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Review

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

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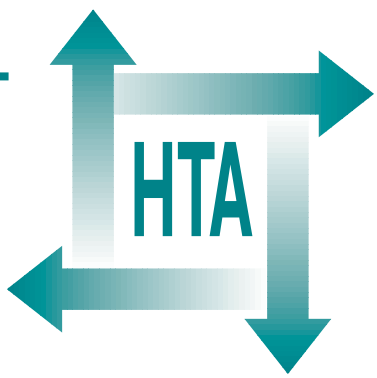
Health Technology Assessment
NHS R&D HTA Programme



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

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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.

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Glossary and list of abbreviations

Technical terms and abbreviations are used throughout this report. The meaning is usually clear from the context but a glossary is provided for the non-specialist reader. In some cases usage differs in the literature but the term has a constant meaning throughout this review.

Glossary

Permanent hearing impairment Average hearing levels measured at ≥ 40 dB HL on all occasions.

Congenital hearing impairment Hearing impairment considered by examination of the case history to be present and detectable using appropriate tests at or very soon after birth. This was the default classification when there was no indication in the case notes of the presence of an acquired impairment or evidence of a progressive or a late-onset element to the impairment.

Acquired hearing impairment Hearing impairment acquired post-natally or of late-onset or progressive nature which, on the basis of case history was not considered to be present and detectable using appropriate tests at or very soon after birth.

Average hearing level The average of the thresholds (in dB HL) measured in the better hearing ear at 0.5, 1, 2 and 4 kHz, or whatever combination was available.

Moderate hearing impairment Average hearing level 40–69 dB HL.

Severe hearing impairment Average hearing level 70–94 dB HL.

Profound hearing impairment Average hearing level ≥ 95 dB HL.

Cohort That section of a population born during a particular period and identified by period of birth, e.g. born between 1985 and 1990.

Incidence The number of new instances of a specific condition occurring during a certain period in a specified population.

Prevalence The total number of instances of a specified condition in a given population at a specific time.

Prevalence rate The number of people with the condition or attribute, divided by the population at risk at a point in time (or midway through a period).

Family history Permanent hearing impairment present since childhood in at least one of the following family members; parent, sibling, grandparent, great-grandparent, aunt, uncle, nephew, niece or cousin.

Referral The date when the child was referred to an audiologically qualified professional for assessment. This does not include a routine or at-risk referral for a hearing screen.

Confirmation of permanent hearing impairment The date when the first measurement of raised thresholds by an age-appropriate audiological test was made, with the reliability of the results being high.

Prescription of hearing-aid The date when an audiological professional recommended a hearing-aid for the child. (Note: this date may be before the aid is officially authorised by an ENT consultant.)

Fitting of hearing-aid The date when the child is first fitted with and given a hearing-aid, not the date on which moulds are taken.

Sensitivity The effectiveness of a screen (or test, or programme) in identifying cases. Test sensitivity is the percentage of cases failing a single test opportunity; screen sensitivity is the percentage of cases tested referred by a screen (which may be more than one test opportunity); programme

continued

contd
sensitivity is the percentage of cases referred by a screening programme taking into account the coverage of the target population.

Specificity The ability of a programme to screen out individuals who are not cases (e.g. in a screening programme, the percentage of unaffected individuals who pass the screen).

List of abbreviations

ABR	auditory brainstem response
ACSHIP	Advisory Committee on Services for Hearing Impaired People
AR	At-risk*
ARC	auditory response cradle
ATO	Assistant Technical Officer
BACDA	British Association of Community Doctors in Audiology
BATOD	British Association of Teachers of the Deaf
BeST	Behavioural Screening Test
CFA	cranio-facial abnormality
CI	95% confidence interval Note: confidence intervals should only be used to compare independent samples, for example, when comparing severities it is correct to compare the CI for those with 40–69 dB HL impairments with those who have 70–94 dB HL impairments. It is not correct to compare those with ≥ 40 dB HL with those with ≥ 70 dB HL.
CMO	Clinical Medical Officer
dB HL	decibel, log-scale measure of hearing level using pure tone average or an estimate of dB HL made using alternative scales (e.g. dB(A))
DPOAE	distortion product otoacoustic emission
HVDT	Health Visitor Distraction Test
HVS	Health Visitor Surveillance
IDT	Infant Distraction Test
IODC	International OAE Data Centre
JCIH	Joint Committee on Infant Hearing
MRC	Medical Research Council
MTO	Medical Technical Officer
NCHAM	National Center for Hearing Assessment and Management (USA)
NDCS	National Deaf Children's Society
NICU/SCBU	neonatal intensive care unit or special care baby unit
NIH	National Institutes of Health (USA)
OAE	otoacoustic emission
OME	otitis media with effusion (glue ear)
PARC	portable ARC
PCHI	permanent childhood hearing impairment
PPV	positive predictive value
RCT	randomised controlled trial
RNID	Royal National Institute for Deaf People
SES	school-entry screening
TEOAE	transient evoked otoacoustic emission
ToD	Teacher of the Deaf
UNS	universal neonatal screening
VRA	visual reinforcement audiometry

* Used only in figures and tables

Executive summary

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[1] likely to affect their own and their family's quality of life. Present services will miss about 400 of these children by $1\frac{1}{2}$ years of age, and about 200 of these children by $3\frac{1}{2}$ 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

[1] 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.

Chapter 1

Introduction and main review questions

Scope

A summary of the major effects of permanent childhood hearing impairment (PCHI) are presented in this chapter, together with an outline of the historical arguments for early identification and screening. A brief history of childhood hearing screening in the UK is followed by an outline of current concerns about identification and service provision in the light of new developments and, hence, the need for this critical review. The main review questions, an outline of the review procedures, and the structure of the report are summarised.

Introduction

In the UK, about 800 children are born each year with a significant permanent hearing loss (Davis, 1993a). The impact of hearing loss upon both child and family can be substantial. Most obviously, hearing loss in childhood may disrupt the process of communication and normal language acquisition, leading to poor language, communication and literacy skills (see, for example, Conrad, 1979; Bench & Bamford, 1979; Gregory, 1995; Levitt *et al,* 1987; Moeller *et al,* 1986; Markides, 1986; Gregory & Mogford, 1981; Gregory *et al,* 1995; Gallaway *et al,* 1994). However, other areas of development may also be affected. Studies have documented the possible effects of PCHI on educational achievement (Powers, 1996), mental health (Laurenzi & Monteiro, 1997) and self-esteem (Batchava, 1993), and on long-term opportunities for training and career development (Downs, 1994; Gregory *et al,* 1995). The costs to society are also likely to be substantial in terms of audiological, otological, paediatric and education provision.

It is argued, with some support, that these potentially negative effects of PCHI may be reduced by the introduction of language support (signed or spoken), the use of amplification (hearing-aids, cochlear implants), and family support and habilitative interventions of suitable type and quality, and that the earlier this is done the better. Thus it is widely held that "early detection and management of hearing impairment will help to lessen the impact of the condition on the child's social, emotional, intellectual and linguistic development. The child and family will benefit from such early detection and management" (NDCS, 1994).

While parental observation and good, responsive services are likely to identify some of these children reasonably soon after birth, particularly those with more severe impairments, satisfactory identification rates will not be achieved without systematic population screening programmes. These arguments underpin the long history of screening for childhood hearing impairment in the UK, dating back to the introduction of the School Medical Service at the turn of the century (Watkin, 1991). By the 1930s, hearing screening by various methods was being implemented at school entry, although the quality of these early screens was doubtful (Fisch, 1981). In 1957, following a Medical Research Council (MRC) request and studies by Midgely (1957), it was recommended by Ewing (1957) that all children should undergo school-entry hearing screening using the pure-tone 'sweep' test (pure tones presented via headphones at supra-threshold levels). To this day this screen is universally implemented throughout the UK and, despite limited published discussion of its costs and effectiveness, and lack of clarity in its aims, there is a widespread consensus that "the pure-tone sweep test has value educationally and as a safety net to catch any deficiencies of the earlier screening system in the overall public health provision" (Haggard, 1993).

When the 1944 Education Act gave local authorities the means to implement preschool as well as school-entry screening (SES), a number of influential workers in education and health argued strongly that identification of permanent congenital hearing loss by school entry was far too late (Ewing & Ewing, 1944; Whetnall, 1955). The Ewings had demonstrated that testing children's hearing at 9 months was possible, and described the Distraction Test (Ewing & Ewing, 1944). In the early 1950s, a collaboration between the University of Manchester and Dr Berenice Humphreys led to the first implementation of the Health Visitor Distraction Test (HVDT) in Leicester. This was shortly followed by the introduction of the HVDT as a screening test in Birmingham under the guidance of Dr Jean Mackintosh. In 1957, Irene Ewing published recommendations concerning training

and implementation of the HVDT and, by the early 1960s, its use as a universal screen of babies' hearing had become widespread and was supported by training courses in London (Galbraith, 1976), Manchester (Ewing, 1957) and elsewhere. The HVDT is currently undertaken in infants aged 7–8 months; it consists of localisation responses to low-level sounds presented to the child by a tester while the child's attention is suitably manipulated by a second tester. Thus, in the UK, universal pre-school and school-entry screening are well-established procedures that have received continued, if qualified, endorsement (Hall, 1996).

However, as early as the 1970s, doubts were being expressed about the effectiveness of the HVDT and its ability to identify PCHI early. Thus, for example, Boothman and Orr (1978) demonstrated low coverage and low yield; Martin and colleagues (1981) showed the mean age of identification to be about 3 years; the National Deaf Children's Society (NDCS, 1983) argued, as a result of a parental survey, that the screen had low yield, low sensitivity and poor credibility with parents. Indeed, some evidence for these assertions was provided by a parent whose two children passed the screening test, despite severe to profound permanent hearing loss (Robinson, 1983). In 1975, the (then) DHSS Advisory Committee on Services for Hearing Impaired People (ACSHIP) set up a subcommittee to consider matters relating to services for hearing-impaired children "with particular reference to screening. This had become a matter of some urgency because evidence had been received ... suggesting that in some areas not all children were being screened ... [and] doubt about the effectiveness of screening ..." (ACSHIP, 1981).

The final ACSHIP report (ACSHIP, 1981) made 56 recommendations and endorsed the UK's public health approach to the assessment of PCHI, giving clear support to the HVDT and SES, both applied universally. The extent to which these recommendations were acted upon remained, however, largely a matter of local policy and, although the use of the HVDT remains near-universal, with some notable attempts to implement it effectively (McCormick, 1983; McCormick *et al,* 1984a; Watkin, 1991), there are still serious concerns about current arrangements for detecting hearing loss in young children (NDCS, 1994) and about the focus, performance, coverage, yield and cost-effectiveness of the HVDT (as identified by the NHS HTA Programme Commissioning Document in 1994).

Against this background, a number of other key developments were taking place in the 1980s and early 1990s. Increased knowledge of the development of early pre-verbal language and communication and its importance (see, for example, Oller, 1991), some outcomes evidence (Markides, 1986), and widespread belief (Clark, 1989) encouraged the view that the age of identification using the HVDT (which cannot be implemented before a developmental age of 6/7 months) was less than optimal, even if the screen were found to be highly effective. This stimulated the development of behavioural test devices which could be used for the screening of hearing in neonates – notably, in the UK, the Auditory Response Cradle (ARC) (Bennett, 1979; Bennett & Lawrence, 1980). In 1985, a Department of Health initiative funded the provision of a dozen ARCs to maternity units in hospitals with well-provided Audiology Departments; this led to data which supplemented that already emerging from Hillingdon on the ARC and neonatal screening (Davis, 1984; Tucker, 1987; Tucker & Bhattacharya, 1992). The initiative also stimulated further epidemiological work on PCHI, and many good-quality data have been published since 1980, including information on risk factors and prevalence rates (for example, Newton, 1985; Davis & Wood, 1992). One aspect of these epidemiological data which attracted particular interest was the identification of populations-at-risk for PCHI, the simplest of which is those newborn babies needing special care for more than 48 hours. The relative risk of PCHI for babies in Special Care Baby Units or neonatal intensive care units (NICU/SCBUs) was reported to be of the order of 10:1 (Davis & Wood, 1992), and has led to cost-effectiveness arguments for 'targeted' neonatal screening of the at-risk population. It has been argued that screening some 5% of the total birth cohort might identify 50% of the PCHI population (Davis & Wood, 1992). Indeed, targeted neonatal screening is now fairly widespread in the UK (see chapter 4).

Behavioural neonatal hearing screening using the ARC is probably not optimal for at-risk neonates (Davis, 1984; McCormick *et al,* 1984a; Davis *et al,* 1991); hence, the move towards targeted screening depended on technological advances and new devices. Since the early 1970s, auditory brainstem response (ABR) testing had been developed as a powerful diagnostic tool but, by the 1980s, simplified automated or semi-automated devices became available for neonatal screening (see, for example, Mason, 1984; Mason *et al,* 1987; Herrman *et al,* 1995). Otoacoustic emissions (OAEs) were discovered in the late 1970s (Kemp, 1978) and there is now considerable experience of and data on the

use of OAEs for neonatal hearing screening (see, for example, Kemp & Ryan, 1993).

Concern about late identification of PCHI, the theoretical and practical arguments for early identification, and thereby, early intervention, epidemiological knowledge and the advent of potentially suitable screening tests have led to the publication recently of guidelines for quality standards. In the UK, the NDCS (1994) set relatively ambitious early-identification targets (40% of children with moderate or greater PCHI to be identified by 6 months of age, 80% by 12 months) and, in the USA, the National Institutes of Health consensus statement (NIH, 1993a) set very ambitious targets (identification of all congenital PCHI by 3 months of age) and called for universal neonatal screening (UNS) throughout the country. Curnock (1993) neatly defined the service questions of importance for the UK following the NIH consensus statement.

These various influences appear to have affected UK screening services in a somewhat haphazard way. Provision is highly variable. UNS is offered in two localities; neonatal screening targeting at-risk populations (variously defined) is spreading, using a variety of protocols; in some areas attempts are being made to improve the implementation and audit of HVDT (see, for example, Plant & Pick, 1995), while in others it has been abandoned (Scanlon & Bamford, 1990; Watkin, 1996). Surveillance procedures have become more explicit, and the Hall reports (1989; 1992; 1996) on child health surveillance have been influential at some locations but have led to planning blight at others. Screening protocols and procedures are variable, and often unspecified; audit and data-monitoring are increasing but still not widespread (Allen & Wallace, 1996). A critical appraisal of the lack of justification for identifying fluctuating non-permanent hearing loss in infants (Haggard *et al*, 1992) has altered views on the aims of the HVDT in some places. The additional uncertainty resulting from NHS reforms and the service-purchasing consequences has produced a climate in which there is a danger of inappropriate and inconsistent policy decisions being made, and in which service variability is likely to be incompatible with evidence-based decisions and cost-effectiveness.

Thus there is a need to consider "whether available evidence is sufficient to justify a reassessment of policy and practice ... in particular, to identify and review the evidence for the effectiveness and cost-effectiveness of neonatal and targeted screening for the early detection of hearing loss in young children" (NHS HTA Programme Commissioning Document, 1994).

This critical review was undertaken in 1995–96, augmented by a determination of current practice and developments in the UK. Its aim was to enable a set of recommendations on policy and practice to be made that should help:

(i) to reduce the current confusion surrounding screening for PCHI
(ii) to provide access to data which can inform better practice for purchasers and providers of health care, and thereby help to deliver earlier detection, better outcomes and improved cost-effectiveness of screening
(iii) to identify specific areas for further research and service development.

The review has two main strands: a critical review of available literature and a survey of current practice in the UK. These complementary strands are designed to gather information on epidemiology, outcomes and benefits, costs, coverage, feasibility, sensitivity and specificity of screening programmes, service structures, impact and acceptability of screens, and implementation issues. The hearing screening programmes in question are those designed to deliver early identification of at least moderate congenital PCHI – neonatal screening, and the HVDT screen. The evidence for and issues surrounding SES are not examined except insofar as the survey of current practice furnished information. The survey provides the context for any policy and practice recommendations and was supplemented by the use of focus groups (Kitzinger, 1995). Finally, visits to a number of key research groups and/or key service providers in the UK and North America provided further up-to-date contextual information on research, service, and service development.

Discussion of hearing screening programmes and their effectiveness involves not only internal performance measures (sensitivity, specificity, coverage, etc.) but also external performance measures, the chief of which is age of identification or confirmation of PCHI. Age at identification and consequent age of hearing-aid fitting have therefore become important indicators of screening and service performance (NDCS, 1994). However, identification of PCHI and hearing-aid fitting is actually the start of a lifetime process of multi-agency care, with the family and child at the centre, involving Education, Health, and Social Services. Outcomes of early identification and support occur in many different domains and

periods in this lifetime process. Age of identification is merely a surrogate outcome measure for later outcomes (such as quality of life, quality of family life, employment and academic achievement). The case for very early identification may be that not only do parents want it but also that it has a beneficial effect on both lifetime process and lifetime outcomes.

Some relevant life events from birth to the start of adulthood are shown schematically in *Figure 1;* those that are (or would be, if implemented) immovable in time are indicated. The timing, quality and success of the other events may, potentially, be significantly affected by the earliness (or lateness) of identification of the hearing impairment.

There are several opportunities available for screening for hearing impairment. The three most opportune times would seem to be:

(i) at birth
(ii) at a developmental check during the first year of life
(iii) at entry to full-time education.

This review advances considerations about the effectiveness and cost of screening in the first year of life but this cannot be viewed in isolation from the continuing care required by hearing-impaired children. The NIH consensus statement was possibly too concerned with the screening process *per se* and saw 'rehabilitation as a part of screening'. We emphasise that a more natural approach is to see screening as part of (and the start of) the continuing care process that should involve both health and education. NHS priorities are to "ensure, in collaboration with local authorities and other organisations, that integrated services are in place to meet needs for continuing healthcare..." *(Priorities and Planning Guidance for the NHS.* NHS Executive, 1996). The key to effective service delivery, echoed in our discussion and focus groups, is better integration of the services offered to hearing-impaired children and their parents, with parents at the centre of provision and decision-making. The interdependence of these services, which are needed to support the screening programme, as well as throughout the development of the child, are illustrated in *Figure 1*.

Main review questions

In accordance with our terms of reference, this review has been undertaken to examine the

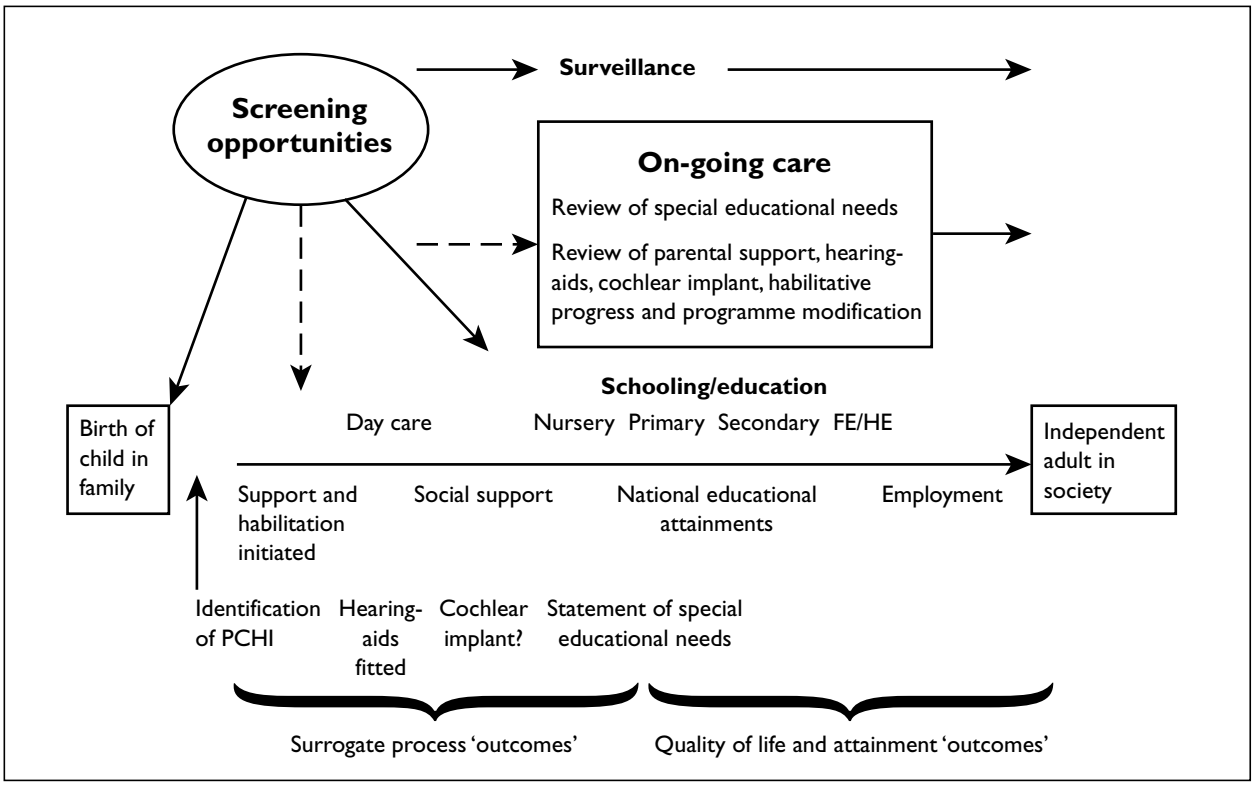

FIGURE 1 *A schema covering birth through to adulthood for the major services and life events in children with PCHI. Events above the line are fixed in time or are on-going. Events below the line could be affected, in terms of timing, quality or success, by the age of identification (FE, further education; HE, higher education)*

evidence for the effectiveness and cost-effectiveness of early screening for PCHI. The remit covered those babies born with PCHI; however, evidence on both the numbers of infants with and the time course of delayed-onset or progressive PCHI is clearly relevant to the overall yield of early screens, and has therefore been included.

Two decisions were necessary about the target group for screening:

(i) that the severity of impairment was at least moderate (≥ 40 dB HL (decibels, hearing level averaged over 0.5, 1, 2 and 4 kHz))
(ii) that the impairment was permanent.

This is not to deny possible subtle but real effects of mild or unilateral PCHI on speech perception in noisy backgrounds (e.g. nursery, reception classes) and on some aspects of educational achievement (see, for example, Bess & Tharpe, 1986). However, at some point on the severity continuum, evidence in a number of domains becomes equivocal, open to multiple interpretation, poorly-controlled or just non-existent, largely because the sub-population of mild or unilateral PCHI may be difficult to identify with the requisite certainty. Thus, while we now have good rigorous data on the epidemiology of moderate or greater PCHI, this is not the case for milder levels of PCHI. It follows that evidence on, for example, screening sensitivity and outcome benefits for mild PCHI is also highly equivocal or non-existent.

Most published work on epidemiology, screening effectiveness and outcomes associated with PCHI has for these reasons been performed with populations with moderate or greater degrees of PCHI. It is therefore sensible to pose the questions of prevalence, screening tests performance, costs and outcome benefits with respect to this population. If the evidence indicates benefits or cost-effectiveness of very early screening for moderate or greater PCHI, future research will need to consider the point on the severity continuum at which early identification and intervention is no longer of sufficient cost-effectiveness to justify service intervention.

The definition of 'moderate or greater' PCHI as the target condition for hearing screening in the first year of life is therefore determined by published evidence and data that are available from audits and research programmes. The bulk of the evidence (on screen performance, and outcomes) is on bilateral PCHI of ≥ 40 dB HL (averaged across mid-frequencies), and sometimes ≥ 50 dB HL. In some studies, the latter point on the severity continuum has been chosen because it largely (although not completely) avoids the confusion of including cases of non-permanent childhood hearing loss. The prevalence of non-permanent childhood hearing loss is relatively high, largely caused by fluctuating middle ear conditions associated with otitis media with effusion (OME). The literature on OME and its effects is considerable and has been reviewed recently elsewhere (Haggard & Hughes, 1991). The current review does not, therefore, include this population which is the subject of an on-going MRC-funded randomised controlled trial (RCT). Current opinion is that screening at an early age for OME is not justified and work is needed on what might constitute a viable system at a later age.

In light of these considerations, evidence on the following questions were sought in this review.

- What is the current epidemiology of PCHI in the UK? In particular, what is the prevalence of PCHI? What is its severity distribution? What is the prevalence of congenital versus late-onset PCHI? What is the time course of late-onset or progressive PCHI? What are the identifiable risk factors which could be used to justify targeted screening? What is the extent of other potentially handicapping conditions (intrinsic or extrinsic) which might have implications for service provision and intervention? What is the current pattern of identification ages for PCHI? (see chapter 2)
- What are the outcome benefits attributable to early identification (e.g. in the first 12 months of life) of and/or early intervention for the effects of PCHI? Early identification itself is not an outcome measure but is often used as a surrogate for short- and medium-term outcomes. Since PCHI can have effects on a child's social, emotional, intellectual and communicative development, interest centres on the extent to which early identification/intervention can be shown to lessen such effects. Other longer-term outcomes (e.g. academic achievements, employment) and overall measures of quality of life may also benefit from early identification, and evidence is sought on these. Particular interest centres on the extent, if any, of additional outcome benefit associated with very early identification (e.g. in the first 6 months of life). The neural or cognitive factors (e.g. the existence of early critical periods for development) which might have implications for early identification and the psychological costs, if any, to child, parent or family, associated with very early

screening are also summarised. Evidence on the extent to which parents welcome early screening and identification of PCHI is examined. (see chapter 3)

- What is the current practice in the UK in relation to screening for PCHI at birth, at 6–8 months, and at school entry? (see chapter 4)
- What are the likely costs associated with existing screening programmes (universal neonatal, targeted neonatal and HVDT) when properly implemented? (see chapter 5)
- What are the beliefs and opinions of groups of consumers, providers, purchasers, and educationalists about early identification and screening? (see chapter 6)
- How efficacious (in 'ideal' conditions) and effective (in current practice) are the existing screens and screening programmes (universal neonatal, targeted neonatal and HVDT) for early identification of PCHI? In particular, what is the coverage, sensitivity, specificity, yield and age of identification associated with these screens? (see chapter 7)

The quality of evidence associated with these questions has been assessed. Good quality evidence has been fed into the review to allow a rational evaluation of screening options on the basis of that evidence, leading to evidence-based recommendations for service development (see chapter 9). Such recommendations should be judged on the basis of **efficiency** (providing patients with treatment and care which is both clinically effective and a good use of resources) and **responsiveness** (meeting the needs of individual patients). In addition to these measures, judgements of **equity** (improving the health of the population as a whole and reducing variations in health status) are highlighted in the *Priorities and Planning Guidance for the NHS* (NHS, 1996); any recommendations could, therefore, form the basis for a national programme for screening for PCHI (see chapter 9).

Finally, there remain areas where the evidence is equivocal or non-existent, and for which the major outstanding research requirements need therefore to be specified (see chapter 10).

Chapter 2

Epidemiology and public health perspective

Scope

A public health perspective on PCHI is presented in this chapter. Epidemiological data relating to the UK are presented and reviewed: the prevalence of PCHI as a function of severity, onset and demographics; the natural history of PCHI, that is, its onset and development; the major risk factors for PCHI; the differential aetiology of PCHI, and the current service indicators or proxy outcome measures, for example, the age of referral, confirmation and fitting of hearing-aids for PCHI.

Introduction

Cochrane and Holland (1971) suggest that screening is ethically different from other aspects of everyday medical care. They argue that for screening to take place there must be conclusive evidence that "screening can alter the natural history of the disease in a significant proportion of those screened". Holland and Stewart (1990) take this further, contending that "screening by itself can provide no answer to anything. Only if it is carried out efficiently and humanely, leads to an improved outcome in those concerned, and is properly monitored and evaluated should it be contemplated." In the survey of current practice presented later in chapter 4, the extent to which present screening is conducted efficiently and humanely is examined, together with the extent to which screening activity is monitored (audited), and an evaluation is made of present services.

In order to explore whether PCHI is a good target for screening, in the sense implied by Cochrane and Holland, it is crucial to understand its epidemiology and to know the outcomes of current service provision for PCHI. The importance of continuing care for PCHI is emphasised in *Figure 1*. For that continuing care to be cost-effective, services need to be accessed in the first place (for example, by screening or referral) and to be targeted at groups who would benefit most. The proxy measures used to indicate 'health' outcomes and access in the short term have been service performance indicators, such as age of referral for audiological assessment, age of confirmation of hearing impairment, age of hearing-aid fitting, and age at which an individual child's educational plan was enacted (see, for example, Joint Committee on Infant Hearing (JCIH), 1994; NDCS, 1994; 1996). These proxy measures are the somewhat arbitrary ages that have served as service indicators and as measures of outcome. It has not been possible to assess, even by proxy, whether services are provided for those who would benefit most.

Davis (1993a) provides an epidemiological model and public health context in which PCHI can be evaluated against the stringent criteria outlined above. Although there is presently very little that can be done in terms of primary prevention for PCHI, it is possible, by intervention for the child with impaired hearing and support for the family, to lessen the consequent disability and handicap that inevitably follows a lack of language and communicative development due to impaired hearing. Davis' model reflects this and the evidence is reviewed in chapter 3. Information on the appropriateness of screening in more general terms is provided in the present chapter.

Wilson and Jungner (1968) and Holland and Stewart (1990) review the principles of screening in general; the preconditions and principles needed for screening for PCHI are further explored by Davis and Sancho (1988) and Haggard and Hughes (1991; Haggard, 1993).

These principles and preconditions adapted for hearing screening are presented in *Table 1*, together with some comments on where evidence relating to each may be found in this review. Those aspects that relate to the epidemiology of childhood hearing impairment, namely the prevalence, severity distribution and natural history of the condition, are examined here.

Fortnum and colleagues (1997) review the recent literature concerning the epidemiology of PCHI. The need for a current, accurate and bias-free evidence-base in terms of prevalence, aetiology and risk factors is stressed. Research by this group has shown that there can be substantial differences in prevalence over districts and in the risk profiles of different populations (Davis & Parving, 1994; Davis *et al*, 1995) even when prevalence rates are broadly similar; this emphasises the need for a

TABLE 1 *The principles of screening adapted from Wilson and Jungner (1968) and Haggard (1993) for the case of screening for congenital hearing impairments*

Principle/precondition	Comments
1 The hearing impairment to be screened for should be an important health problem	Prevalence – see chapter 2; effects on life – see chapters 1 and 3
2 There should be an accepted rehabilitation means for cases of PCHI identified by the screen	See chapters 1, 3, 4 and 8. Knowledge of impairment regarded as of considerable value to parents
3 Facilities for assessment, diagnosis and rehabilitation should be available	See chapters 4 and 8
4 The hearing impairment should be recognisable at an early stage	Present at birth by definition
5 A suitable hearing screening test should be available at the proposed age for the screen (it should be quick, with good sensitivity, good specificity, and easy to interpret)	See chapter 7 for available tests
6 The hearing screening test should be acceptable to both child and parents	See chapters 3, 4 and 7 for tests used
7 The natural history of childhood hearing impairments should be known and understood	See chapter 2 for different aetiologies and onset patterns
8 There should be an agreed policy on whom to treat as patients with hearing impairment	Moderate hearing impairment and worse are target groups for screening and early intervention. See chapters 1, 8 and 9
9 The cost of hearing screening (including all assessments consequent on screening) should not be disproportionate to other healthcare costs incurred by a hearing-impaired child	See chapter 5 for costs of screening
10 Finding cases of childhood hearing impairment should be viewed as a continuous process	This underlies the Hall report philosophy for hearing screening – see chapters 3 and 4
11 The incidental harm of hearing screening programmes, e.g. stress to parents, should be small in relation to overall benefits	See chapter 3
12 There should be guidelines on how to explain results of hearing screening, together with transitional counselling support for those parents of children who have been screened and are concerned	See chapters 3 and 4
13 All hearing screening arrangements should be reviewed in light of changes in demography, epidemiology and other factors	The reason for this review
14 Costs and effectiveness of hearing screening should be examined in a stratified manner, and benefit maximised in each stratum	See chapters 2, 4, 5 and 7

local evidence-base to inform local policy. Previous studies of childhood hearing impairment reported that between 30% and 50% was of unknown (usually presumed genetic) aetiology (Parving, 1984; Newton, 1985; Davis *et al*, 1995; Parving, 1996). However, recent advances in molecular genetics (Steel, 1995) are yielding exciting possibilities for the determination of this large population of genetic impairments which will allow better evidence-based genetic counselling to be offered to families. Identifiable risk factors for permanent hearing impairment potentially enable screening and prevention to be targeted. Originally a list of some seven risk factors (JCIH, 1994), three criteria are now most commonly used: a stay of 48 hours or longer in an NICU/SCBU; a family history of permanent hearing impairment since childhood, and any abnormality of the head or face (Davis & Wood, 1992).

On the whole this earlier work has been carried out with small populations (Davis & Wood, 1992)

using solely clinic-based lists (Newton, 1985) or registers that were not uniformly ascertained across all levels of severity (Martin *et al,* 1979; Das, 1988; Dias, 1990; Shiu *et al,* 1996; Sutton & Rowe, 1997). Reviews of some of these data are also found elsewhere (Peckham, 1986; Davidson *et al,* 1989; Mauk & Behrens, 1993; Davis & Parving, 1994). The epidemiological data in this chapter are not presented as a review of our own previous work or that of other groups. This is because, on the one hand, our most recent published work has superseded our previous work, having a much wider scope and broader population base and, on the other hand, work from other groups has some of the faults mentioned above and was difficult to use because the required primary data were often unavailable in a form that was optimum for this review. Hence, we have opted to take the epidemiological data from the one large recent study that provides most of data required, consolidating and enhancing it with detail from other studies as appropriate.

Thus, recent work carried out at the MRC Institute of Hearing Research,[1] which is possibly the most extensive study of the epidemiology of PCHI in the UK, is reviewed here. The full details of the methods are described elsewhere (Fortnum *et al,* 1997), with only the essential aspects of the study being presented here. The results presented concentrate on hearing impairments that are at least moderate in nature (i.e. ≥ 40 dB HL in both ears). This is not to deny that milder impairments, either unilateral or bilateral, may have an effect on outcome measures such as quality of life and educational attainment, but rather these impairments are not presently legitimate aims for very early screening because:

(i) they do not constitute a serious public health hazard
(ii) they would be very costly to identify early compared to the benefit that might accrue
(iii) there is no evidence about whether intervention would be effective.

Case definition

All children with a permanent bilateral hearing impairment of ≥ 40 dB (average of 0.5, 1, 2 and 4 kHz, if available) born between 1 January 1985 and 31 December 1993, and currently living in the area covered by the Trent Regional Health Authority, were the target for the study conducted between 1994 and 1996. The Trent Region has a population of about 5 million, with ethnic minorities totalling 5%, and an annual birth cohort in the region of 61,000 per annum between 1985 and 1990. It may be considered reasonably typical of the UK in demographic terms. However, in terms of service provision for hearing impairment, it may be slightly atypical, due to the higher-than-average proportion of audiological staff in the region despite lower-than-average NHS expenditure overall. This makes the area ideal for an ascertainment study on PCHI but, in terms of some of the service provision data, it may give a slightly optimistic account.

Search strategy

Multiple sources were used to minimise the number of missed children. Ethical approval to search the records in community child health, audiology and education departments was obtained, when required. Information was extracted on demographics, timing and results of hearing screens and assessments, relevant medical history including possible risk factors and associated disabilities, and stated aetiologies with a measure of certainty. The risk factors were the most difficult data to obtain, and it appeared initially that there were far fewer children in the study period with a history of NICU/SCBU of more than 2 days than in other studies. This problem was addressed in a number of ways, not all of which have, as yet, been finalised. The data on NICU/SCBU and family history are therefore interim and will change over the next few years; currently they provide a picture of the number of hearing-impaired children with such histories that is probably on the low side.

Analysis

The children in the 1991–93 cohort would have been aged between 21 months and $4^{1}/_{2}$ years when data collection finished (September 1995). Examining the data using a GLIM (or general linear) model with Poisson error distribution, the number of children born in these later 3 years was found to be significantly smaller than other cohorts and thus would give a substantial underestimate, if used.

[1] The work was partially funded by the Trent NHS Research Scheme and the final report (Fortnum *et al,* 1997) was accepted by Trent after peer review. The *British Journal of Audiology* has accepted a paper for publication based on this report.

Therefore, prevalence rate data for the cohort of 487 hearing-impaired children born between 1985 and 1990 are presented, and the 166 children born between 1991 and 1993 are omitted. Confidence intervals (CIs) for prevalence have been calculated using the appropriate logistic 95% CIs. The audiometric data used was the most recent audiogram, using an age-appropriate method. For most of the children in the population born between 1985 and 1990, this would have meant a fairly accurate pure-tone audiogram. If younger children had been included there would have been much greater variation in assessment of severity.

Prevalence of hearing impairment

All data presented here have been aggregated over the 11 districts in the Trent Region and over the birth cohorts for 1985–90, since there were no substantial secular trends in the prevalences (Fortnum *et al*, 1997).

The data given in *Table 2* show the prevalence of PCHI, that is, a hearing impairment that is considered to be permanent irrespective of origin. It includes those children with sensorineural and permanent conductive impairment, for example, bilateral atresia. Children with mixed pathologies were included provided that the estimate of their permanent impairment was ≥ 40 dB HL on the better hearing ear. These children form the major group who require their hearing impairment to be detected at the earliest opportunity for early intervention. The data overall give the prevalence for all types of onset and represent the best estimate of prevalence in children aged between 3 and 9 years. The congenital hearing impairment group are those children who are presumed to have had a hearing impairment pre- or perinatally. Assignment into this category is undertaken retrospectively from case notes from the audiology clinic or other sources. Unfortunately, assignment into this category is usually due to an absence of evidence for an acquired hearing impairment rather than to the presence of such evidence. This category may, therefore, be overstated. The third category is that of 'acquired' hearing impairment. This category is actually three groups combined into one and includes:

(i) the group with postnatally-acquired hearing impairment, for example, children who have had meningitis
(ii) children with progressive hearing impairments, usually diagnosed as such because their hearing impairment post-assessment had deteriorated; however, the status of their hearing in the neonatal period is unknown; for example, it could have been mildly, moderately or severely impaired, but not recognised
(iii) 'late-onset' childhood hearing impairment, usually diagnosed when a child is assessed as hearing-impaired but with no evidence of progression; however, there is some evidence that the child may have been able to hear earlier.

TABLE 2 *The cumulative prevalence (with 95% CIs) of PCHI as a function of severity in 10 dB bands. Data are presented for all PCHI, congenital and 'acquired' groups, and refer to a population of 487 children with permanent hearing impairment born between 1985 and 1990 and residing in the Trent Region in 1994/95*

	Overall		Congenital		Acquired	
Severity (dB HL)	**Prevalence per 100,000**	**95% CI**	**Prevalence per 100,000**	**95% CI**	**Prevalence per 100,000**	**95% CI**
≥ 40	133	122–145	112	101–123	21	17–26
≥ 50	110	100–121	90	81–100	20	15–24
≥ 60	81	72–90	64	56–72	16	12–21
≥ 70	59	52–67	47	41–55	12	8–16
≥ 80	47	41–55	37	31–44	10	7–13
≥ 90	35	29–42	28	23–33	8	5–11
≥ 100	24	19–29	18	14–22	6	4–9
≥ 110	11	8–15	9	6–12	2	1.3–4
≥ 120	4	3–7	3	2–6	1	0.4–2

The uncertainty over the onset of deafness certainly extends both ways and few studies have looked at this in depth (see, for example, Stevens *et al,* 1991) in a sizeable population. Even in prospective studies it is often difficult to separate out the groups.

The data in *Table 2* are compiled from 487 children with valid audiograms showing the better hearing ear to have an averaged hearing impairment (over the frequencies 0.5, 1, 2 and 4 kHz) of at least 40 dB HL. At the other extreme there were just 17 children with a hearing impairment of at least 120 dB HL (13 congenital, four acquired) from a birth cohort of 366,480 children. The overall prevalence rate of PCHI shown in *Table 2* is highly dependent on the severity of the impairment and the time of onset. Overall, there are 133 (95% CI, 122–145) per 100,000 children with hearing thresholds of at least 40 dB HL (1 in 752 children). This decreases to 110 per 100,000 (1 in 909) at ≥ 50 dB HL, to 47 per 100,000 (1 in 2127) at ≥ 80 dB HL, to 24 per 100,000 (1 in 4167) at ≥ 100 dB HL, and to 4 per 100,000 (1 in 25,000), with a 95% CI of 3–7 per 100,000, for total/profound hearing impairments at ≥ 120 dB HL. The range 40–120 dB HL is not a homogeneous category, with different management options at each broad level of impairment and consequent, variably expressed, disability.

