Screening for hearing loss in children

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Abstract

Assessment of hearing in children is important because early identification of hearing loss results in better developmental and educational outcomes. In the UK slightly more than 1 in 1000 children have significant permanent hearing loss diagnosed by the Neonatal Hearing Screening Programme (NHSP). This is based on Otoacoustic Emission (OAE) testing and Auditory Brainstem Response Testing (ABR).

OAE testing is performed in the first few weeks of life and identifies infants who warrant further testing with automated ABR. Automated ABR uses an encephalogram to monitor response to sounds. Infants who meet the 'high risk' criteria will be referred directly for automated ABR testing. If automated ABR suggests abnormality the child is referred for diagnostic ABR testing, which is a more detailed investigation capable of giving actual hearing thresholds and differentiating between conductive and sensorineural hearing loss.

Childhood hearing screening in the UK is also performed at school age, with the School Entry Hearing Screen (SEHS) at the age of 5. Outside of screening programmes there are a variety of hearing assessments available for children of all ages. This review article will outline the screening pathways, methods of assessment, and follow up of children from neonates to school age. The management of hearing loss in children is detailed in an accompanying article.

Keywords auditory brainstem response; deafness; hearing loss; Otoacoustic Emissions; screening

Introduction

Assessment of hearing in children has long been recognized as important, because early identification of hearing loss results in better outcomes in speech and language development, and in educational achievement.

Since the 1950s, screening of children for hearing loss has been routine in the UK, utilizing the health visitor distraction test. In March 2002, this was replaced by the more reliable Neonatal Hearing Screening Programme (NHSP), based on Otoacoustic Emission testing and Auditory Brainstem Response Testing. Childhood hearing screening is also performed at school age, with the School Entry Hearing Screen (SEHS) at the age of 5. Because many children with sensorineural hearing loss (SNHL) are now picked up by NHSP before the school age, the SEHS identifies fewer children with SNHL than previously; however, there is a comparably higher detection of conductive hearing loss at school entry, typically due to Otitis Media with Effusion (Glue Ear). Outside of screening programmes there are a variety of hearing assessments available for children of all ages. This article will highlight the current structure for provision of screening services in the UK.

Screening for hearing loss

Permanent congenital hearing impairment, with a hearing loss of $\geq$40 dB HL (as an average in the better ear), has a prevalence of approximately 1.1 per 1000 live births, with around 840 children in the UK affected each year.

Hearing is measured in decibels (dB). It is a logarithmic measurement as intensity of sound is exponentially related to loudness perception. There are various decibel scales but the one commonly used in clinical practice is decibels hearing level (dB HL). The reference 0 dB HL is an internationally agreed standard of hearing for each frequency of sound and represents the average standard of hearing of a group of normal young adults. So measurements of hearing are relative to this standard, with 0 dB HL indicating normal hearing, and levels worse than 20 dB HL (i.e. higher numbers) for each frequency considered abnormal.

In the past, the health visitor distraction testing was used for screening. This involved distracting the infant with toys and then periodically presenting a noise to either ear from behind. It was assumed that, if hearing was normal, the child would turn to the side of the sound. Historically, health visitor distraction testing was performed on all children aged 7–8 months old (when they could sit up and turn their head). There was a high degree of variability in pick-up rates and the test was potentially subjective and operator-dependent. In 1997 a Health Technology Assessment raised concerns over the high rate of missed diagnoses, and this resulted in the introduction of a new way of screening, termed the Neonatal Hearing Screening Programme (NHSP). It is estimated that of 840 UK children born with significant hearing loss each year, distraction testing would miss around 400 of these cases by 1.5 years of age.

NHSP is based on Otoacoustic Emissions testing and automated Auditory Brainstem Response (ABR) testing. The exact tests used in each neonate depend on the underlying risk of hearing loss. Screening is performed within a few weeks of birth, either prior to discharge from hospital, or in the community. The test can be administered by trained hearing screeners or by the health visitor, depending on local arrangements. For purposes of neonatal hearing screening, babies are divided into high risk and low risk groups and each has a specific testing pathway, as detailed below.

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Screening for hearing loss satisfies all of the Wilson's Criteria for a screening programme, as per World Health Organization recommendations (Box 1). The impact of not detecting hearing loss at a young age can be significant for both the child and the family, the pathophysiology is usually well understood and a variety of interventions are available at different stages of development.

