

CURRENT TOPIC

Neonatal screening for hearing impairment

Colin R Kennedy

Scope of this discussion

Hearing impairment in childhood may be conductive or sensorineural or a combination of the two (mixed) with additive effects. Impairments affect one or both ears, vary from mild to profound in degree, and may be congenital, acquired, transient, fluctuating, recurrent, progressive, or permanent. Screening positive for hearing impairment at any age leads firstly to the need for follow up tests, including an estimation of hearing threshold levels, and secondly, if these are elevated, to a medical evaluation to determine whether or not the hearing loss is permanent. Bilateral congenital permanent childhood hearing impairment (PCHI) of moderate or greater degree can be expected to lead to major deficits in the development of language as well as secondary effects on the child and the family. This is largely avoidable with early intervention,^{1,2} thus providing the rationale for neonatal screening. Consideration of this rationale and its practical implications is the focus of this article. This is timely as Yvette Cooper, UK government minister for Public Health, announced on 22 June 2000 that the government had decided in principle to introduce universal neonatal screening (UNS) and that districts are invited to apply to be part of a 20 site pilot starting perhaps at the end of 2000.

Epidemiology of childhood hearing impairments

PCHI is sensorineural except for a small number of cases of atresia. Bilateral PCHI ≥ 40 decibels relative to hearing threshold level (dB HTL) affects 133 (95% confidence interval 122 to 145) per 100 000 children, of whom about 55% have moderate losses and the remainder are divided equally between severe (70–94 dB HTL) and profound (≥ 95 dB HTL) losses.^{1–3} About 112 per 100 000 of these are congenital, which in this context means prenatal or perinatal in onset with the remainder being either postnatally acquired, most commonly after meningitis, progressive, or late onset, the last two categories accounting for about 10% of all PCHI.¹ PCHI is thus one of the most common congenital disorders. If mild and unilateral PCHIs are added (as in most reports of screening programmes in the United States), the total prevalence rises to 2–3 per 100 000.^{4–6}

Many risk factors for PCHI have been identified but they can be summarised as (a) a his-

tory of neonatal intensive care unit/special care baby unit admission for 48 hours or longer, (b) a family history of PCHI, and (c) craniofacial abnormality noticeable at birth. One or more of these is present in about 60% of all babies with PCHI. However, almost half of positive family histories of PCHI are only uncovered by the family after diagnosis of the index case.⁶ This reduces the percentage of all PCHI that could be identified with a screen targeted on babies at high risk to 45–50% of all PCHI.^{1,7} In practice, most neonatal screening programmes targeted on high risk infants in the United Kingdom succeed in testing only about one third of all high risk babies.¹ The prevalence of PCHI in the population of babies without risk factors is around 54 per 100 000.¹ Another disability in addition to PCHI is present in 30% of hearing impaired babies with no risk factor, 20% of those with a positive family history, and 60% of those with a neonatal intensive care unit/special care baby unit history.¹

Conductive hearing impairments are about two orders of magnitude more common than sensorineural impairments, being almost exclusively mild (< 40 dB HTL) or moderate (≥ 40 –69 dB HTL) and non-permanent. Middle ear effusion is the most common cause, affecting 36% of a large prospectively studied sample of normal children for more than 20% of days in the first three years of life.⁸

Current screening for hearing impairment in infancy in the United Kingdom

Screening for hearing impairment in infancy in most districts in the United Kingdom is currently undertaken using firstly the infant distraction test (IDT) administered by health visitors at 7 to 8 months of age, and secondly a targeted neonatal screen for babies at high risk. The IDT has been reported to have a low specificity, low sensitivity, and poor yield for PCHI in many districts¹ but is the predominant source of audiological and otolaryngological referrals for the much more common conductive impairments and middle ear effusion. This is confusing because the case for neonatal screening has been made in relation to PCHI but is also inextricably linked with the fate of the IDT which currently functions primarily as a conductive hearing impairment screen, the benefits of which are unclear.^{9,10} The criteria for targeting infants at high risk are often

Department of Child Health, Southampton General Hospital, Southampton, UK
C R Kennedy

Correspondence to:
Dr Kennedy, Mailpoint 21,
Child Health, Southampton
General Hospital,
Southampton SO51 0OJ,
UK
crk1@soton.ac.uk29/06/00