For congenitally hearing-impaired children, the cumulative prevalence rate distribution given in *Table 2* suggests that the best estimate for congenital PCHI ≥ 40 dB HL is 112 per 100,000 children (95% CI, 101–123) or 1 in 893 children. Again, this decreases with increasing severity to 90.5 per 100,000 (95% CI, 81–100) (1 in 1105) at ≥ 50 dB HL, to 37 per 100,000 (1 in 2703) at ≥ 80 dB HL, to 18 per 100,000 (1 in 5555) at ≥ 100 dB HL, and to 3 per 100,000 (1 in 33,333) at ≥ 120 dB HL.

The discrete distribution of the prevalence rate for PCHI is shown in *Table 3* for congenitally hearing-impaired children and those with 'acquired impairments' as a function of three severity bands (moderate, severe and profound[2]). It can be seen from *Table 3* that for moderate congenital hearing impairment the prevalence rate is 64 per 100,000, which is more than the severe and profound categories combined. However, this relationship does not apply for 'acquired' impairments, with 23% of profound impairments being 'acquired' (mainly as a result of meningitis). Overall, 16% (95% CI, 13–19) of impairments were thought to be 'acquired'.

TABLE 3 *The discrete prevalence (per 100,000 children) of three broad severity categories of PCHI as a function of onset (congenital versus 'acquired') for the birth cohort 1985–90 in the Trent Region*

	Severity (dB HL)	Prevalence per 100,000	95% CI
Congenital	40–69 (moderate)	64	56–73
	70–94 (severe)	23	19–29
	≥ 95 (profound)	24	20–30
'Acquired'	40–69 (moderate)	9	7–12
	70–94 (severe)	5	3–8
	≥ 95 (profound)	7	5–10

Published data on what proportion of children actually **acquire** impairments compared with those in whom they are congenital are scarce. The Sheffield group of researchers has examined this longitudinally (Stevens *et al,* 1991; 1997), and has followed-up all the children tested in a targeted neonatal screen. They showed that, at the 5-year follow-up point, of 24 children fitted with hearing-aids, with permanent hearing impairment of at least 50 dB HL, 14 had been detected by the neonatal screen. This gives a figure of about 42% (95% CI, 24–62) for the percentage of impairment that may be 'acquired'. The group examined here were mainly at-risk children; however, in the overall Trent data such a higher prevalence of acquired impairment in the at-risk children was not seen; indeed, quite the opposite was found. Thus, for children who were in NICU/SCBUs the proportion of 'acquired' cases was 12% (95% CI, 7–19); for those with a family history it was 10% (95% CI, 6–17); and for those with no risk factor it was 22.6% (95% CI, 18–29). If those with a true acquired impairment, mainly caused by meningitis, are excluded then the proportions become 8.8%, 5.6% and 11.2%. Thus, although the overall

[2] Whenever the terms moderate, severe or profound hearing impairment are used in this report they always refer, unless specifically delimited, to the ranges identified in *Table 2,* viz. the average of the thresholds for frequencies 0.5, 1, 2 and 4 kHz in the better ear subdivided at 40–69, 70–94 and 95+ dB HL.

proportion of 'acquired' (that is, true postnatally-acquired, progressive and late-onset) impairments was 16%, the proportion with progressive and late-acquired impairments appears to be about 10%. If we adhere to the recommendations that children with meningitis should be referred for assessment (Fortnum & Davis, 1993; NDCS, 1996), then a neonatal screen would possibly 'miss' about 10% of the children with hearing impairments at ages between 3 years and 9 years. However, it is possible that a screen at 6–9 months of age would also miss the majority (maybe three-quarters) of these children if it is assumed that onset is uniformly random.

Taking the prevalence estimates derived from Trent Region (Fortnum *et al*, 1997), the number of children who might be expected to be hearing-impaired in the UK can be calculated. The data for the number of live births in 1994 (OPCS, 1996) for the UK have been convolved with the cumulative prevalence rates from *Table 2* and are presented in *Table 4*. This shows that we might expect to find just under 1000 hearing-impaired children in the UK per annual birth cohort with at least a moderate hearing impairment. Just under 84% of the hearing-impaired population of children with at least a moderate impairment will probably have a congenital hearing impairment.

Thus, UNS will have the potential to find 840 children per year with congenital hearing impairment in the UK, if the screen is aimed at those with at least a moderate hearing impairment. There must, however, be some provision for case-finding for the remaining 160 per year (see NDCS, 1996). If the target for screening is all children with hearing levels > 50 dB HL at birth (see NDCS, 1994), then a screen would have the potential to detect 675 children at birth. Case-finding would then have to locate up to 323 children at some stage, preferably before age 2 years. The estimate of the number of children with congenitally severe and profound hearing impairment born each year in the UK is about 353. If we take the UK as a whole, with about 128 Health Districts, then the mean number of congenitally hearing-impaired children per District per annum is 6.6, with 2.8 being severely or profoundly impaired and, of these, 1.4 would be profoundly impaired. Thus, evaluating a screening programme in the way that Holland and Stewart (1990) suggest is really quite difficult on a District basis and appropriate aggregation is required. This can be achieved either in a temporal sense or by widening the geographical basis. Obviously the latter would give more timely information, while local relevance would be supplied by the former.

TABLE 4 *The estimated number of children with PCHI in the UK per annual birth cohort, estimated from the 1994 birth cohort, using prevalence data from* Table 2, *showing congenital and acquired combined and congenital alone*

Severity of hearing impairment (dB HL)	All PCHI	Congenital PCHI only
≥ 40	998	840
≥ 50	825	675
≥ 60	608	480
≥ 70	443	353
≥ 80	353	278
≥ 90	263	210
≥ 95	233	180
≥ 100	180	135
≥ 110	83	68
≥ 120	30	23

The data in *Table 5* have been disaggregated to show England, Wales, Scotland and Northern Ireland data separately; the table shows the overall number of children per year who might have PCHI by ages between 3 years and 9 years for each country and for a range of severity. An approximate 95% CI is derived and the number of hearing-impaired children for England alone is estimated to be in the range 767–912 per year.

Data in the same format are presented in *Table 6* but for congenital PCHI only. This shows that for England alone, it is estimated that there will be 704 children per year (approximate 95% CI, 635–774). At ≥ 50 dB HL, this would decrease to 566, at ≥ 70 dB HL to 296, and at ≥ 95 dB HL to 151. In Wales, Scotland and Northern Ireland, the number of expected congenital PCHI patients is quite small – 69 for Scotland, 40 for Wales, and 27 for Northern Ireland.

Although the range of hearing impairments presented here is wide, this is necessary to understand what the implications might be for setting severity criteria in different types of screening programme. If a screen uses transient evoked otoacoustic emissions (TEOAE), it is fairly insensitive to the degree of hearing impairment over and above 30 dB HL (see, for example, Lutman *et al*, 1997). It may therefore detect most of the moderate or worse impairments. However, if a more targeted approach is used, for example, with an ABR test or with a

TABLE 5 *The estimated number of children (with 95% CIs) with PCHI in the four countries of the UK per annual birth cohort estimated from the birth cohort of 1994 using the prevalence data from* Table 2, *showing congenital and acquired combined*

Severity of hearing impairment (dB HL)	England	Wales	Scotland	N Ireland
≥ 40	837 (767–912)	47 (43–51)	82 (75–89)	32 (30–35)
≥ 50	692 (629–761)	39 (35–43)	68 (62–75)	27 (24–29)
≥ 60	509 (453–566)	29 (25–32)	50 (44–56)	20 (17–22)
≥ 70	371 (327–421)	21 (18–24)	36 (32–41)	14 (13–16)
≥ 80	296 (258–346)	17 (15–19)	29 (25–34)	11 (10–13)
≥ 90	220 (182–264)	12 (10–15)	22 (18–26)	9 (7–10)
≥ 95	195 (151–245)	11 (8–14)	19 (15–24)	8 (6–9)
≥ 100	151 (120–182)	8 (7–10)	15 (12–18)	6 (5–7)
≥ 110	69 (50–94)	4 (3–5)	7 (5–9)	3 (2–4)
≥ 120	25 (19–44)	1 (1–2)	2 (2–4)	0 (0–2)

TABLE 6 *The estimated number of children (with 95% CIs) with congenital PCHI in the four countries of the UK per annual birth cohort estimated from the birth cohort of 1994, using the prevalence data from* Table 2

Severity of hearing impairment (dB HL)	England	Wales	Scotland	N Ireland
≥ 40	704 (635–774)	40 (36–44)	69 (62–76)	27 (25–30)
≥ 50	566 (509–629)	32 (29–35)	56 (50–62)	22 (20–24)
≥ 60	403 (352–453)	23 (20–25)	39 (35–44)	16 (14–17)
≥ 70	296 (258–346)	17 (15–19)	29 (25–34)	11 (10–13)
≥ 80	233 (195–277)	13 (11–16)	23 (19–27)	9 (8–11)
≥ 90	176 (145–208)	10 (8–12)	17 (14–20)	7 (6–8)
≥ 95	151 (120–182)	8 (7–10)	15 (12–18)	6 (5–7)
≥ 100	113 (88–138)	6 (5–8)	11 (9–14)	4 (3–5)
≥ 110	57 (38–75)	3 (2–4)	6 (4–7)	2 (1–3)
≥ 120	19 (13–38)	1 (0–2)	2 (1–4)	0 (0–1)

behavioural test then the level for which the test is set, for example, 50 or 80 dB HL, will determine how many children might be detected and how many would have to be identified in different ways at a later date.

Although there are only about 700 children per year born with congenital PCHI in England, of whom 151 may be profoundly deaf and, hence, candidates for a cochlear implant at a young age, if the number of children of school age (from approximately 5 to 15 years of age) are considered, there are about 11,200 hearing-impaired children and about 2400 with profound deafness. The commitment to educational provision for that number of children is quite large, given that the children will be spread throughout the country. The British Association of Teachers of the Deaf (BATOD) survey (M Eatough: personal communication, 1996) conducted in 1994 found that there were about 1700 qualified teachers of the deaf (ToDs), about 200 untrained teachers and over 800 classroom assistants working with hearing-impaired children. The total of annual salaries for the teachers was in excess of £65 million.

As part of the BATOD survey, the ToDs were asked to give aggregated numbers for 'deaf

and hearing-impaired children'. These data are a potential source of validation for the prevalence estimates that we have derived. In *Figure 2*, the number of hearing-impaired children from the 1994 BATOD survey for England alone are shown as a function of age and also severity. There are some limitations to these data because a proportion of the children were not classified in respect of either age or severity of impairment. However, the trend is clear, showing that the number of hearing-impaired children known to ToDs rises during the first 5 years of life, is asymptotic for the next 9–10 years, and then declines again. This is to be expected, as the number of children with hearing impairments known to the health services may only increase slowly for the first 5 years, and those that drop out of education are 'out of the school system'. If an average is taken over the middle years that is equivalent to the 1985–90 birth cohort, then the BATOD survey found a mean annual figure of 845 children with moderate impairment or worse. This compares closely with our estimate of 837, thus providing some validation of the data from the Trent Region on prevalence of hearing impairment, allowing generalisations, as in *Tables 5* and *6*, to be made with some confidence.

Risk factors for hearing impairment

There could be many factors that predispose towards a high risk of congenital PCHI. The JCIH (1994) has developed several individual factors, and Gerber (1995) has summarised some of the major factors that influence PCHI. However, many of these risk factors can be summarised under a small number of headings (Davis & Wood, 1992; Davis, 1995a; Shiu *et al*, 1996; Sutton & Rowe, 1997):

(i) history of NICU/SCBU for 48 hours or longer
(ii) family history of permanent childhood deafness
(iii) cranio-facial abnormality (CFA) noticeable at birth.

Fortnum and colleagues (1997) discuss these risk factors, their distribution among the congenital PCHI population and their prevalence in the population. Overall, in Trent, 59% (95% CI, 54–64) of the congenital PCHI population have one or more of these three risk factors. Taken individually, 29% (95% CI, 25–33) have NICU/SCBU histories, 31% (95% CI, 26–35) have family histories, and 12% (95% CI, 9–15) have a CFA. The percentage with CFA decreases to about 3.7% when the other two factors are taken into account; hence, history of NICU/SCBU and family history are the two major risk factors that need high coverage in a targeted screening programme. Obtaining a high coverage for both these risk factors entails considerable effort to ensure that, on the one hand, all appropriate NICU/SCBU babies are tested before discharge back to referring hospital, for example, and that, on the other, the babies with a family history are located and tested before discharge home.

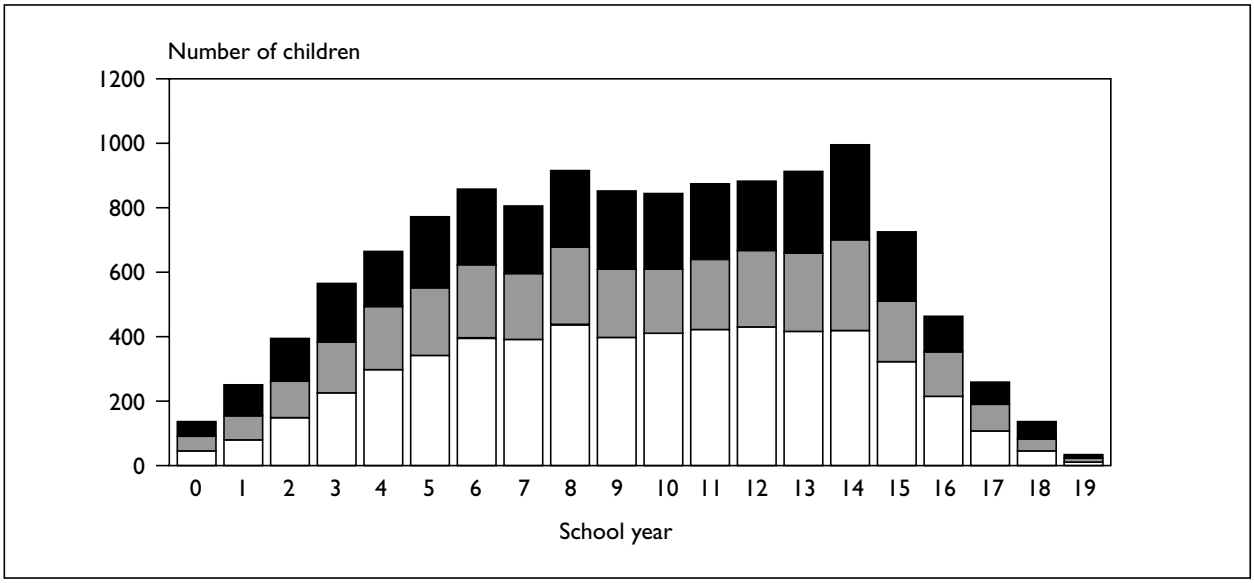

FIGURE 2 *The approximate number of children who are deaf or hearing-impaired known to the education authorities in England as a function of age and severity. Data are from the BATOD survey, 1994 (Note: Some children are omitted because they were not classified.) (■, profound; ▩, severe; □, moderate)*

One problem with these data is that they are retrospective and gathering the data from different sources, for example, parents, health services and education services, gives different results even for the first of these risk factors. It is likely that the data underestimate the prospective risk factors by a small percentage for both the NICU/SCBU factor and for family history. Shiu and colleagues (1996) have suggested a figure for the three risk factors that is somewhat higher but there seems to be general agreement that the percentage of congenital PCHI children with at least one of these three factors is near to 60%. However, if these three risk factors are used to affect a 100% sensitive neonatal screen we would not expect to achieve a yield of 60% of congenital PCHI, because there are operational problems in identifying those with a significant family history of hearing impairment. There are two parts to this problem. The first is that the number with a family history known at the time of, or before, birth is lower than the number who report a family history after confirmation of hearing impairment (Wood *et al,* 1995). The second is that there needs to be an effective system of registering the risk factor in the maternity unit and acting on it. For these reasons, it is probable (Fortnum *et al,* 1997) that the upper limit of a targeted neonatal screen would be a 45–50% yield of the expected number of congenital hearing impairments, when programmes are running effectively.

There were three sub-populations in Trent where there were significantly raised odds ratios. These were the NICU/SCBU, family history and the Asian sub-populations. The prevalence estimates for overall PCHI (40 dB HL or greater) were 301 per 100,000 live births with an NICU/SCBU history, 733 per 100,000 live births with a family history, and 249 per 100,000 live births for an Asian ethnic background. The prevalence rate for those children without an NICU/SCBU history or a family history was 54 per 100,000 live births, reducing to about 47 per 100,000 live births allowing for ethnic background as well.

The increased odds ratios are shown in *Figure 3,* on a log scale, for the three risk factors, taken separately, as a function of severity of hearing impairment. From this figure it can be seen that the increased risk for NICU/SCBU history is between 4.4 and 7.1, for family history between 10.6 and 20.5, and for Asian background between 1.5 and 2.6. In addition to these risk factors, there was also evidence for a socio-economic component to the risk factors, with those living in more deprived areas having a raised risk of PCHI, but the effect was small compared with the other risk factors (Shiu *et al,* 1996; Sutton &

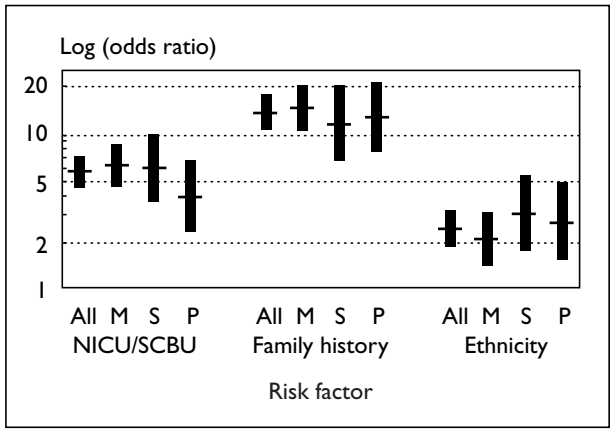

FIGURE 3 *The increased odds ratio (with 95% CI) for three major risk factors associated with PCHI. The risk factors are NICU/SCBU history, family history of childhood deafness and ethnicity (Asian). The odds ratios are given for all PCHI, and then separately for moderate (M), severe (S) and profound (P) hearing impairment severity categories*

Rowe, 1997). The assumptions made here should be explicitly acknowledged (see Fortnum *et al,* 1997, for a detailed discussion) and are discussed briefly here.

Firstly, we have used the whole of the NICU/SCBU population in determining this risk, despite the operational risk factor being NICU/SCBU for 48 hours or more. The NICU/SCBU population in Trent was 10.7% of live births. In previous work (Davis & Wood, 1992) in Nottingham, the NICU/SCBU population was much smaller – about 7% – and about a further 2% were discharged back to maternity wards before 48 hours. The prevalence and odds ratio derived here is therefore an underestimate (perhaps by a factor of between 1.5 and 2, that is, the upper range of the odds ratio 95% CI). The reported population with a family history of childhood deafness was estimated from the work of Wood and colleagues (1995) to be in the region of 4.5% of the total live birth population, based on questionnaires to mothers in maternity units. This population, as was shown, depends on the extent and type of questioning involved. So, although the prevalence and increased odds ratios are estimated (and given 95% CIs) there is a considerable potential for bias to occur, depending on the definition of family history. At present there is no reason to suppose that it is much less than the estimates provided above, which are the first attempt to characterise the scale of the risk involved. There is also considerable scope for bias in interpreting the ethnic background risk factor. Ethnicity was taken from case notes, where entered, and from the names of the children (using a community paediatrician with expert local knowledge). The scope for under-inclusion was great. Thus, the estimates

we have derived should be treated with caution, perhaps as lower bounds for the true odds ratios and prevalences.

Aetiology of hearing impairment

It is difficult to make a judgement about the major aetiologies of PCHI in prospective studies, and in retrospective studies this is even more true. It is particularly difficult if more than one factor is thought to be responsible. The major aetiological classification suggested by Davidson and colleagues (1989) has been that used in most studies in the 1990s. The categories adapted for use here, in *Table 7*, are genetic, prenatally acquired, perinatally acquired, postnatally acquired, CFA, other (e.g. chemotherapy) and missing. The major problem with most studies is that there are many children who do not have an ascribed aetiology. In Trent (Fortnum *et al*, 1997), 41% of the PCHI population do not have an aetiology. This is not atypical; a review of this area has been undertaken by the EU Concerted Action on Hereditary Deafness (HEAR), who are concerned with the genetics of deafness, and this comes to a very similar conclusion (see Parving, 1996). If the aetiology is imputed from other data, for example, NICU/SCBU history, family history or CFA, then the percentage who have no aetiological information at all decreases to about 25%.

The overall percentage of children falling into each of seven 'aetiological' categories are presented in *Table 7*. Overall, 40% of children have been ascribed to have a genetic aetiology (Davis *et al*, 1995) but the largest group is the missing category. When this 'missing' group is reassigned to the probable imputed category, based on the risk factor information, then the data for the two birth cohorts, 1985–90 and 1985–93, are in very close agreement. Overall, about 45% of PCHI can be ascribed to genetic aetiologies, of which half have a family history. Also within this genetic group are one in five with a history of NICU/SCBU of more than 48 hours. This highlights the problem of aetiological attribution, and it is by no means certain if both are important contributing factors or not for some children. Viewed from the risk factor perspective, about one in three hearing-impaired children with an NICU/SCBU history may also have a genetic predisposition to hearing impairment.

There is a difference in aetiological classification between the congenital group and those who acquired their hearing impairment at a later date. There are more genetic and perinatally attributed aetiologies, but also more missing data. The proportion of genetic aetiologies was 49% (of whom 10% had an NICU/SCBU history) and of perinatal aetiologies was 17%. One-third of prenatal cases were caused by rubella, which gives it about a 1.2% prevalence within the PCHI group, many times less than children born 30 years ago (Martin *et al*, 1979; Davis *et al*, 1995). The next congenital group of children who should be the target for further

TABLE 7 *Classification of aetiological groups for children with PCHI in the Trent Region born 1985–1993, as a function of birth cohort, and separately for those who are thought to be congenitally impaired and those who are not (see text for how data are imputed). The figures in each column are percentages in each group*

	Overall % 1985–93 (n = 653)	Overall % imputed 1985–93 (n = 653)	Overall % imputed 1985–90 (n = 487)	Congenital % imputed 1985–93 (n = 556)	'Acquired' % imputed * 1985–93 (n = 97)
Genetic	39.7	44.7	44.6	48.2	24.7
Prenatal	3.7	4.0	3.7	4.0	1.0
Perinatal	6.7	16.7	16.4	17.6	11.3
Postnatal	6.1	6.0	6.2	0.0	41.2
CFA	1.2	2.5	1.4	2.9	0.0
Other	1.7	2.0	1.4	1.0	3.0
Missing	40.9	24.6	26.3	25.7	18.6

* *'Acquired', as defined here, includes impairments that are progressive or late-onset, hence the inclusion of aetiologies other than postnatal.*

primary prevention is the perinatal group, the majority of whom have had NICU/SCBU care. However, from the data shown here, the major cause of PCHI is genetic, particularly that linked to familial childhood deafness. The major public health emphasis should therefore be in terms of early identification to prevent major developmental disability and handicap. In time, with the discovery of those genes that may be responsible for PCHI (Steel, 1995), the emphasis may change. However, such work is at present clearly basic science and has little input to the public health priorities for congenital deafness and hearing impairment.

Additional disabilities

Many children with hearing impairment have additional disabilities. Overall, 39% of PCHI in the Trent Region was associated with another disability. Two from three of those with additional problems had more than one additional problem, that is, there were multiple disabilities. The largest group of disabilities were additional cognitive deficits (36% of those with additional disabilities). Visual problems were identified in 10% of all children with PCHI and about 13% had a systemic disorder as well as their hearing impairment.

One of the major risk factors for another disability was NICU/SCBU history, as might be expected. Thus, 45% of all children with additional problems also had a history of NICU/SCBU intervention, while 60% of hearing impaired children with an NICU/SCBU history had at least one other clinical or developmental problem, 30% of those with no major risk factor, and only 20% of those with a family history of hearing impairment had additional problems.

There were 89 children with a named syndrome – about 14% of all the hearing-impaired children. However, there is obviously scope to view children with additional clinical and developmental problems in the same manner because, at the lowest level, a named syndrome is a systematic collection of pathological indicators.

Children with clinical and developmental problems are an important consideration when planning a screening service because they must have access to an appropriate screening test and their management should be coordinated according to the nature of their additional disabilities.

Proxy outcome measures

The lack of short- and mid-term outcome measures, coupled with the need to obtain meaningful timely feedback on how well screening and intervention programmes are performing, mean that the service indicators that are available should be examined carefully (for example, NDCS, 1994). These include indicators such as the age at which children are referred for assessment and the percentage of children that are referred. The results that relate directly to the performance of screens are detailed later (see chapter 7). Here, four indicators (the ages of referral for appropriate audiological assessment, confirmation of hearing impairment, prescription and fitting of hearing-aids) are presented, together with the major factors that affect these healthcare indicators. The cumulative distributions of the four indicators are shown in *Figure 4*.

Table 8 shows that the median age of referral for all PCHI was 10.4 months, confirmation of hearing impairment was 17.1 months and age of hearing-aid fitting was 26.3 months. It was clear from the analysis (Fortnum *et al,* 1997) that the major, and highly significant, factor affecting the distributions of all indicators was the severity category of hearing impairment. From *Figure 4* it can be seen that the distributions are highly skewed and non-normal, so the data were transformed (log transform) before analysis of variance. Apart from severity of hearing impairment, there were no factors that systematically affected all indicators. An ENT operation significantly delayed the median age of hearing-aid fitting by up to 6 months for those with severe impairments, and this was a cause for concern. There was also substantial variation between Districts. This emerges not so much in the rate at which the first 40–50% of hearing-impaired children are referred and fitted with hearing-aids but in the tails of the distribution. The quality of services is probably therefore better monitored not by the median but by the higher percentiles such as the upper quartile. It is worth noting in *Table 8* that the variability as shown by the standard deviation is much lower for the profoundly impaired group, becoming significantly larger for the severely impaired and largest for the moderately impaired, for all four measures.

As is shown in *Figure 4,* the referral rate distribution for all hearing-impaired children is in two parts. The first part of the curve is fairly steep and is followed by a shallower curve indicating a slowing down of referrals. The age of confirmation of hearing impairment lags behind the age of referral

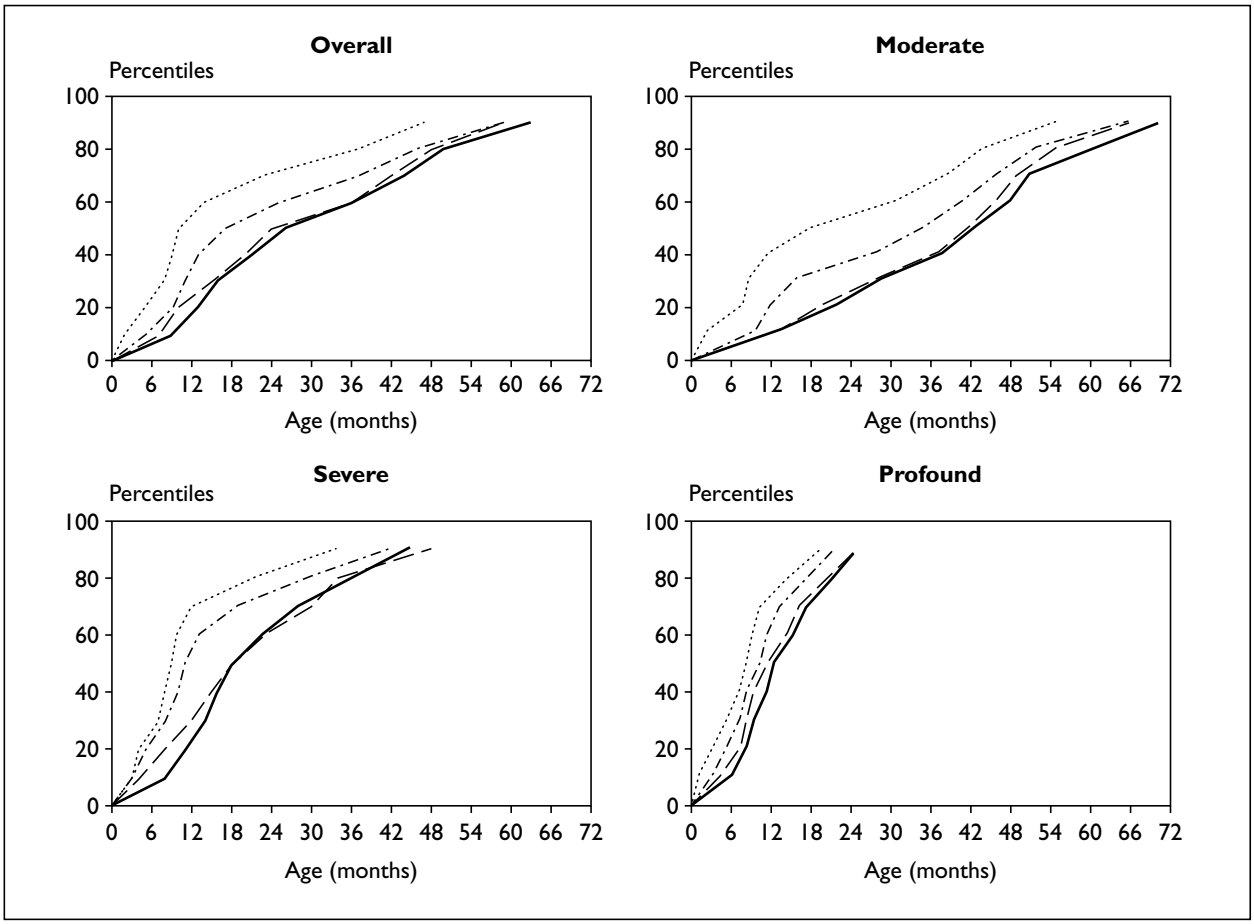

FIGURE 4 *Distribution of age at referral (n = 284), ..., confirmation of permanent hearing impairment (n = 309), – · – · –, prescription of hearing-aid (n = 223), — — —, and fitting of hearing-aids (n = 336), ——, for children born 1985–90 with presumed congenital onset (n = 350) who were born in their District of residence. The four panels show the overall data and separately for moderate, severe and profound. Age of hearing-aid fit was imputed from age of prescription if not available (see Fortnum* et al, *1997 for details)*

TABLE 8 *The mean, median, standard deviation (SD) and quartiles of the distribution of ages of referral, confirmation, prescription and hearing-aid fitting as a function of severity of hearing impairment for children with congenital PCHI, who resided in their District of birth*

Indicator	Severity	Mean age (months)	SD	Number (n)	25th percentile (months)	Median age (months)	75th percentile (months)
Referral	All	18.8	18.1	284	6.8	**10.4**	30.8
	Moderate	25.5	20.5	155	8.5	**18.0**	41.8
	Severe	13.4	12.7	61	5.6	**8.7**	17.0
	Profound	8.6	6.2	68	4.1	**7.6**	10.7
Confirmation	All	26.0	20.8	309	9.9	**17.1**	41.6
	Moderate	34.7	21.4	181	13.0	**35.1**	48.6
	Severe	17.1	15.8	56	7.4	**11.2**	22.8
	Profound	11.1	6.7	72	6.8	**9.7**	15.6
Prescription	All	30.3	20.6	223	12.7	**24.4**	45.7
	Moderate	40.1	19.7	127	22.3	**42.3**	50.8
	Severe	22.4	16.3	44	10.1	**17.7**	31.5
	Profound	12.8	7.4	52	7.7	**11.0**	17.4
Fitting	All	32.3	21.3	336	14.0	**26.3**	47.5
	Moderate	42.3	20.5	195	25.0	**43.2**	55.7
	Severe	23.5	15.7	67	13.2	**17.7**	32.6
	Profound	13.9	7.0	74	9.0	**12.0**	17.7

by about 12 weeks at the lower quartile but the delay increases as the distribution gets more extreme. It is interesting to compare all three levels of severity of hearing impairment. For profound impairment the rate of referral is very high, with 75% of referrals being completed by 11 months. The HVDT does not appear to have an effect on referrals at 6–9 months, which suggests the children are identified by professional and parental concern in the first year of life. However, for severe impairments the upper quartile is only reached after 17 months and for moderate impairments it takes a very long time, 41.8 months. The change in slope of referral comes at about 70% for severe impairment. This reflects the incremental yield (Haggard, 1993) from the HVDT, which is quite marked, ceasing at about 12 months. It is not until later, when these children are noticed to have delayed language, that the remaining 30% start to trickle in for hearing assessment. There is a small change in the rate of referral for moderately impaired children due to the HVDT. However, the slope is much shallower than that for the severe and profoundly impaired, and is a trickle of referrals most of the time.

The difference between referral, confirmation and hearing-aid fitting is small for the profound, larger for the severe, and very large for the moderate impairment groups. This reflects partly the urgency for those profoundly impaired and also the problems in assessing accurately the severely and moderately impaired children. For children with a hearing impairment in excess of 50 dB HL, the current wait for hearing-aid fitting is longer than recommended (see NDCS, 1996). It is not the impetus of screening that lies behind the incremental yield patterns, it is the severity of the hearing impairment, with the profoundly impaired cases being similar to good service provision and moderate cases being similar to poor service provision. The challenge is to lift the severely and moderately impaired detection rate to a level similar to that for the profoundly impaired. It should be recognised that the paediatric audiological services in the Trent Region are regarded as having been reasonably well developed and quite considerable effort has been put into making the HVDT an effective screen. Hence, these data can be thought of as being towards the better end of the quality distribution.

How do these data compare with the NDCS targets (NDCS, 1994; Hall, 1996), which derive in some ways from those used previously in the USA (JCIH, 1994) but also arise from the desire to introduce targeted neonatal screening? The NDCS target is to have a confirmed assessment of bilateral permanent hearing impairment of at least 50 dB HL for 40% of cases by 6 months of age and for 80% by 12 months of age. For the birth cohort 1985–90 only 13.8% (95% CI, 10.0–18.7) in the Trent Region had an assessment confirmed by 6 months, and for the birth cohort 1989–93, 25.3% (95% CI, 19.6–31.8). The latter figure will decrease slightly as the children in the latter part of the birth cohort who were missed (particularly the moderately impaired) by the age of 24 months are discovered later. For the 12-month target, the figures for 1985–90 were 41.5% (95% CI, 35.5–47.7) and for 1989–93, 57.7% (95% CI, 50.7–64.5). These performance indicators are nowhere near the targets, which is not surprising as only three of the 11 Districts had targeted neonatal screening as a service, and that only since about 1990. However, if the data are analysed with respect to severity of hearing impairment, then there is an interesting but expected result, as shown in *Table 9*. In the 1985–90 cohort, at 6 months about 20% of the profoundly and severely impaired group have been identified but only 5% of the moderately impaired. In the later cohort, the data appear better but time will tell how far the 17% figure for the moderately impaired group will fall. Again, in the 1989–93 cohort, the 12-month target is almost met for the severely and profoundly impaired groups but, for the moderately impaired group, it is nowhere near being met, although the proportion found in this group has increased substantially. However, as always, there is need for caution because of the long time-span in finding all those with moderate impairment. On the benefit side, as discussed later in chapter 7, the introduction of targeted neonatal screening does seem to benefit the age of identification substantially, especially those with moderate impairment.

These data suggest that there needs to be continued aggregated monitoring of the health service performance indicators, such as ages at referral, identification, prescription and hearing-aid fitting, so that changes in service patterns can be monitored. However, there is ample evidence that the service is under-attaining the NDCS targets and, indeed, it confirms that it is rare for a moderately impaired child to be 'diagnosed' by age 12 months. Only one in four moderately hearing-impaired children has hearing-aids by the age of 24 months, and only one in four severely hearing-impaired children by the age of 12 months. Although these data are the best that can be used, nevertheless, they should be treated with caution because they reflect the services in place in the late 1980s, and

TABLE 9 *The percentage of children with PCHI who had had a confirmed audiometric assessment ('diagnosis') by age 6 or 12 months, as a function of birth cohort and severity of hearing impairment*

	6-month NDCS attainment target = 40%			12-month NDCS attainment target = 80%		
	%	95% CI		%	95% CI	
Birth cohort 1989–93						
Moderate	16.5	11.3	23.7	39.6	31.8	47.9
Severe	30.8	19.8	44.5	69.2	55.5	80.2
Profound	36.2	23.8	50.7	74.5	60.2	84.9
Birth cohort 1985–90						
Moderate	5.0	2.6	9.3	21.0	15.7	27.5
Severe	19.6	11.2	32.0	57.1	44.0	69.3
Profound	22.2	14.1	33.2	65.3	53.6	75.3

may not accurately reflect the effectiveness of services since 1994. However, it must be said that there were three major, fairly mature, targeted neonatal screening programmes introduced in the Trent Region during the late 1980s.

Opportunity for neonatal screening

The coverage of the HVDT and of UNS is discussed in the survey of current service provision in chapter 4 and later in chapter 7. One of the challenges that UNS would have to face would be to achieve a coverage approaching 95%. While services are piecemeal, children who move from one District with one set of screening programmes to another with a different set will need to be catered for in any proposal. The Trent study showed not only that 2.9% of hearing-impaired children have moved District within the Trent Region but also that 9.2% had transferred into the Region. Some transfers into the Region were from other parts of the UK but a sizeable minority were from outside the UK. The needs of children transferring into a District, in terms of screening, therefore need some careful thought, especially as to whether there might be existing information systems that might help with this problem. Of course, if a uniform screening system was used country-wide the problem would be minimised.

A second problem for UNS coverage are those babies who might leave hospital before the opportunity to screen occurs. There has been increasing interest from a cost-effectiveness point of view, and also from parents, in discharging mother and child home earlier and earlier. The number of babies born in England and Wales in the financial years 1990/91 to 1993/94, the last year for which data are available, are shown in *Table 10;* also shown are the number of children discharged on the day of birth, 1 day after birth or 2 days or more after birth. In 1993/94, there were just over 50,000 children discharged on the first day. The expected trend towards earlier discharges and decline in numbers staying in hospital for 2 days or more are shown in *Figure 5*. There are no data available on the pattern of discharges, for example, whether certain times of the day/week are more likely to lead to earlier discharges. Health Districts conducting UNS have found a higher false-alarm rate for the earlier tests and there have been some problems over coverage. This is discussed later but further research is needed to establish the exact pattern of discharges and, hence, the best staffing and testing arrangements that UNS might need to obtain good coverage and an acceptable false-alarm rate. A more detailed pilot study carried out in Nottingham showed that for two maternity units, over a period of 4 weeks, only 18 of 800 babies were discharged on the same day as birth. However, some Districts may have marked deviations from

TABLE 10 *The number of children discharged from hospital after 0, 1 and 2+ days between 1990 and 1994, from routine activity analysis supplied by the Department of Health*

	Numbers of children discharged from hospital at		
Days from birth to discharge	**0 day**	**1 day**	**2+ days**
Financial year			
1993/94	52,543	15,1273	412,172
1992/93	44,492	13,5159	416,409
1991/92	36,984	12,3017	435,313
1990/91	35,817	11,6430	484,811

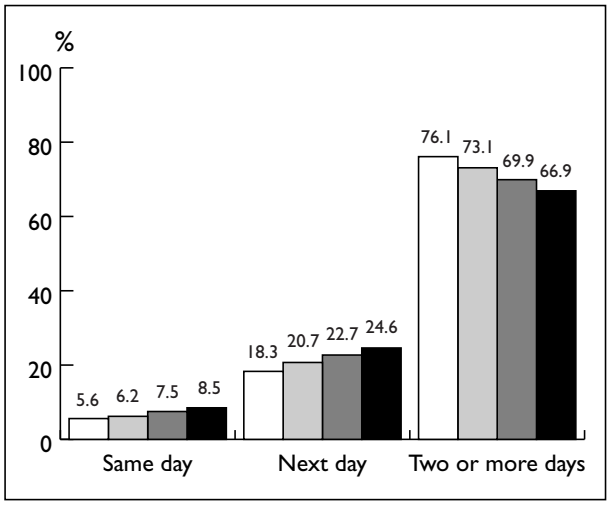

FIGURE 5 *Percentage of babies discharged at different times* post-partum *(□, 1990; □, 1991; ■, 1992; ■, 1993/94)*

the national figures. For example, although the number of home births is small nationally (about 1.4%), in some areas, such as Bath, more than one in three babies may be born outside District hospitals (T Williamson: personal communication, 1997).

