Executive summary

A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment

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Background
This review was commissioned because of the increasing doubt about the ability of existing screening programmes (mainly the health visitor distraction test (HVDT) at 7–8 months) to identify children with congenital hearing impairment, and technological advances which have made neonatal hearing screening an alternative option.

Objectives
- To review the available literature on the screening of permanent childhood hearing impairment.
- To provide commissioners and providers of health care with information about how to deliver a more uniform service, better outcomes, and more cost-effective screening.
- To identify areas for further research and service development.

How the research was conducted
The research involved a review of the available published and unpublished literature, and a comprehensive survey of current pre-school hearing screening provision in the UK coupled with a health economics study of hearing screening costs. The research also included a number of focus groups and visits to key centres in the UK and North America.

Research findings
Epidemiology of permanent childhood hearing impairment
There are approximately 840 children a year born in the UK with significant permanent hearing impairment† likely to affect their own and their family’s quality of life. Present services will miss about 400 of these children by 1½ years of age, and about 200 of these children by 3½ years of age. Such late identification of hearing impairment greatly reduces the responsiveness of the services for individual children.

Evidence for improved outcomes with earlier identification
Hearing-impaired children identified late are at risk of substantial delay in their acquisition of language and communication skills, with consequent longer-term risk to education achievement, mental health and quality of life. Theoretical arguments on neural development support the limited evidence for the increased benefit for child and family associated with very early identification. In general, parents and professionals want very early identification, which, if implemented properly, does not cause undue anxiety.

Current UK practice
The survey of current practice indicated a major problem with poor information systems. This problem was further highlighted as a major concern by the multi-disciplinary focus groups. Practice varies. There are two District-wide programmes in which all newborn babies are neonatally screened, a large number of ad hoc programmes for neonatal screening of ‘at-risk’ babies, a variety of early surveillance programmes, and widespread use of the HVDT.

Intervention and habilitation for the majority of those screened neonatally is routinely undertaken within 6 months of birth. For those screened only by the health visitor, identification was on average at about 26 months of age with intervention at about 32 months on average.

The effectiveness of existing screening programmes
The published evidence on screening performance indicates poor sensitivity and relatively poor specificity for the HVDT, with relatively low yield. Median age of identification via the HVDT varies from 12 to 20 months.

Neonatal screening shows high test sensitivity and reasonably high programme sensitivity, with high specificity. The limited number of universal neonatal screening programmes implemented at present give yields of the expected order.

† This is defined as being a hearing impairment on the better ear of ≥ 40 dB HL over the frequencies 0.5, 1, 2 and 4 kHz.
(1–1.3 per 1000), with a median identification age for those screened of about 2 months.

The costs of different programmes
The cost comparisons within the different implementations of hearing screening in the first year of life were encouragingly uniform. Universal neonatal screening appeared to have lower associated initial costs than the HVDT on a cost per child screened basis. Additionally, the cost per case found would be several orders of magnitude lower with universal neonatal screening.

Conclusions
Neonatal hearing screening in the UK has been successfully implemented for targeted screening (in over two-thirds of Districts) and universal screening (in two Districts).

Universal neonatal screening has a lower running cost and much lower cost per child detected than HVDT. Coverage can be greater than 90% and specificity about 95%. Sensitivity has not yet been assessed but may be greater than 90% as indicated by the yield from the universal screening trial.

Recommendations
Nine screening options in different categories (no screen, HVDT, at-risk neonatal screening and universal neonatal screening) were evaluated in terms of their running costs, incremental yield, efficiency, responsiveness and equity. A number of recommendations are made in three areas – service development, implementation and research. The major recommendations are as follows.

• The National Screening Committee should urgently consider whether there should be a national screening programme for congenital hearing impairment. We have shown that a programme based on universal neonatal screening, followed at 7 months by a targeted screen using an infant distraction test (mainly for those who have not had the neonatal screen), is the most equitable and responsive, and gives best value for money.
• An information system strategy should be developed to facilitate the coordination of the services needed for screening and following-up hearing-impaired children. Such a system would involve the development of a local shared list/register of hearing-impaired children, leading to the establishment of regional and national lists, and linked to local child health-record information systems.
• A model screening programme, with appropriate targets, is proposed around which the preferred option of universal neonatal screening might be based. Such a programme should have as its main aim the early identification of all children with a permanent hearing impairment of at least 40 dB HL (average in the mid-frequencies for the better ear). Responsibility for implementing and monitoring the programme should be explicit. Habilitation should be initiated early and be provided within a seamless service (within health services, and between health and education services) for parents and their children. Service links with education are likely to be crucial and need to be well coordinated.
• There are a number of research and development needs:
  – to find the best methods of habilitative management of children identified by neonatal screening
  – to identify optimum models for service coordination, including joint commissioning
  – to further refine screening techniques, both neonatal and infant
  – to estimate prevalence and identify risk factors for late-onset and progressive permanent childhood hearing impairment.

Publication
The overall aim of the NHS R&D Health Technology Assessment (HTA) programme is to ensure that high-quality research information on the costs, effectiveness and broader impact of health technologies is produced in the most efficient way for those who use, manage and work in the NHS. Research is undertaken in those areas where the evidence will lead to the greatest benefits to patients, either through improved patient outcomes or the most efficient use of NHS resources.

The Standing Group on Health Technology advises on national priorities for health technology assessment. Six advisory panels assist the Standing Group in identifying and prioritising projects. These priorities are then considered by the HTA Commissioning Board supported by the National Coordinating Centre for HTA (NCCHTA).

This report is one of a series covering acute care, diagnostics and imaging, methodology, pharmaceuticals, population screening, and primary and community care.

The views expressed in this publication are those of the authors and not necessarily those of the Standing Group, the Commissioning Board, the Panel members or the Department of Health.