

Auditory neuropathy: three cases among a group with sensorineural hearing loss

Mohd Khairi M D, Normastura A R, Wan Zaharah A W

ABSTRACT

The prevalence of auditory neuropathy is not known, although the majority of cases are felt to lie within the population of neonatal intensive care unit graduates. We report three cases of auditory neuropathy, out of 211 children with sensorineural hearing loss, seen at our audiology clinic from April 1, 1999 to December 31, 2003. Two patients did not have a risk factor for hearing impairment. Screening policies based solely on transient evoked otoacoustic emissions testing will not detect auditory neuropathy effectively, and may falsely reassure parents and professionals unaware of this condition.

Keywords: auditory neuropathy, deafness, hearing loss, sensorineural hearing loss, transient evoked otoacoustic emissions

Singapore Med J 2009; 50(9): e324-e325

INTRODUCTION

The means of early identification and diagnosis of hearing impairment in the neonatal population have formed the subject of numerous studies and investigations. Selection of an adequate and specific screening protocol is difficult, and has been an issue of consideration in many centres. Transient evoked otoacoustic emissions (TEOAEs) fill most protocols as a noninvasive, time-saving, easy-to-apply and nearly 100% accurate method of screening the newborn.^(1,2) However, by using this instrument alone as a screening tool, we will miss the patient with a retrocochlear lesion. Auditory neuropathy (AN) is a hearing disorder characterised by the lack of auditory brainstem responses (ABRs) but with normal otoacoustic emissions. The prevalence of the disease is not known, but the majority are felt to lie within the population of neonatal intensive care unit (NICU) graduates.⁽³⁾ Therefore, ABR testing is used in combination with TEOAEs for screening at the NICUs in some centres.⁽⁴⁾ Nevertheless, it is also possible for AN to occur in the general population.

CASE REPORTS

We reviewed the medical records of the children who had been referred to our audiology clinic for hearing problems and speech delays, from April 1, 1999 to December 31, 2003. Out of 211 children with sensorineural hearing loss, three of them had AN. In all three patients, TEOAEs were present but their ABR results showed profound hearing loss in both ears. Therefore, the prevalence of AN in a group of patients with sensorineural hearing loss was 1.42%. Only one child had a high risk factor for hearing impairment. This child had severe neonatal jaundice secondary to G6PD deficiency. He underwent exchange transfusion twice. Apart from his speech, he also had gross developmental delay, and could only walk with support at two and a half years of age. The other two patients had no risk factors for hearing impairment. Both of them, a Malay and a Chinese, had been referred to us because of speech delay. They were born full term via normal delivery and had normal motor developments. Assessment by the speech pathologist showed that all three patients had good detection skills, but very poor discrimination skills.

DISCUSSION

The prevalence of AN is not well established. We report three cases of AN in a group of patients who presented to our clinic with sensorineural hearing loss, giving an AN prevalence of 1.42%. Rance et al and Madden et al reported a higher prevalence of 11% and 5.1%, respectively,^(5,6) while Berg et al found a much higher incidence of AN among the population at risk for hearing impairment at 24%.⁽⁷⁾ In two other studies on hearing-impaired children, the incidences were 2.5% and 4.0%, respectively.^(8,9) Two of our patients did not have risk factors for hearing impairment, while one had a history of severe neonatal jaundice. It is known that AN can occur in the general population although it is more common in the group of infants with high-risk factors for hearing impairment. Foerst et al found that out of 32 children with AN, five did not have a high-risk factor for hearing impairment.⁽¹⁰⁾ Hyperbilirubinaemia was the second commonest risk factor for AN (11 children) after prematurity with postpartal complications (15 children).

Department of
Otorhinolaryngology
– Head & Neck
Surgery,
School of Medical
Sciences,
Universiti Sains
Malaysia,
Health Campus,
Kubang Kerian,
Kota Bharu 16150,
Malaysia

Mohd Khairi MD,
MMed
Surgeon and Senior
Lecturer

Wan Zaharah AW, BSc
Speech Pathologist

School of Dental
Sciences

Normastura AR,
MCommMed
Senior Lecturer

Correspondence to:
Dr Mohd Khairi Md
Daud
Tel: (60) 9 766 4098
Fax: (60) 9 766 4093
Email: khairi@kck.
usm.my

All of these hearing-impaired children were not detected by the screening programme using TEOAE alone, and therefore an ongoing surveillance following screening at birth is critical to assist with the diagnosis of these cases.⁽¹⁰⁾

In the present study, it was noted that all of the children with AN had a profound ABR impairment. Starr et al and Raveh et al reported similar findings in the majority of their patients, though a milder form had also been noted.^(11,12) The disorder occurs bilaterally in most of the cases, even though unilateral cases have been described.⁽¹²⁾ All the patients in our case series had problems in both ears. As far as behavioural audiometry is concerned, it is interesting to note that the results vary and can range from normal to profound levels.^(5,11,12) The disruption of auditory nerve activity may not result in a significant loss of sensitivity, but usually causes difficulties in understanding speech. The patients in our case series conformed to this typical presentation of AN. They had good detection skills but very poor discrimination skills. In general, conventional amplification does not improve speech understanding when the auditory nerve is compromised. Therefore, the method of management for this group of patients remains unclear.