Public health implications

The data reviewed here can be used to inform an effective screening programme for bilateral PCHI in the country as a whole and in individual Districts. The target for this screening programme would be children with congenital hearing impairments ≥ 40 dB HL. These children constitute the majority of those with PCHI and the prevalence rate is about 110 per 100,000 live births. If only those with 50 dB HL or greater were considered, the prevalence would be about 90 per 100,000. Taking the former prevalence rate, this equates to a detection rate of 6–7 children per year in an average District. For a maternity unit with 3000 births per year, the expected number with hearing impairment is very small (i.e. three children) and the variance year on year is consequently very large. At a District level, therefore, PCHI is rare and there is quite a strong argument for some form of service aggregation to ensure that the quality of the screening and follow-up services are adequate. This is because the skills needed are specialised and should have appropriate use rates and quality control. This applies across all the professional areas involved in screening and follow-up, for example, screening, health visitor surveillance (HVS), audiological assessment, hearing-aid fitting, ENT involvement, educational support, family support, speech-language therapy. In contrast, at a local level, because of the need to integrate services from these diverse providers, the whole of continuing care for PCHI might be considered as a habilitative package. Thus there might be considerable benefits in aggregating resources over the traditional boundaries of health and education (and social services).

Most congenital PCHI does not appear to have a progressive component; however, when available medical records are consulted, 16% of PCHI is either postnatally acquired (e.g. after meningitis), progressive or late-onset. The best estimate for the two latter categories is about 10% of all PCHI (although it should be remembered that Stevens and colleagues (1991; 1997) found a much higher figure, most of which seems to occur after the 6–8-month screen). There is no evidence that these impairments selectively occur in the first year of life and they do not, therefore, indicate a considerably better yield from a screening system based in the first days of life compared to one in the period of 6–8 months. Finally, there is no evidence that the overall prevalence of congenital hearing impairment has changed over time (Davis, 1995b; Fortnum *et al,* 1997) or that it is changing rapidly.

Thus, in terms of prevalence, congenital PCHI is rare but, with 840 new cases expected each year in the UK, it is an important health problem for the country as a whole (see *Table 1,* Screening principle 1), because of its potentially substantial effects on the development of language (see chapter 3) and its impact on educational outcomes for affected children (see chapter 1). As well as being an important public health issue, it should be considered a priority, in the sense that the data reviewed here (and in the current practice chapter) show how inadequate the services have been in the recent past in detecting substantially disabling congenital hearing impairment by 1 year of age, which is broadly considered to be an important aim (NIH, 1993a; JCIH, 1994; NDCS, 1994; NDCS, 1995; NDCS, 1996; Hall, 1996). Most profoundly deaf children do have a quicker passage through the health system, so the present task is to improve the services for children with severe and moderate hearing impairments (see *Table 1,* Screening principle 14).

The distribution of PCHI is well understood, the natural history less so. The data from Stevens and colleagues (1997) show a much higher proportion than other estimates for those with progressive or late-onset impairments, although their data are derived solely from at-risk cases. However, two larger studies (Lutman *et al,* 1997; Fortnum *et al,* 1997)

show a smaller proportion, which implies that a majority (e.g. 80–90%) of all moderate impairments or worse could be detected in the first week or the first year of life (see *Table 1,* Screening principle 7).

The two major risk factors, NICU/SCBU history and family history, that presently drive the majority of targeted neonatal screening programmes in the UK (see chapter 4) are found in 55% of all congenital hearing impairments. When combined with the presence of a CFA at birth, the three risk factors cover almost 60% of the target population. However, problems of implementation, particularly for the family history risk factor, mean that effective yield from targeted screening may be in the region of 45–50%. (This is supported later in chapter 4 (see also, Fortnum *et al,* 1997; Wood *et al,* 1997; *Table 1,* Screening principle 14.))

In assessing the appropriateness of screening activity it is important to consider also the aetiology of PCHI in order to see the scope for primary prevention. An outline consideration of the aetiology of congenital hearing impairment suggests that health investment for primary prevention would not have great benefit at the present time, given that the majority of impairments are considered to be genetic in origin. Health investment should be in a screening system (including appropriate genetic counselling) that not only can conform with the 14 MRC principles of screening, but also takes the guidance of Holland and Stewart (1990) into consideration in providing a screening programme that is conducted "efficiently and humanely, leads to an improved outcome in those concerned, and is properly monitored and evaluated".

Chapter 3

Early intervention and outcomes

Scope

In this chapter the evidence from studies on the effects of early identification on outcomes for children with permanent hearing impairments are reviewed. In addition, the implications of work on sensitive periods for development are considered and evidence on parental attitudes to neonatal screening reviewed.

Early intervention

Screening programmes for the identification of PCHI, specifically the HVDT and SES, were introduced in order to lower the age of identification of PCHI. The justification for this is that early identification tends to improve outcomes for hearing-impaired children in a number of domains and that parents want identification to be early. The evidence for these assertions is assessed in this chapter.

The literature on the potential benefits of early intervention for children with disabilities has grown rapidly in recent years. Nevertheless, there are still few high quality studies on early intervention for congenital hearing impairment which permit reasonably unequivocal data interpretation. Many studies are limited by small sample sizes, a lack of controls and the ambiguous use of terminology (e.g. variable definitions of 'early' and of categories of hearing impairment). In addition, there is variability in determining appropriate outcome measures and inadequate specification of the interventions used. These inconsistencies combine to make it difficult to separate out the effects of **early** intervention from those of the intervention itself. The key studies in this area are listed and summarised in *Table 11*.

TABLE 11 *Outcomes of early intervention*

Study (year)	Sample characteristics	Report type*	Statistical analysis	Outcomes measured	Definition of 'early'	Comments
Downs (1995)	69 children; all impaired, from neonatal screening programme. Earlier sample from Yoshinaga-Itano *et al.* (1996) – see below.	G	None reported	Minnesota Child Development Inventory	2 months (habilitated before 3 months).	Earliest identified had best scores compared with norms. This more marked in 'severely' hearing impaired, but no statistical verification. Lack of definition of population.
Eilers & Oller (1994)	131 infants, range of hearing impairments – mainly severe to profound.	R	Appropriate	Canonical babbling; age of onset	As early as possible.	Infants with normal hearing babbled earlier than those who were deaf; age at hearing-aid fitting related to onset of babble.
Feinmesser *et al* (1982)	65 children; 26 profound, 27 severe, 12 moderate.	R	None	Educational placement	7–9 months.	No late-intervention 'control' group.
Imai (1983)	24 children, hearing impairments > 70 dB HL; education started from 12 months.	R	None	Speech sounds	Fitting from 2 months onwards.	Purely qualitative with no control data.
Markides (1986)	153 children; severe to profound hearing impairments; 4 groups matched across range of variables.	R	Appropriate	Speech intelligibility	Aided at less than 6 months compared with those aided later.	Earliest intervention judged to have best speech by teachers.
Musselman *et al* (1988)	118 children, severe to profound hearing impairments.	R	Appropriate	Language	Earliest habilitated at less than 24 months (48%).	Raises possibility that effects of early intervention may only be short-lived.

* *G, general review, see edited volume or chapter; R, published in peer-reviewed academic journal; P, preprint.*

continued

TABLE 11 contd *Outcomes of early intervention*

Study (year)	Sample characteristics	Report type*	Statistical analysis	Outcomes measured	Definition of 'early'	Comments
Musselman *et al* (1988)	139 children, severe to profound hearing impairments; ages 33–100 months during study.	R	Appropriate	Language, education, social development	Uses median split – age of intervention before or after 24 months.	Found that type of intervention programme interacted with age of intervention.
Musselman & Kircaali-Iftar (1996)	20 children.	R	Appropriate in part	Speech	Groups similar on age of fitting.	Worked from outcome backwards, i.e. children with good and poor speech compared on other characteristics.
Parving (1992)	138 participants (hearing-aid sample).	R	None	Schooling	Not applicable.	Shows that children identified earlier more likely to be found in schools for deaf.
Parving & Christensen (1993)	288 participants (hearing-aid sample).	R	Appropriate	Employment	Not applicable.	Those with congenital hearing losses more likely to have manual jobs, whereas those with acquired losses more likely to be 'professionals'. Problem: no data for hearing individuals reported, so comparisons difficult.
Ramkalawan & Davis (1992)	16 children; ages 27–79 months; mild to profound hearing impairments; hearing parents.	R	Appropriate	Spoken language	Continuous variables of ages of intervention, earliest 2 months.	Earlier intervention brings linguistic benefits.
Ramkalawan (1997)	33 children; ages 32–85 months; mild to profound hearing impairments; hearing parents.	P	Appropriate	Spoken language and communication	Continuous variables, earliest 2 months.	Unpublished PhD – more complex interactions found than Ramkalawan & Davis (1992).
Ruben *et al* (1982)	72 children.	R	Appropriate	Speech and language quotient (SLQ)	Diagnosed at 5.3 years on average and fitted with aids at 6.3 years.	Disadvantaged black and Hispanic Americans showed average SLQ delay corresponding to over 3 years.
Robinshaw (1995)	10 infants; severe to profound hearing impairments; hearing parents; matched controls with normal hearing; studied from 6 to 21 months.	R	None	Gestural and vocal communication	Aided between 3 and 6 months.	Early-aided infants showed slightly delayed but similar development to normally hearing infants. Lack of late-aided controls.
Robinshaw (1996a)	10 infants; severe to profound hearing impairments; hearing parents; matched controls with normal hearing; studied from 6 to 21 months.	R	None	Gestural and vocal communication	Aided between 3 and 6 months.	Progression of communicative development in deaf infants might be similar to that of hearing infants if aiding early enough. Lack of late-aided controls.
Robinshaw (1996b)	1 child; profound hearing impairment; cochlear implant at 2 years; hearing parent.	R	None: single case study	Spoken language	Not applicable.	Details of communicative development of child with implant.
Vernon & LaFalce-Landers (1993)	57 participants; IQs of 130+; hearing levels not specified; aetiologies detailed; some participants had 'additional problems'.	R	None	Education, employment, mental health	Not applicable.	Intellectually-gifted deaf persons do not have positive outcomes despite gifted status. Lacks control group. Results confounded by inclusion of participants with 'other additional problems'.
Yoshinaga-Itano *et al* (1996)	109 children; mild to profound hearing impairments; all enrolled in same intervention programme; hearing parents.	P	Appropriate	Vocabulary expressive and receptive language	Before 6 months.	Interim report showing clear benefits for early identification; detailed protocol and analysis not yet available.

* *G, general review, see edited volume or chapter; R, published in peer-reviewed academic journal; P, preprint.*

In the most pertinent literature the outcome benefits of early intervention derived from neonatal hearing screening are considered. To date, there are two such reports from the USA (Downs, 1995; Yoshinago-Itana *et al,* 1996). In a short account of the neonatal screening programme in place in Colorado, Downs (1995) asserts that a group of infants habilitated before 3 months of age scored 87% of normal on the expressive language section of the Minnesota Child Development Inventory. This performance compared favourably with that of children habilitated between 3 and 12 months, who scored 66% of normal. The results suggested that children with severe hearing impairment showed the greatest benefit in expressive language when habilitated early. Trends in this direction were also reported for all other sub-tests of the Inventory, although these were non-significant.

Yoshinaga-Itano and colleagues (1996) compared the language abilities of hearing-impaired children whose hearing impairments were identified before 6 months of age (n = 46) with those identified after 6 months of age (n = 63). The children in the study had confirmed bilateral hearing impairments ranging from mild to profound, had hearing parents and were all enrolled in the same early intervention programme. Unfortunately, no information is provided on the actual ages of identification of the later identified group. Measures of vocabulary size and of expressive and receptive language were compared between the two groups (early versus late identification). Non-verbal cognitive skills were controlled using analyses of covariance. The results indicated that the children identified early had significantly larger lexicons than the children identified later, along with significantly better expressive language scores and comprehension scores ($p < 0.05$). Although the study was cross-sectional rather than longitudinal in design, a suggested trend towards increasing group differences over time was apparent.

A study by Markides (1986) considered the speech intelligibility of 153 children as subjectively assessed by their teachers. Children were stratified into four groups:

(i) those children fitted with hearing-aids before 6 months
(ii) those fitted with hearing-aids between 6 and 12 months
(iii) those fitted with hearing-aids in their second year of life
(iv) those fitted with hearing-aids in their third year of life.

Groups were matched for age, sex, age at onset of deafness, degree of hearing loss and schooling. Teachers rated the speech intelligibility of their pupils using a 7-point scale representing the range 'normal speech' to 'no speech'. The speech intelligibility of the first group (fitted by 6 months) was reported to be significantly superior to that of any of the other groups. This study provides relatively strong evidence that early intervention may be related to outcome benefits for hearing-impaired children, at least in the domain of speech intelligibility. However, it should be noted that teacher ratings of speech intelligibility may be subject to bias effects which could have been avoided by using naive listeners.

These three studies point to the potential benefits of identification and intervention received within the first six months of life. Studies of very early development in normal populations provide further, though indirect, support for the importance of very early intervention. Kuhl (1994) and colleagues (1992) have investigated the early development of categorical perception and the perception of native-language phonology. Using a standard behavioural dishabituation paradigm, Kuhl and colleagues (1992) conditioned infants to turn their heads discriminatively in response to computer-synthesised variants of the vowel sounds English /i/ and Swedish /y/. They found that, as early as 6 months of age, infants were responding more accurately and frequently to sounds from their 'native' language; that is, the infants were able to differentiate between phonemes from their own language and a foreign language. This suggests that, during the first 6 months of life, infants have already begun to distinguish language sounds, having become 'tuned' to those they hear spoken around them.

In a study on the spoken language of a group of hearing-impaired children, Ramkalawan and Davis (1992) provided evidence that language can be affected by age of intervention. The authors studied children with a range of hearing impairments (mild to profound) and ages of intervention (2–36 months). Age at hearing-aid fitting ranged from 11 months to 54 months. Video-recorded samples of the children's language were analysed to consider the effect of age at detection, referral, first appointment, and hearing-aid fitting on a range of spoken language measures. The authors found that, controlling for age, "the lower the age of intervention the better the outcome measures for language". A further study by Ramkalawan (1997) indicated that a complex interaction of factors – including earlier referral for hearing

assessment – influenced spoken language production for the hearing-impaired children in her study.

Eilers and Oller (1994) compared hearing-impaired and hearing infants longitudinally to assess the onset and development of canonical babbling. While the normal infants initiated well-formed babbling at no later than 10 months of age, the hearing-impaired infants did not begin to babble until 11 months of age, many of them somewhat later. In addition, age at hearing-aid fitting, which ranged from 2 months to 30 months, was found to be significantly related to the age of babbling onset ($r = 0.68$).

Sutton and Stokes (1994) raise an issue about the interpretation of data from early intervention studies. They argue that the benefit of early hearing-aid fitting is demonstrated only if it is **additional** benefit. That is, in the case of the study by Eilers and Oller (1994), additional benefit from the early use of hearing-aids would only be shown if a baby who was fitted with a hearing-aid at a young age babbled after a shorter interval than a baby fitted at a later age, all other things being equal. In such a case, the slope of the regression between age at onset of babbling and age at hearing-aid fitting would be greater than unity.

In a re-analysis of their data, Eilers and Oller (1994) showed "...a relatively constant relationship between age of fitting of a hearing-aid and onset of canonical babbling". They concluded that Sutton and Stokes' suggestion that earlier fitting might not result in earlier babbling was confirmed. On average, it appeared that, regardless of the age at hearing-aid fitting, it took about 8–10 months for canonical babbling to appear in the children studied.

Despite Sutton and Stokes' argument, it is difficult not to see such results (i.e. a regression slope of unity) as evidence in favour of early intervention. Eilers and Oller make the point that "anything that can be done to provide the child with an earlier opportunity to babble is likely to help reduce cumulative deficits. This is the only sense in which we argue for the benefit of early fitting of a hearing-aid, but it is based on the best developmental models we know and it is logically independent of the statistical issue raised [by Sutton and Stokes]."

In a detailed examination of the habilitation of a small number of profoundly hearing-impaired children identified and aided early (between 3 and 6 months), Robinshaw (1996a) and Robinshaw and Evans (1995) present data which suggest that it is possible for some deaf children to follow a normal pattern of communicative development provided that appropriate and timely intervention is received. Robinshaw (1996b) presents a case-study of a cochlear-implanted child, known as Adam. She asserts that his progress after implantation at 2 years of age, although slightly delayed, was in line with that of hearing infants in quality. Adam's adaptation to the post-aural hearing-aids that he had used prior to implantation was poor, but after implantation and switch-on he was able to use his own speech feedback to moderate his speech output. By 22 months **hearing age** (i.e. time from cochlear-implantation), Adam was able to deal with some voicing contrasts and could differentiate between fricatives. After 2 years of experience with the implant device, he had reached the same level of phonological competence and performance that his hearing cousin had reached at the same hearing age.

In studies looking at the verbal and non-verbal interaction between infants and care-givers, Robinshaw (Robinshaw, 1995; Robinshaw, 1996a) found that language acquisition (whether spoken or signed) for hearing-impaired children aided by 6 months of age followed a similar course as that for hearing infants (matched for age, sex and cultural background). The author compares these findings with a similar study (Tait, 1987) in which hearing-impaired infants who were not aided early (after 24 months) showed significant language delay.

Robinshaw suggests that care-giver attention is particularly important, since the stimulation provided by the care-giver can offset hearing problems to some extent – fluctuations in the hearing levels of the infants in her studies were accompanied by changes in care-givers' sensitivities to the infants' language skills. Therefore, the interaction between care-givers and hearing-impaired infants must be appropriate for the learning situation to be effective. Although the lack of a control group fitted with hearing-aids at a later age limits the conclusions of these studies, Robinshaw's work provides evidence that at least some infants who are severely and profoundly deaf can, given appropriate care-giver stimulation after early hearing-aid fitting, develop language and communication abilities in a similar progression to hearing infants. However, the data are insufficient to determine whether or not children fitted with hearing-aids early continue to exhibit less delay in language development than those fitted with hearing-aids later. The Eilers and

Oller argument is sufficient to justify the need for early intervention: it is beneficial to facilitate communicative development as early as possible, otherwise cumulative delays, and hence deficits, are likely to occur.

The results of one study presented in *Table 11,* (Musselman *et al,* 1988) suggest that the benefits of early intervention might actually be short-lived. In a piece of longitudinal research on the effects of hearing impairment on language, Musselman and colleagues followed-up early habilitated children (0–36 months) at ages between 3 years and 9 years. The children were given a battery of tests for intelligence, receptive and productive spoken language, communicative competence with the mother and social development. The effect of age of intervention on spoken language production was not found to be significant but the effect on receptive language was. There were no effects of age of intervention on mother–child communication or on social development.

However, although age of intervention was significantly associated with receptive language in the first year of testing, by the time the children had been administered their final assessment, after 3 years, the effect had disappeared. This has important implications for studies on the potential benefits of early intervention, suggesting that it may produce short- to mid-term rather than long-term gains. The data are far from conclusive. Musselman and colleagues suggest that "it is possible that long-term benefits do result from early intervention, but that the measures used ... have not been sufficiently sensitive to detect them". Furthermore, since the outcome benefits are likely to depend on the quality of habilitation as well as the age of intervention, a weakness in their study is the failure to examine the qualitative aspects of the programme under which habilitation was effected and the nature of parental involvement. As suggested earlier, this is a weakness of a number of studies and indicates an urgent need for basic research to develop systems for coding and including in analyses the type, extent, range and quality of intervention provided for hearing-impaired children and their families.

The recognition that **type** of habilitation and intervention is as important as age of intervention is addressed in further longitudinal work by the same authors (Musselman *et al,* 1988). Children enrolled into their study were educated in one or more of a number of programmes: a hospital programme (where parental instruction was emphasised), home visiting (which was also heavily parent-oriented), itinerant programmes (provided within regular schools), segregated classes and provincial schooling (including both segregated and integrated schooling). Hearing loss, intelligence, age of intervention and language variables were considered. It was found that the type of programme in which children were enrolled had a significant effect on their speech reception. Children in individual programmes were found to achieve better scores than those in group programmes. However, the results were found to be modified by significant higher order interactions, thus confusing the picture. Further analysis showed that, while those with profound hearing impairments scored higher in individual programmes, severely hearing-impaired children fared best in group educational situations. Age of intervention also showed a similar interaction with programme-type on four language measures: late-starters in individual programmes scored better than those in group schemes, whereas early-starters showed the reverse pattern. The authors say, "Unequivocal statements about the value of particular approaches or the consequences of not following one approach or another are unwarranted".

While there are benefits which accrue from early intervention and the strengths of particular programmes, the matching of habilitation to child is not easy, and the success of early intervention must depend, at least in part, on dedicated intervention. It should be noted, however, that studies such as this one may be compromised by the selection biases which result from the non-random allocation of children to particular intervention programmes. In addition, as with other studies, the problem may arise that those children whose hearing impairments are identified later may be identified late precisely because they do better than their earlier-identified hearing-impaired peers. These potentially biasing factors are difficult to disentangle.

Musselman and Kircaali-Iftar (1996) compared deaf children stratified into two groups according to speech skill – those determined by assessment to have 'good' speech skills and those with 'poor' speech skills. Although the sample size was small (ten in each group), the groups were matched for a range of other factors, including IQ. Good speakers were found to be more likely to wear ear-level aids, whereas poor speakers tended to wear body aids, although the average age at initial hearing-aid fitting was similar for both groups (about 22 months). The main difference found was the higher level of education in

mothers of good speakers. Although a model to explain the differences between good and poor speakers is offered by the authors, age of identification, hearing-aid fitting or habilitation does not feature. However, this does not suggest that early identification did not affect speech outcomes, because the groups were not significantly different in terms of age of identification. What this study does suggest is that, when age of identification is controlled, other factors may affect outcome. Unfortunately, regression analyses relating age of identification to outcome were not performed in this study.

These studies have used measures of speech, language and communication as the outcome measures of interest, reflecting the widely-held view that measures in these domains are of crucial importance (a view supported by parents, see Gregory, 1995). Arguably, however, these outcomes are stepping stones towards longer-term outcomes such as quality of life, psycho-social adjustment, academic achievement and employment. Studies focusing specifically on these longer-term outcomes are sparse and subject even more to intervening biases which are difficult to control.

Some investigators have used school placement as a surrogate outcome measure, arguing that good communication skills are more likely to give rise to a mainstreamed placement. Again, this ignores the large number of other important variables which may affect decisions on placement. A follow-up study of 65 deaf infants initially tested between 7 and 9 months at clinics in Jerusalem (Feinmesser *et al,* 1982) *(Table 11)* showed that severely and profoundly deaf infants who were aided early were successfully mainstreamed in school. Although the absence of a late-aided control group prevents pertinent comparisons, it was argued that the satisfactory mainstreaming of severely and profoundly deaf infants was a success for the early screening system used.

At a Japanese centre, Imai (1983) reported good language development in a small group of early-aided infants receiving good educational follow-up. Examining the quality of speech sounds, it was found that the mainstreamed children had better word intelligibility scores than children attending a school for the deaf. Again, however, selection biases in school placement are not accounted for and probably confound findings. Parving (1992) conducted a study of 138 hearing-impaired Danish children showing that age of intervention is **not** a good predictor for later type of schooling. Parving argued that referral to a particular type of school depends largely on the child's preferred method of communication. She found that attendees of schools for the deaf were identified earlier than those in mainstream classes.

Two studies have looked at the long-term outcomes of hearing-impaired children into adulthood. It is probable that these would have been identified relatively late, given the services available at the time. Parving and Christensen (1993) found that those individuals with an early-acquired or congenital hearing loss were more likely to work in manual occupations than those with a late, progressive loss. The latter group were also found most likely to be university educated. Vernon and LaFalce-Landers (1993) showed that even intellectually-gifted deaf people are at a disadvantage compared with their gifted but hearing peers. In their American sample, 30% of gifted deaf people (characterised by IQs of 130 or more) were found to be unemployed, and 40% had required some form of mental health intervention. It is worth noting, however, that the study lack detailed audiological profiles for the subjects, some of whom are described as 'deaf-blind' and 'multiply handicapped'.

We can conclude that, although the evidence on early identification is limited and complex, there is a definite indication that, in terms of language and communication outcomes, earlier identification may be beneficial. It is reasonable to conclude that deferred identification might result in cumulative delays and/or deficits. The reviews of the literature which have been written in the last two decades concur with this summary. Meadow-Orlans (1987), for instance, concludes that early intervention is beneficial, even though most of the studies discussed in her work involved intervention at about 3 years of age compared with late habilitatory commencement at about 5 years.

Bess and Paradise (1994a) have argued against the trend for UNS in the USA. One of their central claims is that there is insufficient evidence to justify neonatal screening in terms of outcomes and therefore it should not be carried out. However, responses to the published article voicing this perspective (i.e. letters in a later issue of *Journal of Pediatrics*) clearly challenged this view, citing much of the evidence that has been discussed here.

Robinette (1994) pointed out that Bess and Paradise argue in favour of early identification and intervention for at-risk babies, even though evidence for beneficial outcomes of early identification is not derived from this particular population. In a further reply, Bess and Paradise

(1994b) admit that "...unarguably, all infants with handicapping degrees of hearing impairment should ideally be identified as early as possible".

Sensitive periods for language acquisition

Evidence relating to the existence of sensitive periods for language development provide another area of insight into benefits that may be derived from early intervention for PCHI. A sensitive (or critical) period consists of a specific period during which an organism maximally responds, or shows heightened sensitivity, to aspects of the external environment in relation to some feature of its development. It has been defined by different experimenters in various ways, with the main emphasis being that external sensory stimuli have an important effect on the formulation of neural connections. Eggermont produced the following definition of 'critical period' from a combination of more restricted definitions:

> "...a period during which the action of a specific stimulus is required for normal development of the system, and during which the organism is maximally vulnerable to environmental manipulation" (Eggermont, 1986).

Critical periods of this kind have been extensively researched and have been demonstrated to be a common feature of sensory mechanisms in a number of animals. In particular, studies on the visual system (Weisel & Hubel, 1963; Blakemore, 1978) have demonstrated how deprivation of specific stimuli can result in the retarded development or abolition of certain developmental features. More recently, similar but subtler (experience-sensitive) mechanisms have been demonstrated to exist for the auditory system (Rubel, 1985). However, it has remained difficult to extrapolate from animal studies, and thus it has been more difficult to establish experience-sensitive periods of this kind for humans. Nevertheless, these findings have often been used to support the suggestion that there exists some similar restricted time-frame for elements of human development.

More recently it has been suggested that staggered development of different brain regions occurs, resulting in different sensitivities and maturational time courses for developmental features (Greenough *et al*, 1987). Various time-frames have been posited in relation to speech and language acquisition processes. Some researchers have suggested that the first 2 years of life constitute the 'sensitive period' for language development. As such, it has been suggested that auditory deprivation within the first 2 years may impact most significantly on aspects of language and cognitive development (Webster, 1983). The NIH consensus statement (1993a) proposes that the first 3 years of life are generally regarded as 'the most important period for language and speech development'. Others propose that developmental changes occurring between ages 2 years and 4 years are of particular importance (Corballis, 1991), while some investigators propose periods of sensitivity which more closely parallel the protracted period of postnatal neural development observed from birth to puberty (Neville, 1991).

Certainly some behavioural evidence from cochlear implant studies suggests that a sensitive period for spoken language development may not be so narrowly defined, as some success is being demonstrated in spoken language perception and production with profoundly hearing-impaired individuals, born deaf, who receive implants in late childhood (Summerfield & Marshall, 1995). In addition, while the development and progress of spoken language in children implanted at older ages might not be as swift as that observed in children implanted at younger ages (Tye-Murray *et al*, 1995), results are still encouraging.

While it appears that information from the linguistic environment is an essential element for normal language acquisition to occur, and that our ability to acquire a first language diminishes with age, the length of the period of sensitivity, and whether or not it is the same for all individuals, is undetermined. The potential existence and length of such a period (or periods) obviously has important consequences when first language acquisition is considered for hearing-impaired children. The question of how such a sensitive period might vary depending on the modality of language development (i.e. spoken/signed/both) is also of importance, although a study conducted by Newport (1990) suggests that early exposure to sign language (before age 6 years) is essential if it is to develop proficiently. If the period of sensitivity were to be narrowly defined as being confined to early childhood, as has been suggested, greater emphasis would need to be placed on early detection and diagnosis of hearing impairment. This evidence is, however, far from consistent or conclusive.

The first language acquisition of sign by deaf children, or by hearing children born to deaf parents, has shown itself to be similarly subject to experiential sensitivity. Neville (1991) provided

evidence pertaining to the influence of early experience on language and cognitive processing. She showed that acquiring sign language early as a first language, for both deaf and hearing subjects born to deaf parents, resulted in left cerebral asymmetry for the detection of the direction of motion. This asymmetry was opposite to that found for hearing non-signers (where the right hemisphere mediated the detection of motion direction). Thus early language experience demonstrably influences cerebral development and specialisation.

The implications of a sensitive period for spoken language acquisition in aided hearing-impaired children lacks detailed evaluation. This information may be invaluable in determining the optimal period from which a child might benefit from aiding and intervention (or from implantation, should that be considered a viable option). More urgently, there is a need to assess the potential benefits and the time of maximum benefit for hearing-impaired children who wear conventional hearing-aids and who constitute the majority of hearing-impaired children. Particular emphasis may need to be placed on those children with moderate-to-severe hearing impairments for whom conventional aids are potentially of greatest benefit. Even for those with more severe hearing losses, the evaluation of the possible benefits of receiving hearing-aids, cochlear implants and/or early exposure to sign language is essential.

Neural plasticity, the auditory system and language

Lenneberg (1967) proposed that the acquisition of language is governed by a critical period which relates to the maturation of other developmental systems. Coupled with the probable existence of critical or sensitive periods for aspects of development, is the concept of plasticity. For individuals with hearing impairment, issues relating to the development and plasticity of the auditory system and how this might impact on experiential sensitivities for language acquisition are of relevance. Some of these issues are briefly reviewed below.

The auditory system is cochleotopically organised, that is, a projectional map exists whereby the cochlea is topographically represented by structures of the central nervous system. These maps may be retained, and possibly maintained, by neural processing. Both normal developmental, and abnormal sensory, experiences can result in changes to these maps (the cochlea map and the neural map) (Rubsamen, 1992; King & Moore, 1991). These types of changes within the auditory system are what are commonly referred to as 'plasticity'. Tsukuhara (1981) provides a general definition of plasticity as:

> "...any persistent change in the functional properties of single neurons or neuronal aggregates" (cited by Irvine and Rajan, 1995, p.351).

Black (1995), however, provides a more flexible and broader definition in proposing that plasticity:

> "...refers to brain mutability and flexibility, which underlies alteration of structure and function over time in response to environmental change" (Black, 1995, p.5).

Black further specifies that this plasticity rests on molecular and cellular determinants which, in turn, fundamentally underpin **all** aspects of cognitive development.

Neural plasticity is not restricted to early development and is a feature that can be observed in adult animals. As such, a large body of the experimental work on neural plasticity has been conducted on adult animals (see Robertson & Irvine, 1989; Rajan *et al,* 1993; Irvine & Rajan, 1995, for reviews). Animal studies have indicated that mechanical lesions result in dynamic frequency reorganisation within the auditory cortices of adult animals. However, reorganisation of the auditory cortex following neonatal lesions produces a developmental plasticity which differs from the plasticity found in studies on adult animals. As such, it has been proposed that developmental plasticity may differ from adult plasticity. Developmental plasticity in the period where axons are growing to their targets can involve the axons growing to new targets; adult plasticity is generally believed not to involve axons growing to new targets but to involve changes in the efficacy of existing synapses and (possibly) axonal sprouting, and the generation of new synapses such that the axon provides stronger input to an area in which it previously terminated but only had weak effects (DRF Irvine: personal communication, 1997).

It has been found that during foetal development one set of factors affecting auditory system functioning relate to cochlear development and innervation. Developmental changes observed after (full-term) birth involve auditory nerve myelination, changes in the brain-stem tracts and inter-cellular connections in the cortex. While reorganisation following peripheral damage to

the adult auditory system demonstrates that cortical and subcortical sensory structures have the capacity for modification if altered sensory input is received, the benefits of the reorganisation are difficult to surmise. It seems to afford no compensatory function, although there may be functional consequences that have not been identified by commonly employed audiological procedures. Existing anatomical constraints make it unlikely, however, that the mechanisms that account for neonatal auditory system plasticity also account for adult auditory system plasticity. It seems probable that peripheral damage to the neonatal auditory system may be quite different in that reorganisational changes may afford some compensatory function in these situations. Differences observed between children and adults in recovery from various forms of acquired brain damage lends some support to this theory (Lenneberg, 1967).

Evidence has also been derived from studies on learning-induced plasticity. As the term implies, studies of learning-induced plasticity are concerned with plastic changes in the nervous system that are associated with (and might be responsible for) the behavioural changes in which learning is manifest (DRF Irvine: personal communication, 1997).

Learning-induced plasticity has been demonstrated in adult animals trained to make subtle frequency discriminations over a number of weeks (Recanzone *et al,* 1993). These behavioural changes have been found to be stimulus-specific and to be associated with neuronal changes in the form of increased areas of frequency representation. Other studies have demonstrated that training – conditioning with specific stimuli – can lead to changes in the frequency selectivity of cortical neurone clusters (Weinberger, 1993) and that these changes can be permanent.

The specific mechanisms involved in auditory plasticity have not been identified. In addition, whether or not the same mechanisms underpin these differently induced examples of plasticity is unclear. It has been suggested that the mechanisms may be intrinsic to the neocortex, are self-organising and are responsible for the refinement of cortical maps related to higher-order cognitive processes (learning, memory) during development. Cortical plasticity has been invoked to explain data on frequency discrimination and visual perceptual learning but there may also be plasticity at subcortical levels. Alternatively, it has been suggested that neuronal groups and synaptic connections corresponding to previously dominant inputs (which have now been eliminated by peripheral lesions) result in inputs that were previously expressed weakly, if at all, becoming more effective. This implies that alterations in input influence the stabilisation of certain connections and the elimination of others.

Ryals and colleagues (1991) discuss the implications of neural plasticity for cochlear implantation in children. They suggest that developmental changes affect tonotopic neuronal organisation in the auditory pathway, leading to a progressive apical shift along the cochlear partition and central auditory pathway. As a consequence, frequency organisation is not fixed but dynamic during development. Neurones along the central auditory pathway change characteristic frequency during development, and it has been postulated that parallel anatomical changes occur. This developmental feature of the auditory system was first referred to as the 'shifting-place' principle (Rubel *et al,* 1984).

The fact that the development of hearing begins with low to mid-range frequency discrimination is well documented (Rubel, 1978; Lippe & Rubel, 1983). High-frequency discrimination is not demonstrable until later in development. Rubel (1978) showed that behavioural and electrophysiological responses to low-frequency sound could be elicited before responses to higher-frequency sounds (~ 3 kHz) in foetuses of 24–30 weeks gestation. These observations are paralleled by anatomical data. As a result of the findings in behavioural and electrophysiological responses, it might be anticipated that the apical turn of the cochlea should mature first, that is, that part of the cochlea corresponding to low-frequency sounds. However, the organ of Corti develops from base to apex; thus, the base (which corresponds to high-frequency sound) develops first and responds to low-frequency sound during the earliest periods of development, gradually shifting as the rest of the cochlea develops to respond to high-frequency sound.

Early sensory stimulation is imperative for normal physiological, neural and perceptual development, and it has been found that foetal environments are typically rich in low frequency sound (from ~ 24 weeks gestation) (Hepper & Shahidullah, 1994). This low-frequency sound provides stimulation to all tonotopic regions in the central auditory nuclei. These observations would suggest that low-frequency stimulation is of importance during the earliest stages of development. The question then arises as to how important low-frequency input might be in the presence

of neonatal hearing impairment. It may be that low-frequency stimulation is essential for higher-frequency neurones to mature.

One implication is that the same sound may be perceived differently at different stages of development. This possibility is supported by Hyson and Rudy (1987) who demonstrated that the auditory perception of rats changes during ontogenesis. Rats conditioned to respond to a specific low-frequency tone would, 72 hours later, behave in such a way as to suggest that a higher-frequency tone now corresponded to the earlier perception of a low-frequency tone. In explanation, they proposed that the basal end of the cochlea is the first to mature and, as such, early in its development, responds only to low frequencies. As the cochlea develops and matures, low-frequency resolution moves progressively closer to the apex while the base responds maximally to high frequencies. They proposed that this shift in frequency-encoding along the basilar membrane will be mirrored by corresponding changes in central auditory system tonotopic organisation. The implication of this is that, if recordings are made from the same neurone throughout development, the characteristic frequency to which that neurone maximally responds should increase with maturation.

These findings have implications for the way in which early auditory stimulation may affect neuronal development in the auditory system. In turn, there are implications for the way in which the development of processes such as speech, which rely on the accurate encoding and repetition of direct and incidental auditory information, are affected and for the way in which early cochlear implants are assessed and evaluated in children. Thus, Kuhl and colleagues (1992) suggest that speech stimuli may be represented in the form of 'perceptual maps' by infants as young as 6 months of age and that these maps form a basis for the later development of spoken language. However, they fail to discuss how these maps may be established in the first place or how they may be represented neurally. Others have specified that delays in the maturation of cortical areas concerned with the acoustic analysis of speech might in turn result in the impaired development of environmentally dependent mechanisms of auditory processing (Kurtzberg *et al,* 1984) thus emphasising the interdependence of developmental processes.

In summary, it is known that the human brain has a protracted postnatal period of development and maturation, extending from birth to at least puberty (Neville, 1991). Evidence suggests that plasticity is a characteristic of the human brain that exists to some degree throughout adulthood as well as during early development. The precise nature and time-frame of such plasticity has implications for the inter-dependent development of auditory perception, speech and spoken language. That is, evidence would seem to suggest that a deficit in the stimulation of one of these areas will have an impact on the development of another, and that this pattern of interference is reciprocal.

These three sections on early intervention, sensitive periods for language acquisition and neural plasticity are complementary. The hard experimental evidence from early intervention studies needed to conclude that very early identification and habilitation are better than at, say, 12 months is still emerging. Hence, we have included a review of the other two areas (see also Ramkalawan, 1997). This has added some depth to the limited material available. Thus, the major evidence currently available, while not conclusive, points to:

- early sensitive periods for aspects of language acquisition, that suggest earlier intervention to be better than later intervention, other factors being equal
- substantial and long-term detrimental effects of the lack of sensory input on neuronal pathways that suggest that the earlier the lack of input is overcome, the less detrimental the effects will be.

These two conclusions help to buttress the findings from our review of the outcome evidence from early intervention which were that:

(i) there is a potential for more successful language acquisition with early intervention for children with moderate to profound hearing impairments
(ii) better short- and medium-term outcomes in the communication domain are achieved for children with moderate to profound hearing impairments who are identified earlier.

Parental views

There is considerable evidence that outcomes are significantly dependent on the extent to which services for hearing-impaired children are family-centred and the extent to which parental wishes and anxieties are taken into account (Moeller, 1996). There is a growing interest, therefore, in the extent to which parents want early identification and,

conversely, the extent to which parental anxieties are raised by early screening programmes.

A survey of parental views of hearing screening conducted in the USA by Sweetow and Barrager (1990) found that parents' satisfaction could have been increased had the parents been given more information about hearing impairment prior to the screen. The fact that 8% of them were not allowed to see the tests being performed was a source of some disquiet. About 20% of parents were displeased at the audiologist's manner and over a quarter did not have the results explained to them.

The NDCS published details of the replies from 246 parents of deaf children to a questionnaire on views of diagnosis and audiologist services in the UK (NDCS, 1983). Given that many parents expressed dissatisfaction about 'late' diagnoses, the unstated implication is that the earliest possible diagnosis is favoured. Furthermore, most remaining parental distress was associated with the lack of information and poor treatment of parents who were anxious.

