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CASE REPORT

Cochlear microphonics in sensorineural hearing loss: Lesson from newborn hearing screening

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Sensorineural hearing loss;
Enlarged vestibular aqueduct

Summary The diagnostic dilemma surrounding the presence of cochlear microphonics (CM) coupled with significantly elevated auditory brainstem response (ABR) thresholds in babies failing the newborn hearing screening is highlighted. A case report is presented where initial electo-diagnostic assessment could not help in differentiating between Auditory Neuropathy/Auditory Dys-synchrony (AN/AD) and sensorineural hearing loss (SNHL). In line with the protocol and guidelines provided by the national Newborn Hearing Screening Programme in the UK (NHSP) AN/AD was suspected in a baby due to the presence of CM at 85 dBnHL along with click evoked ABR thresholds of 95 dBnHL in one ear and 100 dBnHL in the other ear. Significantly elevated thresholds for 0.5 and 1 kHz tone pip ABR fulfilled the audiological diagnostic criteria for AN/AD. However, the possibility of a SNHL could not be ruled out as the 85 dBnHL stimuli presented through inserts for the CM would have been significantly enhanced in the ear canals of the young baby to exceed the threshold level of the ABR that was carried out using headphones. SNHL was eventually diagnosed through clinical and family history, physical examination and imaging that showed enlarged vestibular aqueducts. Presence of CM in the presence of very high click ABR thresholds only suggests a pattern of test results and in such cases measuring thresholds for 0.5 and 1 kHz tone pip ABR may not be adequate to differentiate between SNHL and other conditions associated with AN/AD. There is a need for reviewing the existing AN/AD protocol from NHSP in the UK and new research to establish parameters for CM to assist in the differential diagnosis. A holistic audiological and medical approach is essential to manage babies who fail the newborn hearing screening.

1. Introduction

The hearing of all babies is now screened in the UK through the national universal Newborn Hearing Screening Programme (NHSP). Despite some studies unable to show superior language outcome between those detected with hearing loss at a

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mean age of 6.6 months following newborn screening and those not screened and hearing loss detected at a mean age of 16.5 months [1], it is generally accepted that newborn hearing screening is an effective screening programme [2] with the primary goal of early detection and management of significant permanent hearing impairment. The early detection of hearing impairment has introduced issues that are distinctly separate from the hearing screening per se but intimately related to the ultimate success of the screening programme such as the challenges of fitting and managing hearing aids in young infants, support for such children and their families from education and social services, medical assessment and aetiological investigations and so on. These issues are evaluated and addressed in the UK through a NHSP quality assurance mechanism.

One of the issues that have significant impact on the audiological as well as medical diagnosis and management in the post-screen failure phase include a pattern of electro-diagnostic test results where otoacoustic emissions (OAE) and/or cochlear microphonics (CM) are present along with absent or abnormal morphology of auditory brainstem response (ABR) at high stimulus levels. The term Auditory Neuropathy or Auditory De-synchrony (AN/AD is used for such patterns of test results that occur in a small number of children with different conditions requiring different management and with different outcomes [3—6]. The NHSP in the UK has a protocol and policy for assessing and managing AN/AD which suggests that either AN/AD or Sensorineural hearing loss (SNHL) should be suspected when either OAE or CM is present along with absent/abnormal ABR response to air conduction click of 80 dBnHL or above [4,5]. In such a situation the distinction between SNHL and AN/AD is made by 1 kHz tone pip ABR, with a diagnosis of SNHL if response is better or present and a likely diagnosis of AN/AD if the response is still poor or absent [4,5]. It is crucial that a distinction between AN/AD and SNHL is made soon after the screen failure as the management and outcome in terms of audiological, early support and medical intervention will be significantly different.

We present the findings from a baby that suggests that the mere presence of CM in the presence of significantly elevated click and 1 kHz tone pip ABR thresholds do not always rule out SNHL. The diagnostic dilemma and subsequent findings will reiterate the importance of a holistic audiological and medical assessment and management of children who fail the newborn hearing screening, a review of existing AN/AD protocol from NHSP and further research.

Table 1 Diagnostic ABR thresholds following failing hearing screening

Mode and side of stimulus	Stimuli	Threshold (dBnHL)
Bone	Click	>55
Right air conduction	Click	95
	1 kHz tone	95
	0.5 kHz tone	95
Left air conduction	Click	100
	1 kHz tone	95
	0.5 kHz tone	95

2. Case report

2.1. History

Child AB was born at 38 weeks of gestation following an uncomplicated pregnancy and normal vaginal delivery. She was a healthy baby without any postnatal problems and was referred to our centre after failing her newborn hearing screening test. The electro-diagnostic assessments in the post screen failure period included air conduction ABR test using click and tone pips of 0.5 and 1 kHz and bone conduction ABR using click (Table 1) and measurement of CM from both ears. The CM traces from the left ear is shown in Fig. 1. OAE responses were absent but both the ears showed CM that reversed with changes in the stimulus polarity. ABR thresholds were way above 80 dBnHL for clicks and 0.5 and 1 kHz tone pips. In view of the results and the existing NHSP protocol/policy for AN/AD we adhere to in our centre the likelihood of AN/AD had to be considered in the initial clinical management and aetiological investigation [4,5].

AB, with the above electro-diagnostic test results, was seen along with her parents in the multidisciplinary tertiary paediatric audiology clinic involving Audiologist, Health Visitor, Audio-vestibular Physician and Teacher for the hearing impaired when she was about 6 weeks of age. She had no apparent behavioural response to sounds. Examination showed no dysmorphic features or any other abnormal physical findings. Both the tympanic membranes looked healthy and were mobile with Type A tympanograms.

2.2. Family history

Father has unilateral moderate SNHL. There were no other known medical conditions that ran through the family. The cause of the fathers hearing loss was not investigated in the past. Examination at the time of baby daughter's consultation it was found that he had synophrys and very blue irides.