Accepted 17 July 2000

Table 1 Major criteria for assessing a screening programme⁴³

-
- Does the screening programme do more good than harm and at acceptable cost?
 - Is the impairment sufficiently common to justify screening all children?
 - Does the impairment cause significant disability or handicap?
 - Is there agreement about what is meant as a case?
 - Is there a screening test that accurately identifies children who may have an impairment?
 - Is there an agreed and available effective intervention with which to treat the impairment or reduce the disability after identification?
 - Is there an advantage in detecting and/or treating the impairment earlier, before it becomes clinically observable?
 - Is the cost of screening justified by the net benefit?
-

applied inconsistently or only to the subgroup who are admitted to a neonatal intensive care unit/special care baby unit and, as mentioned in the preceding paragraph, typically succeed in screening only one third of high risk infants or well under 20% of babies with congenital PCHI.¹

What to screen for and why

An agreed case definition is one of the prerequisites for a screening programme (table 1). In the United States, mild and unilateral permanent losses have been included in the case definition, but it is not yet clear how beneficial early identification of these losses is. It also requires the threshold for screening positive to be lower to detect, for example, all unilateral failures, and therefore also increases the number of false alarms. Moderate or greater bilateral congenital PCHI is the case definition for which the argument for screening is strongest because it is well established that the target condition causes significant delays in language development and academic achievement (see “When to screen” below), and it can be effectively treated by providing advice and support to the family and amplification or cochlear implantation. Additional novel interventions are on the way.¹¹

Who to screen

The impairment is sufficiently common (still 50 per 100 000 in “low risk” infants) to justify the solution that is most equitable, namely screening all children and not just the high risk group; the latter strategy requires the identification of risk factors, the need to put in place other arrangements for the low risk children which account for 50% of PCHI, and a higher staffing cost per patient tested. In practice, these costs reduce the difference in total cost between targeted and universal screening.¹² It is important to bear in mind that most screen failures and false alarms will inevitably occur in the 90% of the population not at high risk, so the context of universal screening adds to the importance of using a screen of high specificity and high positive predictive value.

When to screen: the effect of early intervention

There is evidence that early intervention is more effective than later intervention in improving outcome. Some linguistic input seems essential for language acquisition, and the ability to acquire a first language diminishes with age, suggesting an upper age limit for the ability to acquire certain aspects of speech and language. This has also been suggested by

analogy with sensitive periods shown in animals in relation to the visual¹³ and auditory¹⁴ systems. However, analogy with sensitive periods in animals was used to justify the introduction of vision screening in children but this has since been strongly challenged.¹⁵ Some ability to develop spoken language is demonstrable in infants with profound congenital PCHI who do not receive cochlear implants until late childhood.¹⁶ The length of any sensitive period for language acquisition and whether it differs between individuals remain to be determined.

The benefits of early intervention in congenital PCHI have been difficult to assess, until very recently, because of the absence of an effective strategy for early detection. Most studies have therefore been limited by small sample sizes, lack of comparison groups, and variable definitions of early.¹ Markides¹⁷ reported that the teachers’ ratings of speech intelligibility among 153 children matched for age, gender, age at onset of deafness, and degree of hearing loss and schooling was significantly better among those with hearing aids fitted before 6 months than among those with aids fitted later.

Analysis of video recordings of the language of 33 children with mild to profound congenital PCHI aged 32–85 months suggested that earlier referral for hearing assessment improved language production.¹⁸ Robinshaw¹⁹ compared three groups of children for the development of gestural and vocal production: five children with severe and profound PCHI who received amplification before the age of 6 months, 12 children with similar losses identified at an average age of 2 years and 3 months that had been reported in another similar study,²⁰ and a comparison group with normal hearing. The early treated group showed slightly delayed but similar development to normally hearing infants whereas development in those in the late identified group was delayed by two years.

In Colorado, USA, the receptive and expressive language abilities, assessed at a mean age of 26 months, of 72 children with congenital mild to profound bilateral PCHI whose loss was identified between birth and 6 months were compared with those of 78 children with a later (median age 16 months) identified loss.² Fifteen of the total of 150 had only mild (26–40 dB HTL) bilateral losses. The children received a family focused home intervention programme for one hour a week, with goals and activities determined according to an analysis of a videotaped parent-child interaction. The groups were similar with respect to sex, ethnicity, socioeconomic status, degree of PCHI, mode of communication, and cognitive status (assessed by a questionnaire on play). Language quotients, assessed by a well validated questionnaire completed by the primary care giver, were significantly higher in the early identified (mean (SD) 79.0 (20.9)) than the late identified (63.8 (19.3)) group ($p < 0.001$), irrespective of severity. Children identified between 6 and 12 months, however, did not show improved outcomes compared with chil-

dren identified after 12 months. Identification of PCHI in children with “normal cognitive ability” (defined as a cognitive quotient above 79) before 6 months of age reduced the difference between cognitive and linguistic quotients from 24–26 points to 5–7 points.