Strikingly, there are only two published papers available on the specific issue of attitudes and anxiety towards neonatal-hearing screening, both from the same group. In the first of these, Watkin and colleagues (1995) sought the views of the parents of 356 hearing-impaired children. Responses were received from 208 of these. Participants were asked if they were happy with the age at which their child was identified as having a hearing impairment; two-thirds of them were not. They were also asked if they thought a neonatal screen desirable, to which 89% of respondents answered affirmatively. The report does not give reasons for the negative response of the remaining 10% of the sample. As Watkin and colleagues state, the "...high level of satisfaction should not cloud the anxieties and concerns expressed by the small minority".

In a more detailed study, Watkin and colleagues (1997) asked 288 mothers about their feelings toward **universal** neonatal-hearing screening. Anxieties were investigated *post hoc* but the results suggested that neonatal screening generates very little maternal anxiety (no more than 15% of mothers had **any** anxiety, with less than 1% being 'very worried'). However, there was a recruitment bias within the study against parents from ethnic minority groups, since mothers who were not fluent in English were excluded because of a lack of availability of interpreters. Levels of parental anxiety may therefore have been slightly underestimated.

Gregory (Personal communication, 1997) has data from studies of young deaf people and their families (Gregory *et al,* 1995), indicating that 90% of parents of hearing-impaired children wanted identification to be as early as possible. Results from a survey by Ramkalawan (1997) also suggest that earlier and prompt intervention for childhood hearing impairment was viewed as beneficial and essential by the parents of hearing-impaired children.

Some of the efforts needed to allay anxieties associated with screening are described by Marteau and colleagues in a number of articles (Marteau *et al,* 1989; 1990; 1992; 1993; Marteau, 1994). In one study (Marteau *et al,* 1992), the interaction between patients and professionals was examined when a routine screen was being explained to mothers in the antenatal period. It was evident that different professionals gave different information to patients and that, generally, the length of consultation was not related to the quantity of information conveyed, showing a poor use of contact time. The authors argue that professionals need more training in giving information about screens to parents. Marteau and colleagues (1990) also stress that before establishing a screen, a protocol should be drawn up to deal with all aspects of the screen in operation. In addition, guidelines should be established which are based on empirical research, where appropriate.

In summary, and with regard specifically to first-year screens for hearing impairment, the few studies there are suggest that most parents:

- want earlier identification of hearing loss
- want neonatal hearing screening, given that it is technically feasible
- show little anxiety associated with neonatal hearing screening, **provided that proper procedures are in place**.

Chapter 4

Survey of current practice

Scope

The methodology and findings of a survey of current practice concerning hearing screening in the UK are presented in this chapter. The data reported fall into three broad categories:

- Neonatal screening
- HVDT
- SES.

An indication of the organisation, purchasing, implementation and effectiveness of these screens is obtained that informs the context in which choices between future options may be made.

Introduction

There is a clear need to find out about hearing screening practice in the UK as a whole. Since the last survey of current practice (Stewart-Brown *et al,* 1986a; 1986b; Stewart-Brown & Haslum, 1987) and the work of Haggard and Hughes (1991), there has been considerable and unplanned expansion in terms of neonatal screening.

In addition, there have been recommendations (Haggard & Hughes, 1991) about screening between 1 year and 5 years of age (i.e. on removing such screens) and considerable debate concerning the HVDT itself (see, for example, Scanlon & Bamford, 1990). There is also concern that Health Districts do not have the data to be able to evaluate their actions or, if the data do exist, they are often uninterpretable unless aggregated with similar data from elsewhere.

This survey of current practice had, therefore, three aims; these were to collect data that indicated:

(i) what is being done
(ii) what programme information is readily available
(iii) whether that which is available can be aggregated to provide useful information on screen performance, such as coverage, yield, false alarm rate, sensitivity and cost.

The methodology of the study is presented here in full rather than as an appendix to point-up the general problem of lack of coordination, knowledge and information that is the antithesis of what an integrated national screening programme should be.

The aim of the survey was to gather information about existing children's hearing screening services in the UK and the coordinator(s) of those services. The survey was divided into two parts.

- Part one consisted of a short questionnaire sent to purchasers, in this case Directors of Public Health or equivalent in each District Health Authority, to gather some initial information about which services were provided and by whom.
- Part two consisted of two more detailed questionnaires sent to the coordinators of the different parts of the children's hearing screening services to gather more detailed information.

The term 'District' is used throughout to denote the purchasing agency, that is, District Health Authority, Health Board, Commissioning Agency, Health and Social Services Board or other equivalent term currently in use.

Questionnaire to Directors of Public Health

The aim of this part of the survey was to gather initial information about which children's hearing screening services were commissioned by Districts and the names of those people responsible for running these services. Directors of Public Health were also asked to provide some basic demographic information about their District. This would provide background information on the range of services provided in Districts and information about the potential for change.

A 4-page questionnaire was designed to gather:

(i) basic demographic information for the year 1993/94
(ii) information on which children's hearing screening services were currently in place in the District
(iii) information about any Audiology Working Party (if one existed) and if services were purchased from outside the District

(iv) the names of those responsible for the different parts of the hearing screening services and their contact addresses
(v) information about audits and costing of such services.

The questionnaire was piloted by members of the project's Key Advisors Group and circulated to the HTA programme manager at the Department of Health for comment. Changes were made following this feedback (see Appendix 2 for questionnaire).

Details of Directors of Public Health were obtained from the Regional Handbooks for seven of the 11 health Regions. Further names were obtained from the 1994 *Medical Directory*. At the time of the survey there were 128 Directors of Public Health representing 128 Districts in the UK, 109 in England and Wales, 15 in Scotland and four in Northern Ireland.

The questionnaire was sent to Directors of Public Health with a covering letter, an information sheet giving further details about the aims of the project and a reply-paid envelope on 13 and 14 June 1995. They were asked to return the information preferably by 4 July 1995.

Questionnaire returns

By 4 July 1995, 46 of the 128 questionnaires had been returned. A reminder letter was sent on 13 July requesting the return of the questionnaire, and those who had not returned the questionnaire by the beginning of September were contacted by telephone.

Eventually, by September 1996, questionnaires had been returned from 108 Districts in England and Wales, and from all Scottish and all Northern Ireland Districts – an overall response rate of 84%.

Some Districts returned more than one questionnaire for one of two reasons. First, where there had been a recent merger of Districts into a larger District, questionnaires were completed for each of the 'old' Districts. Second, separate questionnaires were completed for each trust commissioned by that District both within and, in some cases, outside the District boundary.

Most respondents returned demographic information for 1993, since 1994 data were not available until the August of that year. Also, a complication arose over the figures given for the numbers of live births. Some respondents gave two figures – those relating to live births of mothers resident within the District and those for live births of mothers both resident in the District and outside of it. (The questionnaire did not make it clear which figures were required.) Where two figures were supplied, the numbers of live births of mothers born in and outside the District were entered.

As is typical of such surveys, in some cases only partial information was returned. For example, information was given for only one provider trust in the District; all or some of the demographic information was missing. The largest omission of information was of numbers of children discharged from NICU/SCBUs, with 20 Districts not returning these figures. No other source of information was found to provide national figures for the numbers of children discharged from NICU/SCBUs. The names and addresses of the coordinators of services were often omitted, even though this was highlighted as being the most important element of what was required.

The names of 310 coordinators were obtained from the returned questionnaires. Further names were obtained from either:

(i) the Directors of Public Health for these Districts
(ii) the list of names of audit coordinators supplied by the BACDA (British Association of Community Doctors in Audiology) National Audit of Hearing Loss in Children
(iii) contacting key people known to the research team, such as audiologists and educationalists, working in these Districts.

This realised a total of 474 names of coordinators. Some people were responsible for more than one service in a provider trust and, frequently, one or more people were said to be responsible for the same service provided by a trust in a District. The next stage of the survey involved sending out two further questionnaires to the coordinators of these services.

Selected results

In cases where more than one questionnaires was returned for a District, the information was collated to form one entry for that District. Only partial information was supplied by some Districts. The findings from the 108 Districts with good data are summarised as follows.

- The total number of live births for 1993 was 614,508, of which about 97% were born in Hospital Maternity Units.

- There were 263 Hospital Maternity Units and 199 NICU/SCBUs from which 46,678 children were discharged in 1993.
- 50.9% of Districts have Audiology Working Parties.
- 23.4% of Districts thought they purchased hearing screening services from providers outside their District.

Some of the findings reported from the Directors of Public Health survey are shown in *Table 12*. These must be regarded as what the Public Health team **thought** was in operation, because the reality turned out to be different in some cases when the coordinators were contacted at a later stage. Eight of the Districts thought that they had UNS already but this was the case in only two of the Districts; hence, the table has been amended accordingly.

This finding threw some doubt on the validity of this part of the survey. However, we went on to contact the actual providers of services and it is from their returns that the survey of current practice has been built up.

Table 12 presents the estimates from the Directors of Public Health of the numbers of Districts offering the different children's hearing screening services, the numbers of provider trusts involved, the number of these carrying out audits and having contracted costs for these services. Note that very few Districts report that they have cost data written into their contracts for hearing screening. Only two Districts reported not having an HVDT and seven not having SES. About three-quarters of respondents replied that they had a targeted neonatal screen/assessment of some sort.

Questionnaire to coordinators of children's hearing screening services

The questionnaires were designed to gather information about Neonatal Screening Services, the HVDT, HVS between birth and 12 months of age, and the SES carried out at about age 5 years. The end result was condensed into two questionnaires, one detailing Neonatal Screening Services, the other covering the HVDT, surveillance services and the SES, in order to reflect the joint responsibilities of many coordinators for these services.

A decision was made to omit questions about intermediate screening and surveillance between 12 months and 5 years of age because of the small number of providers offering this service (35.5% of Districts still offered intermediate screens despite previous recommendations by Haggard and Hughes (1991); 68.5% had surveillance services for children aged between 12 months and 5 years) and because of concern over the number of questions being asked of coordinators, some of whom were responsible for all post-natal children's hearing screening services.

The neonatal screening/assessment questionnaire

The questionnaire was designed to gather information about:

(i) the structure and organisation of the service
(ii) the management and funding of the service
(iii) the performance of the screen and assessment service
(iv) assessment following the screen and follow-up services

TABLE 12 *The public health response to hearing screening provision in the UK. The data reflect positive statements only (n = 108/128 Districts)*

Screening/surveillance service	District	Number of provider trusts	Audits	Availability of audit	Contracted costs
UNS	2	2	2	2	1
Targeted neonatal	78	85	24	14	6
Neonatal other	14	14	4	3	0
HVDT	106	118	46	36	3
Surveillance, 0–12 months	61	65	12	7	0
Intermediate screen	38	38	13	9	0
Surveillance, 1–5 years	74	78	14	7	0
SES	101	113	26	26	3

(v) respondents' views about UNS and any general comments.

Five people known by the research team to be providing neonatal screening and neonatal assessment services were asked to complete the questionnaire and comment on the design and content. Questionnaires were also sent to members of the Key Advisory Group and to the HTA programme manager. The design was also discussed at the September 1995 meeting of the Key Advisory Group. Alterations and additions were made as a result of this feedback (see Appendix 2 for the questionnaire).

Questionnaires were sent to 204 coordinators in 106 of the 128 Districts. The first 100 questionnaires were sent out between 13 and 24 October and the remainder between mid-November and mid-December 1995. The delay in sending this second batch reflected the difficulty in obtaining names and addresses of coordinators from those Districts who had omitted this information or who had not returned the first questionnaire.

A covering letter and an information sheet giving details and background to the project were enclosed with the questionnaire, together with a reply-paid envelope. The covering letter asked respondents to return the questionnaire even if they undertook assessments in the neonatal period rather than screening. All were asked to return the questionnaire by early 1996.

According to the returns from the survey of the Directors of Public Health, 22 Districts had no neonatal screening service. These Districts were contacted by telephone (either the Directors of Public Health, the coordinators of the postnatal screens and/or Audiology Departments) to clarify that this was the case. Of these 22 Districts, one provided neonatal assessment, one neonatal screening, and another screened children at 6 weeks of age. Coordinators in these three Districts were sent questionnaires.

By early January 1996, 91 of the 204 questionnaires had been returned (40% within the first 6 weeks). Reminder letters were sent to the remaining 113 coordinators in January and February, requesting the return of the questionnaires as soon as possible. Of these, 35 coordinators were telephoned at the end of February and this resulted in 19 further returns.

By the end of April, 134 of the 204 questionnaires had been returned (a response rate of 65%). A further 46 questionnaires would not be returned because a coordinator in the same provider trust was already completing one or because that particular trust did not offer neonatal screening or assessment. This left 24 questionnaires to be returned.

The 134 questionnaires covered 96 of the 108 Districts sent neonatal screening questionnaires. Two of these Districts offered regional neonatal screening services, covering their own District in addition to providing neonatal screening services to a further three Districts (bringing the total number of Districts to 99). This left ten Districts not covered by the questionnaire returns. Of the 24 questionnaires not returned, ten covered these ten Districts. Coordinators of these services were contacted again asking them to return the questionnaire. The remaining outstanding 14 questionnaires covered provider trusts in Districts from which at least one other provider had returned a questionnaire.

In some cases more than one person was responsible for a service and more than one questionnaire was returned for one neonatal screening service. The data from all the questionnaires returned and discussed here were finally gathered together in September 1996, although there were still some notable exceptions which are discussed below.

The HVDT/surveillance and SES questionnaire

This questionnaire was designed to cover three services, the HVDT between 6 and 9 months of age, hearing surveillance services between birth and 12 months of age and the SES service at about 5 years of age. Surveillance was defined as any routine task or observation, including the use of a questionnaire, performed by staff to monitor a child's hearing. The design was similar to the neonatal screening questionnaire and covered five areas:

(i) the structure and organisation of the services
(ii) the management and funding of the services
(iii) the performance of the screens and the surveillance services
(iv) information on assessment and follow-up services
(v) any comments coordinators wished to make about these services.

Coordinators were asked to indicate which of the three services they coordinated.

Four pilot questionnaires were sent to people known to the research team to be running at least

one of these services. The Key Advisory Group were asked to give feedback on the design and content of the questionnaire as was the HTA programme manager. The questionnaire was amended as a result of these comments (see Appendix 2).

Questionnaires were sent to 308 coordinators in 128 Districts in the UK; 236 were sent on 1 and 2 November 1995, the remainder from mid-November to early January.

An information sheet giving details and background to the project was enclosed with the questionnaire together with a covering letter and reply-paid envelope. All were asked to return the questionnaires by early 1996.

By early January only 32 questionnaires had been returned. Reminder letters requesting their return were sent out between mid-January and mid-February 1996.

By mid-1996, 207 of the 308 questionnaires had been returned covering 111 Districts (a response rate of 66.9% of coordinators and 87% of Districts). Of the 101 questionnaires not returned, only 24 covered the remaining 15 Districts. The other 79 covered providers from Districts which had already returned at least one questionnaire either from the same or a different trust.

In many cases more than one person was responsible for the same service and this resulted in more than one completed questionnaire for that service. In these cases, information was collated to form one entry for that service from that provider. However, for one District the returns covered different geographical areas and the services were structured and managed differently. In this case each questionnaire was entered separately. Information was collected on 164 postnatal screening/surveillance services covering 111 Districts. Several further questionnaires have subsequently been returned and these were still arriving in December 1996; however, these have not been taken into account here.

There are advantages in obtaining a national response, particularly when trying to characterise the quality of screening practice. In view of the length of time taken to obtain responses, it may have been better, in retrospect, to sample Districts. However, as most of the data might be thought of as routine, the need for better information systems is highlighted.

Results – neonatal screening questionnaire

In all there were 171 replies to the neonatal questionnaire as a result of either a posted return or a telephone follow-up, representing 126 of 128 Districts. Of these replies, 43 providers essentially included no data, although 16 were covered by other respondents. This left 128 providers, covering 96 Districts, who provided data either on neonatal hearing screening that was on-going or on real plans to develop such a service (21 providers). Fifteen providers undertook neonatal assessment for very-high-risk cases, which were included in the data-set analysed. Thus, the questionnaire accessed a wide number of services that were working (mid-1996) or are planned for the near future, and the replies can be seen as being reasonably representative of the UK as a whole. However, at the individual question level, much data was unavailable, for example, specificity of programmes, costs and contract information. Several replies were received after the cut-off date for analysis and these have not been included here.[1]

The Neonatal Screening Questionnaire is presented in Appendix 2 to provide information on what questions were asked and in what context. The descriptive analysis of the questionnaire is superimposed on the questionnaire itself, with some of the more qualitative data being summarised either on the questionnaire or in the text here. Only the major points are dealt with in the text.

Structure and organisation

This section of the questionnaire shows that there are a large number of neonatal screening programmes in the UK carrying out targeted screening; for example, 84 providers in 65 Districts have a targeted screen for neonates in SCBU/NICUs (see Appendix 2: the number of Districts

[1] The major centre initially omitted was Southampton. Due to the special circumstances that applied to the trial of UNS run by Dr Colin Kennedy and his team, the service providers in the Districts taking part in the trial replied that they did not have a service, but a research project for half of the time. Since then the position has become clearer, as there is no neonatal screening service in Southampton *per se*, because (we were told) the purchasers await the outcome of this report. The preliminary data from Southampton are therefore dealt with in chapter 7, rather than in the results of the survey.

contributing data is given in parenthesis). The time for which these programmes have been in operation is relatively short and is summarised in *Figure 6*. Of the 84 programmes, 30 (36%) have started within the last 3 years (bearing in mind the survey started in mid-1995 and finished in mid-1996). This is attributable in great part to the NDCS targets and recommendations (NDCS, 1994).

Overall there are 107 providers of neonatal screening or neonatal audiological assessment, carried out in 80 Districts. In 59 Districts, 76 providers say that their service aims to target all three major risk factors, with ten providers using the two major risk factors (NICU/SCBU and family histories) and five targeting NICU/SCBU alone. Screening for criteria other than those on the questionnaire was provided in 18 Districts; these included cytomegalovirus, rubella, professional concern, ototoxic drugs, and jaundice.

Only two Districts currently provide (funded) UNS as a service: Hillingdon, which has been providing it since 1985, and Whipps Cross/Waltham Forest, which has been providing UNS since 1992. The work of both of these Districts is discussed in chapter 7. Interpretation of the remaining data is difficult because the coverage (see below) of the targeted screening programmes is quite low and of assessment programmes much lower. Nevertheless, more than half the Districts have a targeted neonatal screening programme of some sort. This corresponds well, although not exactly, with the information from the Directors of Public Health.

Screening techniques and equipment used

Two major techniques are currently being used. The use of TEOAE is available to, and probably used by, 68 providers in 51 Districts, predominantly using the ILO® hardware and software provided by Otodynamics Ltd. Slightly more providers, 79 in 66 Districts, are using ABR.

The distribution of the levels at which ABR was used is shown in Appendix 2. This shows that 15 providers actually test to threshold (these are predominantly those who are carrying out small numbers of neonatal assessments on children at very high risk), and that among other providers there is a very wide spread of thresholds being used, 20–70 dBnHL. However, 20 providers use 40 dBnHL and 16 use 50 dBnHL as screening thresholds; these constitute over half of the providers and would generally represent a sensible threshold for a neonatal screening programme giving better sensitivity for moderate impairments at 40 dBnHL but possibly much better specificity at 50 dBnHL.

Location of neonatal screening services

The survey showed that the NICU/SCBU was the predominant location for routine neonatal hearing screening (54 providers) prior to discharge, with hospital outpatient departments being the most preferred site for post-discharge services. The responses came from 104 providers in 79 Districts and showed that 74 providers gave a service both pre- and post-discharge, with 30 providing either a pre-discharge service (5 providers) or a post-discharge service (25). Five service providers gave a service in four locations, 28 in three locations, 46 in two locations and 25 in only one location. Rooms close to the NICU/SCBU were used by some providers, and others provided a service from the neurophysiology department.

A total of 56 providers (in 52 Districts) either had a separate sound-proofed room (e.g. in audiology) or an adapted room (e.g. close to the NICU/SCBU) in which to undertake screening, with only 24 providers giving a service at the bedside.

The pattern given here is of a heterogeneous service provided at a number of time-points and in a number of locations. Of course, it must be remembered that the predominant

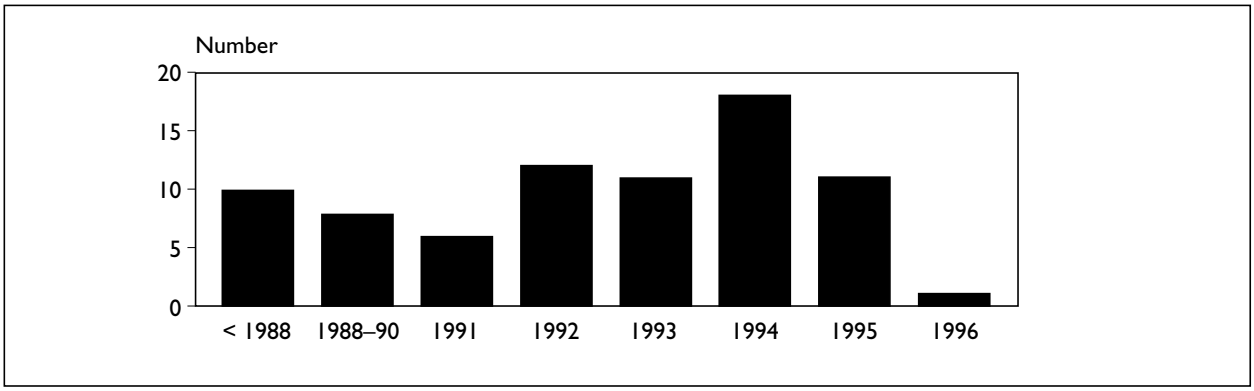

FIGURE 6 *Number of targeted neonatal screening programmes starting each year that include NICU/SCBU children*

implementation of neonatal screening is presently concentrated on at-risk babies, the major/most readily accessible group of children being those in NICU/SCBU, and this is confirmed by the 85 service providers (in 68 Districts) who state that there is a policy to test premature babies.

Parental involvement

The majority of providers have engaged parents in the screening process by giving written and verbal material about the screening procedure and service. Parental consent was sought by 76 providers (in 63 Districts) but not by 27 providers (in 25 Districts). The major reason given for requesting parental consent is that the programme may be considered to be at the research stage. A major reason given for not so doing is that screening is a part of the normal NICU/SCBU pre-discharge procedure. Just over 60% of the service providers who responded supply written information to parents, with over 85% encouraging the parent(s) to be present while their child was being tested. However, only 60% gave out information after the test.

It is important to involve the parents from the outset (as discussed in chapter 1, see *Figure 1*) but the results obtained here suggest that full parental participation is not currently being sought by all providers. This is a cause for some concern, but further examination is needed to pinpoint exactly what is being done by those service providers and Districts who did not respond positively to this section of the questionnaire. Nevertheless, while 23 providers (out of 101 who responded, 27 missing) responded 'Yes' to each question, only three providers answered 'No' to each question.

Records

How and where records of screening are kept is a complex issue, and the extent to which they are accessible is not clear. Only nine service providers record the outcome of neonatal screening on the child-health computer database, with 45 providers (in 38 Districts) having the data stored in another computerised database (ranging from their own spreadsheet to the Biologic Navigator's information system). Only two Districts had all four record systems (parent-held, paper, child-health computer plus own database), with 22 providers having three out of four and 42 having two out of the four systems. Forty providers had a parent-held record system and another type of record system.

This is an area where providers and client organisations are dissatisfied with the *status quo* and there is a need for guidelines, and possibly some development work (see Focus Group reports and recommendations).

Neonatal screening staffing and training

Some questionnaire problems had been noted in this area during the pilot stage of the study, but which we thought had been overcome. However, the heterogeneous nature of the responses means that only the most qualitative of analyses are possible. A more detailed analysis of particular selected Districts is shown under the costs section in chapter 5.

From the 101 providers who replied in this section, most of the staff involved in neonatal screening (and neonatal assessment) at present are technical audiologists (predominantly Medical Technical Officers (MTOs) grades 3/4). Thus 71 providers, in 56 Districts, use an audiologist to undertake most of the testing. Considerable use is also made of audiological scientists (graded at B or C) to do some testing but for the most part they provide the business case and monitor the quality of the programmes.

A wide range of nursing staff (ranging from nursery nurse to Grade G) are involved in the 14 programmes that are predominantly based in NICU/SCBU. The input from Clinical Medical Officers (CMOs), paediatricians, ENT and other consultants is acknowledged in just 15 programmes.

There is evidence of the use of relatively junior (less qualified) staff in the 13 programmes that use Assistant Technical Officer (ATO) or MTO appointed staff (not audiologists). These tend to be in the larger programmes, including the universal programme organised by Dr Watkin at Whipps Cross/Waltham Forest.

Staff training was commented on by a smaller group of providers, with 75 giving details of relevant professional training and 58 of specialised training. The need for refresher courses for those involved in the neonatal screening service was recognised by 60 providers.

In summary, a wide variety of professionals are providing predominantly targeted neonatal screening services, with the major time investment coming from audiologists. However, a cause for concern was that only 58 providers (in 49 Districts) had a written protocol to follow when screening neonates and there is a lack of information systems.

Neonatal screening structure

In previous work, Haggard and Hughes (1991) had conducted a survey of screening providers and

asked them to provide a summary diagram of their service structure, or to judge which of a set of given structures best described the service. In this survey, we provided an example, similar to the structure used by the Nottingham Neonatal Unit, and we expected that many providers might have a similar structure. There were abundant attempts in completing the questionnaire to enter a lot of detail about the screening service provided, which was very encouraging. However, it became clear after initial attempts to analyse the structures that no overall pattern was emerging. Each service (unless replicated within a District) had a fairly individual structure that related to:

(i) the person organising the service
(ii) the testers that were available
(iii) the room and testing facility available
(iv) who was paying (research versus trust, as well as which directorate).

The combinations are large and no easily discernible pattern emerged at this stage.

Management of the neonatal screening service

Contracts with purchasers/funding

The contract to provide neonatal screening was a specific part of a contract for only eight providers in seven Districts. Both universal screens were part of a larger block contract. Only 70 providers, in 59 Districts, answered the questions concerning funding (reflecting that the other 58 providers were either still negotiating or unsure). The major directorate in terms of numbers of contracts was ENT/audiology with 27/70, with the NICU/SCBUs providing contracts for only eight providers, although a further 12 contracts were with child health. A total of 24 providers had contracts with other hospital departments, such as medical physics or clinical neurophysiology.

Although only 70 providers gave information on contracts, 82 providers in 64 Districts were able to give information of service funding. Thus 72 providers, in 56 Districts, said that the NHS trusts paid the salaries of those involved in testing. A sizeable proportion of the equipment money, as detailed by 28 providers, has come from charitable donations (e.g. the Hearing Research Trust).

Aims and plans

Although there were over a 100 providers of neonatal screening and assessment, only 50 of these, in 44 Districts, had written or agreed aims for the neonatal screening service. In terms of developing the service, there were 82 replies, in 69 Districts. The replies were relatively easy to divide into three major categories:

(i) those Districts who said they were waiting for further advice, either from this current report or from *Health for All Children* (Hall, 1996)
(ii) those who said that they hoped to develop UNS (24 providers in 23 Districts)
(iii) those who wished to further develop, maintain or monitor targeted neonatal screening.

Quality control, audit and costs

There were 87 replies concerning the quality control that was presently exercised. In all, 38 providers said that they either carried out an audit of the service as a means of quality control or monitored the screening process in some way. In fact, 35 providers backed this up by saying that they have performed an audit and 26 have said that these audits could be made available to us, with ten being sent to us with the completed questionnaire. In general, these reports support and endorse the conclusions that emerge from chapter 7.

Performance of the neonatal screening service

Here our aim was to discover what information the typical service actually had available on numbers of children tested, failure rates, false-positive rates, yield and sensitivity of the screening programme. There was a serious design flaw in the questionnaire at this stage concerning the number of children tested. We should have asked for the total number of children tested rather than the number of children tested from NICU/SCBU, and should **then** have asked how many were from NICU/SCBU. However, we have re-analysed the data using additional information available from other sources, for example, the survey of the public health departments, and also by going back to providers where the figures appeared anomalous.

To put the figures here into perspective, some data from the survey of the public health departments is first reported. Replies were received from 159 Districts or sub-districts representing 127/128 Districts. However, not all data were supplied, or supplied accurately. A total of 116 purchasers, covering 108 Districts reported a mean of about 5300 births per purchaser and a mean of 2311 births for each of the 266 maternity units. Given these rates, it might expected from the data for the Trent Region (Fortnum *et al,* 1997) that there would be 815 babies with congenital permanent hearing impairment born in the UK per year. This is slightly lower than

reported in chapter 2 and reflects the bias of those completing the questionnaire.

The Directors of Public Health also reported that there were 93 NICU/SCBUs with a mean discharge rate of 456 babies (median 359) per year. We were able to link these data into the return for the number of children tested.

The number of NICU/SCBU children who were reported in the questionnaire as having had a neonatal screen in 1993 was 6303 by 46 providers. In 1994, this number increased by 57% to 9883 by 64 providers. In terms of the birth populations in the Districts in 1994, this represented 2.5% of the birth cohort, or 36% of the children that were discharged from the NICU/SCBU in those Districts. For 41 from 64 providers, the figures given were the actual numbers tested and for the remainder they were estimates (usually to the nearest 10). These data on coverage are reasonably available for many Districts but data on failure rates and yield are not. However, an overall pattern can be constructed from the data that are available.

A total of 47 providers indicate that their mean fail rate on first neonatal screen was 10.5% in 1994 (using a mean that was weighted to the number tested in the NICU/SCBU), with a second test fail rate (only supplied by 29 of the 47 providers) of 6.4% of those failures. For the two screens that were universal, the fail rate was 5% (approximated) for Hillingdon and 13.8% (actual data) for Whipps Cross/Waltham Forest on first test, and 1% (approximated) and 1.6% (actual) on second test, respectively. The false-positive rate would be very low for both sites if a test/retest regime was used, for example, the TEOAE fail rate would be about 0.2%. However, not every child who fails the first test can be tested a second time; thus, for 1994 the false-positive rate was 5.6% of babies tested. Caution is necessary here, as only 29 providers gave false-positive rates, and both UNS programmes gave approximated false-positive rates of about 1% (based on about 8000 babies tested in 1994).

Data on yield and sensitivity were collected for 1990–94. However, in terms of yield the data are only shown here for the 2 final years of this period because of the introduction of so many programmes in recent years. In 1993, the neonatal screening and assessment programmes yielded 63 permanently hearing-impaired children with average hearing impairment in their better ear of ≥ 50 dB HL. In 1994, this increased to 100 babies from 52 providers in 42 Districts. Of the 100 babies, 11 were detected by the universal screening programmes and 89 through the remaining targeted screening and neonatal assessment programmes. The 100 babies represent about 36% of the 275 who might be expected to be bilaterally hearing-impaired at the 50 dB HL or greater criterion[2] for these Districts, based on the estimates presented in chapter 2. For providers who use targeted neonatal screening, the yield was about 35%. This is consistent with data presented in chapter 7. In terms of point prevalences for 1994, the universal screens gave a prevalence of 1.37 per 1000 children tested (95% CI, 0.76–2.48 per 1000).These are also consistent with data reported in chapter 2 and imply that this 30% of the Districts in the UK who supplied data are reasonably representative.

The targeted screens are more difficult to interpret as we only asked for the numbers of children tested in NICU/SCBU. As can be seen from the analysis above, the screens presently in place are highly targeted and only test about one in three of those from the NICU/SCBU, with one in two of those in NICU/SCBU > 48 hours. If the number tested were used as the denominator it would give a prevalence in the tested population of 1.2% children (95% CI, 0.98–1.49). This estimate is much too high. The second major risk factor used is the presence of a family history of childhood deafness, which has a prevalence of about 1 in 25 families (Wood *et al,* 1995). It is unlikely that more children with a family history risk are being tested than NICU/SCBU children, as they are more difficult to find; hence, a low-side estimate of yield would be 0.6% of children tested rising to perhaps 0.9% (i.e. 1.2% allocated between an equal number of NICU/SCBU and family history children). Both estimates of yield, that is, the 1.4 per 1000 for universal screening and 0.6–0.9% for targeted screening seem a little high but not unreasonable compared with other data reported in chapter 2. A further reason that the targeted screening yield may be high is that it includes high-risk children tested from outside the District. It seems, therefore, that the screens reported on here, which represent about 30% of the Districts in the UK, are working effectively.

[2] It tends to be the larger centres that have introduced targeted neonatal screening, and, of course, these numbers are fairly crude approximations, because the babies tested in NICU/SCBUs tend to include 10% or more extra-District referrals, some being regional centres for neonatal intensive care.

It is difficult to calculate the sensitivity over a short period, because it takes 3–5 years to find children missed by screening. With PCHI the picture is further complicated by the time of onset of the hearing impairment; that is, a late onset, progressive, or congenital impairment. Data for the years 1990–94 were requested but data from years 1993–94 are too recent to make any judgement. It was possible to make a provisional judgement on the sensitivity for the years 1990–92. In these years, there were 141 children who were tested (by 17, 21 and 26 providers in 1990, 1991 and 1992, respectively) who were confirmed to have PCHI of 50 dB HL or greater in the better ear. From these providers, we have an estimate that 17 passed the neonatal screen, giving an operational false-negative rate of 12% (95% CI, 7.6–18.5). This compares with a false-negative rate of 15% (95% CI, 9.0–24.6) for the years 1990–91 alone. These figures compare well with those provided by Lutman and colleagues (1997) which are discussed later in chapter 7.[3]

Interpretation and communication of the screen result

This section of the questionnaire lacks detail about who communicates the result of the screen to the parents, how that is done and where. It is good that in 79 out of 87 programmes expert opinion is on hand on the day of the test to help interpret the results. This is the same as the number of programmes that use personal interpretation of the display by the screener to determine a pass or fail. This is not unreasonable, given that the predominant use is low-volume targeted screening, mainly undertaken by audiologists (MTO 3/4s). However, for higher volume screening, such as UNS, a more automated method would be needed.

There were 89 replies to the question about responsibility for explaining to parents the result of the screen. There were several mixed responses, often with the responsibility depending on the test outcome. However, there was a strong tendency for the tester to have the initial responsibility; thus 12 of 89 replied it was the 'tester', 27 of 89 replied it was the audiologist, 22 of 89 replied it was the 'consultant' or 'doctor'. It is clear that there is considerable heterogeneity of practice.

Details of follow-up services

Assessment and hearing-aid fitting

ABR threshold assessment is the major assessment technique (83 of 99 replies mention use of ABR). However, almost everyone commented that it depended on the age at which the child was coming to them for assessment. So 17 of 99 would use behavioural or observational assessment or (in the case of 11) VRA (Visual Reinforcement Audiometry), if appropriate.

Follow-up assessments are carried out in audiology departments (42), the Children's Hearing Assessment Centre or children's hearing department (38), with a smaller number being undertaken in medical physics (8) or clinical neurophysiology departments (2).

These assessments were carried out 'as soon as possible' or 'immediately' by 45 providers. A total of 25 providers waited until about 4 weeks post-screen, with 11 routinely waiting 8 weeks or more. However, the comments indicated that it really depended on the urgency of the case and the gestational age of the baby, as well as on other factors.

Of some concern was that although most providers (94) were able to undertake aided threshold testing, only 36 had probe-tube microphone facilities; these are essential if a systematic approach to hearing-aid fitting in very young children is to work (Westwood & Bamford, 1995).

Just under 50% of the respondents (91 providers) fit hearing-aids as soon as is 'practicable'. This seemed to mean about 2–4 weeks after confirmation of the hearing impairment for those who commented further. However, 13 providers suggested 2–4 months, 12 suggested 4–6 months, three suggested before the age of 1 year, while eight providers said it depended on the age of confirmation of the impairment, for example, if the child was very young when the hearing impairment was confirmed they might wait longer.

Once hearing-aids are fitted (in the first 3 years of life), only 17 providers review the fitting monthly, with the largest number (56) of providers carrying out a review every 3 months. A significant number of providers (10) said that they could in fact only carry

[3] Interpretation of these data is complicated by two aspects of the data collection. First, we do not know what tests were being used, that is TEOAE or ABR or threshold assessment. Second, the data reported here are not independent of the studies conducted by Fortnum and colleagues (1997), and by Lutman and colleagues (1997) and Mason and colleagues (1997), as a subset of the data presented in those papers is included in the responses to the questionnaire.

out such a review once a year or less often. Obviously, for very young babies, a more frequent review (e.g. four times per year, depending on clinical practicalities) of their progress with the aids would be highly desirable and, indeed, is necessary for the recursive processes of converging on a detailed audiological profile and optimum hearing-aid fitting.

Educational services, voluntary organisations and other family support

The family support services that are available through the gateway of the health services have not been probed in depth. It is important that education services are informed promptly of the confirmation of hearing impairment in young children. It is worth noting that a majority of providers (54 of 106) inform education services within 24 hours, with a further 16 providing information routinely within 48 hours. It is surprising that as many as 27 providers say that they inform education services only 'within a week'.

Education services provide a predominantly (57 of 84 replies) term-time service. The peripatetic service includes a visit to the family once a week in 68 of 105 areas and twice a week in only 13 areas. The inability of many Education Services to provide year-round support for the families of very young, newly identified, hearing-impaired babies is of major concern, since early successful habilitation is likely to be heavily dependent on early family involvement. Families in these circumstances face major emotional and practical challenges for which home-based support is essential.

Voluntary organisations play an increasing role in family support, and it is interesting to note that 5 of 97 providers do not routinely give information about those organisations which can provide further advice and support. The organisations that were mentioned by name are the NDCS, Royal National Institute for the Deaf (RNID) and the Elizabeth Foundation.

Further information was given by 90 providers (out of 99; 29 did not fill in the section) on health, education, social and voluntary organisations services/support for parents. A total of 35 providers (in 30 Districts) provided extra information for all four types of service/support.

The NDCS quality standards and other reports

Six respondents (out of 103) said they were not aware of the NDCS standards (NDCS, 1994) but 75 out of the 79 replies described how the NDCS report had influenced their service. For 13 of 75 there was no influence at all, and for five further providers there was only minimal impact. On the positive side, nine providers said that it had helped them focus better on what needed doing. For a further 41 providers, the report had helped them to plan their services better, for example, by enabling them to draw up business plans. There were seven providers who said that the NDCS quality standards had been instrumental in securing the staff to implement targeted screening. Several providers commented that they had great difficulty in achieving the targets set out in the quality standards document. This is not surprising, as the time-lag has been too small to see any real change unless the national data are routinely aggregated. However, perhaps the targets should be differentially interpreted for moderate, severe and profound impairments.

A total of 69 providers (in 57 Districts) commented that reports other than the NDCS quality standards had influenced their services. Among these were the Nottingham screening workshops, the NIH consensus statement, the *Health for All Children* report, BAAP (British Association of Audiological Physicians) and BACDA policy documents, *Screening Children's Hearing* (Haggard & Hughes, 1991), and various published articles.

The comments given at the end of the questionnaire supported the information given in the questionnaire. While there were many services in operation, they were by no means all fully operational, and exist on a minimal budget. In some cases, there were Districts who had had to discontinue services, in the period covered by this review, that had been put in place partly for research purposes and with research money (a prime example of this is the UNS operated at Southampton, but other examples exist for targeted screening). The situation needs clarification and a more consistent and systematic approach would seem to be highly warranted.

Results – HVDT and SES

Questionnaire returns

By early January only 32 questionnaires had been returned. Letters requesting their return were sent out between mid-January and mid-February 1996.

By mid-1996, 207 of the 308 questionnaires had been returned covering 111 Districts (a response rate of 66.9% of coordinators and 87% of Districts). Of the 101 questionnaires not returned, only 24 covered the remaining 15 Districts. The other 79

covered providers from Districts which had already returned at least one questionnaire either from the same or a different trust.

In many cases, more than one person was responsible for the same service, which resulted in more than one completed questionnaire for that service. In these cases, information was collated to form one entry for that service from that provider. However, for one District, the returns covered different geographical areas and the services were structured and managed differently. In this case, each questionnaire was entered separately. Information was collected on 164 post-natal screening/surveillance services covering 111 Districts. Some questionnaires were still arriving in December 1996 but these have not been taken into account.

Structure and organisation

The full questionnaire with annotations is shown in Appendix 2. The major results in each section are reported here.