NHSP has been shown to have a higher level of sensitivity and specificity than distraction testing, is cheaper to run and gives objective results. NHSP aims to screen neonates within the first 4 weeks of life if screened in hospital (but usually occurs within the first day) and within 5 weeks if screened in the community; the diagnosis is thus made much earlier than the 7 months of age with health visitor distraction testing.

Otoacoustic Emissions

In NHSP, initial screening is done with automated Otoacoustic Emissions Testing (OAE). This test is suitable for use with neonates as it is minimally invasive and (unlike distraction testing) does not require assessment of behavioural response.

OAEs are sounds produced by the outer hair cells of the cochlea, and can be either spontaneous or evoked. Spontaneous emissions produced by the cochlea can be detected in up to 50% of the population but only a small minority of people are aware of these sounds. The screening OAE aims to detect evoked otoacoustic emissions. To perform the test a small ear-piece is placed into the child's ear and a microphone emits a 'click'. This click is detected by the cochlea, and the cochlea in turn itself produces a sound (the otoacoustic emission), which is detected by the ear-piece. If the expected OAE is not detected, this implies dysfunction in the cochlea. The infant gets two attempts at the test on a pass/refer basis, although when explaining to parents it is important not to state that baby has 'failed the test' but that further testing is required. Approximately 15% of cases will be referred for further testing. Abnormal test results can often be due to other factors, such as a noisy testing environment, background noise, or amniotic fluid in the ear canal.

Automated Auditory Brainstem Response testing

If unsatisfactory responses are obtained after two attempts at OAE testing in a well baby that has no risk factors for hearing loss, the child is referred for secondary testing with automated Auditory Brainstem Response (ABR) testing. Infants falling into the 'at risk' category (see below) will all require automated ABR testing as well as OAE, regardless of OAE test result.

Automated ABR testing is usually done in a hospital environment, in a quiet room, by a trained audiologist. Ideally it should be performed within the first 12 weeks of life as the required sleep state becomes less predictable after this age. Headphones are placed over the infant's ears, or insert earphones are placed in the ears, along with three small sensors on the infant's head which are attached to a computer. A series of 'tone pips' are played through the headphones and the brainstem response (electro-encephalogram) is detected via the sensors. The system relies on there being a functioning system to transmit sound from the outside world, through the inner ear and along the acoustic nerve to the brain. Automated ABR is performed on a pass/refer basis at a level of 45 dB HL.

If automated ABR testing suggests any abnormality, then further assessment is required with diagnostic ABR testing; this is a more detailed investigation capable of giving actual hearing thresholds and differentiating between conductive and sensorineural hearing loss, not just a pass/refer answer that the automated ABR gives. Approximately 3% of children undergoing automated ABR will require referral for full audiological assessment to characterize the hearing loss.

Risk factors

Screening programmes are divided into two pathways, for "well" babies (Figure 1) and "at risk" babies (Figure 2). The criteria that define "at risk" babies are listed in Box 2; these infants are screened with OAE and automated ABR routinely and are followed up at 8 months regardless of previous test results.

Previously, the following risk factors were also included in the "at risk" group, but these factors have been removed following a systematic review:

- Family history of hearing loss
- More than 5 days intermittent positive pressure ventilation
- Jaundice at exchange blood transfusion level
- Neurological developmental disorders

Any children that missed their routine neonatal screening are also reviewed by audiologists at 8 months of age.

There are some notable exceptions from the screening system as described above. Infants with microtia or atresia of one or both external ear canals are not primarily screened with OAE and progress directly to full audiological assessment. This is due to the fact that there will always be a degree of hearing loss in these infants and so do not need initial screening testing. Infants with bacterial meningitis or meningococcal septicaemia should also

Wilson's Criteria for the Principles of Screening

Wilson's Criteria for the Principles of Screening (WHO)

1. The condition should be an important health problem.
2. There should be a treatment for the condition.
3. Facilities for diagnosis and treatment should be available.
4. There should be a latent stage of the disease.
5. There should be a test or examination for the condition.
6. The test should be acceptable to the population.
7. The natural history of the disease should be adequately understood.
8. There should be an agreed policy on whom to treat.
9. The total cost of finding a case should be economically balanced in relation to medical expenditure as a whole.
10. Case-finding should be a continuous process, not just a "once and for all" project.

Box 1