All these studies suggest that intervention must begin well before 1 year and possibly before 6 months to maximise the potential benefit. It is unclear why, in the Colorado study, great benefit was found in babies with PCHI identified before 6 months but none in those identified from 6 to 12 months nor why the benefits were just as great for mild losses, the effect of which on outcome would be expected to be less, as for greater degrees of PCHI. These findings need to be confirmed in other populations and in longitudinal studies. Nevertheless these studies taken together clearly support the case for a specifically neonatal screen. UNS therefore seems to be the logical solution.

Is universal neonatal screening really effective?

There have been many reports that a number of PCHIs come to light with UNS but these have been open to the criticism that the condition may have come to light just as quickly without the screen. The added value of the screen has, however, been directly measured in a controlled trial in the United Kingdom.²¹ A total of 53 781 babies born in four maternity hospitals in Wessex from October 1993 to October 1996 comprised the target population for screening. Two teams of four part time testers and equipment moved between the two pairs of hospitals to achieve four periods with neonatal screening and four without neonatal screening in each hospital, each of four to six months duration. Babies did or did not undergo neonatal screening depending on during which period they were born. A two step neonatal screen was used (see below). The IDT screen continued throughout all periods of the study.

Sixty two more babies with moderate, severe, or profound congenital bilateral PCHI per 100 000 target population (95% confidence interval 19 to 105 per 100 000) were referred before age 6 months during periods with neonatal screening than during periods without. Early confirmation and management of PCHI were also significantly increased. By April 1998, 53 infants with bilateral PCHI of moderate or greater degree had been identified from this birth cohort, including 33 identified before age 6 months. An additional four such cases of moderate or severe PCHI were identified before the age of 6 months for every five expected in the population, during periods with UNS compared with periods without. Detection of moderate PCHI this early was achieved in 93% of babies born during periods with UNS compared with 17% of babies born during periods without. Thus detection before the age of 6 months was closely associated with UNS.

In this way, the effectiveness of UNS for the early identification of PCHI in the United

Kingdom has been firmly established. The benefit of early treatment to speech and language development of children with PCHI was discussed in the preceding section, but the more cautious observer can still point out that such benefit has never been demonstrated in a controlled trial (which would be ethically impossible to conduct in moderate or more severe PCHI) and has not been measured in the context of UNS in the United Kingdom. A proposed study of the outcome at school age in babies enrolled in the Wessex trial,²¹ and the UNS programmes in Waltham Forest²² and Hillingdon²³ in the United Kingdom in the early 1990s aims to determine the benefits and costs of early provision of treatment to children with congenital bilateral PCHI for subsequent language development, behaviour, family relationships, quality of life, and related use of resources.

Which screening test

A screening test for any condition is valid if it detects the majority of people with the target disorder (high sensitivity) and excludes most people without the disorder (high specificity) and if a positive test usually indicates that the disorder is present (high positive predictive value). The best measure of the usefulness of a test for the infant being screened is probably the likelihood ratio—how much more likely a positive test is to be found in someone with, as opposed to someone without, the disorder²⁴—values above 10 being likely to be useful. It is also important to consider the “programme” performance of a screen, which relates to the target population, and not just the “test” performance, which relates to those infants actually screened. The two stage UNS used in the Wessex trial (see below) had a positive likelihood ratio of 63.9. The programme performance of that screen, including the run in period of the trial when coverage was poor, was a specificity of 98.8% and a sensitivity of 85.2%.^{21 25} Although the performance of UNS is often not reported in this way, those data that are reported suggest equally good performance in other implementations of two stage screens referenced below.