There were 148 providers, in 104 of 111 Districts, who said that they provided a universal HVDT at 6–9 months, with a further three replying that they offered a targeted HVDT. A smaller number, 94 providers over 70 Districts, provided universal HVS at 0–12 months, with seven offering targeted HVS. At a later age, there were 144 providers, over 102 Districts, who provided a universal SES, with a further seven giving a targeted SES.

The range of ages for the HVDT was 6–9.5 months, with a median of 8 months. The range of ages for the SES was somewhat wider at 3.5–6.5 years of age.

The question regarding test technique for the HVDT was fairly uninformative, with 149 providers reporting that they used the 'distraction' method. However, it was interesting to note the variation in test level that the different providers were trying to achieve, as indicated in *Table 13*. The major finding here is that the most used levels are at 35 dBA, for 61 providers, and 30 dBA, for 44 providers. However, although this is the stated aim, only 66 providers were routinely using warble tones for the distraction test, while 105 were using live voice. There was some overlap, with 38 providers using voice and warbles. Thus, there were 67 providers (from 141) who relied on live voice (sometimes backed up with rattles, hums, chimes, etc.) at a 'calibrated' level, the problems of which are well documented (McCormick, 1993).

The SES is predominantly accomplished using an adaptation of pure tone audiometry, that is, the stimuli are pure tones at specific frequencies. The distribution of screening levels that different providers try to achieve is shown in *Table 14*. This indicates that several providers (19) are screening at 30 dB HL at low frequencies but that the majority are screening at 20 or 25 dB HL for mid and high frequencies.

The levels set by the providers show the problems encountered with the screens if used for the detection of PCHI at the 40 dB HL or 50 dB HL level. The tests are trying to be too sensitive to the lower levels of impairment, which are generally associated with other conditions, for example, OME. An immediate improvement in the test's cost-effectiveness for moderate, severe and profound (unilateral or bilateral) hearing impairments would be achieved by increasing the test levels to 40 dB HL in the HVDT and 30 dB HL in the SES. Other, more focused methods could be used to find the mild, unilateral or transient conditions.

The HVDT is usually performed at a health visitor clinic (144 providers giving a median of 80% of tests carried out in a health visitor clinic) but it is also carried out in a child's home by 103 providers (who estimate that 5% of tests may be carried out at home). Increasingly, general practice is the location where the HVDT is undertaken and this accounts for the remaining 15% of tests.

The SES is carried out predominantly in schools (142 providers) although some screening is undertaken at school clinics (18), health visitor clinics (18), child's home (7) or general practice (4).

TABLE 13 *The number of providers who reported the level they were attempting to screen at for the HVDT – some providers gave two figures, one for low frequencies and another for higher frequencies*

	Number of providers reporting level of screening for HVDT						
Level (dB A)	**20**	**25**	**30**	**35**	**40**	**45**	**50**
Low frequencies	2	5	3	11	19	1	2
High frequencies	3	4	44	61	17	0	0

TABLE 14 *The number of providers who indicated the levels at which they screened for each frequency at the SES stage*

Frequencies (kHz)	Number of providers reporting levels of screening for SES			
	20 dB HL	25 dB HL	30 dB HL	35 dB HL
0.25	44	33	19	1
0.5	44	33	19	1
1	45	42	1	0
2	50	36	1	0
4	42	39	1	0

Hints for Parents sheets are given out by 115 providers, usually at birth (21) or at the first home visit (66). Involving the parents is thought to be beneficial at this stage; however, no District has audited the use of such a handout in the period covered by the present study.

Staffing details

The answers provided regarding staffing details on the questionnaire proved too inconsistent to draw firm conclusions; however, they help to put the HVDT and SES undertaking into perspective.

As might be expected, 152 providers used trained health visitors for the HVDT, with a trained assistant being part of the test team for 74 providers. In addition, 37 providers used trainee health visitors, 28 used nursery nurses, 21 used general practitioners (GPs), 20 used CMOs and 18 used nurses. A similar pattern emerged for the HVS, except that GPs had a greater role. The SES was carried out mainly by school nurses (99 providers), with the second most numerous staff being 'others'. These were mainly staff grade named 'audiometricians' or MTOs.

The staff numbers and time estimates returned were patchy and this is reflected in the numbers of providers who have filled in each question (see Appendix 2).

The messages that can be drawn from the table in the questionnaire are:

(i) that the mean number of health visitors involved in HVDT is 42 full-time (from 90 providers) and 25 part-time per District, that is, between 60 and 70 individuals are involved in testing in each District
(ii) that only half of the Districts entered in a time estimate. The mean of this was 40 weeks per year testing (taken from 45 Districts). Thus, an estimate for overall staff time per provider for all unit staff involved might be about 2 person-years per year, as testing involves two or three people for each District (this is lower but not incompatible with the estimates from chapter 5)
(iii) that even less detail emerged about the SES staffing, on average 14 full-time and 14 part-time school nurses were involved in the screening, with an estimate of testing time for hearing being 31 person weeks (average taken over only 24 Districts).

The data within the training sections were sparse. There was a considerable amount of time for health visitors in-house, in service, training for the HVDT, HVS and SES.

Many more providers said they had written protocols for the HVDT (129) and the SES (123) compared with the HVS (61). It is noteworthy that written protocols were more abundant for the HVDT than for neonatal screening. However, more work is needed to see exactly how available the protocols were to the staff testing. (NB: there are far more staff testing in HVDT compared with, say, targeted neonatal screening where there is probably only one tester per District rather than over 60).

The HVDT is a well-developed test system which is reflected in the fact that 129 providers enter the screen result in parent-held records and 112 on the child-health computer system. The SES has predominantly a paper-based record but, in addition, 77 providers enter the data on the child-health computer system. Further work is needed to see what exactly is stored on the child-health computer system and whether this could be used for quality assurance.

The diagrams for referral routes worked well in the pilot study but, because of the variability in the systems used, it has been very difficult to extract any generalities.

Management of the screening services

Only 54 providers, in 47 Districts, indicated that they had written aims for their services, with 42 indicating that they had a service development plan for the next 5 years (and 67 indicating that they did not!). Those that did indicate a service development plan mentioned:

(i) that they would be auditing their service
(ii) that they were watching carefully for the impact of neonatal screening

(iii) that they wished to improve their training
(iv) that they awaited technical developments such as the behavioural screening test (BeST) (see chapter 7).

Similarly, for SES only 57 providers, in 51 Districts, had written aims and only 37 providers indicated that they had a service development plan. When there were comments about the plan, they were fairly non-specific, for example, 'continue as before', 'await school health review'.

The main attempt towards a quality-controlled methodology/assessment has been by means of audit, which has been undertaken by 67 HVDT and 51 SES providers. The availability of the audit reports has been a very useful source of information which, again, supports the conclusions of the published data reviewed in chapter 7.

We did attempt to obtain costs for all the services but the data are also very patchy. The HVDT data are presented in chapter 5.

Only five providers costed SES, although one of the these, in Scotland, provided a recent detailed costing which was very helpful. This indicated that the running cost of the screening/testing service was £19,300 for salaries and £22,300 in total. This District has about 6000–7000 births per year. Another District estimated its expenditure on testing alone as £12,000–13,000 for a District with 3000 births per year. Thus, there is some consistency in these figures. A third service, with 3000 births per year, reported time estimates of 0.55 school nurses plus 0.43 MTO audiologists plus 0.27 of an (S)CMO. This includes follow-up at secondary level and is consistent with the previous two estimates.

Performance of the screening services

The coverage and referral rates have been analysed using all the data reported because, when incorporating either actual or approximated data, there was little difference between the means that were obtained. Data for 1994 were available for 99 providers of the HVDT (in 79 Districts) and gave a coverage figure of 90.5%. This agreed well with the coverage for 1993 of 90.1%. The HVS coverage was only completed for 44 providers and gave a figure of 92.7%. The SES gave a slightly higher coverage, 93%, for a total of 91 providers (in 73 Districts). However, it is not clear if the pupils who might be at independent schools are tested or not.

In terms of referral rates, there were some large numbers reported for some Districts (e.g. 40% claimed as actual referral rates for the HVDT). The figure for 1994, given by 63 providers, is a mean of 9.3%. If the figures of those who supply actual referral rates are used instead then the mean referral rate (after re-screens) for hearing assessment would be of the order of 8.4%. This is highly comparable with that reported by Wood and colleagues (1997).

The referral rates for the HVS and SES are remarkably similar for 1994 and these procedures seem to have an inherent referral rate of between 8% and 11%. Whether this a property of the tests or the population is unclear.

The yields from the different tests were collected for the years 1990–94. Here, we have concentrated on 1993–94 for which more data were available. In 1994, data were given by 51 providers in 47 Districts and, in 1993, there were 47 providers in 44 Districts. This is quite a low proportion; two out of three of the Districts did not have the data available, even approximately, which possibly indicates that the screen is not being monitored appropriately.

The yield from the HVDT is shown in *Table 15* to be of the order of 1 per 4000 births, using the assumption that the number of children to screen is the number born in that District. The further assumption is that the prevalence of congenital hearing impairment is 0.905 per 1000 (derived from data on PCHI ≥ 50 dB HL by Fortnum *et al,* 1997). The 50 dB level is used here because that was the level used in the questionnaire to minimise confusion between PCHI and OME. Using this assumption, we can calculate that HVDT probably yields about 26–28% of the children with PCHI.

The data have also been analysed for those Districts that did and did not also have a neonatal screen at the time *(Table 16)*. The data are complicated because not all Districts that have a neonatal screen have given a yield from that screen, and not all Districts who have given a yield for neonatal screening have given a yield for the HVDT. However, we can see from the table that there were 13 Districts in 1993 and 15 in 1994 who had both screens in place and who have given yield figures for both. The data for the providers with neonatal screening and the HVDT have been presented twice, first using all providers who have neonatal screening and second by omitting the District that had UNS and supplied all data concerning neonatal screening, the HVDT and SES.

TABLE 15 *The yield of the HVDT in 1993 and 1994, expressed as a crude yield (amalgamated over approximate and actual yields) and as a standardised rate, using the 1993 birth-rate in each District*

	Yield of the HVDT	
	1993	**1994**
Yield (cases)	72	71
Providers	47	51
Providers giving actual data	30	33
Districts	44	47
Births (approximate number)	276,128	297,073
Yield rate	2.6 per 10,000 births	2.3 per 10,000 births
Expected number of hearing-impaired children (50 dB HL +)	250	296
Yield of HVDT as a percentage of expected number of children	28%	26%

TABLE 16 *The yield of the HVDT in 1993 and 1994, expressed as a crude yield (amalgamated over approximate and actual yields) and as a standardised rate.This uses the expected number of hearing-impaired children derived from the annual birth rate and the prevalence of congenital PCHI that is ≥ 50 dB HL, for those providers who have neonatal screening and those who have not implemented targeted neonatal screening or assessment at the time of the survey*

	1993	**1994**
Providers giving neonatal screen yields and HVDT yields	13 84,785 births	15 99,931 births
(a) Yield from all neonatal screens	24 (31% expected)	27 (30% expected)
Yield from HVDT	21 (27% expected)	27 (30% expected)
Combined yield	45 (58% expected)	54 (60% expected)
(b) Yield from targeted neonatal screens only	16 (22% expected)	21 (24% expected)
Yield from HVDT	21 (29% expected)	26 (30% expected)
Combined yield	37 (51% expected)	47 (54% expected)
Providers not having a neonatal screen and giving HVDT yield	23 133,799 births	25 144,022 births
Yield from HVDT	26 (21% expected)	25 (19% expected)

It is of interest that, contrary to expectations, the yield of the HVDT from the Districts that also had targeted neonatal screening was higher than the yield for those Districts where no neonatal sceening was in place. Of course, the number of Districts included here is small but the major confounding aspect is that these Districts are those that have been doing the neonatal screening for the longest time – the median year for starting neonatal screening programmes was 1991 for those reported here, compared to 1993 overall. Despite this, however, there was a lower yield from the neonatal screening progammes for this group of 13–15 Districts than shown overall (35% – see above).

The effect of whether just the targeted neonatal screens or all neonatal screens are considered is minimal, with the evidence suggesting there is little effect on the comparative yield of the HVDT of having a neonatal screen in place. Other evidence is considered in chapter 7 for the Trent Region only (Fortnum *et al*, 1997) However, in the Trent Region the non-neonatal screening Districts (either in time or place) had a higher yield of nearer 40% for the HVDT for 1985–90.

The data on yields from the HVS and SES are not so reliable, with far fewer providers having the appropriate data for 1994 and 1993. Taking just the 1994 data, there were 68 children reported as being identified through SES by 31 providers, which is a rate of about 4.3 per 10,000 children. There are two factors that caution the interpretation of these findings. Firstly, although it was stipulated that the children had to be impaired at the level of ≥ 50 dB HL, there may have been some children entered here with milder impairments. It is also unclear whether the children identified by SES have had a congenital hearing impairment, a late-onset or progressive impairment. Secondly, we cannot interpret the data as a system because there are so few Districts where all the yields have been recorded. There were just 19 (out of 164) providers who recorded the yield from the HVDT and SES. In these 19 areas, the yield from the HVDT was 32 children and from the SES was 45 children. There were 13 areas with the HVDT, HVS and SES that yielded 20, 24 and 34 children, respectively. There were only five areas that also had neonatal screening data in addition to these three yields. Thus, it is possible to make statements concerning the approximate yield of each screen, but not statements concerning the incremental yields of the different screens. Another factor to bear in mind is that the children were reported by year of screen and not year of birth (because those having neonatal screens between 1990 and 1995 would not

have been at school). Thus, SES tags the neonatal/HVDT screen that was in place 5 years previously and about which we have even fewer details.

How many screening tests are carried out before an onward referral is made determines to a large extent the false alarm rate for the screen. Most Districts do one further screen for both the HVDT (n = 133) and SES (n = 101). However some providers refer on the basis of the first test only (HVDT, n = 5; SES, n = 26), while nine providers do three screening tests before referring on. This accounts for the large variation in referral rates from 40% to 0.1% for these tests.

Follow-up services

The assessments that are carried out at identification, their location and timing are consistent with previous work, and are not presented in great detail here. Again, there were only about a quarter of providers who had access to probe microphone facilities.

It was not possible to analyse the data about the age at which hearing-aids were fitted after identification by the HVDT and HVS, as half of the respondents replied 'as soon as possible' and the other half correctly replied with an age. The most popular age given was 'about 18 months'. This contrasts with the neonatal screening replies of 12–15 months earlier.

The availability of educational support, family support, or voluntary organisations were all similar to that found on the neonatal screening questionnaire. There were 22 respondents who were not aware of the NDCS standards, which was a higher proportion than in the neonatal screening questionnaire, as might be expected.

Summary and implications

The major points that arise from the survey of current practice are summarised for the two questionnaires separately.

Neonatal screening

1. Just under two-thirds of all Districts have a neonatal hearing screening or assessment programme of some sort. Only two Districts have universal programmes; the remainder are mainly targeted at high-risk children with a history of NICU/SCBU or a family history of hearing impairment. In those programmes that target the NICU/SCBU children, about one in three children at risk are tested, so coverage of this group is focused mainly on those at very high risk possibly due to lack of funds for extending the testing.
2. The two UNS services are both running successfully and providing the data needed with no difficulty.
3. The targeted screening programmes are relatively young, median year of start being about 1992–93.
4. Both TEOAE and ABR are in use, and some Districts use the NIH recommended method of screening with TEOAE first and then re-screening the failures using ABR.
5. The screens are carried out by a number of professionals, the largest number being audiologists. The screens are carried out at a number of locations, depending partly on whether it is done pre- or post-discharge.
6. Funding is provided by more than five different directorates and the trusts pay the salaries and equipment costs for the programmes currently running.
7. The failure rate for the predominantly at-risk group at first test is of the order of 10–12%, with false-alarm rates of about 5–8%, which compares very well with the referral rate from the HVDT and SES tests – both about 10%.
8. The yield is quite high already, with 100 babies being detected with PCHI of at least 50 dB HL in 1994. This represents about 35% of babies who might be expected to have a congenital hearing impairment of this level in these Districts, and about 16% of all such children in the UK.
9. The overall field sensitivity of neonatal screening seems to be of the order of 85% in the at-risk population.
10. The age at hearing-aid fitting for those screened neonatally is routinely before 6 months of age, with a median age of referral of about 1.6 months (range 0–8.1).
11. There is some cause for concern in that there is no systematic approach to the family and parents, both in terms of information given before or after the test and in terms of support post-diagnosis. There are, for instance, several professional groups with the responsibility of explaining the test results to parents, and there is no information about the training that such groups have had for this.
12. There is some cause for concern over the availability of educational services for children identified by neonatal screening. This manifests itself in the availability of the

educational support out of term-time, the timing of educational support and the ability of the service to provide effective support to families with early identified hearing-impaired children.

13. Many Districts report that they wish to extend their targeted screening or to introduce UNS at an appropriate time.

The HVDT, HVS and SES

1. The HVDT and SES are universal in their application, both across the country and within Districts. HVS plays a part in identification for about two-thirds of all Districts.
2. The professionals responsible for carrying out the tests come from a more homogeneous group than for neonatal screening. However, where there might be one person responsible for targeted neonatal screening for a whole District, there are more than 60 full- and part-time staff who might perform the HVDT. For SES there are, again, many fewer testers. This has considerable implications for the different training requirements and for systems of quality assurance.
3. The HVDT attempts to use a level for testing that is too acute for the room conditions and for the aim of the test (to discover as young as possible those children with hearing impairments that put their development at risk).
4. SES probably uses screening levels that lead to a greater number of referrals than is efficient.
5. The *Hints for Parents* sheets are used quite regularly but, from this survey, there is no direct evidence of their role in family support or in identification of hearing impairments.
6. The coverage of the HVDT is about 90%, which some may think is low for this type of universal screen (Johnson & Ashurst, 1990) and others may not (Torgerson & Donaldson, 1994). SES and HVS are nearer to the 95% of the stated children's population which would be an appropriate target for service attainment.
7. The referral rates for all three post-neonatal screens is nearly 10%. This is not atypical (see chapter 7; Wood *et al,* 1997).
8. The yield from the HVDT is lower than we might expect from other work (Davis & Wood, 1992; Fortnum *et al,* 1997) and is consistent at 26–28% of the expected number of congenitally hearing-impaired children for the 50 or so providers (one in three) who supplied data.
9. Perversely, there was no sign in the small number of Districts (n = 14, in 1994) for which the data for both targeted neonatal and HVDT screens were available that the neonatal screen depressed the yield from the HVDT. This was probably because the yield from the HVDT was quite low anyway.
10. Both HVS and SES gave yields for bilateral hearing impairment close to 4 per 10,000 children. This means that, within the present system, they play a large role in identification overall. However, it also means that there are long delays in identification. There is also a need to find out whether children identified by SES were truly as severely affected as the questionnaire required and to determine whether these children were thought to be congenitally hearing-impaired or had acquired hearing impairment subsequently.
11. The costs for SES were only available for five Districts; however, these enabled us to estimate the testing cost for SES at between £3000 and £4000 per 1000 children entering school, without any overheads being included.
12. The age at which hearing-aid fitting following the HVDT was thought to be routinely carried out was 18 months, which is earlier than reported elsewhere (see chapter 7; Fortnum *et al,* 1997).

Implications

There are important implications from the data of the survey for the role of neonatal screening in the early identification of children with PCHI. However, caution must be applied given that:

(i) the data are questionnaire-based
(ii) there was a large element of non-completion of the yield and false-positive data
(iii) the data have not been validated except for four of five sites where we have personal knowledge.

Nevertheless, the following conclusions are consistent with the other strands of this review, and with those audits that were sent to us.

1. There is much evidence that neonatal screening is presently playing a substantial role in identifying PCHI. However, it is organised in a diverse manner, with funding not being overtly identified. This may not be surprising as much targeted screening is presently on a very small scale.
2. There ought to be appropriate written aims for the neonatal screening services, and service development plans need to be updated regularly.
3. The HVDT is not playing as large a role as it has been thought to do. The evidence from this survey suggests that its coverage is not

acceptable in about half the Districts (it falls below 90%), its referral rate is not acceptable in about half the Districts (it raises over 10%) and its overall yield (about 26–28%) is less than that of targeted neonatal screening (at about 35%).

4. SES may be playing a larger role than expected, although the data concerning yield were only from 31 of 128 Districts and further work is needed.
5. The data concerning coverage, referral rates, yield and sensitivity were not available for a very large number of providers. Considerable effort is needed here to standardise and co-ordinate such data so that they can be routinely available to help develop policy at a national and local level.
6. A key concern that emerged was that there was little coordination and thought given to the involvement of parents and the role of other organisations such as the education and voluntary sectors.
7. A more systematic approach to quality assurance is needed for all the areas of hearing screening considered in this survey. Some Districts have shown an exemplary level of quality assurance, but these are very much in the minority.

Chapter 5

The costs involved in neonatal and HVDT hearing screening

Scope

Costing data from the survey of current practice and from an additional more in-depth survey of selected Districts are presented in this chapter. The HVDT and neonatal screening are compared on a number of cost measures.

Introduction

There are many ways in which costs may be considered, for example, health costs, individual costs, family costs, social costs. Our primary concern here is with the health costs incurred by the different hearing screening techniques. In analysing health costs, two major approaches are usually taken. The first is the 'bottom-up' approach, which includes individual time components for a specific service. This approach collects data on all subcomponents of the service and considers its implications for further services. The second is the 'top-down' approach in which the overall cost components of the service are considered as structural entities. This approach collects aggregated data on costs, and estimates individual cost by factoring in activity levels for particular services. The two approaches are complementary; however, it is easier to obtain rigourous data using the bottom-up approach but easier and cheaper to get appropriate top-down information. As our main purpose is to make a relative comparison of costs associated with different screening options, the choice of method is less important than the need to make similar assumptions for both techniques.

In the survey of current practice, two different attempts were made to discover the resource implications for the four screening opportunities (the neonatal screen, the HVDT, the HVS and SES). The first was to obtaining the bottom-up costs and the second the top-down costs. There were, however, very few providers who could tell us how many staff were involved in the screening, their grades and how many person-weeks they spent on hearing screening activity. The forms (see Appendix 2) were not easy to complete and probably deterred both coordinators and managers alike. However, such data should be available, even if only approximate. In the neonatal screening questionnaire there were 27 providers (out of 90 respondents) who were able to cost the service in any way; the number was even lower for the HVDT. The verbatim responses to the question on costs are shown in *Tables 17* and *18*. As can be seen, there were 29 written replies, some services making good attempts, to the question in the neonatal screening questionnaire. However, replies to the HVDT and SES questionnaire were much poorer, with only four reasonable responses being received.

Results

Even though there was a more reasonable response to the questions on the neonatal screening questionnaire there was such a mixture of responses that it was not really possible to aggregate the information to provide a unified view. It had been noted that this might be a problem when the questionnaire was piloted and two members of our Key Advisory Group designed a study to give more systematic data in a small number of provider units.[1] The data items collected at each of ten centres are presented in a report by Stevens and colleagues (1997), which can be consulted for further methodological information.

The ten providers who were asked to participate in the study were identified because:

(i) nine had an on-going neonatal screening service which was reasonably stable
(ii) all had a post-neonatal screen through HVDT or HVS
(iii) all could identify staff costs
(iv) all could provide audit type data about their service, for example, how many tested, referred, how many visits per referral

[1] Our thanks to Dr John Stevens, Professor David Hall, Mrs Catherine Davis and Mr Simon Dixon for helping us in compiling the data presented here and for allowing us access to these data for this report.

TABLE 17 *The 29 written replies to the cost question for neonatal screening within the questionnaire*

0.1 wte nursery nurse + equipment + servicing.
After capital costs 1 day per week for screen, 1 day per month for follow-up.
Annual cost £2700, 0.1 wte nursery nurse salary £1200, consumables £900, 4 hours audiology scientist per month £600, no costing for follow-up BSER testing given (including electrodes £500).
We have provided a cost analysis for the proposed changes to the programme.
ABR screener – £6450 + VAT, OAE – £10,000 + VAT.
Cost of machine, £7000.
Professor David Hall now has this information. Total cost of screen and follow-up to 3 months per 1000 births, cost = £13,747. No data on numbers tested.
Approximately £55 per test.
Approximately £12 per baby. This will be less in future because we will use less doctor time and limit more babies per session.
Provided by existing funds except 1 day per month of clinical/clerical staff.
£627 for 94 based on time for testing (does not include postage, administration, etc.).
£73.19 per test with follow-up costs to 3 months and 40% overheads.
No contract.
About £50 per case tested in 1992 (see report for details).
Midpoint MTO3 1 session per week and cost equipment, travel and on-costs.
Approximately £40 per screen with 80–90 screens annually.
£7441 per year including pay and non-pay costs.
Anticipate revenue cost of £15,000 per year for targeted screening of 350 babies per year from test time of 5000 live births.
£124.33 per test with follow-up costs to 3 months and 40% overheads.
£131.88 per test plus follow-up to 3 months of age with 40% overheads (£89.25 if numbers tested are 314 plus 150 others).
£26,400 for equipment, £7500 per year for salaries, £8000 per year additional.
£4800 per year (consists of salary, overheads, and depreciation of equipment).
The service is not costed as separate to the overall audiology service provided.
£20.25.
£4000 for equipment.
Cost of F grade nurse 20 hours per week, cost of Medelec® screener and servicing, cost of small ultrasound bath for cleaning earpieces (plus costs of consumables).
£60.62 per test with follow-up costs to 3 months and 40% overheads.
(Part of the Paed Audiol Services). £30.40 per head – does not include cost of testing of at-risk group at 6 months in secondary centre. £25.49 per test with follow-up costs to 3 months and 40% overheads.
Reported in audit report and also to John Stevens. £14.03 per test with follow-up costs to 3 months and 40% overheads.

TABLE 18 *The 13 written replies to the cost question for the HVDT within the questionnaire*

Together 36.5 weeks x £500 per week = £18,250.
No, not exactly.
Impossible to cost as part of surveillance, would have to set up data collection specifically for this to get accurate figures.
No – cost for 1 tier service only, not broken down.
Formal costing of each of these services has not been carried out.
No – will be done as part of study on BeST test.
Information on this was provided to Professor David Hall/John Stevens.
£17,156 salary and equipment.
Part of health visitor service.
No idea.
Not available.
Total cost per year and 40% overheads = £46,644 or £18.14 per child screened.
Awaiting details from nurse management.

(v) they were known to have a research or audit interest in hearing screening.

The neonatal screen was in many ways much easier to cost because of the small numbers of staff involved in the testing compared with the HVDT (see comments above). However, the follow-up was less easy to cost for both types of screening because of the larger number of alternatives, for example, including ENT, and the difficulty in knowing how many follow-up appointments were made on average. Missing data were taken from the mean of the other respondents when the data missing played only a small part of the overall total. If a large amount of data was missing, then there was no alternative but to drop that centre from the costing of that particular service. This was done most frequently for the HVDT follow-up data.

Salary and other costs were estimated at the level current on 1 April 1994 and included London weighting where appropriate; 12% was added for employer's costs and then an additional 40% to cover other overheads, such as central staff costs, accommodation, and equipment depreciation. No allowance was made for start-up costs such as training and equipment.

The estimated standardised costs for the neonatal screening programmes are presented in *Figure 7*, including sufficient follow-up to establish the correct state of false-positive referrals. Eight out the nine centres performing neonatal screening supplied data that was of use (the letters associated with the provider trusts are as presented in Stevens *et al*, 1997).

The three universal screening sites supply data that are very similar (range £13,100–14,800 per 1000 births), bearing in mind that one is a research programme using nursing staff with a coverage of 89%, the second is a routine service using mainly ATO staff with a coverage of 92%, and the third uses a behavioural test with an estimated coverage of 99%. There is also remarkable agreement between the

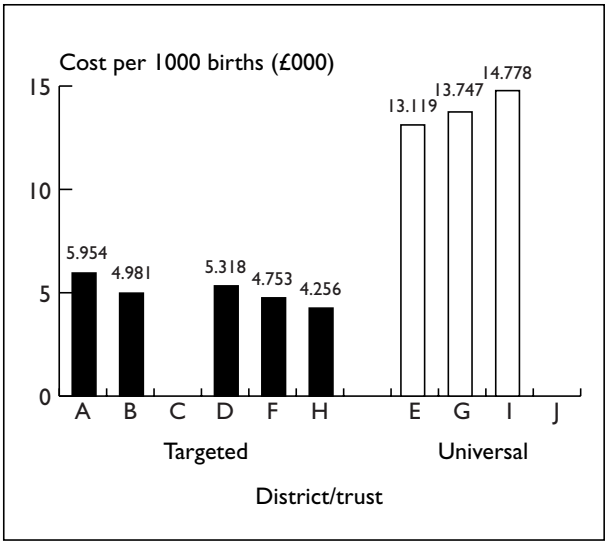

FIGURE 7 *Cost of neonatal screening in eight Districts with 'mature' programmes expressed as a cost per 1000 babies born for targeted (■) and universal (□) screening*

programmes that undertake targeted neonatal screening, with a range from about £4200 (testing 528 of 3000 babies per year) to £5900 (testing 471 of 6955 babies per year) per 1000 births.

There is much greater variation in the HVDT data, as shown in *Figure 8,* which presents, for nine Districts, the core testing costs, with the follow-up costs added to those for which the data exist. Again, the follow-up costs are those associated with appointments sufficient to establish the correct state of false-positive referrals. The follow-up costs are not that dominant in determining the overall costs; the variability comes from the actual screening testing. District J (with the lowest birth-rate of the providers – 2600) has the highest cost and does not have neonatal screening. It uses two health visitors per session for its testing, has a moderate percentage (14%) of children tested at home and tests ten children per session. The cost per 1000 children in the target population of District J, a highly urban area, is nearly £27,000. District E, which has UNS, has the second lowest HVDT cost at £17,000 per 1000 target population. This District uses one health visitor and one trained assistant at each session, seeing an average of 9.6 children, for a birth cohort of about 3700; very few children are seen at home. District B has the lowest cost, about £14,000 and uses one health visitor per session, seeing an average of eight children, in order to carry out a structured surveillance of a birth cohort of 6000 children. In general, the Districts with higher costs tend to use two health visitors per session and a higher proportion of children are visited at home. All programmes, apart from Districts carrying out surveillance, aimed to screen at about the 35 dBA level.

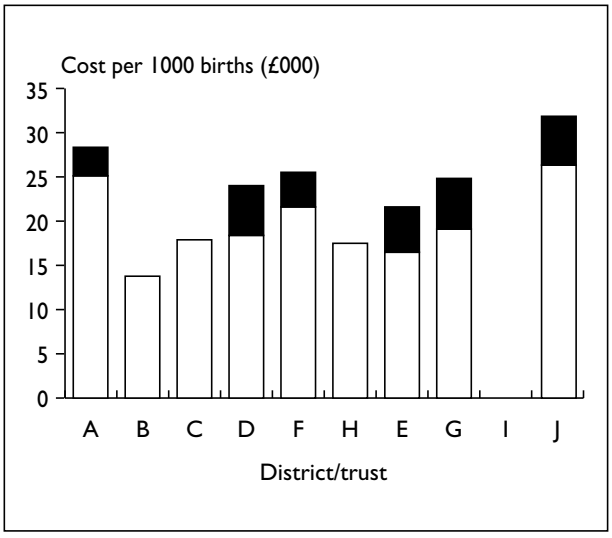

FIGURE 8 *Cost of post-neonatal screens in nine Districts (as for the neonatal screens) in terms of cost for the screen (□) and in some Districts a separate cost for the follow-up (■)*

The approximate average costs for the different types of screening programmes investigated here are shown in *Figure 9*. Inevitably, many assumptions had to be made to average the data in this way but it does give a fairly clear picture to a prospective purchaser of the standardised costs involved. Thus, for a birth cohort of 1000 children, the targeted neonatal screen, which is attempting to test about 8% of the birth cohort, costs about £5000, averaged over five Districts. The universal screen, which will achieve a coverage in the region of 92–95% will cost in the region of £14,000, averaged over three Districts.

Post-neonatal services were conducted in nine Districts, with the average standardised cost (for 1000 births) being about £19,800 for screening and

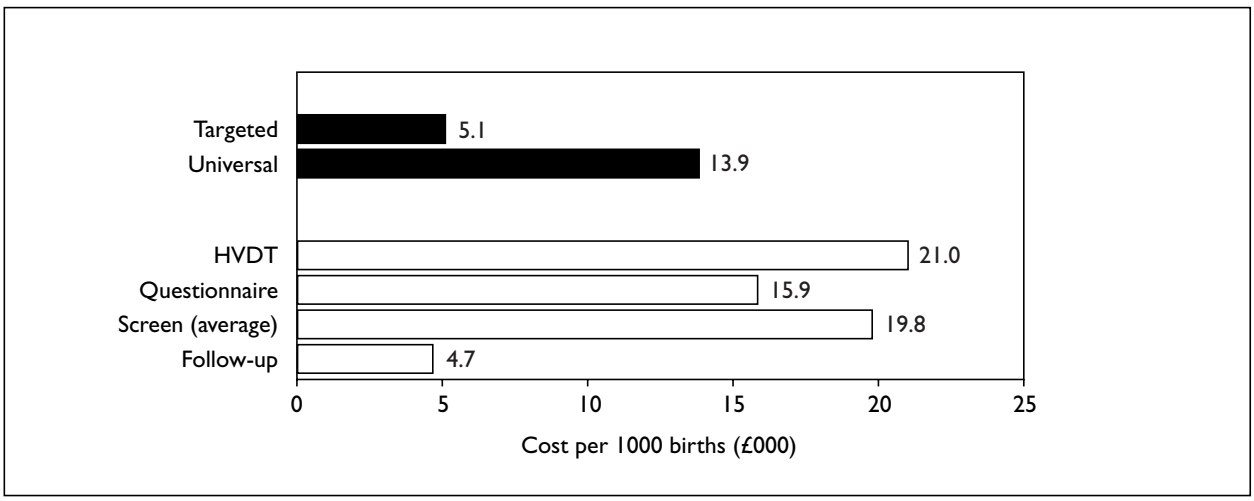

FIGURE 9 *Average cost of different types of screening programmes expressed as a cost per 1000 births (1994) in £000 (Note: the neonatal screens include follow-up costs) (■, neonatal; □, post-neonatal)*

£24,500 when follow-up is included. The HVDT was conducted in seven of the Districts at an average standardised cost of about £21,000 per 1000 births. The two Districts which used surveillance questionnaires instead of the HVDT had an average cost of £15,900 per 1000 births, about £5100 less per 1000 births but with no difference in follow-up costs.

The difference in the cost of UNS and the HVDT (excluding surveillance methods) is larger than was expected and is of the order of £11,000 for an annual child birth cohort of 1000. Taking the mean birth-rate per District, 5300, the overall cost of a universal HVDT would be about £130,000 and that of the UNS about £56,000. The difference is of the order of £68,000 for the average District in terms of cost alone. Another way of looking at the HVDT costs is that it is equivalent to between four and five full-time health visitors (when all overheads are included as explained above). From our survey of current practice, we estimate about two full-time per provider or three full-time per District. A difference of this magnitude and direction might be expected between the top-down and bottom-up methods of cost calculation. The cost of targeted neonatal screening for such a District would be £27,000. At present, Districts using targeted neonatal screening tend to run universal HVDT screening. Thus, the overall cost to a District of this size running a targeted neonatal screening programme would be about £157,000.

Targeted neonatal screening reduces the age of detection for children at risk who are included in the targeted programme. The evidence from our survey that the screens are complementary to each other is presented above – each seems to have an approximately equal yield in the range of 25–35% of those expected to be congenitally hearing-impaired in 1993–94. However, there is some other evidence, from the study of all Districts in Trent (Fortnum *et al,* 1997), that the yield of the HVDT was actually diminished by the introduction of targeted neonatal screen. One factor is that, during 1989–93, the Trent Region did seem to have a higher yield from the HVDT than the Districts that filled out the questionnaires in our survey. The Trent study was conducted in a much more thorough way, ascertaining individual children rather than asking for aggregate figures from the programme coordinators. It suggested that targeted neonatal screening might account for up to 40% of cases, with the HVDT adding another 20%. Without targeted neonatal screening, the HVDT was probably yielding about 40%. The view is, however, confounded by the fact that the providers from 'well funded' teaching Districts, who might do best with the HVDT, are those who have implemented neonatal screening as well.

It is often useful, instead of standardising on the birth cohort, to standardise on the children intended to be tested. This is shown in *Figure 10*. Some comparisons are included from the USA where

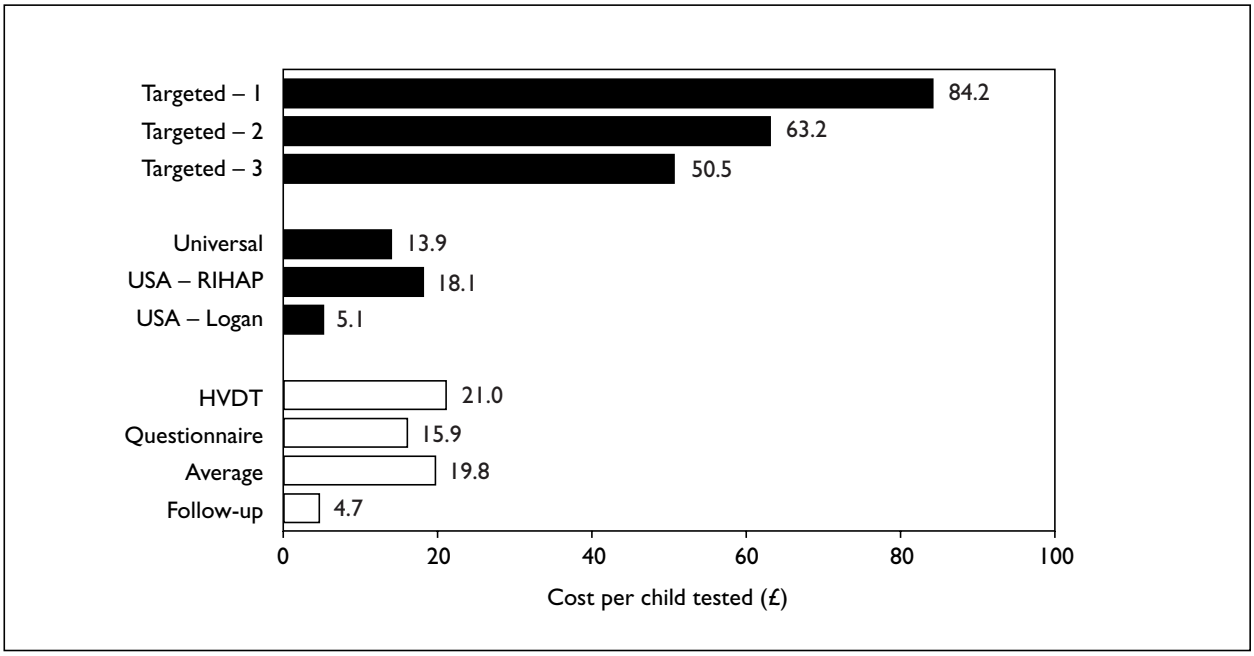

FIGURE 10 *Cost per child tested (£) in different programmes, assuming that about 6% (1), 8% (2) or 10% (3) of births are tested in targeted screening programmes, US data are from White & Maxon, 1995. UK data for neonatal screening includes follow-up costs (■, neonatal; □, post-neonatal)*

programme costs are centred around activity levels more than in the UK. A low estimate has been taken from Logan, where the screening is performed by volunteers or non-technical staff as fill-in activity and the 'cost' of salaries for screening is absorbed by others; at the other end of the spectrum we have taken White and Maxon's estimate for the Rhode Island screening project. Thus, for those Districts who have implemented UNS in the UK, the cost per child screened is about £13.90. This falls right in the middle of the two estimates from the USA. The post-neonatal screens are broken down into those that use surveillance, £15.90 per child, and those that use the HVDT, £21.00. The figures for targeted neonatal screening are calculated assuming the percentage of the birth cohort to be tested is (i) 6% (ii) 8% and (iii) 10%, (i.e. 6% NICU/SCBU > 48 hours and 4% family history cases). This gives a range of costs between £50.00 and £84.00 per child tested. Three major factors increase the cost for targeted screening:

(i) there is a higher degree of skill needed to test very small babies who are either being discharged back home, after major health concern, or who are transferring from NICU to an SCBU elsewhere
(ii) finding the children/families with a family history or CFA takes extra time and effort, as well as the on-going training of maternity staff
(iii) quality assurance is more often built into the system.

