

CASE REPORT

Screening and follow up assessment in three cases of auditory neuropathy

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Three children with auditory neuropathy are described. Two were detected via a targeted neonatal hearing screening programme based on auditory brain stem response testing, and one via the routine Health Visitor Distraction Test. Auditory neuropathy is an important but poorly understood disorder which has implications on planning future hearing screening policy and management of hearing impairment.

CASE 1

A 2.6 kg male infant was born via emergency section at 35 weeks gestation because of possible fetal distress. Antenatally there was rising maternal anti-D titres. Following birth he required three exchange transfusions because of significant jaundice, reaching a maximum serum bilirubin (SBR) of 520 μmol on day 3. He had no clinical features of kernicterus. He did not receive any aminoglycosides and there was no significant family history of hearing problems. He was noted by his parents to respond to sound during his first few days, but not subsequently. Routine neonatal screening was performed at 1 week of age. He had clear bilateral responses on otoacoustic emissions (OAE) screening but referred on auditory brain stem responses (ABR) bilaterally using a 50 dBnHL click stimulus. ABR thresholds were suggestive of profound hearing impairment; for example, no response to 1 kHz at 100 dBnHL bilaterally. During infancy there was evidence of mild gross motor delay but no athetosis. Magnetic resonance imaging performed at 11 months revealed normal auditory pathways but increased signal intensity in the basal ganglia. Hearing aids were tried during late infancy with no demonstrable benefit. He is currently 5 years of age, attends a specialist school for the deaf, and communicates effectively using sign language and lip reading. Paradoxically speech discrimination performance has on occasion been normal at a conversational level of voice. Recent pure tone audiometry showed a response on air conduction testing at 35 dBHL for 500 Hz and at 30 dBHL for 4 kHz on bone conduction testing. His attention state was, however, variable during testing and these responses could not be replicated.

CASE 2

An 800 g female infant was born at 24 weeks after spontaneous premature labour. She was ventilated for two weeks, requiring oxygen therapy until a corrected age of 36 weeks. During the neonatal period she received six courses of antibiotics which included gentamicin. Trough gentamicin levels were always within acceptable limits. Routine cranial ultrasounds were normal throughout. Her maximum SBR was 198 μmol and there was no family history of hearing problems. Routine neonatal hearing screening was performed at discharge. She referred both ears on ABR with a 50 dBnHL click stimulus (fig 1). ABR thresholds were suggestive of profound hearing impairment, for example no clear response to 1

kHz at 90 dBnHL bilaterally. Subsequent OAEs have shown normal responses. At 1 year of age she has no evidence of persistent respiratory, visual, or other neurodevelopmental problems and is responding well to visual based communication. Her parents feel that at times she appears to respond to the sound of her name and to music. Their observations are supported by recent behavioural audiological assessment using visual reinforcement audiometry. Responses were seen to 500 Hz, and 2 and 4 kHz sound field warbletone stimuli at a level of 55–70 dBHL. She presently does not use hearing aids although a trial of amplification is being contemplated.

CASE 3

A 4.1 kg female infant was born at term via induced vaginal delivery for maternal hypertension. There was no significant family history of hearing or neurological problems and her subsequent neonatal course was unremarkable. At 6 and 8 months of age she failed her routine Health Visitor Distraction Test. Prior to this there had been no parental concern. Further audiological assessment showed normal OAE responses; ABR thresholds showed no response to 1 kHz and 4 kHz tone pip stimuli at 100 dBnHL on the right. The left ear could not be tested at this time. At 13 months of age she shows no behavioural response to sound. She has mild gross motor delay with a normal neurological examination. British Sign Language is being used and cochlear implantation is under tentative consideration at present.

DISCUSSION

The first two cases described are both children whose hearing impairment has been identified through targeted screening using ABR. The third case had no risk factors for hearing impairment and was detected via routine child health surveillance.

Evoked OAEs on all children showed normal responses, suggesting a normal preneural pathway. The ABR was abnormal in all three cases from the point of view of an absent wave V component even at high levels of stimulation. On some recordings it was possible to identify early receptor activity on the waveform which, depending on the stimulus and recording technique, is the result of summing potential or cochlear microphonics originating from hair cells in the cochlea (fig 2). This pattern of abnormality with the OAE and ABR has been classified as anomalous. The abnormality is presumed to lie proximal to the cochlea and has been termed auditory neuropathy.