The performance of several neonatal screens for PCHI has been the subject of many reports.^{5 6 21–23 26–32} Transient evoked otoacoustic emissions (TEOAE) and the automated auditory brainstem response (ABR) have now been tested in several UNS programmes. TEOAE, also known as cochlear echoes, are low intensity sounds originating from the outer hair cells in the cochlea and can be elicited in response to clicks presented to the ear through a light weight probe. Their detection by a microphone within the same probe is a simple and rapid clinical test for the normal functioning of the middle and inner ear. The ABR is an electrical response to auditory stimuli which is usually recorded with three surface scalp electrodes and is a well established measure for assessment of the function of the eighth cranial nerve and the auditory pathway in the brainstem.

Each individual test has drawbacks. TEOAE testing is quick, sensitive to cochlear pathology, and involves only a small probe in the outer ear, which makes it very acceptable to parents and babies. AABR, and not TEOAE, is potentially sensitive to auditory neuropathy (accounting for 1.8% of PCHI in one study⁶) and the equally rare “central deafness”, an ill defined condition that merges into other neurological abnormalities, which are not the target of the screen.

Some 2.0–3.6% of screened infants in UNS programmes in the United Kingdom are reported to screen positive^{21 22 25 33} on TEOAE screening. A similar rate of 3.1–4% on AABR testing has been reported in the context of UNS in “community hospitals” with relatively low birth rates in the United States,^{26 31} but this test involves the placement of three scalp electrodes and, in the Wessex trial, was less readily accepted by families. The need for electrode placement may also decrease the number of screens that can be completed in a day and therefore its suitability for application in busy units. Historically it has more often been applied to the neonatal intensive care unit/special care baby unit population, in which the number of tests to be completed is tenfold smaller than the number in the low risk population and the incidence of abnormalities rostral to the cochlea is higher.

Whichever test is used, variation in test performance between hospitals and testers is manageable and improves progressively over the first year or more after introduction in the United Kingdom and the United States.^{5 6 21 33} Quality standards and consensus statements regarding early identification of PCHI have been put forward in the United States,^{34–36} the United Kingdom,³⁷ and for the European Union.³⁸ Particularly seminal were the recommendations of the National Institutes of Health that all infants should be screened for congenital PCHIs before the age of 3 months and that TEOAE detection and ABR measurement be used sequentially as a two stage universal neonatal screen.³⁴ The task force of the American Academy of Pediatrics on newborn and infant screening “defers recommendation as to a preferred method”,³⁶ but the combination of TEOAE and AABR technologies in a two stage screen, which has the most favourable published combination of specificity, sensitivity, acceptability, and high coverage in hospitals with a wide range of birth rates,^{4–6 21 22 39} has been recommended for national implementation in the United Kingdom. Data collected using such a two stage screen during the Wessex controlled trial suggests that amendment of pass/fail criteria can result in a screen with a high sensitivity and yield, a false alarm rate of less than 0.7%, and a rate of diagnosis of raised auditory thresholds due to non-targeted—that is, mild, unilateral, or conductive—hearing impairments of less than 0.1%.³² This and associated protocols are accessible on the internet at the website address: <http://www.ihr.mrc.ac.uk>.

The national screening committee has rightly been concerned about the possibility of

UNS leading to unnecessary anxiety in parents of babies screening positive. Studies of this issue have reported no increase in anxiety,^{25 40} and there is a striking lack of even anecdotal published accounts of this potential problem occurring in practice, but more studies are needed to provide reassurance about the absence of problems. Clearly the need to minimise the risk of such problems is another reason to choose screening tests and pass/fail criteria that reduce the false alarm rate.^{5 32 33} Delays in achieving differentiation between conductive and sensorineural losses are another avoidable cause of uncertainty and therefore anxiety and delayed treatment. Watkin and Baldwin³³ have reported an effective solution to this problem by adding bone conduction ABR to the test battery used in those screening positive on UNS; this reduced the average age of “audiological certainty” for those with a temporary conductive impairment and 50, 60 or 70 dB HTL thresholds from 35 to 8 weeks.

Which setting

The target population against which performance of UNS has been measured in the United States, Germany, Austria, and also in the Wessex trial in the United Kingdom has been all births in a given hospital or series of hospitals. Coverage of 90–95%, including 95–100% coverage of babies at high risk, has been achievable using a hospital based screening programme before postnatal discharge from hospital, supplemented, in some cases, by a recall system.^{5 6 21 23 26 28–31} This has proved sustainable for periods of years with practical economical staffing arrangements. It is still the case that the published evidence that UNS can be made to work on a large scale has all been obtained in a hospital setting.