If these data are taken as fairly typical of the services that are presently functioning, and if it can be assumed that the data on yield reported above is typical of the screens as used in the UK, two approaches to assessing the cost-effectiveness of the different screening protocols for finding PCHI at 50 dB HL or greater can be made. These assumptions are probably more correct for the HVDT, which has been operating for a longer period than for the targeted neonatal screening, much of the implementation of which is *ad hoc* compared with the centres that have provided the costing data. In the first assessment **(a)** the data gathered in the survey of current practice here will be used, and in the second **(b)** the data gathered in Trent will be used (Fortnum *et al,* 1997; Wood *et al,* 1997).

The data used in **(a)** are fairly optimistic and are derived from the 52 providers who gave details in our survey about their numbers tested and the yield that they had derived for 1994. The yield from the UNS is high compared with that expected from Fortnum and colleagues (1997), which suggested a prevalence of about 0.9 per 1000 for ≥ 50 dB HL, but it has a large confidence interval extending down to about 0.7 per 1000 which has been used as our estimate of yield **(b)**.

The yield for the targeted screen in **(a)** is a mid-estimate from the range between 0.6% and 0.9% yield estimated in chapter 4. This is somewhat high for a number of reasons, the major one being that the screens included in the survey of current practice appear to be highly selective. This yield has been decreased to 50 per 10,000 in **(b)** which is in line with that shown in the Trent study (Fortnum *et al,* 1997) and by Lutman and colleagues (1997) (see chapter 7). The yield for the HVDT is 2.4 per 10,000 (estimate **a**) in the survey conducted here and in the region of 3 per 10,000 in the Trent study (estimate **b**).

The estimates of the sensitivity of the screens are derived from a number of sources (see chapter 7 and the current survey) and are not used directly in the calculations except for UNS (estimate **b**). It should be borne in mind that the coverage for the HVDT is of the order of 90%, for UNS it is a few per cent higher, and that for targeted neonatal screening there is a maximum potential detection level of 60% (i.e. the proportion of the congenital PCHI population that has one of the three major high-risk factors).

The cost per child detected is one figure that can be used to index the cost-effectiveness of the different screens. It can be seen from *Table 19* that the HVDT is currently the least cost-effective. There are probably three major contributors to this high cost. The first is that two trained testers are required; the second is that there is a low yield, partly because by the age of 6–9 months several hearing-impaired children have already been referred through targeted neonatal screening or professional/parental concern; and the third is that there is a low sensitivity and specificity for the test caused, for example, by:

(i) the low level (35 dBA) at which the test is carried out
(ii) the large number of testers that have to be trained.

There may be technical improvements that could be made, for example, the BeST test (see chapter 7) to reduce the number of testers needed, and a higher presentation level could be used. This would reduce the overall cost. An estimate can be derived from the health visitor questionnaire

TABLE 19 *Two estimates for the cost of each hearing-impaired child detected (50 dB HL or greater) based on the survey of current practice and the cost survey. The values for yield have been chosen to show how sensitive the estimates are to varying conditions*

	HVDT	UNS	Targeted neonatal
Cost per child tested	£24.50	£13.80	£70.60
Test sensitivity estimates	60–80%	80–95%	80–90%
Yield per 10,000 children tested (**a**)	2.4	14.0	75.0
Cost (£000) per child detected (**a**)	102.1	9.9	9.4
Yield per 10,000 children tested (**b**)	3.0	7.0	50.0
Cost (£000) per child detected (**b**)	81.7	19.7	14.1

approach, which only requires one person, costs £15,900 per 1000 (or £20,600 per 1000 with follow-up). However, it is difficult to see a change in the relative ranking of the different screening protocols. Perhaps the most cost-effective that the HVDT could become, based on present data, would be a highly optimistic £40,000 per hearing-impaired child.

The cost-effectiveness differential between targeted and UNS is less than previously reported, (see, for example, Davis, 1993b; Davis & Parving, 1994). This difference is due to the higher costs of targeted screening than assumed previously and the higher yield of UNS found here.

In terms of flexibility for improvement, there is probably scope for targeted neonatal screening to become more cost-effective. Better targeting of children with a family history and more streamlined automated testing would clearly increase its cost-effectiveness.

Summary

This costing exercise has been useful in pinpointing some of the differences that exist between the screening protocols. In terms of running costs (excluding start-up and equipment costs but including employers' full cost plus 40% overheads), in the ten Districts surveyed the HVDT screening costs about £24,500 per 1000 target population. This is reduced to about £20,600 when surveillance is used instead of the distraction test. The mean standardised cost of UNS is about £13,900 in the three Districts who are presently conducting such screening in England. Targeted neonatal screening is costing a mean of about £5100 per 1000 births.

The cost per test and the cost per hearing-impaired child detected were investigated to examine the relative differences between the protocols. Under different sets of assumptions, the relativities favoured neonatal screening over the HVDT quite substantially. However, for the neonatal screening programmes, purely in cost terms, there has been no allowance made for the **additional** 12–18 months health and educational support that hearing-impaired children would need because they had been found earlier. This is estimated at direct health costs of £2000 per hearing-impaired child detected, in terms of hearing-aids (£800), earmoulds (£200) and assessment/rehabilitation time (£1000). If there were 50 sessions of peripatetic educational support also associated with this extra period, that might add in an extra £4000–6000, giving a total of perhaps an extra £8000 per child. This would still represent an overall saving but it should be emphasised that there will be substantial additional costs to education if, say, even 80% of all hearing-impaired children were found and habilitation started by 6 months of age. Overall, identifying congenital PCHI by 6 months of age would amount to about a mean of 27 months[2] extra educational support for each hearing-impaired child, or 36 months extra for moderately impaired, 19 months extra for severely impaired, and 9 months extra for profoundly impaired children. In the long term, the earlier expenditure may in fact enable savings from possible less intensive support later in life. However, at present, that is speculation and can only be addressed by a proper prospective study.

It must be emphasised that these costs, especially education costs, are highly speculative compared with the detailed work reported above and by Stevens and colleagues (1997). Further research is needed to examine the cost-effectiveness of the different screening models in further detail and to predict the cost implications (including variations) that might arise from different strategies. The capital costs of the different approaches have not been discussed, nor the opportunity costs,

[2] These data are difficult to interpret as it is unclear what an appropriate baseline might be; that is, should it be the present system or how the present system is **supposed** to work?

social costs or start-up costs in any great detail. Start-up costs are discussed further in chapter 9 but the other issues were considered to be outside the scope (and budget) of the present study. One weakness in these costing exercises is that they may underestimate the costs of running a programme and of keeping all contributing departments informed of the progress of the programme.

Chapter 6

Focus Groups and visits

Scope

The largely qualitative findings from the meetings of the five Focus Groups are summarised in this chapter, together with those from the review team's planned visits to services and colleagues in North America and the UK.

Focus Groups

During this review, five Focus Group meetings took place (in Glasgow, Manchester, Nottingham, Bath and London) involving professionals from health and education services, parents of hearing-impaired children, voluntary groups and purchasers. Details of the composition and methodology of the Focus Groups are given in Appendix 1. Discussion was allowed to flow freely to some extent but the leaders of the Group (members of the review team) guided the discussion through a series of topics/question that had previously been identified by the review team and sent to Focus Group invitees prior to the meetings (see Appendix 1).

The Focus Groups do not provide a representative sample of professionals or parents, and such was not the intention (Kitzinger, 1995). Nevertheless, a total of 108 people, mostly health and education service professionals, together with some parents of hearing-impaired children, attended the whole-day meetings, thus providing the review team with a valuable opportunity to explore preconceptions and expectations. In particular:

(i) to identify and gauge the extent of any consensus on service and screening issues
(ii) to provide a source of partial validation for some of the data presented in chapters 2 and 4
(iii) to confirm that the review and the questionnaire survey were free of any serious omissions in subject matter
(iv) to build on existing networks both for input into the review process (in, for example, the search for unpublished studies), and for building up ownership for any later dissemination efforts.

Those areas/issues/concerns which emerged from the Focus Group discussions with a degree of consensus are discussed here. Note-takers were present at all Focus Group meetings and their near-verbatim notes were summarised by a team member into six digests of 3–6 pages. (The extra summary was devoted to the comments of a group of nine parents of hearing-impaired children who made up a separate discussion group at the Manchester Focus Group meeting; their views will be dealt with separately.)

The satisfaction of the Focus Group participants with current services in their particular areas was (not unexpectedly) variable; however, it was generally low enough, on average, on a scale of 1–10 to give some cause for concern. Services and quality of screening for hearing impairment were thought to be patchy; a very few participants expressed satisfaction, with ratings of up to 9 on the scale, but most ratings fell in the range of 4–7. Ratings of satisfaction with services tended to be high when they:

(i) were well-resourced
(ii) included good follow-up
(iii) had good information systems
(iv) had effectively coordinated hospital, community and education services.

Large metropolitan areas face particular problems in clinic attendance and screening coverage caused mainly, it was argued, by population movement. Dissatisfaction and low rating were associated with poor service coordination (between audiology and ENT, hospital and community, health and education services), the perceived low quality and effectiveness of the HVDT, late identification of permanent hearing impairment, and concern about the identification of late-onset hearing impairment.

Apart from varied and detailed concern about the implementation and effectiveness of the HVDT, there was widespread support for the introduction or continuation of targeted neonatal screening, on the grounds of apparent cost-effectiveness, and for the value of SES, which was seen as an easy-to-implement and useful 'longstop' for any late-onset cases not identified and for earlier false-negatives (largely, though not exclusively, mild high-frequency or unilateral losses).

Questioned about the 'ideal' age for identification of moderate or greater congenital PCHI, there was generally strong **belief** – on the basis of parental rights to significant knowledge, outcome benefits and ease of introduction of aspects of early intervention – that such children should be identified well within the first year of life and, more specifically, within the first 6 months. The distinction between identification of a hearing loss and first fitting of hearing-aids was frequently acknowledged, and there was a strong (though not universal) consensus that identification by 2–4 months of age and appropriate hearing-aid fitting (a process to be approached with care and accurate assessment) by 6–7 months of age would be highly acceptable. Proper diagnostic procedures (as suggested, for example, by the EU Concerted Action on Genetic Deafness (Parving, 1996)) following identification were often stressed, such investigations being easier at earlier ages. The same point was made about unilateral losses although, this aside, it was considered that the case for very early identification of unilateral and mild hearing loss was difficult to make (and consideration of screening of these is not covered by this review – see chapter 1).

Notwithstanding the widespread consensus between the Focus Groups for identification of moderate or greater hearing loss within the first 6 months of life, there was no overwhelming support for the **immediate** introduction of UNS, although most participants expected it to become more common over the next decade. The reasons for the hesitation to endorse UNS had largely to do with priorities for service improvement – the most obvious consensus across all aspects of all Focus Groups' concern was about service fragmentation and lack of adequate coordination within health services and between health and education services: "Both the NHS and local authorities have responsibilities for arranging and funding services to meet peoples' needs for continuing care. Collaboration is crucial to ensuring the effective and integrated delivery of care" (*Responsibilities for Meeting Continuing Health Care Needs* (NHS, 1995)). In the light of this, the implementation issues associated with the introduction of UNS (rather than targeted screening) were seen by Focus Group participants as too great, giving rise to badly-managed cases and high levels of parental anxiety. Since outcomes are likely to be crucially affected by quality of intervention, and not just age of identification, Focus Group participants remained largely sceptical. However, some participants (more likely to be educationalists, parents or voluntary sector members) acknowledged the implementation issues, but argued that the introduction of UNS would tend to precipitate solutions, and they were therefore in favour of UNS.

Thus, there was widespread support for targeted neonatal screening (targeted at defined at-risk groups), followed by universal HVDT (despite widespread doubts about its effectiveness) and SES. There was widespread support for identification of moderate and greater congenital PCHI within the first 6 months of life. There was some limited support for UNS but more widespread concern about the implementation of such a potentially sensitive screen on top of existing poorly organised and poorly coordinated services. There was widespread agreement that, for (a) epidemiological and (b) implementation reasons, UNS would still have to be followed during the first year of life by further effective screening or surveillance measures. This raised interesting issues as to what such measures might be, and whether they might be universal or targeted. The idea, for example, of targeting the HVDT (following UNS) was raised.

These views permeated the priorities for service improvements. Overwhelmingly, participants identified the need for better coordination and better information systems, with joint care plans, effective team support, family-centred services, greater responsiveness and easier access to services high on the priority list. The need for **year-round** high-quality education service support via ToDs was frequently referred to, with the implications for ToD training and recruitment stressed.

The NDCS Quality Standards were well-known and welcomed. Participants felt that they were useful in promoting discussion between purchasers and providers, and that the targets set in the first volume (NDCS, 1994) were ambitious but appropriate. There was widespread awareness that a second volume was about to be published (NDCS, 1996).

While parents of hearing-impaired children were participants in all five Focus Groups, the numbers involved in the Manchester Group gave an opportunity to set up a discussion group comprised of parents alone. In general, this group felt let down by the services and wanted standards to improve. The HVDT was given an average rating of 2.5 (on a scale of 1–10) and the responsiveness of the service to parental concerns an average rating of 4.5. Parents did not feel that they were treated as partners and claimed that they often felt dismissed and ignored. They noted 'huge' differences in

service and service quality from place to place (in both health and education) and several had considered or even been advised to consider moving in order to access better quality services.

The parents considered that the earlier they knew about their child's hearing loss, the better. All agreed that identification should occur by 6 months of age and many considered that by 6 weeks of age would be appropriate. For a few, however, age 6 weeks was considered too early because of bonding and issues concerned with post-natal depression. However, neonatal screening (targeted or universal) was not seen as the answer to early identification, since the parents were unconvinced by the sensitivity of potential screens. As might be expected, parents were less aware of technological advances than professionals, and their scepticism may owe something to their negative experiences with other screens (particularly the HVDT). They doubted that the support services were in place to deal effectively with neonatal screen failures. Interestingly, however, they put more faith in their own observations of their children, and felt that good surveillance and high levels of responsiveness to parental concern (including open-access referrals) could be made more of than at present. Although this may be true of some parents (such as these, who were able and willing to attend a Focus Group meeting), the literature is not particularly encouraging towards parental concern as the major route to early identification (see, for example, Watkin *et al,* 1990; Sutton & Scanlon, 1996).

With regard to post-diagnostic services, there was a general feeling in the parental group that they had received very little support and very little follow-up care. Information was sparse and choices limited. Experiences of education services were mixed; some spoke positively about their ToD support, others were highly critical. Overall, the parents expressed the following views of the follow-up services.

1. Too little choice is offered; services were not perceived as being family-centred.
2. The different services should be better integrated.
3. ToD support should be at least weekly until the child is 2 years old and should not cease during school holidays.
4. ToDs need to work more **with** the parents rather than 'taking over'.

Asked to consider other desired improvements, the parents were in complete agreement that early and accurate 'diagnosis' (that is, identification) was an overwhelming priority, followed by an increased choice of good quality hearing-aids. There were also calls for treating the child as a whole, improving interdisciplinary working, nominating a key worker, and developing family-centred rather than 'ear-centred' services – that is, services which take into account the dynamics of the family, and which provide both parents and families with full information with which to make management decisions, rather than have them imposed by professionals.

In summary, the highest priority for participants in the Focus Groups was for better information systems and better integrated services. Early identification was also a high priority and a majority of participants would welcome UNS, but there was not a consensus on this issue because of the concerns raised above, which mainly stemmed from present inadequate coordination and lack of appropriate back-up services. There was widespread agreement that a 'shared list' of hearing-impaired children would be helpful.

Focus Groups remain a debatable source of information, with conclusions that are difficult to calibrate and which may lack scientific rigour. Nevertheless, in this instance they provided:

- clear evidence of consensus in some areas
- further confirmation of data presented in chapters 2 and 4
- confidence in the appropriateness of the survey questionnaires
- access to some unpublished information
- an expanded network for dissemination.

Visits

The purpose of the visits to a number of services in the UK and North America was to provide up-to-date contextual information on research, service, and service development that would not be found in the literature. Visits to North America were particularly helpful in gauging the impact of the NIH consensus (NIH, 1993) and to observe first-hand the organisation and implementation of universal neonatal hearing screening programmes. Although there are many examples of targeted screening programmes in the UK, universal programmes are rare.

Visits to North America were made in July 1995, as follows below. We were highly grateful for the contributions made by Karl White and Harry Levitt, both in arranging the meetings and providing us with much material.

(i) ***Seattle, Washington, USA: University of Washington and Children's Hospital.*** Discussions were held with a leading ENT specialist (Gates), auditory physiologist (Rubel), and with the research audiologist responsible for managing the current large-scale NIH-funded project designed to examine the efficacy of different protocols and techniques for neonatal screening using primarily a high-risk targeted population of babies (Norton). Useful meetings were also held with Werner and Kuhl. In particular, the latter furnished articles concerning auditory plasticity.

(ii) ***Rhode Island, USA: Women and Children's Hospital, Providence and Newport Hospital, Newport.*** Discussions were held with the manager, audiologist, paediatrician, Director of Public Health, ToD and those screeners involved in the implementation of the statewide mandatory universal neonatal hearing screening programme. Routine screening sessions were observed in both Providence and Newport.

(iii) ***Nashville, Tennessee, USA: Vanderbilt University.*** Discussions were held with the audiologist (Hall) responsible for a research team investigating aspects of distortion-product otoacoustic emissions (DPOAE) for screening.

(iv) ***New York, USA: Albert Einstein Memorial Hospital.*** A multicentre neonatal screening project to assess efficacy across NY state is managed from here (Gravel). Screening procedures were observed and discussions on the project held; further discussions were held (Stapells) on aspects of electrophysiological follow-up assessments of babies referred by screening.

(v) ***Toronto, Ontario, Canada.*** Discussions were held with Hyde and Riko from Mount Sinai Hospital on their long experience with aspects of neonatal screening, and service policy and plans within Ontario.

(vi) ***Toronto, Ontario, Canada.*** Discussions were held with Picton concerning the progress with neonatal screening in Canada (particularly the Ottawa programme) and the possibility for rapid ABR screening/assessment in the future using frequency-specific ABR techniques. This discussion also elicited much new material on auditory plasticity, and a context for on-going research in behavioural-evoked responses using multiple electrode recordings.

(vii) ***New York, USA.*** Discussions were held with Jont Allen and Pat Allen about the technology of emissions, in particular the prospects for DPOAE.

(viii) ***Portland, Oregon, USA.*** Several people's views on neonatal screening were sought at a conference on tinnitus held in Portland; Naunton (NIH), Tyler (Iowa), Vernon (Portland), Dobie, (Texas), and Eggermont (Canada) were particularly helpful.

The visits were valuable in providing **context**; in terms of **evidence**, the visits also provided us with background material (e.g. training and implementation material) and literature references not identified in our literature search. The key problems concerning implementation were highlighted several times.

Given the necessarily limited nature of our visits they were open to the charge of being unrepresentative and giving merely an impressionistic view of the impact of the NIH consensus. However, subsequent visits (to major conferences on early identification issues in Iowa and South Carolina) have confirmed the major points that emerged, which were as follows.

1. The NIH consensus has brought the issue of age of identification of PCHI to the fore as a public health priority, and several important on-going research projects are currently being funded. Implementation of UNS is increasing, but still accounted for only 2–3% of the total birth cohort in 1994. In some states UNS is mandated but generally it appears that 'bottom-up' implementation – by providers and provider consortia – will prevail. Implementation will continue to increase but is more likely in urban than rural areas because of ease of access to the population and general level of healthcare advancement in urban areas. Costs of screening and follow-up are non-trivial obstacles to implementation and the spread of screening programmes. However, the benefit arguments are strong since, without a well-developed Community Health Programme in the USA (although there are exceptions (Blackman & Hein, 1985) the USA has no equivalent to the HVDT screen), age of identification of PCHI tends to be later than in the UK. There was some feeling that if and when a cheap hand-held device (for emission screening) became available, universal screening would take off. At this stage it would not matter whether the screening was done at a hospital or at home during a post-natal visit.
2. Current research, in particular, the NIH-funded multicentre project on screening

protocols coordinated from Seattle, Washington, by Norton, is likely to indicate that the particular technology chosen for universal screening is less important than the details of and control over service organisation. Of particular importance are monitoring, information and tracking systems, information for parents, and training and supervision of those undertaking screening. Some aspects of some maternity practice in the USA may assist in neonatal screening; for example, longer maternity stays (currently) than in the UK, routine separation of mothers and babies at certain predictable times.

Visits were made in the UK to the only two services which routinely provide UNS: Hillingdon (Tucker and Bhattacharya, using the portable ARC (PARC)), and Whipps Cross/Waltham Forest (Watkin and Baldwin, using OAEs). Both services were characterised by detailed records, close monitoring and a commitment to high standards; they are led by senior practitioners who regard audiology as their primary discipline, and who are aware of relevant research developments. Both services have published the results of their programmes and these are dealt with in chapter 7. It is enough to remark here that both examples of implementation of universal screening in the UK appeared effective and well-monitored, were welcomed by parents, and involved close co-operation between health and education services.

During the period of this review (1995–96) there was a funded trial of UNS led by a group in Southampton, and implemented in Southampton, Portsmouth, Swindon and Bath. This team were visited in March 1996. Interim data have been presented by Kennedy (1996) and the team agreed to provide the review with their findings; these are dealt with later in chapter 7. This programme has now ceased and the children tested are being followed-up to find the false-negatives and to finish the comparison between the HVDT and the neonatal screen.

Finally, a visit was paid to Otodynamics Ltd, manufacturers of the ILO® range of OAE equipment. This company has contributed a statement on screening developments, aspects of which are considered in chapter 8.

Further visits were not undertaken, since our extensive consultations using the Focus Groups and the senior authors' research and service contacts in the UK would have rendered them less valuable.

Summary

The focus groups were invaluable in gauging the opinions and beliefs of a range of professionals involved in hearing screening and habilitation. A major lack of coordinated information was highlighted. The groundwork was undertaken for later dissemination of the review's recommendations. The visits, both in the UK and the USA, demonstrated the feasibility of neonatal screening in several different contexts, as well as the challenges of implementation and follow-up that have to be overcome by training and appropriate management.

Chapter 7

Performance of hearing screens

Scope

The evidence from studies of the performance of both neonatal and HVDT screens is reviewed in this chapter.

Introduction

The HVDT has been in place as an almost universal screen across the UK since the 1960s. While protocols and test details vary from area to area, the most common format involves a two-person distraction test, using frequency-specific stimuli presented at quiet levels (e.g. 35 dBA) to the side and slightly behind (at 45 degrees) the 8-month-old infant, who is seated on the parent's knees. A full localisation response to all stimuli on both sides represents an acceptable pass. Failures are usually re-tested once at a later date, except in the case of obvious concern, and two test failures constitutes a screen referral. This may be made to a secondary or tertiary audiology department, to a GP for onward referral, or to an ENT department. Further details are available in McCormick (1993) and Hall (1996).

The arguments which were marshalled in the 1940s and 1950s (Ewing & Ewing, 1944; Whetnall, 1955; Ewing, 1957) to support the introduction of the HVDT screen have, since the 1970s, been used to argue for neonatal screening for hearing impairment. These arguments – broadly, that parents want very early identification and that earlier identification will be beneficial – have interacted with four further significant developments:

(i) consumer concerns about the quality of the HVDT screen, (see, for example, NDCS, 1983)
(ii) the development of apparently viable techniques for neonatal screening
(iii) the trend towards hospital birthing giving a 'captive' test population
(iv) an increasing knowledge of the epidemiology of PCHI upon which rational service provision options can be based.

The result, in terms of service provision, has been somewhat patchy across the UK. The HVDT screen is still very widespread, almost universal. Some providers have abandoned it, however (e.g. Scanlon & Bamford, 1990; Watkin, 1996a). Surveillance or programmes stressing early professional and parental vigilance have burgeoned (McCormick, 1988; Scanlon & Bamford, 1990; Hall, 1996). Some degree of neonatal 'screening' of at at-risk babies is now in place in perhaps two-thirds of the Districts in the country (see chapter 4), and UNS is provided in at least two hospital trusts in England.

The evidence for the performance of these screens is examined below. There has been much fundamental and semi-applied research on the test techniques, particularly for ABR and OAE, which served to establish the biophysical bases for them, optimal recording parameters, and so on; however, these studies are not within the remit of this chapter. Our starting-point is that the HVDT, OAE, ABR and PARC are being used as the basis for first-year screening programmes in the UK and the aim is to review those key studies by which an evaluation of screen performances can be made. Since the public health context is that found in the UK, there is a selection bias towards UK publications and the UK context, although other studies are included where relevant. For methodological reasons there are no published RCTs in this field, and the published studies that exist are somewhat mixed, both in terms of methodology and quality. Decisions on which papers to include in this part of the review were made on the basis of size, relevance and quality, and how recently they were published (see Methodology, Appendix 1); publications up to the second quarter of 1996 (and sometimes beyond) are included.

Evidence on neonatal screening

Early techniques for neonatal hearing screening were based upon behavioural responses to sounds, such as the Crib-o-Gram in the USA (McFarland *et al,* 1980) and the ARC (Bennett, 1979) in the UK. The most recent version of the latter is the PARC (Tucker & Bhattacharya, 1992). This device presents 70–80 dB SPL high-pass noise to one or both of the baby's ears, via an earphone or probe assembly. The cradle monitors possible behavioural

responses, such as head turn and body movement, via sensors in the cradle, for both sound and no-sound (control) trials. Software-based decisions are made comparing responses to sound and control trials, culminating in a statistically-based pass or refer decision.

Early ABR technology required trained audiologists to make on-line decisions but, in due course, automated devices were devised for screening by non-specialists, thus reducing staffing costs. Herrmann and colleagues (1995) report one such development. In their study, the ABR screening device was assessed across five published studies for its ability to mirror the results of full non-automated ABR tests on neonates. The combined data on 1187 ears have a specificity estimate (against full ABR) of 96% and a sensitivity estimate of 98%, indicating the efficacy of automated approaches to ABR screening.

In this technique, wide-band clicks are presented to one ear at a predetermined screening level (e.g. 50 dBnHL). Far-field electrical activity generated by the whole nerve action potential of the VIIIth (vestibulocochlear) cranial nerve and the auditory brain-stem pathways are recorded via surface electrodes, and averaged over a large number of stimulus presentations (typically 2000); machine-based decisions are made on the presence or absence of 'Wave V', resulting in a pass or refer outcome. In some cases, hard-copy output is also available for double-checking later by an experienced audiologist.

OAEs (Kemp, 1978) are generated by an active physiological mechanism in the healthy cochlea and can be elicited in response to wide-band clicks presented to the ear via a lightweight ear canal 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 presented, and the responses averaged and repeated to generate an ear-specific, but repeatable, 'waveform'. These are known as TEOAEs and are not apparent with mild or greater cochlear hearing loss, or if there is significant middle-ear pathology. Pass–refer decisions are usually made by the screener on the basis of a combination of displayed statistics. Double-checking by an experienced audiologist is, again, possible later. There are other OAEs, notably DPOAE (Brown *et al,* 1989), but these have found only limited use in neonatal screening, as yet.

It is important to note that these devices assess different functions. While PARC reflects auditory function and consequent reflex responses up to and including the auditory cortex, ABR reflects the integrity of the outer, middle, inner ears and lower auditory pathways, and OAE reflects, primarily, the integrity of outer, middle and inner ears. These distinctions are important diagnostically, and may have implications for screening. There have been some reports (J Stevens: personal communication, 1997; J Gravel: personal communication, 1997) of cases without a recognisable ABR but with a reliable TEOAE response. This would imply functional cochlear activity but damage or immaturity in higher auditory pathways. Such cases are rare but are more likely to be found in NICU/SCBU graduates than in the normal-birth population. Until more is known about these cases, it has been argued that at-risk screening of NICU/SCBU graduates should be done with ABR techniques, with TEOAEs included in the follow-up assessments. Such a testing strategy would avoid the risk of 'false-negative' TEOAE passes in such cases. PARC testing should also refer these cases, and is not (in theory) subject to this risk.

Aspects of the main studies on neonatal screening are summarised in *Table 20*. Some useful studies in which aspects of OAE testing efficacy have been reviewed are not included (Prieve *et al,* 1993; Smurzynski *et al,* 1993; Norton, 1994) because they do not address screen implementation; however, all three present compelling evidence of the potential of OAEs for screening.[1]

The studies in *Table 20* can be divided into those directed at UNS trials, and those dealing with neonatal screening of babies at-risk for hearing-impairment – low birth weight, NICU/SCBU admissions, family history, CFAs, and/or other indicators (see chapter 2).

Of those dealing with universal screening, the (interim) data from Kennedy and colleagues (Personal communication, 1997) (see also Hunter *et al,* 1994) concern the only controlled experimental study in the field. In this 'Wessex trial', all births in four areas were screened by two teams of neonatal screeners, each team moving between two areas in 4-month blocks. Babies born during a 4-month block in one area underwent neonatal screening (TEOAE as in-patients, with immediate screening ABR for those referred) followed by the

[1] The report of the multicentre semi-randomised trial of screening methods (Professor Susan Norton and colleagues) is due in 1997/98. No preliminary data have yet been released from this 3-year programme.

TABLE 20 *Neonatal screen studies*

Study	Sample size	Type of report[1]	Population[2]	Screening method[3]	Aim of screen[4]	Coverage (%)	Test sensitivity (%)	Test specificity (%)	Screen sensitivity (%)	Screen specificity (%)[5]	Yield	Comments
Bhattacharya *et al*, 1984	5000	R	NAR	$ARC_i \rightarrow ARC_i$	40					98.9		Early report on ARC screen
Davis *et al*, 1991	1502	R	AR	ARC_i					50 20			Low sensitivity of ARC for AR cases (50% for severe/profound, 20% for moderate)
Fonseca *et al*, 1996	104	P	AR	Various					91			Retrospective ascertainment study of PCHI cases across a number of centres
Galambos *et al*, 1994	5901	R	AR	A_i	30	50		80 88			2%	Argues for 'diagnostic' testing of NICU graduates. Large retrospective study, but not of 'screening'
Hall *et al*, 1996	1784	R	All births	A_i	35	35		99.7 98.4				Feasibility study of A_i with new device and simultaneous bilateral stimulation. Corresponding specificities for NICU babies still high at 95% (B) and 87% (U)
Hunter *et al*, 1994	213	R	All births	$O_i \rightarrow A_i$	40	90		91.5		99.5		Used to estimate resource needs for UNS and confirm feasibility
Hyde *et al*, 1991	1367 (ears)	R	AR	A_i	20				90.4	95.2		Rare follow-up study allows estimation of sensitivity of A_i for this population; threshold elevation at 2 and 4 kHz was screen aim
Kei *et al*, 1997	568	P	All births	O_o				92				Trial of TEOAE screening in community setting at 8 weeks. 8.1% untestable, fail rate 8%. Results affected by activity state, gender. Total test time 11–17 minutes
Kennedy *et al*, 1997 (Personal communication)	21,190	P	All births	$O_i \rightarrow A_i$ and HVDT	40	91		90.2		98.4	1.2:1000	Only controlled trial of UNS; screen specificity falls to 90.8% in 24-hour-discharge babies; no incremental yield of HVDT when neonatal screening in place; yield of HVDT excluding parental/professional referrals because of concern only 0.1:1000
	29,281	P	All births	HVDT	40						0.7:1000	

[1] *P, preprint; R, published in peer-reviewed academic journal; A, unpublished audit report; G, general review for edited volume or chapter.*
[2] *AR, at-risk only; NAR, not at-risk only.*
[3] *O, OAE; A, ABR; P, ARC; → route for those failing; i, in-patient; o, out-patient; R, at-risk register.*
[4] *Indicates target level of hearing loss (and greater).*
[5] *Sometimes the failure rate, as an acceptable approximation.*

continued

TABLE 20 contd *Neonatal screen studies*

Study	Sample size	Type of report[1]	Population[2]	Screening method[3]	Aim of screen[4]	Coverage (%)	Test sensitivity (%)	Test specificity (%)	Screen sensitivity (%)	Screen specificity (%)[5]	Yield	Comments
Kramer *et al*, 1989	667	R	AR	$R \rightarrow A_o$	30 40			88.2 93.9				Claim 100% sensitivity of A for severe losses, but full ascertainment doubtful
Lutman *et al*, 1997	7500	R	AR	O_i	50				80	92		Large-scale, multicentre trial of Programmable OAE Measurement System (POEMS) device/ screen; of 45 cases, nine passed OAE screen. Reasons (test interpretation, progressive, acquired) uncertain
Mason *et al*, 1997	6983	R	AR	A_i	50 bilateral 50 unilateral				90 82	93 98		Large-scale, multicentre trial of an automated ABR device. Of 51 cases, five passed screen (reason uncertain) on both ears and nine passed on at least one ear. Machine plus visual scoring most effective
Maxon *et al*, 1993	1850	G	All births	$O_i \rightarrow O_o \rightarrow A_o$	20			73				Early report from large-scale US trial, indicating poor test specificity; improves to 85% if 'partial pass' counted as pass
McClelland *et al*, 1992	405	R	AR	ABR_i	aided				100	88		Detailed follow-up after 4 years of age. Median identification age 1 month
McCormick *et al*, 1984a	396	R	AR	$ARC_i \rightarrow ARC_i$	moderate			81	84	96.5		
Rowe, 1991	243	R	AR	A_o	40	86	100	91			1.4%	Tested feasibility of outpatient screen of AR population
Shepard, 1983	218	R	AR	$R \rightarrow ARC_I$						98.6		Suggests high specificity for ARC
Shiu *et al*, 1996	43	A	AR	$OAE_i \rightarrow ABR_i$	50	56			79			Part of large-scale audit. Sensitivity estimate of 81% if small number of other screened cases included

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[4] *Indicates target level of hearing loss (and greater).*
[5] *Sometimes the failure rate, as an acceptable approximation.*

continued

TABLE 20 contd *Neonatal screen studies*

Study	Sample size	Type of report[1]	Population[2]	Screening method[3]	Aim of screen[4]	Coverage (%)	Test sensitivity (%)	Test specificity (%)	Screen sensitivity (%)	Screen specificity (%)[5]	Yield	Comments
Stevens *et al*, 1991	723	R	AR	$O_i \rightarrow A_i$				0 = 80 A = 90		93		Feasibility study of O and A tests for neonatal AR screening. Suggests optimal screen would be O → A for this population. Some outpatient testing
Stevens *et al*, 1991; 1997 (Personal communication)	1430	R	AR	$O_i \rightarrow A_i$	moderate			0 = 86 A = 97				Specificity of tests now suggests ABR as method of choice for this population. Emerging evidence of significant number of apparent false-negatives; further data awaited
Sutton & Rowe, 1997	145	P	AR	R_1 R_2			60 65					Detailed retrospective analysis of 145 cases and potential sensitivity of two AR registers
Tucker & Bhattacharya, 1992	6000	R	All births	$ARC_i \rightarrow ARC_i$	40				75–88	92–98		95% CIs (not given) would be large. One of two UNS services in UK at present
Watkin *et al*, 1991	322	R	AR	Ai	moderate							AR screen identified (only?) 43% of retrospectively identified cases from cohort
Watkin, 1996a	11,606	R	All births	$O_i \rightarrow O_o \rightarrow A_o$	moderate	92		87.0			2:1000	One of only two UNS services in UK at present
Watkin, 1996b	11,606	P	All births	O_i	moderate			97.0				Change of criteria improved test specificity without sensitivity loss
Watkin, 1996c	14,353	P	All births	$O_i \rightarrow O_o \rightarrow A_o$	moderate	92		87.4		99.5	1.5:1000	Too recent to ensure full ascertainment; three false-negatives arguably 'explainable'.
	8172	P	All births	HVDT	moderate	87		80.5	86.4	92	0.1:1000	Study subgroup who had both neonatal screen and HVDT
White & Maxon, 1995	4253	G	All births	$O_i \rightarrow O_o \rightarrow A_o$	20			93.0		99		Later report of screen now implemented as service shows improved specificity of test and screen. Aim of screen questionable, prevalence data and screen sensitivity (100% claimed) doubtful

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[2] *AR, at-risk only; NAR, not at-risk only.*
[3] *O, OAE; A, ABR; P, ARC; → route for those failing; i, in-patient; o, out-patient; R, at-risk register.*
[4] *Indicates target level of hearing loss (and greater).*
[5] *Sometimes the failure rate, as an acceptable approximation.*

HVDT screen at 8 months; in the next 4-month block, while the team moved to its other area, babies received only the HVDT at 8 months.

A total of 40,471 babies were screened, of whom 21,190 underwent the neonatal screen and the HVDT, and 29,281 the HVDT only. Overall, the neonatal screen referral rate was 1.6%, giving an approximate specificity of 98.4%, with an achieved coverage in the latter stages of the trial of 87–95% of hospital births. An important caveat here is that the coverage rates refer only to babies born in District general hospitals. While for most Districts this would be almost all births, in one of the four Districts covered in this study only 62% of babies are born in the District general hospital. Coverage of babies born in smaller, local, units has not therefore been investigated and might be challenging. Mean coverage from October 1994 to July 1996 was 86% (range 79–91%) and with the coverage of NICU/SCBU discharges to home this approaches 91%. Of those tested, 40%, 75% and 88% cumulatively were tested within 24, 48 and 72 hours of birth, respectively. The percentage not screened for lack of time is less than 5%; birth and discharge over the weekend, 'early' discharge (i.e. from delivery suite) and refusal rates account for most of the babies not covered by the screen. In the later stages of the trial, the team introduced a weekly recall clinic for those not covered as in-patients and this increased coverage by 4–5%. Emissions were not detected in 9.8% of ears (either because of incomplete testing or completed testing with no OAE); this figure rose to 15.6% in babies screened within 24 hours of birth, reducing to about 4% by 72 hours. Overall screen referral rates (emissions fails were followed by immediate automated ABR) stabilised at a median of 1.6% (range 0.6–2.8%).

Other studies have noted the higher failure rate in babies tested within 24 hours of birth; this could become an important issue for neonatal screening if the proportion of discharges home within 24 hours were to rise significantly from present levels. El-Refaie and colleagues (1996) and Sutton and colleagues (1996) tested at-risk (NICU/SCBU) babies and concluded that transient middle and outer ear obstructions gave rise to significant failure rates, especially for OAE screening. Thornton and colleagues (1993) serially tested normal birth babies for 3 days *post-partum* with OAEs and concluded that the higher failure rates on day one were only partly accounted for by middle ear state and outer ear debris, and that a maturational mechanism was also implicated.

The key aspect of the interpretation of the Wessex trial concerns the comparison of data from the two arms of the trial – one with neonatal screening plus HVDT, the other with HVDT alone. For the group tested neonatally, the PCHI yield for moderate or greater hearing loss was 1.2 per 1000 (95% CI, 0.8–1.7). The yield of cases identified before 9 months of age in this arm of the trial was 1.1 per 1000 births (95% CI, 0.8–1.7). These figures for absolute yield are of the order that would be expected if screen sensitivity is high (see chapter 2).

The yield from the group without neonatal testing was 0.7 per 1000 births (95% CI, 0.4–1.0). However, not all these cases were identified as a result of HVDT screen failure, giving a HVDT screen yield of only 0.10 per 1000 (95% CI, 0.03–0.31). There were no cases found by the HVDT when the neonatal screen was in place (i.e. incremental yield of zero). The remaining hearing-impaired babies in the HVDT-only arm were identified following parental or professional concern, often relatively early: all but one were identified under 9 months of age and referred by 10 months of age. Most of these did not have the HVDT, since they were already in the system. Only one case (severe loss) passed the HVDT. The referral of some cases as a result of parental or professional concern, often before the HVDT, reflects a general trend to earlier identification resulting from listening to parental observations and concerns, encouraged by *Hints for Parents* forms (used in both arms of this trial). Nevertheless, while about half the cases in this group were identified at under 9 months of age, in the group screened neonatally the proportion was, not surprisingly, much higher at 96% (95% CI, 78–99).

It is highly likely, given the relatively low absolute yield in the non-neonatally-tested group, that HVDT false-negative cases will emerge in due course. While this may also occur in the neonatally-tested group, the existing broadly expected yields suggest the number of such cases is low. Continued data monitoring and case ascertainment for this study are crucial in order to complete the picture.

