

Screening brief

Screening infants for congenital deafness

The disorder

- Bilateral permanent hearing impairment ≥ 40 dB in the better ear. Most cases are due to cochlear disorders.
- About half of cochlear disorders are due to genetic factors; at increased risk are infants who need admission to a Neonatal Intensive Care Unit (NICU) for >48 hours, and infants with cranio-facial abnormality.¹

Prevalence

- About 1.4 per 1000 live births in the UK by the age of five¹⁻³ of which 1.1^{1 2} may be congenital and detectable at birth.
- This corresponds to about 1000 new cases per year in the UK.^{1 2}

Natural history

- In the UK, few cases of permanent childhood hearing impairment are diagnosed before age six months (average 26 months).
- Early diagnosis and intervention appears to improve communication and language skills. Studies of deaf children indicate that language performance at age three is substantially higher for the children whose hearing impairment was identified before six months than in cases not diagnosed early.^{4 5} Long term follow up of these children is ongoing.
- There is little evidence on whether impaired language performance results from unilateral deafness or mild (<40 dB) bilateral deafness

Screening tests

- The Infant Distraction Test is currently used, scheduled usually at 6–9 months of age. One tester distracts an alert baby seated on a parent's lap whilst a "standard" sound stimulus is presented by the other tester, out of sight of the baby. The response is a clear head turn to the sound. Both sides are tested. The test has a poor detection rate, high false positive rate and low cost effectiveness.²
- Transient evoked otoacoustic emissions (TEOAE). A series of transient "clicks" is emitted by a soft probe inserted in the ear canal. An intact cochlea returns acoustic energy monitored by a microphone in the probe and the average response is calculated.
- If no clear TEOAEs are obtained or if the child has been in a NICU for >48 hours, automated auditory brain stem response (AABR) is also used. Clicks are presented to the ear, and the electrical activity in the auditory brainstem deriving from a healthy cochlea is monitored via three scalp electrodes and the average response calculated.
- A randomised trial of screening neonates has confirmed the efficacy of a stepwise approach testing first for TEOAEs and, in babies who fail this, for AABR.⁶

Screening performance

- Detection rate is at least 80%.⁷⁻⁹
- In screening neonates, the combination of TEOAE and subsequent AABR gives a relatively low false positive rate of about 0.6% with a positive predictive value of 17%; false positives were mostly cases of non-permanent conductive loss and were identified by retesting the babies when they were older using TEOAE, AABR, and other tests.¹⁰

Interventions

- Hearing aids. Direct electrical stimulation of the cochlear nerve via cochlear implants for more severe impairment.
- Help in developing signed (BSL) or spoken language as appropriate.

Costs of screening

- In the UK the cost of TEOAE based screening per 1000 live births (and the cost per case detected) is about £19,000.¹¹
- The Infant Distraction Test, currently used but less effective, costs more (about £24,500).

Overall assessment

- This approach to screening is effective in reducing long term language impairment due to deafness. The current pilot study in 20 areas of England is expected to identify worthwhile improvements to the screening programme.

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4 Yoshinaga-Itano C, Sedey AL, Coulter DK, et al. Language of early- and later-identified children with hearing loss. *Pediatrics* 1998;102:1161–71.

5 Mayne AM, Yoshinaga-Itano C, Sedey AL, et al. Expressive vocabulary development of infants and toddlers who are deaf or hard of hearing. *Volta Review* 1998;100:1–28.

6 Wessex Universal Neonatal Hearing Screening Trial Group. Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment. *Lancet* 1998;352:1957–64.

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