Implementation has been taken one step further by Watkin and colleagues who have published a series of reports showing that an equally high level of coverage can be achieved not only for babies born in a given hospital but also for babies resident in a geographically defined area (the district of Waltham Forest in Essex) using, once again, a hospital based screen managed, in this case, by the same audiology service that also assesses the babies who screen positive.^{7 22 33 41} High coverage has also been achieved in health authority districts in Denmark.²⁷ Ten years later it still remains to be seen if other districts in the United Kingdom will be able to do as well.

There are obvious attractions to a screen at home rather than in hospital, especially as rates of early postnatal discharge from hospital are rising. Furthermore, midwives are already visiting newborn babies at home within a week of birth, achieving coverage with heel prick blood spot screening of 95–100% of babies (excluding those still in neonatal intensive care units and special care baby units) for other conditions, and recording the same in a database. However, any neonatal hearing screen delivered at home may involve several novel steps, possibly including management and delivery of UNS by a community based team, neonatal testing by many testers rather

Table 2 Definition of family friendly services (from website: <http://www.ihr.mrc.ac.uk>)

Family friendly service is:

1. Service provision by all professional sectors in a positive family friendly culture that encourages
 - collaboration that is "seamless"
 - responsiveness that meets families' real needs
 - provision of appropriate information between all agencies and for parents that enables families to make informed choices about services for their children
2. Paediatric audiology that exceeds a minimum standard in terms of quality and accessibility
3. A culture of service evaluation, including peer review with an element of feedback from parents and their hearing impaired children

than a specialist team, the use of new more portable technology, and so on. These innovations are largely untried and must, in the first instance, be compared with or at least added to hospital delivered UNS, which is the only system that has been shown to be workable on a large scale, and not simply introduced in a haphazard way.

What next in the United Kingdom

The comprehensive critical review commissioned by the Department of Health, published in 1997¹ (and usefully summarised in this journal⁴²) set in train a process which now seems certain to end with a national change in practice.^{15 42 43} The National Screening Committee of the Department of Health recommended to ministers that UNS be introduced across the United Kingdom and, as mentioned in the opening paragraph, an announcement was made on 22 June 2000 by Yvette Cooper, government minister for public health, that the United Kingdom government had decided in principle to introduce UNS and wished to invite districts to apply to be part of a 20 site pilot. A national screening protocol and quality markers such as coverage rates above 90% and maximum false positive rates of 3% have already been specified. Districts will also be required to show that they can provide a well coordinated audiological and educational care service which is patient centred and "family friendly" (table 2) as defined by a number of specific standards.⁴⁴ All this information is currently posted at website address <http://www.ihr.mrc.ac.uk>. Specific proposals for "rolling out" training, the use of information technology, quality assurance, and pump priming funding may follow. The yield of the IDT screen is negligible once UNS is in place²¹ and, although there will be a need for some form of targeted postnatal screen for those not neonatally screened and some others, the role of the health visitor in hearing screening is likely to be redefined more widely in the context of changes in child health surveillance.¹⁵

The expected change in national practice to UNS as the screen of choice for hearing impairment in infancy can reasonably be described as evidence based, although some questions remain. What is the optimum age for intervention? What is the interaction between severity of impairment and magnitude of benefit of early intervention? How will all babies be screened, irrespective of place of birth? Is community based UNS feasible on a large scale? Are the latest commercially available variations on the testing equipment better in ways that matter?

Individual districts will have to decide how to achieve high coverage of their entire population by adapting national recommendations to their particular circumstances, how to react to new untried technology, and how to fund and manage the changeover from the current situation to UNS. If, however, outcomes are to improve, the introduction of UNS will need to be coupled to the provision of early treatment in family friendly form—probably the greater challenge.

Summary

UNS for bilateral congenital permanent hearing impairment has been shown to be effective in the United Kingdom and elsewhere, and the United Kingdom government has decided in principle to introduce it in place of the current arrangements for hearing screening in infancy. This will require changes in the configuration of screening and audiology services and an acceleration of the current trend for health and education to work more closely with the family and with each other. Continuing evolution of the pattern of postnatal care and of screening technology may alter the available options for the precise method of neonatal screening but, in practice, effectively managed screen delivery and early patient centred treatment are more likely to affect outcome. We can now look forward to better language development with all its secondary benefits for these children and their families in the United Kingdom.