The Wessex trial has been a valuable comparative study, and has delivered high quality, detailed and useful data. The study also provides a rich seam of lessons for training and implementation, and those providers moving towards some form of neonatal screening would be well-advised to build upon the experience of this and other teams (see below). Finally, the trial will also provide an opportunity to compare outcomes in early-identified children and late-identified children, which could be a further valuable addition to the literature.

Watkin (1996a) reports on the implementation of a UNS as a service, one of only two at present in the UK. The study cohort was 11,606 babies, from a population of 14,353, with a sub-group of just over 8000 having undergone the HVDT as well as neonatal screening. The targets are those babies with bilateral permanent hearing loss of 40 dB or more, and the neonatal screen involves TEOAE screen as an in-patient, followed by repeat OAE screen as an out-patient for failures, followed by immediate ABR for the second OAE failures. Coverage of the neonatal screen has stabilised at 92%, with **test** specificity at 87–92%. Specificity has been improved recently without sensitivity loss by changing the OAE pass–fail criteria (Watkin, 1996b); neonatal **screen** specificity (i.e. all three tests) is now 97% or above. Yield (Watkin, 1996c) is of the order of 1.5–2 per 1000 births (see *Table 20*), although the study is too recent to be sure of full ascertainment. Thus, sensitivity may decrease but probably only slightly, given the yields already achieved. The sub-group (Watkin, 1996c) who underwent HVDT screen as well as neonatal screen (n = 8172) gave an incremental yield for the HVDT of only 0.1 per 1000 births (see *Table 20*). Specificity of the HVDT **screen** was estimated at 92%, with single test specificity at 80%. Coverage was higher for neonatal screening (92%) than for the HVDT screen (87%).

The Rhode Island group (Maxon *et al,* 1993; White & Maxon, 1995) have implemented UNS first as research, now as service across the State of Rhode Island. Unlike most states in the USA, Rhode Island is relatively compact and populous. The screening system is similar to that used by Watkin (1996a): in-patient TEOAE, leading to referrals to out-patient OAE and (for referrals from that) to immediate ABR screen. Early reports from the group indicated high and unacceptable referral rates of 27% ('refer' and 'partial pass' categories) or 15% (if partial passes counted as passes). More recent changes have brought this down to a reported first test failure rate of 7%, although our own observations (see chapter 6) indicate that it fluctuates between 7% and 12%, being higher in outlying birthing hospitals. These levels probably manageable, however, and the overall **screen** failure rate (possibly after several tests) is nearer 1%.

Clear data on the sensitivity of this screen are difficult to calculate and some doubts remain on this aspect of the group's results. Since their target cases are any hearing loss (20 dB or more), bilateral **and** unilateral, the identification of false-negative cases becomes impossible or imprecise at best. The team claim no false-negatives but this is not statistically valid. Also, their quoted prevalence rates (over 5 per 1000) are far higher than epidemiological evidence leads us to expect; again, this is due to the confusion arising from including mild and unilateral hearing losses. Since the benefits of early identification for such impairments remain to be proven, the approach of having target cases of moderate bilateral hearing loss has to be preferred when reporting the efficacy and cost-effectiveness of screens.

The use of PARC testing for UNS is reported by Bhattacharya and colleagues (1984) and Tucker and Bhattacharya (1992), the latter being the definitive report. Target cases are those with 40 dB hearing loss or greater; PARC testing is undertaken in the maternity hospital, with two test failures defining a screen referral. Screen sensitivity is difficult to estimate from the data given but appears to be between 75% and 88%, although the small numbers involved mean that CIs will be large. Screen specificity is reported to be over 90%, sometimes approaching 99%. Shepard (1983) also reported high specificity (98.6%) in a small feasibility study in which at-risk babies were screened using the ARC (an earlier version of PARC). However, the use of the PARC **with high-risk babies** is questioned by the evidence from McCormick and colleagues (1984a), with a sensitivity of 84%, and particularly from the larger cohort study of Davis and colleagues (1991), where sensitivity was only 50% for severe/profound cases and 20% for moderate hearing losses.

The evidence on UNS, then, points to the achievability of acceptable coverage (although this is largely a matter of resources, particularly adequate staffing) and screen specificity of > 90%. Evidence on screen sensitivity is harder to come by, although Watkin's large cohort study (Watkin, 1996a;b;c) indicates high sensitivity for moderate bilateral losses. Both Kennedy and colleagues (1997) and Watkin (1996b) show the yields expected, 1–2 per 1000 births.

Sensitivity estimates are available, however, from studies of neonatal screening of at-risk babies. The higher prevalence of hearing impairments in this population (odds ratio of 10:1, depending on definitions of the at-risk group) means that fewer numbers need to be screened to achieve stable estimates.

The largest such study is reported by Lutman and colleagues (1997). In this multicentre trial, 7500 at-risk babies were screened using a non-commercial OAE device (POEMS: Programmable Otoacoustic Emission Measurement System). Target cases were those with 50 dB or greater hearing loss. Average specificity was 92%, although this varied widely

between different test centres for reasons that are not clear but which probably reflect training and tester variables. A follow-up ascertainment of known cases from those areas where the screen had been implemented identified over 200 babies, of whom 45 had been screened neonatally and whose hearing loss was unlikely to have been acquired post-natally. Of these 45 babies, nine had passed the screen (false-negatives). The nine cases and screen printouts were examined carefully but the reasons for the false-negative passes – poor test interpretation, progressive loss, later-acquired loss – remain uncertain. However, what the study does supply is a robust estimate, of 80%, of the sensitivity of a neonatal screen (95% CI, 65.8–89.3). There was evidence of progression of hearing loss in two of the nine cases, so this estimate is a 'worst case' estimate. However, from one viewpoint, such cases are still properly regarded as false-negatives.

In a similar study from the same group (Mason *et al,* 1997), parallel data from a large multicentre trial with at-risk automated ABR screening are reported. Between 1988 and 1993 approximately 7000 neonates were screened. Later ascertainment of cases of permanent bilateral hearing impairment of 50 dB HL or greater in the participating areas identified 197 children (born 1988–93), of whom 44 had been screened with automated ABR. Using both machine-scoring and an experienced visual appraisal of waveform printout gave higher sensitivity than relying on the automated score alone. Three of the 44 children had passed the screening for reasons that are uncertain – progressive loss, later-acquired loss, incorrect test result or adequate high-frequency hearing. This gives a sensitivity estimate of 90% (95% CI, 78–96) for the screening test. The difference between this sensitivity estimate and that of the OAE study reported by Lutman and colleagues (1997) is intriguing. It suggests that for at-risk children, the lower sensitivity with the OAE screen may reflect test-specific factors rather than late-onset hearing impairments. However, overlap of the two 95% CIs argues against premature conclusions.

Shiu and colleagues (1996) have conducted a detailed retrospective regional audit for children born between 1984 and 1994. Ascertainment is not very high, there being 331 permanently hearing-impaired children out of 383,206 births, which gives a 'low' prevalence rate of 86 per 100,000 births. One of the eight Districts covered runs a targeted neonatal screen, and 21 of the 26 cases of hearing impairment (> 40 dB) screened were referred by the screen (i.e. five cases passed the screen). This gives a sensitivity estimate of 81% (95% CI, 61–93). However, unlike the studies by Lutman and colleagues (1997) and Stevens and colleagues (1991; 1997) (see below) known progressive impairments were excluded from these calculations. Shiu and colleagues identified 24 cases of progressive hearing impairment – about 8% of the 306 cases (excluding acquired impairments). Note that there is some overlap in the samples studied by Lutman and colleagues and Shiu and colleagues. These estimates of progressive hearing impairment are similar to those reported for the Trent study, about 10%, when known acquired impairments are excluded.

However, Stevens and colleagues (Stevens *et al,* 1991; 1997) have also recently indicated a significant number of potential false-negative cases emerging from their follow-up of an at-risk screening study. These studies investigate the feasibility of both TEOAE and ABR screening with inpatient at-risk neonates, with target cases being moderate bilateral losses. ABR test specificity is higher (90–97%) than OAE test specificity (80–86%) but takes longer. The authors suggest that the optimal arrangement would be OAE followed by ABR screen for failures, giving a screen specificity of 93%. More recently they have suggested that the ABR screen might be the method of choice for at-risk babies, with a specificity of 97%. Hall and colleagues (1996) and Lamb (Personal communication, 1996) also found high specificity for ABR with this population (99.7% for bilateral losses, 98.4% for bilateral or unilateral). Similarly, Hyde and colleagues (1991) found high specificity for ABR at-risk screening (95.2%). This is a study with full and detailed follow-up of all screened ears, giving a probably robust sensitivity estimate of 90.4%, even including mild and unilateral losses. McClelland and colleagues (1992) also undertook full follow-up studies of their babies who were screened with ABR, and found no false-negatives. However, Stevens and colleagues' recent follow-up data (Personal communication, 1997) showed that of 23 neonatally-screened hearing-aided children at age 5 years, with hearing loss > 50 dB HL, only 14 children failed the neonatal ABR screen and 13 the OAE screen. The evidence suggests that these were late-onset cases. Since these were all NICU/SCBU babies, it is possible that the proportion of late-onset cases is somewhat higher in this population. If these cases are called false-negatives (see above) then the neonatal screen programme sensitivity was only 61% (ABR) and 56% (OAE). Even more recent analyses (JC Stevens: personal communication, 1997) put these **programme** sensitivity estimates at 52% (ABR) and 44% (OAE). These figures are much lower than those from other studies and seem to be due to

an unexpectedly large proportion of late-onset cases; all but one of these children also exhibited normal hearing at full testing (behavioural) at 8 months of age, indicating that an HVDT screen would also fail to identify them.

Kramer and colleagues (1989) claim 100% sensitivity for severe hearing losses, although full ascertainment of possible cases is doubtful. Specificity ranged from 88% (30 dB + losses) to 94% (40 dB + losses) for their risk register followed by an outpatient ABR screen. Galambos and colleagues (1994), in a 20-year study of 5901 screened babies, give a specificity of 80–88% for full ('diagnostic') ABR screening, depending upon the level of intensive care.

Some have argued that inpatient screening may not be ideal – babies may be neurologically immature at discharge, for example. For universal screening, early discharge may preclude high coverage. Rowe (1991) and Kei and colleagues (1997) therefore investigated the feasibility of screening babies later, either at outpatient recall (Rowe, 1991) or in a routine community clinic (Kei *et al,* 1997). Rowe found a nearly-acceptable coverage of 86%, with 100% sensitivity and 91% specificity for ABR screening, and target cases of 40 dB bilateral or unilateral. The same group (Sutton & Rowe, 1997) examined the potential sensitivity of two different at-risk registers in identifying 146 cases of congenital PCHI, giving figures of 60–65%. However, retrospective analysis of risk register sensitivity is likely to overestimate sensitivity, since some factors may not be apparent perinatally (see also chapter 2). A study from Holland is reported by van Zanten and colleagues (1995), in which 31% of births take place at home and over 90% of hospital births are discharged within 16 hours of birth. In this study, babies are screened with OAEs either at home (n = 545) at age 5–10 days, or in the well-baby clinic at age 4–6 weeks (n = 487). Both methods were feasible, with high success rates (> 90%) for recorded results. Visit time (and costs) were higher for home visits, and later questioning of a sample of parents (n = 50) indicated positive acceptance of the screen, with a preference for home versus clinic screening.

Finally, the evidence is quite clear that neonatal screening does indeed produce earlier age of identification. This is referred to in chapters 2 and 4 for the Trent study (Fortnum *et al,* 1997). McClelland and colleagues (1992) also gave a median identification age for those screened in an at-risk screening programme of 1 month. Watkin (1996c), reporting on a universal screening programme, has a median 'confirmation age' for severe/profound hearing losses of 9.2 weeks, with hearing-aid fitting at 15.9 weeks. For moderate hearing losses, median confirmation age is 12.9 weeks, with hearing-aid fitting at 41.9 weeks. This 'delay' in hearing-aid fitting is interesting. It represents a service approach in which hearing-aid fitting only takes place when parents are ready to accept it – placing them firmly at the centre of events. For moderate hearing losses, this often takes considerable time. With moderate hearing losses, there may also be delays introduced by equivocal or confusing audiological assessments, or by ENT referral (see Fortnum *et al,* 1997). The data from Kennedy and colleagues (1997) indicate that most neonatally-screened cases are identified below 9 months of age, compared with only half of the group without neonatal screening. The study by Shiu and colleagues (1996) showed that a District with targeted high-risk neonatal screening performed significantly better in terms of the number identified by 6 months of age than other Districts in the region, and was the only one in which high-risk infants were detected significantly earlier than non-high-risk infants. They found high-risk cases at a median age of 7 months over the latter half of the audit period.

In summary,[2] the evidence indicates the following.

1. High coverage (90% +) for universal screening is possible.
2. All methods of neonatal hearing screening show high screen specificity, generally well above 90% after a 'settling-in' period.
3. Evidence on test sensitivity for moderate and greater cases of congenital PCHI is less available but estimates range from 80% to 100%. Generally, **programme** sensitivity (reflecting cases which were not covered, and/or which are late-onset or progressive) may be estimated to be nearer 80%.
4. The two large cohort studies of universal neonatal hearing screening in the UK, including the controlled trial in Wessex, produce yields of the expected order, that is, 1–1.5 per 1000,

[2] In this section and the following one on the HVDT, we have chosen not to perform any formal meta-analyses of the data presented in *Tables 20–22*. We have preferred to synthesise the data for two major reasons: the first is that the definition of populations tested and aims of independent studies are such that only one or two studies fall in each separate category; the second is that it is sometimes not clear when the subject material overlaps and is not independent.

and decrease the subsequent incremental yields of the HVDT to very low levels.

5. At-risk neonatal screening has a potential yield of about 60% of all cases. In practice, however, this is likely to be much lower because of the difficulty of implementing full coverage for all indicators of at-risk cases, perhaps 45–50% at best.
6. The median age of identification for those screened neonatally is of the order of 2 months (McClelland *et al,* 1992; Watkin, 1996c) depending upon follow-up procedures and severity of impairment, and is earlier than for cases not screened neonatally (Kennedy *et al,* 1997; Shiu *et al,* 1996; Fortnum *et al,* 1997).
7. The extent to which the opportunities offered by early identification are capitalised on is affected by aspects of the follow-up services. Fortnum and colleagues (1997) present evidence that involvement of ENT services can delay hearing-aid fitting significantly for cases of moderate and severe but not profound, hearing loss.

Evidence on HVDT screening

McCormick and colleagues (McCormick, 1983; 1988b; McCormick *et al,* 1984b) led efforts in the mid-1980s to improve the performance of the HVDT, which had been the subject of widespread criticism (Boothman & Orr, 1978; NDCS, 1983; Robinson, 1983). McCormick (1983; 1988b) reviewed and updated details of test procedure, introduced electro-acoustic sound generators (warblers) which gave better test accuracy and reliability, and introduced new standards for training health visitors to conduct the test. McCormick and colleagues (1984b) showed that age of identification of congenital PCHI improved under the new system and that 94% of a sample of health visitors were 'pleased' with the changes.

Evidence for the recent performance of the HVDT comes from two types of study. First *(Table 21),* clinic-based retrospective studies of children known to have permanent and congenital hearing impairment are easy and inexpensive to perform, hence are numerous. Typically these studies are concerned with the route by which hearing impairment was identified and, in particular, the role of the HVDT. At their best, these studies will be for a defined geographical area with known birth-rate and for a specified period of births. Ascertainment of all cases is important, since those not ascertained may introduce bias: for example, they may be less conspicuous to services (mild and moderate impairments) or they may not yet have been identified as hearing impaired (in which case, their omission would inflate the sensitivity estimate of screen performance). Some published studies of this type are *ad hoc* or opportunistic, involving a less clearly defined group of children with PCHI. Taken as a whole and with statistical caveats, these clinically-based studies can usefully and qualitatively supplement the quantitative data from properly controlled population studies.

The other type of study *(Table 22),* sometimes combined with ascertainment studies, involves examination of the records of a defined population, not necessarily hearing-impaired, to examine aspects of screen performance such as coverage, failure rate, and positive predictive value (PPV).

Ascertainment studies *(Table 21)* have been used to examine the age of identification of PCHI in the ascertained cases. This is generally better expressed for a group as the median rather than the mean,[3] since the distribution of identification age will be skewed, with a long tail of late-identified mild cases and cases in which other considerations (e.g. severe other disabilities) have delayed confirmation of hearing impairments, perhaps justifiably. Neonatal screening is known to lower the identification age (see, for example, McClelland *et al,* 1992) for those screened, although for at-risk neonatal screening the reduction in median age of identification may not reach statistical significance for the PCHI population as a whole (see, for example, Sutton & Scanlon, 1996).

None of the studies listed in *Table 21* involves UNS and the median ages of identification given can thus be taken as reflecting the identification age delivered by HVDT-screen-led systems, even if some

[3] The median is preferred to the mean as a measure of the central tendency of a distribution because it is a more stable indicator than is the mean of the process underlying the distribution. With only a few very long ages of detection, the mean would be highly influenced. However, as discussed in chapter 2, one of the main characteristics of the distribution is the lack of variability in the lower percentiles compared with the large variability above the median. The median may, therefore, be a conservative (or insensitive) estimate of the service quality as it does not change much in response to service changes. In addition, it seems sensible to weight longer ages of detection more in terms of their consequences (which would depend on the severity of the impairment). At present, no such sensitive and outcome-weighted indicator exists. The distribution as shown in chapter 2 is too cumbersome to use generally.

TABLE 21 *HVDT – retrospective ascertainment studies of PCHI cases*

Study	Number ascertained	Type of report[1]	Base population (where given)	Strength of ascertainment[2]	Degree of loss ascertained[3]	Sensitivity (%)[4]	Coverage estimate (%)	Yield (%)[5]	Other disabilities (%)	Median age of identification (months)	Risk factors present (%)	Comments
Davis & Wood, 1992	25	R	29,317	✓✓	aided	88	95		40		64	Detailed study of cases born 1983–86. CIs of sensitivity estimate 68–96%.
Fonseca *et al*, 1996	104	P		✓	50+	52			36	15	54	Interim unpublished multicentre data. Cumulative identification: 19% by 6 months, 39% by 12 months. Sensitivity of neonatal screen (at-risk) for this group was 91%.
Fortnum *et al*, 1997	556	R	552,558	✓✓	40+	65		43 24	40	15 (12 fails, 32 passes)	59	Large comprehensive multicentre study. CIs given. Yield of 43% reduces to 24% for those areas with (at-risk) neonatal screen. Sensitivity by severity: 54% moderate, 75% severe, 80% profound.
Baart de la Faille, 1991	43	R		?	aided	95						Sensitivity doubtful as ascertainment doubtful. Also, 17 of 43 not tested. Study is of 'Ewing' test in Netherlands.
O'Hare *et al*, 1993	71	R		?	aided					18		HVDT highest source of referral (30% of cases). Large numbers referred (e.g. > 2000 in a year).
Johnson & Ashurst, 1990	62	R	64,881	✓	aided	91	94					Large study of at-risk population across region – high sensitivity unsurprising for this group, **but** high proportion of cases not screened by HVDT; should have been, giving 'effective sensitivity' of 56%.
Kennedy *et al*, 1997 (Personal communication)	85	P	50,000	✓	40+	60					47	Baseline audit before RCT of neonatal screening, covering 1980–89. Only 18 cases identified before 12 months of age.
McClelland *et al*, 1992	119	R	114,240	✓	severe					13		Compares identification age with neonatally-screened group, same study: 1 month.
McCormick, 1990	21	R	36,207	?	aided	86						Reply addendum to Scanlon & Bamford (1990).
Plant & Pick, 1995	53	A	48,000	✓✓	aided	18–43		14		20 (fail) 48 (pass)		Unpublished audit. Identification age distinguished on basis of pass or fail HVDT.

[1] *P, preprint; R, published in peer-reviewed academic journal; A, unpublished audit report; G, general review for edited volume or chapter.*
[2] *Judged as (✓✓ = close to 100%; ✓ = probably high; ? = doubtful.*
[3] *Case definitions, e.g. aided (all those with hearing-aids), 50+ (losses of 50 dB or more), severe+ (severe or greater losses).*
[4] *Generally, test or screen sensitivity independent of coverage.*
[5] *Proportion identified by test or screen.*

continued

TABLE 21 contd *HVDT – retrospective ascertainment studies of PCHI cases*

Study	Number ascertained	Type of report[1]	Base population (where given)	Strength of ascertainment[2]	Degree of loss ascertained[3]	Sensitivity (%)[4]	Coverage estimate (%)	Yield (%)[5]	Other disabilities (%)	Median age of identification (months)	Risk factors present (%)	Comments
Robertson *et al,* 1995	197	R		?	aided	54	65			18	49	Australian study. Sensitivity of parental questionnaire 49%. No apparent effect of screen outcome on identification age, surprisingly. Detailed study, high quality.
Scanlon & Bamford, 1990	41	R	54,000	✓	severe +	50						Study which resulted in HVDT being replaced by vigilance programme, currently being evaluated (Sutton & Scanlon, below).
Shui *et al,* 1996 1995	282	A	383,206	✓	40+	57	87	37				10-year retrospective audit; includes Sutton & Scanlon, 1996, data.
Sutton & Scanlon, 1996	62	P	60,910	✓✓	50+	42	92			12 s/p 19 mod		Unpublished interim data from detailed study. HVDT compared with 'vigilance' programme, incl. questionnaire (sensitivity 42%, coverage 71%, identification age 10 months for severe/profound; 27 months for moderate).
Varghese, 1996	47	P		✓	50+	36	86			13 (fail) 40 (pass)		20-year retrospective study, interim data unpublished. Identification age affected by HVDT pass–fail. Sensitivity for severe/profound 50%. Overall sensitivity of HVDT for Asian cases (n = 15) only 15%. No CIs.
Watkin *et al,* 1990	171	R	51,250	✓	20+	93 s/p 36 mod 17 mild	86					Improvements to screening programme have raised sensitivity for moderate to circa 70% – see Watkin, 1991.
Watkin, 1991	198	R	51,249	✓	20+	90+ s/p 70 mod				12 s/p 18 mod		Study of HVDT in Inner London area after service improvements. Ascertainment ✓✓ for s/p; less certain for mild/moderate. Sensitivity low for mild/unilaterals. Identified 13% progressive losses.
Wood *et al,* 1997	68	P		✓✓	50+	68	80	55 35				10-year retrospective study; most up-to-date data from Nottingham group. Neonatal AR screen in place. HVDT sensitivity 78% (years 1–5), 35% (years 6–10). Fall in yield also, from 55% to 35%, as neonatal screen improved and HVDT sensitivity fell. Yield of neonatal screen = 19%.

[1] *P, preprint; R, published in peer-reviewed academic journal; A, unpublished audit report; G, general review for edited volume or chapter.*
[2] *Judged as (✓✓ = close to 100%; ✓ = probably high; ? = doubtful.*
[3] *Case definitions, e.g. aided (all those with hearing-aids), 50+ (losses of 50 dB or more), severe+ (severe or greater losses).*
[4] *Generally, test or screen sensitivity independent of coverage.*
[5] *Proportion identified by test or screen.*

TABLE 22 *Population studies of HVDT screens*

Study	Sample size	Type of report[1]	Coverage (%)	Sensitivity (%)	Failure rate (%)	PPV[2]	Comments
Brown *et al,* 1989	1990	R	56		5	48	Inner London. Coverage rose to 80% by 20 months. Coverage lower for Asian children.
Baart de la Faille, 1991	33,252	R	86	75	7		As implemented in Netherlands. Training and registration of testers centrally organised.
Haggard *et al,* 1992	2492 (B) 2403 (A)	R			7.3 (B) 6.1 (A)	63 (B) 76 (A)	Compared HVDT referrals before (B) and after (A) improvements to screen. Noted that 1–5% of total birth cohort will be referred with OME: service implications.
Mott & Emond, 1994	230	R				35–66	Sampled case-notes of 100 who missed and 130 who failed HVDT.
Scanlon & Bamford, 1990	24,000	R			4.1	38	Sample size estimated from birth rate.

[1] *P, preprint; R, published in peer-reviewed academic journal; A, unpublished audit report; G, general review for edited volume or chapter.*
[2] *Estimated PPV of the HVDT screen,* ***including*** *cases of non-permanent conductive hearing.*

include identification age data from at-risk cases screened and identified neonatally. The reported median identification ages range from 13 to 20 months. These include children who passed and children who failed the HVDT (i.e. false-negatives and true-positives). The extent of false-negatives will affect the median identification age, since such cases tend to have their identification unduly delayed (although Robertson and colleagues (1995), in an Australian study, surprisingly found no effect of pass versus fail on identification age). Evidence for this general trend is provided by studies from Varghese (1996) and Plant and Pick (1995) which, respectively, showed age of identification to be 13 and 20 months for HVDT screen failures, and 40 and 48 months for false-negative screen passes. The audit reported by Kennedy and colleagues (1996) as an (unpublished) prelude to their neonatal screen trial indicates only 18 from 85 cases identified before 12 months of age.

In populations lacking an effective early screen, median age of identification is also affected by severity of hearing impairment, presumably because fewer false-negatives occur among the more severe cases, and there is less delay in audiological confirmation with more severe cases. Thus Watkin (1991) showed an identification age of 12 months for severe/profound hearing impairment and of 18 months for moderate impairment. Sutton and Scanlon's (1996) study indicates 12 months for severe/profound hearing impairment and 19 months for moderate hearing impairment. The study by Shiu and colleagues (1996) gives a median age of identification of 25 months for moderate and 14 months for severe and profound impairments.

Overall, identification ages are disappointingly late with the HVDT. They are inevitably influenced by the **programme** sensitivity, that is, by the sensitivity of the screen and by the screen coverage. Coverage figures from UK studies, *Tables 21 and 22,* vary from 80 to 95%, with one study from an Inner London area showing coverage of only 56% (Brown *et al,* 1989). Coverage in reported studies is generally, therefore, **fairly** high, although above 90% is the generally accepted quality target (Haggard & Hughes, 1991). There is likely to be a bias introduced by effort in research and service development, hence by publication: poorer services will be less likely to audit, less likely to publish. Some unpublished evidence suggests poor HVDT screen coverage in some areas (see, for example, Wanjohi, 1996). In the study by Brown and colleagues (1989), poorer coverage of the Asian baby population is indicated; in the study by Varghese (1997), lower screen sensitivity for Asian babies is suggested.

Sensitivity estimates for the HVDT screen are more available than they were for neonatal screens. This is probably because, for high ascertainment, the retrospective nature of the studies requires a delay of several years between service and ascertainment and, in most areas where it exists, neonatal screening has only been introduced recently. Davis

and Wood (1992), in a study of one UK service that is likely to be of better quality than the norm, report HVDT screen sensitivity of 88% (95% CI, 68–95). However, in a more recent report from the same group covering a wider area and a 10-year cohort, with high ascertainment, Fortnum and colleagues (1997) indicated sensitivity of only 65%, or 80% for cases of profound hearing impairment, 75% for severe and 54% for moderate. The same group (Wood *et al,* 1997) report the results of a 10-year retrospective study of the same geographical area covered by Davis and Wood (1992). Overall sensitivity of the HVDT was 68% but this had fallen from 78% in the first half of the decade to 36% in the second. The reasons for this are not certain but are probably linked to the challenge of maintaining quality through training and monitoring. This 1997 study supersedes a short report by McCormick (1990) for the same area, in which a sensitivity of 86% was reported. The 10-year retrospective audit by Shiu and colleagues (1996) identified 282 infants with a congenital non-progressive permanent hearing impairment (> 40 dB), from a birth cohort of 383,206. Of these, 75 were referred before 8 months of age and, of the remaining 207, 181 had been tested by the HVDT (a coverage of 87%). A total of 77 of those tested passed the screen; thus, screen sensitivity was 57% (95% CI, 50–65), with programme sensitivity at 50% (95% CI, 43–57). Both screen and programme sensitivity varied widely across Districts with the range being 41–85% for test and 32–77% for programme sensitivity.

Two other studies with high ascertainment also provide evidence of low sensitivity of the HVDT screen. Plant and Pick (1995) report a range of 18–43%, depending upon the inclusion criteria for the calculation. There is evidence from this study that the sensitivity has improved in more recent years, compared with earlier years. Sutton and Scanlon (1996) give a sensitivity figure of 42%. This is a follow-up study to Scanlon and Bamford (1990), in which a sensitivity of 50% for severe/profound hearing impairment was indicated. As a result, this District abandoned the HVDT screen in favour of a surveillance or 'vigilance' approach (*Hints for Parents*; health visitor training stressing good developmental observation and referral at any age on parental or professional concern; a questionnaire to parents at 8 months). At-risk neonatal screening was also in place in this District, as it had been during the previous HVDT regime. Sutton and Scanlon (1996) report on the vigilance programme's performance. While median identification age for cases of severe/profound hearing impairment remained stable at 10 months, the median identification age for moderate cases fell markedly to 27 months, as would be expected from the general relationship between severity and the age of identification in populations without an effective screen. The fact that the programme was not good at identifying moderate cases leaves an important gap. Questionnaire sensitivity for hearing losses of 50 dB or more was 42%, with coverage falling to 71%. These figures are important, since there has been widespread comment that more could be made of parental observations of a child's response to sound. While this is undoubtedly true, and Robertson and colleagues (1995) have shown evidence of the delays introduced by sceptical professionals **not** taking heed of parental concern, the Sutton and Scanlon data indicate the possible limits of this approach. Similarly, Robertson and colleagues report 53% of cases as being first suspected by parents, and Watkin and colleagues (1990) give evidence of the proportion being 44%. However, these studies both report referrals on the basis of parental suspicion in the presence of the HVDT, thus representing a version of **incremental** yield. The Sutton and Scanlon study gives sensitivity for a questionnaire approach **without** any HVDT screen in place (NB: their data are also reported and discussed in the regional audit of Shiu and colleagues, 1996).

Several articles furnish evidence on sensitivity from studies which have probably high ascertainment, although probably not close to 100% (Fonseca *et al,* 1996; Johnson & Ashurst, 1990; Kennedy *et al,* 1997; Scanlon & Bamford, 1990; Watkin *et al,* 1990; Watkin, 1991; Varghese, 1997). Their sensitivity estimates for the HVDT screen range from 36% (Varghese, 1997) to over 90% for cases of severe/profound hearing impairment and 70% for moderate (Watkin *et al,* 1991). The study by Baart de la Faille (1991), in which a sensitivity estimate of 95% is reported, is less relevant because it reports on the 'Ewing' screen (equivalent to the HVDT) in The Netherlands, where the screen implementation is centrally monitored and controlled; also, 17 of his 43 identified infants had actually missed the screen. Furthermore, the population study of Baart de la Faille's report (see *Table 22*) gave a lower sensitivity estimate of 75%. The Johnson and Ashurst study (1990) gives a sensitivity of 91% for the HVDT screen identifying children with PCHI from a register of all at-risk children. The high figure is perhaps not unexpected since such a population sample will have a high proportion of children with other disabilities, less likely to give unequivocal 'pass' responses in the screening test. The Johnson and Ashurst study pointed to a coverage problem, however – a larger proportion of the at-risk PCHI

babies had not been screened, indicating differentially low coverage of this population, and an 'effective sensitivity' of only 56%.

The population studies of the HVDT screen *(Table 22)* show referral rates (failure rates) of 5–7%. This is lower than given in chapter 4 which reports referral rates of about 10%, as does the study by Wood and colleagues (1997). For an average-sized 'District' with 4000 births per year, referrals to a Hearing Assessment Centre or to Community Audiology Clinics will, on this basis, be between 160 and 280 9-month-old infants. At an estimated prevalence for PCHI of, say, 1.1 per 1000, only six cases would be expected (year-to-year variability for such a small number will be high), of whom only 1–2 may be detected by the HVDT. Thus, the PPV of the HVDT screen for this low-prevalence condition is itself very low. However, this low PPV in itself is not problematic, and can be justified on the basis of the importance of detecting the condition and the benefits thus accruing. The unfortunate by-product of the high referral rate and low PPV is the large number of infants to be seen, often creating significant waiting time (Scanlon & Bamford, 1990).

One of the reasons for the referral rate being of the order of 5–10% is the existence, particularly in the second half of the first year of life, of transient mild or moderate hearing impairment due to OME. Haggard and colleagues (1992) showed that the improvements to the HVDT screen introduced by McCormick in the 1980s resulted in little change in the referral rate (if anything, a reduction) accompanied by an increased PPV **for all cases**, including OME cases. They argued, therefore, that the screen had increased sensitivity as a result of the improvements. Others (see *Table 22*) have also sought evidence of the PPV of the HVDT screen for OME, and estimates range from 35% to 76%. There are a number of case-definition issues which surround these estimates, since the hearing impairment associated with OME, and the condition itself, can fluctuate rapidly, and the definition of which cases will benefit from treatment is contentious (Haggard & Hughes, 1991). The point argued by Haggard and Hughes (1991) is well-made, therefore – the HVDT screen, the aim of which is to refer cases suspected of having permanent hearing impairment, will inevitably refer about 5% of the total birth cohort largely because of OME. The service implications of this are considerable. Whether the HVDT screen can be **partly justified** on the basis of these OME referrals is very doubtful. A one-off, 30–35 dB screen for mild hearing loss which fluctuates, for which diagnostic assessment cannot yet predict the cases that will persist such as to justify intervention, especially when the treatment options are contentious, provides poor justification. The HVDT screen performance is to be judged upon its ability to act as an efficient screen for PCHI.

The evidence may be summarised as follows.

1. Coverage of the HVDT screen probably falls in the range of 80–95%, although there may be some urban areas where coverage falls to nearer 60%. There is some limited evidence suggesting that coverage **and** sensitivity is lower in the Asian population.
2. Sensitivity estimates of the HVDT vary widely, from 18% to 88% (all degrees of loss). Recent studies and more powerful studies are suggestive of poorer levels of sensitivity. Severity of impairment affects screen sensitivity substantially.
3. Failure rate is of the order of 5–10%. Many of these cases will have fluctuating non-permanent hearing loss, associated with OME. This referral rate has considerable resource implications for services.
4. HVDT incremental yield may be, at very best, 40% but it falls off for the best Districts to low levels (e.g. 25%) when at-risk neonatal screening is introduced. With UNS, the evidence indicates that the HVDT incremental yield falls to very low levels.
5. Median age of identification via the HVDT varies from 12 to 20 months. Age of referral is severity dependent.

The possibility of publication bias should not be overlooked when comparing studies of neonatal screening with HVDT studies. The better, more committed, more evidence-based services may be those more likely to publish, and also to report on new methods as opposed to old ones. It is possible – although obtaining systematic evidence for this is not feasible – that there may be an interaction between this possible publication bias and type of screen studied, resulting in more positive studies for neonatal screening. Furthermore, only one of the studies reported here is a prospective controlled trial (C Kennedy *et al:* personal communication, 1997), although the data are highly indicative. Other studies vary from those of screening services at their 'best' to those of screening services as found in less than ideal circumstances. Two UK studies (Tucker & Bhattacharya, 1992; Watkin, 1996a;b;c) have shown that UNS **can** be implemented with high coverage, high sensitivity and high specificity. At least some studies in the

past have indicated the same for the HVDT screen (e.g. Davis & Wood, 1992), even though the same group have shown a fall-off in effectiveness for the HVDT more recently. If there were more examples of UNS, perhaps data on screen effectiveness would become more variable. High coverage is probably achievable for both neonatal screening and HVDT screening, although the HVDT failure rate will remain high because of OME referrals. Sensitivity is high for neonatal screens and can approach 80%, although only for severe/profound hearing impairments for the HVDT screen. There are new developments which could deliver a semi-automated version of the HVDT performed by one health visitor rather than two (B McCormick: personal communication, 1996). Trials of this device (BeST) are needed to see if test sensitivity for hearing impairments, including moderate impairments, can be improved. The device, which delivers warble tones from mounted (but portable) loudspeakers, could be designed to deliver signals of, say, 50 dB: this should decrease the number of referrals for non-permanent hear-ing loss (mainly OME), which could, in turn, reduce waiting times at follow-up clinics. For this reason, consideration could be given to raising the screen-fail hearing level to 40 or even 45 dB depending on the aim of the screening programme, both with this new device and with the 'standard' HVDT – although, for the latter, retraining and implementation might be challenging.

Chapter 8

Summary of evidence

Scope

The evidence presented in the previous chapters falls in five major categories: the epidemiology of PCHI, outcomes, current practice, cost and screen performance. This evidence is summarised here and general conclusions are drawn so that the different options available for identification of PCHI can be compared.

Epidemiological evidence (from chapter 2)

About 112 children per 100,000 have a congenital PCHI of ≥ 40 dB HL in the better ear. The number of children with congenital PCHI expected annually in the UK is 840 at a bilateral hearing-impairment level of ≥ 40 dB HL, or 675 at a level of ≥ 50 dB HL, of whom 173 children might have a severe impairment and 180 a profound impairment.

At District level, the annual incidence of children with PCHI is small (< 7 on average) and **highly variable** from year to year, as expected from statistical fluctuations in small numbers.

About 16% of PCHI may be acquired, progressive or of 'late onset', of which one-third is caused by meningitis (following which a hearing **assessment** is essential (see Fortnum & Davis, 1993)). Thus about 10% of cases of PCHI may be currently considered as either progressive or 'late-onset'. These cases would not be detected by a screen at birth, and the majority of them would not even be detectable at 6–8 months of age.

Children not born in the Health District of residence may account for about 13% of those with PCHI, although this is likely to vary considerably from area to area.

In any one District, about 30% of children with impairments that are acquired or progressive, or children who move into the District/country, will not be picked up by UNS. It is unlikely that the HVDT at 6–8 months picks up many of these children either (it is assumed that the onset of progressive/late-onset cases is uniformly random within the first 5 years of life; there are no data showing otherwise). This figure will decrease to about 10–15% if neonatal screens are widely implemented in the UK (such that change of residence ceases to be an issue).

Just under 30% of children with PCHI have a history of treatment in an NICU/SCBU, about 27% have a family history of PCHI and nearly 4% have a CFA. Thus 60% of children with PCHI have specific known causal risk factors associated with their impairment. However, the actual yield of targeted neonatal screening is likely to be nearer 45–50% of cases sought (at best) because of difficulties in implementing targeting on the family history factor (Wood *et al,* 1995).

Although many aetiologies of PCHI are genetic in origin, the most cost-effective intervention in the next 10 or so years will be secondary intervention through effective screening programmes, rather than primary prevention. When more is understood about the molecular basis of PCHI this is likely to change.

The availability of children for neonatal screening is an epidemiological factor that is essential for considering options. The number of newborn babies discharged from maternity units on their first day of life has been increasing over the last few years and is currently about 20%. This does not appear to be an exponentially continuing trend but it does mean that systems to cover this particular group of children have to be optimised in order to maintain coverage at ≥ 90% (preferably ≥ 95%).

Major indicators of service performance, for example, ages at referral, identification and fitting of hearing-aids, vary substantially with severity of impairment. Overall, they fall far short of the NDCS targets (NDCS, 1994) with a median age of confirmation of hearing impairment of 17 months. Half of the children with PCHI do not have access to hearing-aids by the age of 2 years, and a quarter by the age of 4 years.

Outcome evidence (from chapter 3)

Theoretical arguments on auditory and cognitive plasticity suggest that earlier stimulation is better

for developing the individual child's auditory and cognitive potential.

There is some evidence of reasonable quality that earlier identification is associated with improved outcomes in the communication domain, although this evidence remains restricted in scope and magnitude of effect. The extent and precise nature of the benefits are not yet fully explored. Thus, while there is little evidence that habilitation initiated at age 3 months is better than that at age 12 months, most studies show that habilitation initiated before the age of 12 months does have advantages over that initiated at over 2 years of age. Outcome benefits in other domains (e.g. educational achievement, mental health) also remain to be explored.

There is evidence that outcome benefits are affected not only by age of identification but also by type and quality of subsequent intervention.

There is substantial belief among professionals and parents that earlier identification of PCHI is better for the family as a whole, as well as for planning interventions and support. Professionals report that significant aspects of management (e.g. hearing-aid acceptance) are easier to accomplish early and parents claim a right to early knowledge about significant aspects of their children's development.

There is evidence that neonatal hearing screening, properly implemented, does not cause undue anxiety for parents.

