Review of diagnostic hearing problems in childhood

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Abstract

The main focus for identification of permanent childhood hearing impairment is through implementation of newborn hearing screening programmes. Early identification and effective management of hearing loss aims to optimize auditory plasticity for auditory potential through life, by making sound audible from the first months of life through hearing aids. For infants with profound hearing loss or auditory neuropathy the option of cochlear implantation may give improved hearing for speech, typically at around 12 months of age. The onus of responsibility for using hearing aids and providing a suitable sound environment for the child falls on the parents, thus requiring a family-centred model of audiology management rather than the traditional expert- or medical-models of intervention.

For later-onset hearing loss, particularly conductive hearing loss secondary to otitis media or other middle-ear conditions, there is no longer a national screening programme, but instead a greater reliance on surveillance. Many school-entry screens of hearing have been disbanded and there is risk of children with later-onset hearing loss being overlooked despite the recognized secondary effects of untreated hearing loss on speech development, social interaction and subsequent educational achievement.

Keywords acquired; auditory neuropathy; childhood; conductive; early onset; hearing loss; newborn hearing screening; sensori-neural

Learning objectives

After reading this article you should be able to:

- review current knowledge and practice for diagnosis of hearing loss in infants and children
- raise awareness of current hearing screening, assessment protocols and practice with UK NHS services
- update readers on appropriate methods of behavioural assessment techniques and those that are no longer recommended practice
- high-light changes in the traditional roles of professionals and family within the hearing-management team

Early onset permanent childhood hearing impairment

Newborn hearing screening programmes have been introduced around the world to identify infants with bilateral permanent childhood hearing impairment (PCHI). The referral of infants from the hearing screen to audiology services for assessment of hearing ability has been demonstrated to be timely and effective in many countries (Figure 1). However, full definition of individual hearing status and effective hearing management has proved to be challenging for paediatric audiology services. Permanent sensori-neural hearing loss occurs in approximately 1:850 new births, with higher prevalence in pre-term and special care babies than in well-babies. The majority of well-babies with hearing loss are likely to have a genetic cause or congenital CMV.

When a neonate is referred from the hearing screen, electrophysiological testing using auditory brainstem response (ABR) or auditory steady state response (ASSR) testing is carried out to define the extent of the hearing loss in each ear, and whether it is a conductive (outer or middle-ear), sensory (cochlear) or neural hearing loss. Around 10% of children with PCHI are now recognized as having a component of auditory neuropathy or neural hearing loss, with increased complexity of management for these cases.

Auditory neuropathy spectrum disorder

Auditory neuropathy is the term given for hearing loss arising from impairment in the neural function of hearing. The hearing screening technique used for well-babies is the oto-acoustic emission (OAE) test which represents the functional pathway up to and including the outer hair cells in the cochlea, thus identifying babies with conductive or sensory impairment. About 10% of cases of early permanent hearing loss are due to neural hearing impairment. The majority of this population are premature or ill babies, though there are also some genetic conditions that fall into this category, for example otoferlin deafness. For this reason babies from special care are screened using automated auditory brainstem responses (AABR) as well as OAEs.

The diagnosis of auditory neuropathy is made in babies who have normal OAEs but abnormal or absent ABR results. The recording of a cochlear microphonic in the ABR, with the absence of later waves, adds to the diagnostic rigour in identifying cases of auditory neuropathy. Although there may be functional hearing, it is not represented by the ABR traces due to loss of synchrony in neural firing across the fibres of the auditory nerve. Thus intervention is reliant on careful observation of hearing responses in life with real sounds. One of the major challenges is that hearing responses in children with ANSD are very variable on a day-by-day or hour by hour basis. Professionals who are trained in close observation of hearing responses in young babies, teachers of the deaf and auditory verbal therapists, can coach these skills in parents. In recognition of the very wide range of auditory potential and outcomes for the individual children with ANSD, the umbrella term of auditory neuropathy

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Figure 1 Hearing screening for cases with permanent childhood hearing impairment is carried out in the first weeks of life.

spectrum disorder (ANSD) is used. Protocols for hearing aid management in ANSD were updated in October 2012 to recommend early fitting and use of amplification around carefully observing the child's hearing responses in life situations. The outcomes for children with ANSD are very variable. It may seem counter intuitive for a neural impairment, but many of these cases have much better speech understanding following cochlear implantation than they do with acoustic hearing aids. This is thought to be partly due to discrete bursts of electrical stimulation along the neural pathways improving the efficiency of neural transmission of auditory potentials. The same benefits for neural plasticity from timely intervention apply as for sensory hearing loss.

Protocols for hearing management following referral from NHSP

Early diagnosis and hearing intervention (EDHI) programmes aim to provide consistency and equity in services and in the quality of intervention. The Newborn Hearing Screening Programme (NHSP) website (http://hearing.screening.nhs.uk/) in the UK specifies defined protocols for both objective and behavioural test techniques following a number of misdiagnosed cases. There are new protocols and requirements for peer-review of ABR traces to provide an independent review of electrophysiological results. The literature on the hearing aid fittings of babies and infants with early fitting of hearing aids continues to show inadequate amplification in the majority of hearing aid fittings (Figure 2).

Figure 2 Hearing aids are fitted within the first few months of life to maximize auditory plasticity and foundations for speech for the life ahead.

It is important that in the rush to improve methods of hearing assessment and use of technology, a crucial component of intervention, parental grief is not overlooked. Parents are the sole agents for intervention for their baby's hearing loss from the first weeks and months of life. The early identification of PCHI for parents is frequently followed by a grief reaction. When parents are in a state of grief they are less effective in being able to undertake their role in hearing aid intervention, and to maintain the rich communication dynamics of early infancy.

Family-centred management

For the audiology team to be able to provide and use technology to improve a child's hearing potential for life, their primary role is to support the family in a time of grief, so that the parents are able to be active in their child's use of amplification and communication. This has given rise to the concept of "family-centred management" for hearing loss for all practical components of early hearing intervention. Concern about the potential negative impact of early diagnosis of PCHI was expressed prior to the implementation of NHSP in many countries but recent studies have shown that families who have earlier adjustment to loss have better outcomes for their children. The initial appointment that the family has in the audiology department, whether for ABR or behavioural assessment will set the scene for continuing care over the next 18 years. It is, therefore, important that the family feels valued and supported from this very first, critical point.

Early amplification, support and monitoring protocols

Hearing aid fitting is undertaken from between 6 weeks and 5 months of age, depending on individual family circumstances. Through the provision of effective amplification the paediatric