- 1 Davis A, Bamford J, Wilson I, Ramkalawan T, Forshaw M, Wright S. A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment. *Health Technol Assess* 1997;1:1-177.
- 2 Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. *Pediatrics* 1998;102:1161-71.
- 3 Davis A, Wood S. The epidemiology of childhood hearing impairment: factors relevant to planning of services. *Br J Audiol* 1992;26:77-90.
- 4 Mehl AL, Thomson V. Newborn hearing screening: the great omission. *Pediatrics* 1998;101:1-40.
- 5 Gravel J, Berg A, Bradley M, et al. New York state universal newborn hearing screening demonstration project: effects of screening protocol on inpatient outcome measures. *Ear Hear* 2000;21:131-40.
- 6 Vohr BR, Carty LM, Moore PE, Letourneau K. The Rhode Island hearing assessment program: experience with statewide hearing screening (1993-1996). *J Pediatr* 1998;133:353-7.
- 7 Watkin PM. Outcomes of neonatal screening for hearing loss by otoacoustic emission. *Arch Dis Child Fetal Neonatal Ed* 1996;75:F158-68.
- 8 Paradise JL, Feldman HM, Colborn K, et al. Parental stress and parent-rated child behavior in relation to otitis media in the first three years of life. *Pediatrics* 1999;104:1264-73.
- 9 Stewart-Brown S, Haslam M. Screening for hearing loss in childhood: a study of neonatal practice. *BMJ* 1987;294:1387-9.
- 10 Mott A, Emond A. What is the role of the distraction test of hearing? *Arch Dis Child* 1994;70:10-13.
- 11 Steel KP. New interventions in hearing impairment. *BMJ* 2000;320:622-5.
- 12 Stevens JC, Hall DMB, Davis A, Davies CM, Dixon S. The costs of hearing screening in the first year of life in England and Wales. *Arch Dis Child* 1998;78:14-19.
- 13 Blakemore C. Maturation and modification in the developing visual system. In: Held R, Leibowitz HW, Teuber HL, eds. *Handbook of sensory physiology, vol VIII*. Berlin: Springer Verlag, 1978;377-436.
- 14 Rubel EW. Strategies and problems for suture studies of auditory development. *Acta Otolaryngol Suppl (Stockh)* 1985;421:114-28.
- 15 Robinson R. Editorial: effective screening in child health. *BMJ* 1998;316:1-2.
- 16 Summerfield Q, Marshall D. *Cochlear implantation in the UK 1990-94*. London: HMSO, 1995.
- 17 Markides A. Age at fitting of hearing aids and speech intelligibility. *Br J Audiol* 1986;20:165-7.
- 18 Ramkalawan T, Davis AC. The effects of hearing loss and age of intervention on some language metrics in young hearing-impaired children. *Br J Audiol* 1992;26:97-107.

