% children. ABRs were absent/abnormal in 37/3 ears and CMs were detected in all. Acoustic reflexes were absent in all ears. Rehabilitation was managed by CI and hearing aids in 15 and 23 cases, respectively. FM system was given to two cases displaying normal hearing but poor speech discrimination in noisy environments.
Conclusion: ANSD is a relatively challenging problem for the audiology departments because of its various clinical features and difficulties in management. Our patients with ANSD most commonly displayed profound hearing loss. The number of overlooked cases may be minimized by performing ABR and OAE in every case referred with the suspicion of hearing loss.

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PALAVRAS-CHAVE
Espectro da neuropatia auditiva;
Perda auditiva neurossensorial;
Amplificação;
Reabilitação

Tratamento de crianças com espectro da neuropatia auditiva (ENA)

Resumo
Introdução: Espectro da neuropatia auditiva ainda é uma condição clínica desafiadora.
Objetivo: Apresentar nossa experiência no tratamento de crianças com espectro da neuropatia auditiva em relação aos dados clínicos.
Método: Este estudo retrospectivo incluiu crianças menores de 16 anos de idade que deram entrada no departamento entre 2005 e 2013 (com exceção de encaminhamentos para triagem auditiva neonatal). Foram avaliados os dados obtidos a partir dos exames de audiometria tonal, emissões otoacústicas (EOA), potencial evocado auditivo de tronco encefálico (ABR) e outros fatores de risco.
Resultados: Das 1.952 crianças com perda auditiva neurossensorial (2,04%) detectadas dentre os 9.520 candidatos que deram entrada no departamento (0,42%), espectro da neuropatia auditiva foi reconhecida em 74 orelhas de 40 crianças (B/U: 34/6). Os testes clínicos revelaram que uma perda auditiva superior a 15 dB estava presente em ambas as orelhas em 38 casos. O grau de perda auditiva das crianças era profundo em 48%, grave em 12%, moderado em 28%, leve em 10%, e normal em 5%. ABR estava ausente/anormal em 37/3 orelhas e microfonia coclear foi detectado em todas as crianças. Reflexos acústicos estavam ausentes em todas as orelhas. A reabilitação foi tratada com implante coclear e aparelhos auditivos em 15 e 23 casos, respectivamente. Um sistema FM foi utilizado em dois casos que apresentavam audição normal, mas discriminação deficiente da fala em ambientes ruidosos.
Conclusão: Espectro da neuropatia auditiva é um problema desafiador para os departamentos de audiologia, devido às suas várias características clínicas e dificuldades no tratamento. Em nossos pacientes a perda auditiva profunda foi a mais frequente. O número de casos negligenciados pode ser diminuído com a realização dos exames ABR e EOA em todos os casos encaminhados com suspeita de perda auditiva.

Introduction

The hearing loss known as auditory neuropathy spectrum disorder (ANSD) has been described by the presence of otoacoustic emissions despite absent or severely abnormal auditory brainstem responses (ABRs).[1–3] Foerst et al. reported its prevalence as 0.94% and 8.44% for infants at risk for hearing impairment and profoundly hearing impaired children, respectively.[4] Previously, Kraus et al. determined these rates as 1.3% and 14%.[5]

It has been reported that ANSD was related with various clinical and audiological patterns. Pure tone thresholds (PTAs) range from normal or near normal to severe hearing loss, particularly characterized by impaired auditory processing skills in noisy environments. These subjects present very low speech discrimination scores (SDS) which are not associated with the pure tone levels. Acoustic reflexes are absent in the majority of the cases.[1,2,6]

It has been reported that, particularly because of poor SDS in relation to better PTAs, management process of children with ANSD is more problematic than that of children with other hearing loss patterns.[7–10] Furthermore, since the site of the lesion in the subjects clinically collected into the ANSD group is still unknown and there has not been a test to discriminate the lesion site of the given cases,[2] selection of the management option becomes more difficult.[7–10] Major interests at this point have been focused on whether the cochlear implant (CI) is beneficial or not in the given cases. However, in some cases, it could even be difficult to decide between the options of a hearing device and ''waiting and actively observing''.[7–10] The accepted approach toward children with ANSD is to initially provide amplification using hearing aids; however, many ANSD patients demonstrate little functional hearing and speech understanding with conventional amplification. In subjects who demonstrate poor speech understanding and delayed language development with hearing aids, cochlear implantation (CI) may be offered.[2,11]

Our purpose in this study is to present our experience on the management of children with ANSD with respect to clinical data.

Methods

This study has been done in a retrospective manner and included all children younger than 16 years of age, who applied to the audiology department between 2005 and 2013 (Ethical Committee: 446). The subjects were first divided into two subgroups according to presence of sensorineural hearing loss (SNHL) (>15 dB),[11] and then the SNHL group was divided into those with ANSD and the others. Thus, prevalence of ANSD for the children applying to the audiology department and prevalence of ANSD in those with SNHL were calculated.