

CURRENT TOPIC

Screening for congenital hearing impairment: time for a change

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The need for the early identification of children with permanent childhood hearing impairment (PCHI), most of which is congenital, has long been recognised in the UK. The early work of the School Medical Service, and of the Ewings in Manchester,¹ supported by key developments elsewhere in the UK, led in the 1960s to a de facto national screening programme. This was configured around a “distraction test” screen performed by health visitors at 7–9 months, and a back up screen at school entry. Although the introduction of the Infant Distraction Test (IDT) probably resulted in a significant reduction in the median age of identification for PCHI, by the early 1980s doubts were being expressed about the performance of the screen,² despite its endorsement by the government’s Advisory Committee on Services for Hearing Impaired People.³ These doubts led to notable and partially successful efforts by some to improve the IDT by better training, equipment and protocols.^{4–8}

However, doubts remained, particularly as the limited data available indicated that for a large proportion of children the age of identification of congenital hearing impairment continued to be very late. In some developed countries, such as Holland, effort was directed towards a more automated version of the UK IDT, backed up by better information systems. In others, where community services were less well developed—the USA, for example, interest centered on technological developments which might permit hearing screening of neonates.

In the 1980s two neonatal screening techniques were developed, one based on the Auditory Brainstem Response (ABR),⁸ the other on Transient Evoked Otoacoustic Emissions (TEOAE).⁹ In ABR screening clicks are presented to the baby’s ear and the resulting electrical activity generated by the eighth nerve and lower brainstem pathways are picked up by surface electrodes and averaged; in automated versions a machine based decision on the presence or absence of waveforms is made resulting in a pass/refer decision. TEOAEs are generated by an active physiological mechanism in the healthy cochlea, and can be elicited in response to clicks presented to the ear via a lightweight probe. This probe also houses a microphone which picks up the acoustic energy generated by the cochlea and transmitted back through the middle to the outer ear. Multiple clicks are pre-

presented, and the responses averaged to generate an ear specific but repeatable waveform. Such TEOAEs are not apparent in ears with middle or inner ear disease. Pass or refer decisions are usually made by the screener on the basis of a combination of displayed statistics.

In 1995 the NHS Research and Development Health Technology Assessment programme funded a critical review of the role of neonatal hearing screening in the UK. The review was necessary because of the continuing doubts about the ability of the IDT to deliver early identification of congenital hearing impairment, the technological advances which have made neonatal hearing screening an option, and the great variability in service provision that has resulted in major inequities. The final report of the critical review¹⁰ was presented to the National Co-ordinating Centre for Health Technology Assessment in 1997; copies are available from the MRC Institute of Hearing Research. This article summarises the review, its main recommendations, and the consequent service implications.

Structure of the review

The review involved two major strands of work. First, a critical review of the available published findings that related to screening for PCHI; and second, a comprehensive survey of current preschool hearing screening provision in the UK, coupled with a health economic study of hearing screening costs, and a number of focus groups held with professionals, parents, members of relevant organisations in the voluntary sector, and purchasers of health care. The study of screening costs was carried out by Stevens *et al.*¹¹ Finally, visits were made to several key sites to provide background or contextual information.

The evidence was reviewed and summarised in five areas: epidemiology of PCHI; evidence for improved outcomes with earlier identification; current UK preschool screening practice; the effectiveness of different screens and screening programmes; and the evidence on costs of different programmes. On the basis of this evidence, an option appraisal was carried out. Screening options in four different categories were evaluated in terms of their running cost, incremental yield, efficiency, responsiveness and equity. Finally, based on this option appraisal, recommendations were made for service development, implementation, and research.

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Summary of the evidence

About 840 children a year are born in the UK with clinically significant permanent hearing impairment likely to affect their and their family's quality of life substantially (1.12 per 1000 births). Current services do not identify about 400 of these children by 1½ years of age nor 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.

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

The survey of current practice indicated poor information systems and a wide variety of differing practice. There are currently two UK

programmes in which all neonates are 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 IDT. Intervention and habilitation for those screened neonatally is routinely well within six months, but for those screened only by the IDT, this begins, on average, at 18 months. The yield from the neonatal screening programmes is increasing, but the apparent yield from the IDT is low, below 30%.

The published evidence on screen performance indicates poor sensitivity and relatively poor specificity for the IDT, with relatively low yield. Median age of identification using the IDT varies from 12 to 20 months. Neonatal screening shows high screen sensitivity and reasonably high programme sensitivity, with high programme specificity. The limited number of universal neonatal screening programmes implemented at present give yields of the expected order, with median identification ages for those screened in the order of 2 months.