- 19 Robinslaw HM. Early intervention for hearing impairment: differences in the timing of communicative and linguistic development. *Br J Audiol* 1995;29:315–34.
- 20 Tait M. Making and monitoring progress in the preschool years. *Journal of the British Association of Teachers of the Deaf* 1999;11:143–53.
- 21 Wessex Universal Hearing Screening Trial Group. Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment. *Lancet* 1998;352:1957–64.
- 22 Watkin PM, Baldwin M. Confirmation of deafness in infancy. *Arch Dis Child* 1999;81:380–9.
- 23 Tucker SM, Bhattacharya J. Screening of hearing impairment in the newborn using the auditory response cradle. *Arch Dis Child* 1992;67:911–19.
- 24 Greenhalgh T. How to read a paper. Papers that report diagnostic or screening tests. *BMJ* 1997;315:540–3.
- 25 Kennedy CR. Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment: coverage, positive predictive value, effect on mothers and incremental yield. *Acta Paediatr Suppl* 1999;432:73–5.
- 26 Mason JA, Herrmann KR. Universal infant hearing screening by automated auditory brainstem response measurement. *Pediatrics* 1998;101:221–8.
- 27 Parving A, Salomon G. The effect of neonatal universal hearing screening in a health surveillance perspective: a controlled study of two health authority districts. *Audiology* 1996;35:158–68.
- 28 Welzl Muller K, Boheim K, Stephan K, Schlogel H, Stadlmann A, Nekahm D. Optimizing hearing screening by transient evoked otoacoustic emissions in newborn infants. *HNO* 1997;45:227–32.
- 29 Arnold B, Schorn K, Stecker M. [Screening program for selection of hearing loss in newborn infants instituted by the European Community]. *Laryngorhinootologie* 1995;74:172–8.
- 30 Maxon AB, White KR, Vohr BR, Behrens TR. Using transient evoked otoacoustic emissions for neonatal hearing screening. *Br J Audiol* 1993;27:149–53.
- 31 Barsky-Firsker L, Sun S. Universal newborn hearing screening: a three year experience. *Pediatrics* 1997:e4.
- 32 Kennedy CR, Kimm L, Thornton ARD, Davis A. False positives in universal neonatal screening for permanent childhood hearing impairment. *Lancet* 2000;in press.
- 33 Watkin PM, Baldwin M. Implementing a diagnostic audiological test battery as part of a universal neonatal screen. *Unknown* 2000;in press.
- 34 National Institutes of Health. *Early identification of hearing impairment in infants and young children*. Bethesda, MD: National Institutes of Health, 1993.
- 35 Joint Committee on Infant Hearing. Joint Committee on Infant Hearing position statement. *ASHA* 1994;36:38–41.
- 36 American Academy of Pediatrics task force on newborn and infant screening. Newborn and infant hearing loss: detection and intervention. *Pediatrics* 1999;103:527–30.
- 37 National Deaf Children's Society. Quality standards in paediatric audiology. Vol 1: guidelines for the early identification of hearing impairment. *National Deaf Children's Society* 1994;1:1–10.
- 38 Lutman ME, Grandori F. Screening for neonatal defects. European consensus statement. *Eur J Pediatr* 1999;158:95–6.
- 39 Vohr BR. Screening infants for hearing impairment. *J Pediatr* 1996;128:710–14.
- 40 Watkin PM, Baldwin M, Dixon R, Beckman A. Maternal anxiety and attitudes to universal neonatal screening. *Br J Audiol* 1998;32:27–37.
- 41 Watkin PM. Neonatal otoacoustic emission screening and the identification of deafness. *Arch Dis Child Fetal Neonatal Ed* 1996;74:F16–25.
- 42 Bamford J, Davis A, Stevens JC. Screening for congenital hearing impairment: time for a change. *Arch Dis Child Fetal Neonatal Ed* 1998;79:F73–6.
- 43 Sheldon T, Wilson P, Boutle MT, Sharp F. Pre-school hearing, speech, language and vision screening. *Effective Health Care* 1998;4:1–12.
- 44 Davis A, Bamford JM, Baguley D. Family friendly audiology services for children. *Br J Audiol* 2000;in press.

website
extra

Figures to accompany
this article appear on
the ADC website

www.archdischild.com

Commentary

Colin Kennedy's review of universal neonatal hearing screening (UNHS) prompts an important question—how do we move in a systematic and timely manner from scientific evidence on screening, to the formulation and implementation of public policy?

The National Screening Committee (NSC) reviews new and existing screening programmes and advises ministers on policy. It originated as a response to disasters with cervical cytology and breast cancer screening. The NSC now has an antenatal subgroup and a children's subgroup (CSG). Programmes are assessed against an extensively revised set of criteria¹ based on those of Wilson and Jungner.²

The issue of screening for hearing impairment was prioritised by the Health Technology Assessment (HTA) programme in 1994 and a systematic review was commissioned.³ It argued that early identification and habilitation of congenital hearing impairment is beneficial, current methods of detection (the infant distraction test or IDT) give very poor results, and UNHS is better and cheaper. The review was debated in 1998 by the CSG, audiology professionals, and the National Deaf Children's Society. The CSG's priority was to secure government commitment to the principle of UNHS and ensure that, once accepted, it would be available to everyone; but members wanted several issues to be clarified before recommending that a new service should be established countrywide.

First, there were some weak links in the evidence that early diagnosis is beneficial. In the absence of UNHS, children diagnosed early are not necessarily the same as those diagnosed later, so direct comparisons may be misleading, even when severity and cause of hearing impairment are controlled. Both early diagnosis and effectiveness of intervention might, for example, be affected by parents' understanding of infant communicative behaviour. Early deficiencies in language acquisition may disappear over time. Communicative and social skills, employability, and mental health as adults are crucial long term outcomes for which all others are proxies. There are, however, now some preliminary data of superior outcomes in a screened as opposed to a non-screened population.