It has been shown that the hearing level has improved spontaneously in a small subset of patients with AN.^(6,12) In some children, they have been successfully managed with the use of hearing aids. There were a number of them who could obtain reasonable receptive language and speech production.⁽¹³⁾ Cochlear implantation has been shown to be beneficial in many other patients. Madden et al and Raveh et al reported a good outcome from implantation in four and 12 children in their case series, respectively.^(6,12) However, speech perception in children with AN have been found to be poorer than those of the implanted sensorineural group.⁽¹⁴⁾ Study of speech perception in the presence of background noise also revealed similar findings.^(15,16) In conclusion, AN is a rare disease but screening policies based solely on TEOAE testing will not detect AN effectively and may falsely reassure parents and professionals unaware of this condition.

REFERENCES

1. Psarommatis IM, Tsakanikos MD, Kontogianni AD, Ntouniadakis DE, Apostolopoulos NK. Profound hearing loss and presence of click-evoked otoacoustic emissions in the neonate: a report of two cases. *Int J Pediatr Otorhinolaryngol* 1997; 39:237-43.
2. Khairi MD, Din S, Shahid H, Normastura AR. Hearing screening of infants in Neonatal Unit, Hospital Universiti Sains Malaysia using transient evoked otoacoustic emissions. *J Laryngol Otol* 2005; 119:678-83.
3. Dunkley C, Farnsworth A, Mason S, Dodd M, Gibbin K. Screening and follow up assessment in three cases of auditory neuropathy. *Arch Dis Child* 2003; 88:25-6.
4. Cone-Wesson B. Auditory neuropathy: evaluation and habilitation of a hearing disability. *Infants Young Child* 2004; 17:69-81.
5. Rance G, Beer DE, Cone-Wesson B, et al. Clinical findings for a group of infants and young children with auditory neuropathy. *Ear Hear* 1999; 20:238-52.
6. Madden C, Rutter M, Hilbert L, Greinwald JH Jr, Choo DI. Clinical and audiological features in auditory neuropathy. *Arch Otolaryngol Head Neck Surg* 2002; 128:1026-30.
7. Berg AL, Spitzer JB, Towers HM, Bartosiewicz C, Diamond BE. Newborn hearing screening in the NICU: profile of failed auditory brainstem response / passed otoacoustic emission. *Pediatrics* 2005; 116:933-8.
8. Lee JS, McPherson B, Yuen KC, Wong LL. Screening for auditory neuropathy in a school for hearing impaired children. *Int J Pediatr Otorhinolaryngol* 2001; 61:39-46.
9. Duman K, Ayçiçek A, Sargin R, et al. Incidence of auditory neuropathy among the deaf school students. *Int J Pediatr Otorhinolaryngol* 2008; 72:1091-5.
10. Foerst A, Beutner D, Lang-Roth R, et al. Prevalence of auditory neuropathy/synaptopathy in a population of children with profound hearing loss. *Int J Pediatr Otorhinolaryngol* 2006; 70:1415-22.
11. Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. *Brain* 1996; 119 (Pt 3):741-53.
12. Raveh E, Buller N, Badrana O, Attias J. Auditory neuropathy: clinical characteristics and therapeutic approach. *Am J Otolaryngol* 2007; 28:302-8.
13. Rance G, Barker EJ, Sarant JZ, Ching TY. Receptive language and speech production in children with auditory neuropathy/dyssynchrony type hearing loss. *Ear Hear* 2007; 28:694-702.
14. Rance G, Barker EJ. Speech perception in children with auditory neuropathy/dyssynchrony managed with either hearing AIDS or cochlear implants. *Otol Neurotol* 2008; 29:179-82.
15. Zeng FG, Liu S. Speech perception in individuals with auditory neuropathy. *J Speech Lang Hear Res* 2006; 49:367-80.
16. Rance G, Barker E, Mok M, et al. Speech perception in noise for children with auditory neuropathy/dys-synchrony type hearing loss. *Ear Hear* 2007; 28:351-60.