Table 1 Different possible options, their costs, benefits and challenges

Screening option	Marginal costs associated/1000	Incremental yield	Efficiency	Responsiveness
0 No first year screening (responsive service only)	None	Yield estimate uncertain - maybe < 0.2 per 1000 in first year (<20%)	Very poor: Cost per case indeterminate	Poor (with possible exception of profound PCHI)
H0 Universal IDT	£29.2k Total = £29.2k	Present average yield is 0.25 which might be increased to 0.4 with good quality control	Poor: Cost per case £90-£110k	Fair
H1 Hall report recommendations: Targeted neonatal screening: introduce targeted screening where not already implemented and make more systematic where very limited at present Universal IDT	£5.7k £29.2k Total = £34.9k	Yield estimates if both NICU and family history groups can get high coverage and better HVDT quality control 0.5 per 1000 by six months, 0.75 per 1000 by 1 year	Poor: Cost per case about £45k (incremental cost per case for IDT about £110k)	Fair
H2 As for H1, but following R&D implement technologically advanced IDT, with increased levels of stimuli	As for H1, with IDT reducing to about £23k Total = £28.1	Yield as per H1, possibly increasing to 0.8 per 1000 by 1 year	Fair: Cost per case about £35k (incremental cost per case for IDT about £90k)	Fair
H3 As for H1, but replace Universal IDT with 6-8 m universal HV surveillance by questionnaire	As for H1, with HV costs reducing to £23.4k? Total = £29.1k	Yield as per H1 or slightly less, particularly the moderates	Poor: Cost per case about £50k (incremental cost per case for IDT about £95k)	Fair
T1 Targeted neonatal screening: introduce targeted screening where not already implemented and make more systematic where limited at present	AS for H1, but no HVDT Total = £5.7k	Yield estimates, given high coverage of both NICU and family history children, 0.5 per 1000 by six months, but probably poor thereafter	Fair to good: Cost per case about £11.4k	Fair
T2 As for H1, but replace IDT with targeted infant distraction test	As for H1, but IDT possibly reducing to about £9k Total = £14.7k	Yield likely to be more than T1 but less than H1	Good: Cost per case about £19.3k	Good
U1 Introduce universal neonatal screening	£15.8k Total = £15.8k	Yield 0.9 per 1000 by six months	Very good: Cost per case about £17k	Good
U2 Introduce universal neonatal screening Modify infant distraction test to be targeted on those not tested and high risk of progressive PCHI	£15.8k £3.5k Total = £19.3k	Yield 0.9 per 1000 by six months and possibly 1.0 per 1000 by one year	Very good: Cost per case about £19.3k (incremental cost per case for IDT £34k, very approximate)	Very good

It is assumed (i) that there will be Health Visitor Surveillance for all children age 0-5 years (as per Health For All Children, cost unknown) and (ii) that the School Entry Screen will be retained for all options (at a cost of about £3.5-4.5k per live births). All costs standardised to per 1000 live births (not 1000 children tested). The 1000 cost per case identified is a broad estimate based on the programme cost and yield.

The cost comparisons between the different implementations of hearing screening in the first year of life were encouragingly uniform,¹¹ with systematic differences observed between implementations, such that universal neonatal screening seemed to have lower costs associated with it than the IDT on a cost per child screened basis. The cost per case found would be several times lower than that obtained with universal IDT options.

Option appraisal

Options in four different categories are shown in table 1 and were evaluated in terms of their running cost, incremental yield, efficiency, responsiveness and equity. The recommendations are based on this analysis.

The first option (O) was NOT to screen. This was rejected because of the number of children affected and the large impact congenital hearing impairment has on children and their families.

The second set of options (H) was configured around the universal IDT as the major screen, with some options, including targeted neonatal screening, and one option a potentially better (but yet to be developed and tested) hearing test

to be used by the health visitors. These options did not have a high estimated yield, and did have a very low efficiency due to the very high cost per child detected. Using the IDT, as presently implemented, over 50% of hearing impaired children would not have access to a hearing aid by age 2 years. Such children would have a high probability of being substantially handicapped by such late detection. These options are therefore low in responsiveness to these children's needs and low in terms of equity.

The third set of options (T) was configured around targeted neonatal screening. These options are low in terms of marginal cost and are highly efficient. However, even if combined with a targeted IDT, such options score low in terms of equity and responsiveness.

The fourth set of options (U) was configured around universal neonatal hearing screening. Such options are more efficient than H, have a lower marginal cost and greater equity. They permit a greater responsiveness, not necessarily because the age of identification will be very early, but because the children who would not have been identified by the IDT or other procedures in the first two years of life will have a better and more uniform chance of early identification and habilitation. Option U2 has, in addition to universal neonatal hearing screening, a targeted infant distraction test, that is primarily for those who did not have a neonatal screen.

Recommendations

The report makes several recommendations in three categories: service development (1–5); implementation (6); and research (7), where the weight of the evidence strongly supports:

- 1 The introduction of a national screening programme for permanent congenital hearing impairment.
- 2 A screening programme that is based on Option U2, universal neonatal screening, and a targeted screen using an infant distraction test at about 7 months (primarily for those who have not had the neonatal screen). This option is the most equitable and responsive, provides the best value for money, and potentially gives the greatest benefit for hearing impaired children and their families.
- 3 The development of an information system strategy, at the local level, that will facilitate the co-ordination of the services needed for hearing impaired children, in line with NHS priorities 1996/7. The local shared-list (or register) of hearing impaired children that would be the backbone of this strategy will be essential in auditing any option chosen and in maintaining a quality screening service. A subset of the locally available information should form the basis of a regional or national list, that would have a key role in monitoring any national screening programme. This strategy should link into the local Child Health Record Information Systems where possible.
- 4 A systematic appraisal of the role of health visitors in the identification of children with late onset or progressive PCHI. Due consideration and priority should be given to other