Since 1986 Nottingham hospitals have provided targeted hearing screening using ABR. Since 1989 transient evoked otoacoustic emissions (TEOAE) has also been performed

Abbreviations: ABR, auditory brain stem response; NICU, neonatal intensive care unit; OAE, otoacoustic emission; SBR, serum bilirubin; TEOAE, transient evoked otoacoustic emission

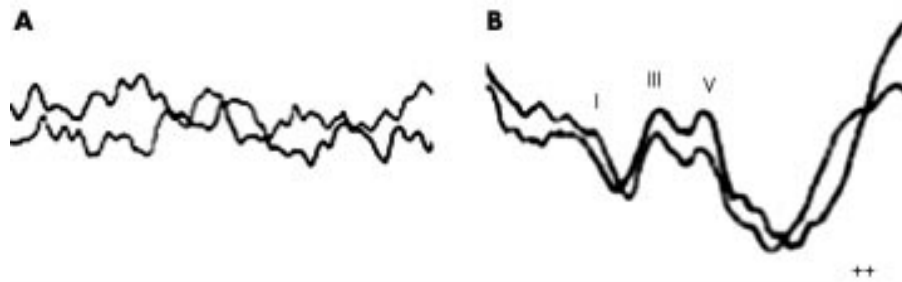


Figure 1 (A) Screening ABR from case 2 left ear, showing no response. (B) An ABR shown for comparison, showing the three typical positive peaks often identified in the neonate with normal hearing.

wherever practically possible. Between 1989 and 1993, 862 neonates who had risk factors for hearing impairment were screened using both tests. Seven neonates had responses on TEOAE screening yet referred on ABR.¹ None of these infants had significant hearing impairment on long term follow up. The three children described presented after publication of this previous study.

This specific pattern of abnormality suggests a dysfunction of the auditory pathway proximal to the outer hair cells of the cochlea. In children genetic causes, hyperbilirubinaemia, "neural immaturity" or other central nervous system pathologies have been suggested as possible causes.²

Estimates of the incidence of auditory neuropathy vary widely. The majority are felt to lie within the population of graduates of neonatal intensive care units (NICU). Rance and colleagues³ detected 109 infants with greater than mild sensorineural hearing loss after screening a population of 5199 at-risk infants. Twelve infants within that group had auditory neuropathy. This correlates to an estimated incidence within the at-risk population of 0.23%, or 11% of children with sensorineural hearing loss. The incidence of auditory neuropathy within a general population without risk factors is not yet established.

Although by definition results of ABR testing in these children are abnormal, results from behavioural hearing tests can be paradoxically normal. The prognosis is unpredictable, varying from complete resolution to permanent hearing impairment to progressive hearing loss. The optimum management of these children is not yet clear. The use of hearing aids has been generally disappointing in children with auditory

neuropathy.⁴ Cochlear implantation seems to improve communication skills in some children, perhaps in those whose defect lies within the cochlea.^{5,6} Phonetic sign language based on cued speech may have advantages as it retains the association between signing and sound, but is not in common usage.⁷

The Newborn Hearing Screening Programme is currently being introduced in 20 sites within the United Kingdom. Their recommendations include two different protocols.^{8,9} Babies admitted to the NICU for more than 48 hours should receive automated auditory brain stem responses (AABR) supplemented by automated OAEs. All other babies should receive automated OAEs as their initial screen. As well as detecting infants with significant hearing impairment at an early stage more effectively, this programme should also detect NICU infants with auditory neuropathy. Screening policies based solely on OAE testing will not detect auditory neuropathy effectively and may falsely reassure parents and professionals unaware of this condition. Children with late onset hearing impairment, progressive hearing impairment, or auditory neuropathy within the non-NICU population may not be detected by this programme, and therefore ongoing surveillance following screening at birth is critical to assist with diagnosis of these cases.

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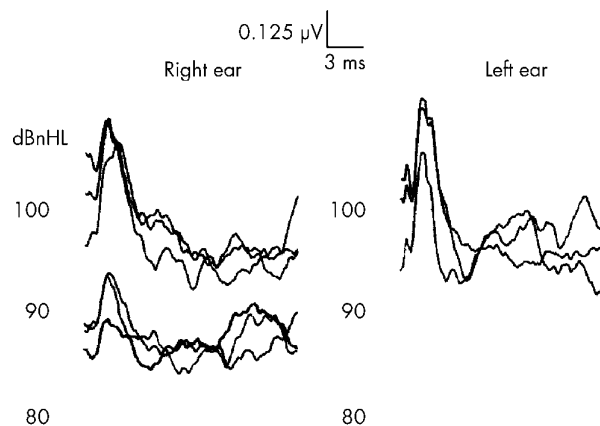


Figure 2 ABR waveforms recorded in case 2, showing typical early receptor responses but no wave V components. An alternately phased high intensity 4kHz tone pip stimulus was used to evoke these waveforms, resulting in enhancement of the summing potential and cancellation of the cochlear microphonics.