The second concern was the potential for harm. Significant numbers of normal babies would be referred for assessment. What is the cost in resources and parental anxiety? The meagre evidence available suggested that most parents value screening and cope with short term anxiety; nevertheless it seemed important to minimise it and protect parents who are vulnerable for other reasons such as depression. Further data were therefore requested on the question of anxiety and harm caused by false positives.

The third issue was whether audiology services could respond effectively to a sudden increase in referrals of very young babies needing investigation, intervention, and support. Many services are under resourced and would probably be unable to cope with UNHS at present.

Finally, cost issues were re-examined. Unfortunately, there was no easy way of recovering resources from discontinuing the IDTs and reinvesting them in UNHS. The costs of implementing and running UNHS were thought to be somewhat higher than previously calculated, though still much cheaper than IDT. An attempt was made to compare the costs and benefits of UNHS with other screening programmes for children. The economic analyses of these programmes turned out to be based on widely varying assumptions and no direct comparison was possible. Further work was commissioned to standardise the presentation of economic data on screening for future projects.⁴

After reviewing the evidence, the CSG and NSC accepted that the balance of probabilities strongly favoured early diagnosis being beneficial, but only if UNHS is linked to an audiology service that offers clear, understandable information with access to personal support and advice, and resolves clinical uncertainty as quickly as possible. Therefore, the “Family Friendly Hearing Service” standards were developed for districts to achieve before embarking on UNHS, but this was only possible because funds could fortuitously be obtained from other sources. The CSG then proposed an implementation group to plan the introduction of UNHS in selected pilot sites, with mechanisms built in for clarifying the concerns raised by the committee. This has now been agreed by the Minister.

The process has raised important questions about how policy is formulated in the UK health service. A highly centralised process has some advantages. It avoids precipitate introduction of new technologies, ensures debate between individuals in a variety of disciplines, and increases equity of access across the country. In principle, it should enable professionals and ministers to make decisions based on sound evidence, although the weakness of current health economics analyses, when deciding on the priority order of new screening programmes, was very obvious.

There are also disadvantages of centralised policy making. When the systematic review was commissioned by the HTA, there was no clarity as to how its findings might influence policy. The reviewers felt that they were expected to treat the process purely as an academic exercise. Their personal investment in implementation of their findings extended beyond their brief. The CSG was (rightly) perceived as lacking the authority to turn policy into action. This in turn was frustrating to clinicians and academics who were eager to innovate and to establish UNHS in their areas, but had to wait for “official” government policy. There was no

related source of funding to address the questions that the CSG felt must be answered before introducing the programme country-wide, and the funding cycles which could support such work move too slowly to support the pace of innovation and research. Similar problems have arisen in connection with proposals for pilot trials on screening for medium chain acyl CoA dehydrogenase deficiency using tandem mass spectrometry.^{5 6}

We agree that screening policy probably is best developed and monitored at national level, but we need a defined pathway from identification and prioritising of an opportunity or problem, to assessment, review, further research, and implementation; better mechanisms for economic appraisal; better ways of responding to the views of the consumer, as represented in this case by the voluntary organisations; and a clear commitment from government formally to review and respond to the advice of professional committees.

DAVID HALL

*Professor of Community Child Health
Sheffield University
Sheffield, UK
d.hall@sheffield.ac.uk*

ADRIAN DAVIS

*Professor of Epidemiology
MRC Institute of Hearing Research
Nottingham, UK
Adrian@ihr.mrc.ac.uk*

- 1 National Screening Committee. Criteria for screening programmes. 1999. National Screening Committee. URL: www.doh.gov.uk/nsc/library/lib_ind.htm
- 2 Wilson JMG, Jungner G. *Principles and practice of screening for disease*, Public health papers no. 34. Geneva: WHO, 1968.
- 3 Davis A, Bamford J, Wilson I, Ramkalawan T, Forshaw M, Wright S. A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment. *Health Technol Assess* 1997;1(10).
- 4 Deverill M. *Comparison of the costs of screening programmes*. Sheffield: SchARR.
- 5 Pollitt RJ, Green A, McCabe CJ, *et al*. Neonatal screening for inborn errors of metabolism: cost, yield and outcome. *Health Technol Assess* 1997;1(7).
- 6 Seymour CA, Thomason MJ, Chalmers RA, *et al*. Newborn screening for inborn errors of metabolism: a systematic review. *Health Technol Assess* 1997;1(11).