Table 1 Continued

Equity	Benefits	Challenges
Very low	Releases time/money to invest in responsive system, improvement of habilitation facilities for severe/profound	Moderate and severe PCHI not identified until >2 y, possible identification if language screen about 2 y
Low	Would consolidate present services and remove uncertainty for HVs	Need better training, facilities and quality standards for IDT ID age suboptimal
Medium	Little change to system, would help build up targeted screening	Need better training, facilities and quality standards for IDT, for targeted screening and for very early habitation in all districts Implementation of family history difficult Overall sensitivity of system poor ID age suboptimal
Medium	Limited change to system, and better test possible for IDT if accepted	As H1, but sensitivity possibly better New equipment needs to be developed, evaluated and bought
Medium Low sensitivity for moderate losses	Limited change to system, well trialed already. More in line with HV mission	As per H1
Low	Better quality control possible	As per H1, tuning the responsive system to find the remaining 0.6 per 1000, possibly needing to spend more on HVS
Medium Likely to miss ethnic minority and low SEGs	Better quality control possible, if less HVs involved, or specialist referral system	As per H1 plus definition of HV target population, crucial to combat inverse care law, needs research to define
Medium	Age ID very good Greater potential for habilitation and education to give benefit	Training, coordination and follow-up pose significant implementation challenges. What to do for those not tested and for late onset/progressive cases National support needed
High	Age of ID best that can be achieved for all PCHI groups Greater potential for habilitation and education to give benefit	Training, coordination and follow-up pose significant implementation challenges, including how to target progressive cases National support needed

- aims included in the health visitor's role, such as detecting the effects or signs of persistent otitis media with effusion (OME). Individual districts will have to appraise the priority they give to the detection of persistent OME and the use of surgical intervention that flow from detection in the first years of life.
- 5 Access to specialist paediatric otological opinion for children who are being assessed for PCHI. This will help reduce the chance of delays in initiating appropriate habilitation that have occurred in the past from ear, nose, and throat referral.
 - 6 The adoption of a model screening programme, including appropriate targets, around which the preferred option 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 (averaged in the mid frequencies on the better ear). It should be the responsibility of an individual (or committee) to implement (including negotiating appropriate transitional arrangements), monitor, and audit the programme. The target should be set such that habilitation for all congenitally hearing impaired children should be initiated by at least 12 months of age and be provided within a service context that is perceived as seamless by parents and their children. Service links with education are likely to be crucial and need to be well coordinated. The relevant groups in the voluntary sector, whose involvement is guaranteed by law, have a significant support and co-ordinating role.
 - 7 Consideration of four priority areas of research and development that flow directly from the review:
 - (i) management of children identified by neonatal screening both diagnostic and habilitative (including three randomised controlled trials that should be considered a high priority);
 - (ii) models for co-ordination of services, including the possibility of developing a joint commissioning model;
 - (iii) development of screening techniques both for the targeted infant distraction test and for early discharge neonatal screening and;
 - (iv) epidemiology, in particular the prevalence and risk factors of late onset and progressive cases, and the development of a national register of hearing impaired children alongside the shared lists that should be developed at a district level.

The recently released European Consensus Statement on Neonatal Hearing Screening (May 1998) embodies the major aims and recommendations of the artical review summarised here.

Practical considerations

The recommendations have considerable policy implications and may result in changes of practice. However, this will not happen overnight. The National Screening Committee considered the recommendations in 1998. The outcome of their deliberations was to convene a working group to consider paediatric and child

health screening and surveillance as a whole, and report back to the National Screening Committee. We have been encouraged to draft proposals that, among other things, will recommend:

- (i) the quality standards that would define adequate performance;
- (ii) how to specify an information system that will allow performance to be measured;
- (iii) what should be done if screening falls below the required level of quality; and
- (iv) a model of good practice for commissioners.

Detailed reading of the report will undoubtedly give rise to further comment and discussion, and we would be very pleased to receive such comment; we intend to monitor the dissemination and discussion and to report back on this during the course of the next year. We also realise that some people or professional groups might find some of the recommendations challenging in terms of their current views or practices. This is, of course, an inevitable consequence of policy recommendations in any area.

It has been argued that the IDT may have a role in the identification of children with persistent otitis media with effusion. However, the evidence does not justify a screening programme based on a one-off 30–35 dB screen for a mild fluctuating and transient hearing loss, for which diagnostic assessment cannot yet predict the cases that will persist, such as to justify intervention, and furthermore when the treatment options are contentious.

The recommendation that there be a national screening programme based on universal neonatal hearing screening, if adopted by the National Screening Committee, will take some time to evolve. The transition period between current services and those envisaged in the detailed recommendations of the report will need careful multiagency planning. During the transition the IDT will continue to play a very important part for some time to come, and will have an important role as a targeted infant distraction test thereafter. It is clearly crucial to ensure adequate quality of such arrangements.²

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