Evidence from the survey of current practice (from chapter 4)

Considerable effort was required to determine the extent of PCHI, and the nature and timing of its management (Fortnum *et al*, 1997). The survey of current practice also showed that services lacked adequate accessible information and that there was no systematic information coordination between the agencies that share responsibility for the continuing care of individual hearing-impaired children. Specific evidence from the survey is presented below for neonatal and other hearing screens.

Neonatal screens

In the UK, two UNS programmes are running as successful services and these were able to provide figures for the present survey with no difficulty. The Wessex trial of neonatal hearing screening also supplied valuable data on a systematic comparative trial of neonatal screening and the HVDT (summarised in chapter 7).

Just under two-thirds of all Districts have a neonatal hearing screening or assessment programme **of some sort**. These are mainly targeted at high-risk children with a history of NICU/SCBU or a family history of hearing impairment.

In those programmes that target the children with an NICU/SCBU history, less than one in three of all children in NICU/SCBU or with a family history of childhood deafness are tested. Coverage is focused on those at very high risk, because of lack of funds for extending the testing.

The targeted screening programmes are relatively recent, the median year of starting being about 1992/93.

Both TEOAE and ABR are used successfully in neonatal screening.

Neonatal screening is undertaken by several different professional groups and in a number of diverse locations.

The failure rate for the predominantly at-risk group at first test is of the order of 10–12%, with overall referral rates of about 5–8%.

The reported yield from neonatal screening is already quite high, with PCHI ≥ 50 dB HL being detected in 100 babies in 1994. This represents about 35% of the babies who might be expected to have a congenital hearing impairment at this level in those Districts using these tests, and about 16% of all such children in the UK.

For the at-risk populations reported in the survey of current practice, the overall field sensitivity of neonatal screening programmes seems to be of the order of 85%.

The age at which hearing-aid fitting is undertaken for those screened neonatally is routinely well within 6 months of age, at least for severe and profound cases of PCHI.

The fact that there is not a systematic enough approach to the family and parents, both in terms of information given before or after the test and in terms of support post-identification gives rise to some cause for concern.

Educational services for children identified very early are not as routinely available as good practice suggests they should be, and the specific skills for working with children and families at the pre-linguistic stage require further development.

Many Districts report that they wish to extend their targeted screening or to introduce UNS at an appropriate time.

The HVDT, HVS and SES

The HVDT and SES are almost universal in their application, both across the country and within Districts. HVS plays a part in identification of hearing impairments in about two-thirds of all Districts.

The professionals responsible for carrying out the HVDT come from a more homogeneous group than for neonatal screening. However, whereas for targeted neonatal screening there might be one person responsible for testing for a whole District, there are more than 60 full- and part-time staff who might perform the HVDT in an average-sized District. In the case of SES there are, again, many fewer testers. This has considerable implications for the quality assurance and training requirements of a screen.

The HVDT uses a level for testing that is too acute for the environmental conditions provided in many Districts.

There is a similar problem with SES, which probably uses screening levels that lead to a greater number of referrals than is efficient.

The reported coverage of the HVDT is about 90%, which some researchers think is low for this type of universal screen (Johnson & Ashurst, 1990) while others do not (Torgerson & Donaldson, 1994). Both SES and HVS achieve nearer to a 95% coverage of their stated child population. However, some reported coverage figures are much lower than these.

The average reported referral rates for HVDT, HVS, and SES are all about 10%. This is slightly higher than shown in chapter 7, but on a par with a recent study by Wood and colleagues (1997).

The reported yield from the HVDT is lower than might be expected from other work (Fortnum *et al,* 1997; Davis & Wood, 1992); it consistently identifies about 26–28% of the expected number of congenitally hearing-impaired children for the 50 or so providers (one in three) who supplied data for this study.

In a small number of Districts (n = 14, in 1994), data for both targeted neonatal and HVDT screens were available. There was no suggestion that the neonatal screen depressed the yield from the HVDT. This was most likely because the yield from the HVDT was low in these Districts, rather than because it was picking up late-onset or progressive impairments. The HVDT yield for the 14 Districts was very close to the yield of 25–28% from the 50 providers who supplied data for this study.

Both HVS and SES gave reported yields for bilateral hearing impairment close to 4 per 10,000 children.

The age at which hearing-aid fitting following the HVDT was thought to be routinely carried out was 18 months, which is earlier than reported elsewhere (see chapter 7; Fortnum *et al,* 1997).

Cost data for SES were only available in five Districts. However, these enabled an estimate to be made of the testing cost for SES of between £3000 and £4000 per 1000 children entering school, without any overhead costs being included.

The data on coverage, referral rates, yield and sensitivity were not available for a very large number of providers. Considerable effort is needed to standardise and coordinate such data so that it can be routinely available to help develop policy at both national and local level.

A more systematic approach to quality assurance is needed for all the areas of hearing screening considered in this survey. Some Districts have shown an exemplary level of quality assurance but these are very much in the minority.

Evidence from costs (from chapter 5)

The cost comparisons within the different implementations of hearing screening in the first year of life are encouragingly uniform, with systematic differences being observed between implementations such that UNS appears to have lower initial cost associated with it than the HVDT on a cost per child screened basis. The estimated cost per case is an order of magnitude lower with UNS. Targeted neonatal screening programmes cost between about a quarter and one-third of the cost of UNS programmes.

In terms of running costs (excluding start-up and equipment costs, but including employers full cost plus 40% overheads) in the nine Districts surveyed, HVDT screening is costing about £24,500 per 1000 live births, including follow-up of false-positives. This is reduced to about £20,600 when structured surveillance is used instead of the distraction test.

The mean standardised cost of UNS is about £13,900 per 1000 live births, in the three Districts conducting such screening in England, including follow-up costs. These data compare well with the costs in the USA for fully-funded services (rather than those which use unpaid volunteers).

Targeted neonatal screening is costing a mean of about £5100 per 1000 live births, testing between 6% and 10% of all live births. However, a yield in excess of 40% of congenital PCHI cases has not been consistently demonstrated. In the service format that is commonly used in the UK, that is, targeted neonatal screening and universal HVDT, the cost per case detected by HVDT is the highest (i.e. the least cost-effective).

The cost per test and the cost per hearing-impaired child detected were investigated to examine the relative differences between protocols. Under different sets of assumptions, the relativities were quite substantially in favour of UNS rather than universal HVDT.

If UNS was introduced, there would need to be transitional funding for two reasons:

(i) there would be a period of at least 6–8 months when the two screens would have to run in parallel because young children just discharged from hospital would not be screened at all if the HVDT was abolished on the day universal screening was introduced
(ii) there would need to be an initial bedding down of the UNS programme.

Further work is needed on the costs of additional services, including education, needed to meet the principles of screening (see *Tables 1* and *24*).

Performance of screens (from chapter 7)

Neonatal

High coverage (better than 90%) for universal screening is possible.

All methods of neonatal hearing screens show high screen specificity, generally well above 90% after a 'settling-in' period.

Evidence on screen sensitivity for moderate and greater cases of congenital PCHI is less available but estimates range from 80% to 100%, except for the PARC when used with at-risk babies, where sensitivity may fall to unacceptable levels. Generally, **programme** sensitivity (including cases which were not covered, and/or which are late-onset or progressive) may be estimated to be nearer 80% than 100%.

The two large cohort studies of universal neonatal hearing screening in the UK, including the controlled trial in Wessex, produce yields of the expected order, that is, 1–1.3 per 1000, but decrease the subsequent incremental yields of the HVDT to very low levels.

At-risk neonatal screening has a potential yield of about 60% of all cases. In practice, however, this is likely to be much lower because of the difficulty of implementing full coverage for all indicators of at-risk cases; perhaps 45–50% at best.

The median age of identification of those screened neonatally is of the order of 2 months, depending on follow-up procedures and severity of impairment; this is earlier than for cases not screened neonatally.

The extent to which the opportunities offered by early identification are capitalised on is affected by aspects of the follow-up services. Fortnum and colleagues (1997) present evidence that ENT referral for possible OME involvement can delay fitting of hearing-aids significantly for cases of moderate and severe, but not profound, hearing impairment.

Coverage of the HVDT screen probably falls in the range 80–95%, although there may be some urban areas where coverage falls to nearer 60%. There is some limited evidence suggesting that coverage **and** sensitivity is lower in the Asian population.

HVDT

Sensitivity estimates of the HVDT vary widely, from 18% to 88% (all degrees of loss). Recent studies and more powerful studies are suggestive of poorer levels of sensitivity. Severity of impairment affects screen sensitivity substantially.

Screen-positive, that is, 'fail' rate is of the order of 5–10%. Many of these cases will have fluctuating non-permanent hearing loss, associated with OME. This referral rate has considerable resource implications for services.

HVDT incremental yield may be at very best 40% but it falls off for the best Districts to low levels (e.g. 25%) when at-risk neonatal screening is introduced. With UNS, the evidence indicates that the

HVDT incremental yield falls to very low levels. (For those Districts who supplied data in the survey of current practice, the average HVDT yield was about 26–28% and at-risk neonatal screening made no difference).

Median age of identification via the HVDT varies from 12 to 20 months. Age of referral is severity-dependent.

Conclusions

There are approximately 840 children per year born in the UK with significant permanent hearing impairment, likely to affect their (and their family's) quality of life substantially. Present services will **not** identify about 400 of these children by the age of 18 months, and about 200 of these children by $3^1/_2$ years of age. Late identification of hearing impairment reduces the responsiveness of the services for individual children by significantly decreasing the flexibility in managing the habilitation of the child and family. Hearing-impaired children identified late will, in addition, be substantially delayed in their acquisition of language and communication with a consequent, longer-term risk to educational achievement, mental health and quality of life.

The universal HVDT has been used as the main screening programme to detect these children and is achieving a yield of less than 30% at a cost of between £60,000 and £125,000 per child detected. The programme is being managed by individual providers, in their own way, and there are no recognised national quality standards. It is very difficult to monitor the programme at District level because of the small numbers and the long wait to confirm missed children. The HVDT is not an efficient or cost-effective service. Furthermore, there are some data that suggest that the HVDT is an inequitable service as its coverage is low for 'non-white' and 'struggling' households (Varghese *et al*, 1996; Johnson & Ashurst, 1990).

In order to reduce median age of identification, targeted neonatal screening has been introduced in many Districts since 1994 in an *ad hoc* fashion, in many places without funding for full coverage of the major at-risk groups. However, this potentially further weakens the effectiveness (and cost-effectiveness) of HVDT. Furthermore, although its effect on the median of the distribution of age of identification may be substantial, its effect on the tail of the distribution may be slight, unless the quality of the HVDT is greatly improved. The yield of targeted neonatal screening, as presently implemented, is somewhat lower than expected (about 35% rather than 50–60%) but its sensitivity for moderate or worse PCHI has been assessed as between 80% and 90%.

Neonatal screening, in the UK, has been successfully implemented for targeted and universal screening. In the USA, there are now a considerable number of successfully operating universal neonatal hearing screening programmes but, as yet, very little data from systematic prospective trials. In the UK, the one research programme with a systematic prospective comparative study of UNS and HVDT has shown a substantial advantage for neonatal screening, with a greater yield and much younger ages of identification and aiding. These results for yield are corroborated by those from two further centres that use UNS routinely. Furthermore, UNS has a lower marginal cost than the HVDT, and a much lower cost per child detected. UNS in the UK has shown that coverage can be in excess of 90%, with a specificity about 95%. It is too early to assess sensitivity yet but, judging by the yields, it should exceed that of the targeted neonatal screens and may be higher than 90%.

Chapter 9

Evaluation of major options and recommendations

Scope

The major options that arise from the summary of evidence and conclusions in chapter 8 are reviewed here. These are then evaluated in the light of the major service principles of effectiveness, responsiveness and equity. The potential yield from each option and the estimated associated costs allow comparative value for money to be broadly assessed.

The options in identification of PCHI

The eight options that are considered are shown in *Table 23*. Several other options might have been considered but these were chosen to represent those we considered were addressed by the data, present practice and Focus Group discussions. Options that would reverse the *status quo* were not considered with the notable exception of Option 0. *Table 23* has eight columns for each option. The first column contains a brief description of the option itself, followed by two columns in which the marginal costs and the potential yield associated with the option are estimated (from the data summarised in chapter 8). The marginal cost is derived from the recurrent costs estimated for the option (including 40% overheads) within the context of the costing exercise described in chapter 5. These costs represent the stable long-term costs that are given in a standardised way for 1000 live births per year at 1993/94 prices. Other costs associated with the option are discussed within the text. The yield is derived from the data given in chapters 4 and 7 for the major components of each option and, where relevant, an incremental yield for each part of the programme is given.

An evaluation of efficiency, responsiveness and equity (see *Priorities and Planning Guidance for the NHS*. NHS, 1996) for each option is presented in the next three columns. Efficiency in this context is a combination of the yield, age of identification and associated potential benefit to the child and family in relation to the on-going costs for making those benefits available. Thus higher yields, giving earlier ages of identification for lower costs per case identified give better efficiency ratings. Efficiency is rated on a 5-point scale: 'very poor; poor; fair; good; very good'. The efficiency column includes an estimated cost per case identified, derived from the yield and the cost. Responsiveness, meeting the needs of individual children and their families, is more difficult to evaluate. However, earlier identification is assumed to enable a better degree of responsiveness and more appropriately formulated individual care plans. Responsiveness is evaluated on the same 5-point scale as efficiency. Equity is the improvement of the health of the population as a whole and the reduction of variations in health status. On the one hand, targeted screening must, in general, be considered less equitable than universal screening but, on the other, universal screening can often be as inequitable if the coverage is less than perfect because of the 'inverse care law' (i.e. those most at risk tend to be in the group least likely to be covered by the 'universal' screen). Similarly, significant variations in screening and follow-up services resulting from demographic factors (e.g. district of residence, socio-economic group, ethnicity) can be seen as inequitable. Equity is rated on a 5-point scale: 'very low; low; medium; high; very high'.

The final two columns contain an evaluation of the benefits of each option considered as a whole, and the challenges that each option presents. The challenges are predominantly those of implementation and apply to each option. The inadequacies in the present system are **substantial** and would benefit from clearer guidelines, better training and better facilities.

Should we screen for congenital hearing impairments?

Option 0 *(Table 23)* addresses the issue of whether the evidence supports screening for PCHI. This issue should be addressed primarily in terms of the prevalence of congenital hearing impairment, its effects, the availability of an appropriate screen and the cost of providing the screen in relation to the potential benefits, including consideration of the disbenefits of not providing a screen.

TABLE 23 *Different possible options, their costs, benefits and challenges*. It is assumed (i) that there will be HVS for all children aged 0–5 years (as per *Health For All Children*, cost unknown) and (ii) that SES will be retained for all options (at a cost of about £3000–4000 per 1000 live births). All costs are standardised per 1000 live births (not per 1000 children tested). The cost per case identified is a broad estimate based on the programme cost and yield

Option	Marginal costs associated per 1000 live births	Incremental yield	Efficiency	Responsiveness	Equity	Benefits	Challenges
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)	Very low	Releases time/ money to invest in responsive system, improve-ment in habilitation facilities for severe/ profound.	Moderate and severe PCHI not identified until > 2 years, possible identification if language screen about 2 years.
H0 Universal HVDT	£25,700 Total = £25,700	Present average yield is 0.25 which might be increased to 0.4 with good quality control.	Poor Cost per case: £80,000–100,000	Fair	Low	Would consolidate present services and remove uncertainty for health visitors.	Need better training, facilities and quality standards for HVDT. Age of identification sub-optimal.
H1 Hall report recommendations: targeted neonatal screening: introduce targeted screening where not already implemented; make more systematic where very limited at present. Universal HVDT	£5100 £25,700 Total = £30,800	Yield estimates if both NICU and family history groups can get high coverage and better HVDT quality control: 0.5 per 1000 by 6 months, 0.75 per 1000 by 1 year.	Poor Cost per case: about £40,000 (incremental cost per case for HVDT about £100,000)	Fair	Medium	Little change to system, would help build up targeted screening.	Need better training, facilities and quality standards for HVDT, for targeted screening and for very early habilitation in all Districts. Implementation of family history difficult. Overall sensitivity of system poor. Age of identification sub-optimal.
H2 As for H1, but following R&D implement technologically-advanced HVDT, with increased levels of stimuli	As for H1, with HVDT reducing to about £20,000 Total = £25,100	Yield as per H1, possibly increasing to 0.8 per 1000 by 1 year.	Fair Cost per case: about £31,000 (incremental cost per case for HVDT about £80,000)	Fair	Medium	Limited change to system, and better test possible for HVDT if accepted.	As H1, but sensitivity possibly better. New equipment needs to be developed, evaluated and bought.

continued

TABLE 23 contd *Different possible options, their costs, benefits and challenges.* It is assumed (i) that there will be HVS for all children aged 0–5 years (as per *Health For All Children*, cost unknown) and (ii) that SES will be retained for all options (at a cost of about £3000–4000 per 1000 live births). All costs are standardised per 1000 live births (not per 1000 children tested). The cost per case identified is a broad estimate based on the programme cost and yield

Option	Marginal costs associated per 1000 live births	Incremental yield	Efficiency	Responsiveness	Equity	Benefits	Challenges
H3 As for H1, but replace universal HVDT with 6–8 month universal HVS by questionnaire	As for H1, with health visitor costs reducing to £20,600 Total = £25,700	Yield as per H1 or slightly less, particularly for moderate.	Poor Cost per case: about £45,000 (incremental cost per case for HVDT about £84,000)	Fair	Medium Moderate impairment has low sensitivity.	Limited change to system, well trialled already. More in-line with health visitor mission.	As per H1
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 = £5100	Yield estimates, given high coverage of both NICU and family history children, 0.5 per 1000 by 6 months, but probably poor thereafter.	Fair to good Cost per case: about £10,000	Fair	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.
T2 As for H1, but replace the HVDT with targeted infant distraction test (IDT)	As for H1, but targeted IDT costing about £8000 Total = £13,100	Yield likely to be more than T1 but less than H1.	Good Cost per case: about £17,000	Good	Medium – likely to miss ethnic minority and low socio-economic groups	Better quality control possible, if fewer health visitors involved or specialist referral system.	As per H1 plus. Definition of health visitor target population, crucial to combat inverse care law; needs research to define.
U1 Introduce UNS	£14,000 Total = £14,000	Yield 0.9 per 1000 by 6 months.	Very good Cost per case: about £15,000	Good	Medium	Identification age 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.
U2 Introduce UNS. Modify IDT to be targeted on those not tested and at high risk of progressive PCHI	£14,000 £3000 Total = £17,000	Yield 0.9 per 1000 by 6 months and possibly 1.0 per 1000 by 1 year.	Very good Cost per case: about £17,000 (incremental cost per case for IDT £30,000 very approximately)	Very good	High	Identification age 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.

In terms of the prevalence/incidence, there are estimated to be 840 congenitally hearing-impaired children born in the UK each year (see chapters 2 and 8) with a moderate, severe or profound hearing impairment. The impact of the hearing impairment on these children and their families is considerable, wide-ranging (see chapters 1 and 3), and changes over time through its impact on the child's development (see chapter 3). Late identification of these children (particularly those with more severe impairments) is clearly associated with poor outcomes (see chapters 1 and 3), while earlier identification is associated with better outcomes (see chapter 3). The **extent** to which good outcomes may be associated with **very** early identification is not yet clear but it does give the greatest responsiveness for habilitation. The most recently available information on age of identification of congenital PCHI (relating to the services provided in the late 1980s and early 1990s) shows that perhaps as many as 200 congenitally hearing-impaired children (out of 840) per annum are not identified until after $3^1/_2$ years of age (see chapters 2 and 8). These children will be substantially handicapped compared with their peers identified in the first year of life. With no screen in place, the situation would be considerably worse. Screens are available (see chapter 7) that are relatively cheap and, given appropriate training and quality control, for example, within a national programme, they could give high sensitivity, specificity and yield. The evidence in favour of a (national) screening programme for congenital PCHI is thus compelling. The question that follows concerns the extent to which present systems (which vary considerably from area to area) should be changed to bring about:

(i) earlier identification for the majority of hearing-impaired children
(ii) greater benefits for the child and the family
(iii) a more cost-effective screening programme
(iv) greater uniformity of service.

Appraisal of different options for screening for congenital hearing impairment

In general, there is no reason to doubt the arguments and evidence in favour of screening for PCHI (Option 0 is not to be recommended). The remaining options in *Table 23* (and other combinations which could be constructed) include the various approaches to screening for PCHI currently found in the UK and elsewhere. This service variability is insupportable in terms of the evidence on cost-effectiveness, however, and is difficult to justify in terms of equity. The terms of reference for this review, which was in part commissioned because of such piecemeal activity and provider creep, included evidence-based recommendations and the development of a more national approach to hearing screening if the evidence was sufficiently clear.

The options presented in *Table 23* are in three categories according to the core screening activity: **H** (HVDT/*Health for All Children*), **T** (targeted neonatal screening) and **U** (UNS). It should be noted that children identified earlier, by any screening programme, would need, on average, 14 months additional audiological care, educational and family support.

Options H0, H1, H2 and H3 are versions of the same basic schema (Hall, 1996). Options H1–H3 covers targeted neonatal screening (constant over these options) coupled to universal HVDT (changing over each option). Option H0 is essentially the current HVDT regime alone, with no targeted neonatal screening. The best yield achieved with this approach is low (about 40%) – most Districts achieve nearer 26–28% – and thus the cost per case is high. The age of identification is non-optimal and, hence, the services are not as responsive as they could be.

Option H1 is essentially the *status quo* **but** with considerable consolidation and improvement consistent with becoming a more uniform national screening programme, with appropriate quality standards that could be audited. Targeted neonatal screening would need to be introduced systematically in each District (there would be a capital cost associated with making equipment and test facilities available) and, in Districts where the targeted group is currently restricted, it would need to be brought up to standard. Considerable effort would be needed to implement the family-history risk criterion. The yield from targeted neonatal screening of 0.5 per 1000 assumes that about 6% of the birth cohort is tested because of stays of > 48 hours in an NICU/SCBU (including CFA) and 4% because of family history (10% overall), with relative risks of congenital hearing impairment (as shown in chapter 2) for the NICU/SCBU history and family history groups. In addition to the neonatal screening, the HVDT would need to be up-rated in line with current best practice, with better training, facilities and quality standards. This might incur a higher cost than given in the short term but this cost is assumed to be similar over all options except for facilities that may require capital expenditure.

For Option H1, the yield, **at best**, would be 45% of the congenital PCHI identified and assessed in the first 6 months, and up to 70% by 1 year of age. This would represent considerable improvement over the provision described in chapter 2, which had a 37% yield by 1 year and a 70% yield by about 3 years of age. Option H1 represents a limited change in philosophy to much of the present system but would entail considerable renewed effort to implement.

Option H2 is a variant of Option H1, in which the organisation is essentially similar but the test component of the HVDT is made more uniform, consistent and cost-effective by using technically more advanced equipment to present the stimuli and help conduct the test. Essentially, the equipment (BeST test) would replace the second tester, and might use a signal at a higher level to overcome poor test facilities (B McCormick: personal communication, 1996). This would increase the sensitivity of the HVDT for the target group (those with at least moderate hearing impairment), reduce false-alarm rates and reduce testing and follow-up costs. At present, the test is at an early stage and needs further research and development. Option H3 also obviates the need for a second person, by replacing the HVDT with a structured questionnaire (Scanlon & Bamford, 1990; Sutton & Scanlon, 1997). There have been trials of such an arrangement and the cost associated with it is less but it is less good at detecting those with a moderate hearing impairment than Option H1 or H2. Most of the comments made about training and implementation effort for Option H1 apply also to Options H2 and H3.

Options T1 and T2 can be considered together. Option T1 is to establish targeted neonatal screening as the major national screening effort for the identification of PCHI, and phase out the HVDT altogether. In its place this option introduces targeted neonatal screening only, as already presented in H1. Thus, we would expect to obtain a yield of 0.5 per 1000 very cost-effectively, but there would be considerable problems in terms of trying to find the other 0.62 cases per 1000. Some children would receive a very much improved service but others possibly a worse service. Thus, Option T1 is particularly low in equity and only 'fair' in terms of the responsiveness in the system. In addition, if the risk factors do change over time and place, the yield would change as well (Davis & Parving, 1994; Davis *et al,* 1995). This would not be a major problem for Option H1 but would be for Option T1. To address this last issue, the risk factors used for targeted neonatal screening might have to be tuned in relation to local epidemiology. Option T2 tries to overcome some of the problems of lack of yield in Option T1 by using a targeted infant distraction test (IDT) (that is, the same test as undertaken by health visitors but not necessarily conducted by health visitors) in addition to the targeted neonatal screen. There are two advant-ages in using a targeted IDT: first, the cost is lower than a universal test and, second, the test could be restructured so that only a few people would conduct it, thus enabling easier training and better quality control. The nature of the targeting for the IDT is something that would need further research to clarify. It might include some of the following groups of children:

(i) those who were included in the targeted neonatal screen population but could not be tested
(ii) those with a borderline test result for the neonatal screen
(iii) some ethnic minority children who have a raised risk of 2.49:1
(iv) children from households with postcodes that are characterised as having very low SES where there is also an increased risk
(v) children moving into a District
(vi) children known to be at risk from late onset or progressive loss, although the evidence that these will have appeared by 6–8 months of age is not encouraging.

The costs for Option T2 are fairly well known for the targeted neonatal screen but difficult to estimate for the targeted IDT, which would vary according to the demographic make-up of individual districts. We have estimated the cost as about £8000 per 1000 live births, to include the test and follow-up, for about 15% of the birth cohort using two testers in well-constructed facilities. The overall cost for Option T2 is in the region of £13,000 per 1000 live births and is likely to be more equitable than Option T1 and more efficient. It would also have a better responsiveness.

Options U1 and U2 both have universal neonatal hearing screening as their core screening activity: U1 includes UNS alone; U2 includes a targeted IDT as well. These two options logically extend Options T1 and T2. The yield from U1 is estimated conservatively at about 0.9 per 1000 (a combination of the programme sensitivity of about 85% derived from the evidence in chapter 7 with the prevalence evidence in chapter 2), of those whose hearing impairment would be confirmed before 6 months of age. This yield is lower than the actual yield from the controlled trial in Southampton, 1.1 per 1000, and the data from Whipps Cross/Waltham Forest, 1.3 per 1000 (see chapter 7). The estimated cost

of running U1 is about £14,000 per 1000 live births. The efficiency of the programme would be very good, and the cost of identification for each hearing-impaired child within the first few months of life would be about £15,000–16,000, at least one-quarter of that for Option H1. The responsiveness of the programme would also be good. However, some children may not be tested and children not tested in screening programmes have a higher probability of impairment. The programme is therefore not as equitable as it might be but is better than T1 and T2 in this respect.

Option U2 addresses the problem of equity by incorporating a highly targeted IDT (possibly run by the paediatric audiology clinic) to test at 6–8 months of age those children who were not neonatally screened and those who have recently moved into the District with no record of a neonatal screen in their parent-held record. In the long term, it might be worthwhile to target those children who were more at risk of a progressive hearing impairment, for example, where there were known relatives with progressive impairments, or relevant perinatal infection. However, more research is needed on this group for this to be implemented effectively. Thus Option U2 represents a high degree of equity, very good responsiveness and would be very good in terms of efficiency, with a cost of identification of each hearing-impaired child of about £17,000. The age of identification would be the best achievable at present, giving a substantial boost to the child's potential to accept hearing-aids, develop language and communication (spoken and/or signed) and giving the widest choice in terms of habilitation. However, this would mean changing the present system substantially. The HVDT would have to be phased out in its present form. A training programme for neonatal screening would need to be implemented together with the framework for quality control. Once established, however, better quality control would be possible since there would be far fewer testers than in most existing schemes.

The main points of each option are summarised in *Table 23*, and it is clear that the best options in terms of the criteria of incremental yield, efficiency (which takes cost into account), responsiveness and equity are U1 and U2 in the long term. Efficiency, which would be very poor for H0, would increase progressively for Options H1, T1 and T2, and is best for Options U1 and U2. Even if Option H1 were very well implemented (which would take considerable training effort) the cost per child detected would be about £40,000 compared with £17,000 for U2, and the median age of identification would be considerably later. The cost per child detected in Option H1 by the HVDT component alone would possibly be in excess of £100,000. Option U2 also allows the greatest responsiveness for the largest number of hearing-impaired children.

We have identified Option U2 as the best overall long-term screening programme. There are several challenges that would face services if such an option were adopted as a national screening programme. Training has already been identified as a major issue for all the options presented in *Table 23*. National support for training would be highly desirable so that it could be undertaken in a systematic and approved manner. The transition between the present system and Options U1 or U2 is challenging and would have to be well-managed. At present, the cost of the HVDT is usually to a community trust and that of neonatal screening to a hospital trust, and it is not a simple matter of transferring funds for one activity to another over time. Furthermore, it would be highly inequitable to introduce Option U2 and immediately discontinue the HVDT. Babies discharged from hospital before Option U2 was introduced and all other children who had not yet had an HVDT (i.e. those under about 6–8 months of age) would not then be screened. The HVDT would therefore need to be continued for up to a year after the introduction of the neonatal screen. This would incur a one-off cost of about £25,700 per 1000 live births if it was for 1 year, and less *pro rata* if the transitional period were shortened. This effectively brings forward the costs of screening for an annual cohort of children.

Other one-off costs would include the start-up costs of providing office space for the testers, a room for testing NICU/SCBU children and storage for testing equipment. UNS programmes are particularly vulnerable at the beginning (e.g. the early coverage in the Wessex trial) and additional testing time may be needed at the very start of the programme. In terms of equipment, each hospital/maternity unit would need to have available automated ABR or ABR equipment for NICU/SCBU and early-discharge children, and OAE/ABR equipment for the remaining children, depending on which equipment was preferred. An overview of such equipment is presented in Appendix 3, supplied by the National Center for Hearing Assessment and Management (NCHAM) programme in the USA. In the UK, an estimate of the purchase cost for such equipment is £6000–9000 per 1000 live births.

In addition to such one-off costs there would be additional health and educational expenditure, if

the children are identified earlier rather than later. To a certain extent this would be true of any option which is an improvement on the present system in terms of age of identification. The extent of such additional expenditure for Option U2 is difficult to estimate but, in the year after identification, if audiological reviews were carried out every 3 months, there might be an expenditure of £2000 per child for hearing-aids, their fitting and assessment, ear moulds and hearing assessments. Appropriate educational input for the family and child might cost in the region of £4000–6000, depending on the frequency of visits by a peripatetic ToD and others, and on the support programme initiated.

Option U2 has been examined in some detail concerning its on-going costs (£17,000 per 1000 live births), its start-up cost (£6000–9000 per 1000 births in the first year, including equipment maintenance and calibration in 4 subsequent years), its transitional costs (£16,000–24,000 per 1000 births in the first year only), further health provision (£2000 per child in the first year) and educational provision (£4000–6000 per child in the first year). However, it should be noted that a sum equivalent to the transitional cost would have to be paid in any case (on a *per capita* basis) to screen these children, and that the £3000 allotted for targeted IDT would not necessarily be spent in the first year if a good quality HVDT was being performed. Options other than U2 would also have inevitable start-up costs; indeed, if Option H1 were taken up by Districts the start-up costs (and catch-up costs) for Districts who do not have targeted neonatal screening would be significant (possibly £3000 per 1000 live births). Only the running costs of the options have been estimated (Stevens *et al,* 1997; see chapter 5), including the follow-up costs. The remaining costs are our own best estimates. The costs for staff to put together the business case, quality control and training, and for liaison between professional groups has not been included. It was considered that these should be in place no matter which option was preferred. However, in many Districts these arrangements are not in place and, in our opinion, such arrangements should be a prerequisite of any screening programme.

It is useful to compare the costs of hearing screening with other neonatal screening programmes currently under review. Such costs (RJ Pollitt: personal communication, 1997; Pollitt *et al,* 1997) have been broadly estimated. Phenylketonuria has an estimated cost of £27,000 per case, congenital hypothyroidism of £14,900 per case and cystic fibrosis of £4700 (but that is the cost marginal to the facilities already being in place). However, it is not possible to compare the **cost–benefits** of screening between these conditions because hearing impairment has a markedly different impact on the individual and family, and appropriate outcome measures have not been developed.

An evaluation of the preferred option in terms of screening principles

If the preferred option involves UNS, we must be sure that it meets the principles of screening and its pre-requisites, as discussed in chapter 2. These 14 points are reviewed in *Table 24*. In general, the principles are addressed by data in this review. There are some instances where there may be cause for concern or a need for further research but these probably apply across all other options in *Table 23*. The first concern is for principle 3, that is, that facilities for assessment, diagnosis and habilitation should be available. From chapter 4 and Appendix 2 it can be seen that there are some Districts with limited facilities to manage very young children with hearing impairment. However, this is not seen as a bar to screening, rather as an opportunity to develop those services, both health and education, that should be in place.

A second cause for concern arises from principles 11 and 12. We are concerned that the role of parents/family is not considered uniformly. Guidelines for best practice need to be developed so that the incidental stress caused by the screen is minimised. Furthermore, in cases where a hearing-impaired child is identified, appropriate family support mechanisms (including support from the voluntary sector) need to be in place at the earliest opportunity.

Two further questions arise from the principles. The screening programme is designed to identify congenitally hearing-impaired or deaf children with at least a moderate (≥ 40 dB HL) hearing impairment. Principle 7 is that the natural history of the hearing impairment should be known. There will inevitably be children who develop hearing impairments, either through acquired or genetic aetiologies. These children will not be identified by a neonatal screen and we have assumed that the proportion in this group is quite small (10–15% of PCHI). Although one study shows a much higher proportion in the at-risk population (Stevens *et al,* 1997), the yield from programmes in place, in the UK and the USA, are more consistent with our estimates. However, further research is needed to identify which risk factors (e.g. genetic) may identify those with late-onset or progressive impairments.

TABLE 24 *Tabulation of screening principles adapted for universal neonatal hearing screening and how evidence from the review addresses each principle/precondition (adapted from Haggard, 1993)*

Principle/precondition	Does UNS affect this principle?	Comments
1 The hearing impairment to be screened for should be an important health problem.	NO	Prevalence shown (chapter 2) to be over 800 children per year. Impact is large (chapter 1) but depends on severity. Delayed identification and rehabilitation is associated with poorer outcomes (chapter 3).
2 There should be an accepted habilitation for cases of PCHI identified by the screen.	NO, but cost may increase.	Early acknowledgement of problem highly desired by parents (chapters 3 & 7), early intervention programmes exist and are acceptable (chapters 3 & 4) and beneficial (chapter 3).
3 Facilities for assessment, diagnosis and habilitation should be available.	Many places may not yet have these facilities.	These exist in the majority of Districts, but there are some where there might be considerable reservations about these facilities and training (chapter 4).
4 The hearing impairment should be recognisable at an early stage.	NO	Hearing is affected from birth and can be detected (chapters 2, 3 & 6)
5 A suitable hearing screening test should be available at the proposed age for the screen (and should be quick, have good sensitivity and specificity, and easy to interpret).	YES	There are suitable tests, with adequate sensitivity, specificity and ability to cover the population. These may differ for sub-populations, e.g. both NICU and < 6-hour discharges may need different protocol to 'healthy babies' who are discharged later (chapter 7).
6 The hearing screening test should be acceptable to both child and parents.	YES	The tests used are acceptable to parents and professionals; none are invasive or cause the child pain (chapters 3, 4, 6 & 7).
7 The natural history of childhood hearing impairments should be known and understood.	Small number of progressives missed.	The different aetiologies and onset patterns are discussed in chapter 2; the proportion of children with progressive/'late onset' hearing impairment varies but is likely to be nearer 10–15% than 40%. More research needed in this area.
8 There should be an agreed policy on whom to treat as patients with hearing impairment.	YES	There is evidence that moderate hearing impairment and worse should be the target group for very early intervention. This may be extended, if tests are sensitive enough; further research needed (chapters 2, 3, 6 & 7).
9 The cost of hearing screening (including all assessments consequent on screening) should not be disproportionate compared with other health and related care costs incurred by a hearing-impaired child.	May reduce health costs.	Cost of screening is discussed in chapter 5, and overall cost of hearing screening is a combination of neonatal, HVDT, HVS and SES. The neonatal screening component is presently small in relation to the overall amount. UNS appears to provide a low-cost package for screening within the first year – but it generates cost in other areas, particularly education because it is effective. Costs and benefits of different screening packages are explored elsewhere in this section (chapters 4, 5,7, 8 & 9).
10 Finding cases of childhood hearing impairment should be viewed as a continuous process.	Needs to be emphasised.	This underlies the Hall report philosophy for hearing screening that should be maintained (chapters 2 & 3).
11 The incidental harm of hearing screening programmes, e.g. stress to parents, should be small in relation to overall benefits.	Stress to mothers of healthy babies may occur exceptionally.	There would be few causes for concern provided national quality standards and information packages in place (chapters 3 & 7).
12 There should be guidelines on how to explain results of hearing screening, together with transitional counselling support for those parents of children who have undergone a hearing screen and are concerned.	Needed	This area is a cause of some concern but there is further scope for agreement on a more uniform approach to this problem.
13 All hearing screening arrangements should be reviewed in light of changes in demography, epidemiology and other factors.	As per any screen.	The need for this review, and for national shared list to audit progress.
14 Costs and effectiveness of hearing screening should be examined in a stratified manner, and benefit maximised in each stratum.	As per any screen.	Addressed in chapters 2, 4 & 5. We have considered severity of impairment, NICU and early discharges as strata over which to look at screen implementation, performance and cost.

Principle 8 is concerned with those we regard as 'cases'. We have justified our decision to regard only those with moderate impairment as legitimate cases for the screening programme in several places in the report (see chapters 1, 2, 7, 8 and 9). We believe that there will indeed be cases of mild or unilateral impairments identified by Options U1/U2 and that this is likely to be beneficial to the individuals so found. Appropriate early management and habilitation programmes need to be developed for these children, and this should be the subject of further research. However, such cases should not be included in the justification or audit of the programme.

Recommendations

Context

The recommendations fall into three categories: service development, implementation and research (the latter is considered separately in chapter 10).

Service development

The weight of evidence strongly supports the introduction of universal neonatal hearing screening, supplemented by a targeted infant distraction test at about 7 months of age (primarily for those children not screened neonatally). This option (U2) is the most equitable and responsive, provides the best value for money and potentially offers the greatest benefit for hearing-impaired children and their families.

We recommend that the National Screening Committee gives urgent consideration to the evidence-based case we have presented for:

(i) a national screening programme for congenital hearing impairment
(ii) implementation based upon Option U2
(iii) the development of an information system strategy that will facilitate the coordination of the services needed for hearing-impaired children in line with NHS priorities 1996–97.

There are two further service recommendations which concern the roles of health visitors and paediatric otolaryngologists:

(iv) a systematic appraisal is required of the role of health visitors in the identification of children with late-onset or progressive PCHI. Due consideration and priority should be paid to other aims included in the health visitor's role, for example, detecting the effects or signs of persistent 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
(v) children who are being assessed for PCHI should have access to specialist paediatric otological opinion. This will help reduce the chance of delays in initiating appropriate habilitation that have occurred in the past from ENT referral.

These recommendations stem from the evidence reviewed in chapters 2–7 and from the on-going service context. If a hearing screening programme were being set-up *ab initio* a further large-scale RCT might be required. However, with neonatal hearing screening there is a very strong case for recommending setting-up a national programme without further long-term research. First, there is good trial evidence for universal neonatal hearing screening from the completed Wessex trial (see chapter 7 for a review of this evidence which is currently being prepared for publication by Kennedy and colleagues); second, an existing although inadequate screen has been in place for 30–40 years and to withdraw it without replacement would be unacceptable; third, the potential harm from treatment consequent on detecting cases is not a major factor in the case of screening for a hearing impairment.

Implementation

The components and targets that might make up a model screening programme are discussed below, followed by consideration of management, coordination and purchasing issues before a discussion of the transition from the present to the recommended option. Finally, some implementation issues are discussed in more detail.[2]

[1] 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 play a key role in monitoring any national screening programme. This strategy should, where possible, link into local Child Health-Record information systems.

[2] Some of these implementation issues will be the subject of an NIH (USA) report, presently being prepared by Professor Susan Norton and her colleagues for possible publication in 1998.

Model screening programme and its targets

A proposed model is recommended with the following components, as outlined in Option U2, with verifiable levels of performance:[3]

(i) UNS with the following quality markers – initial coverage > 90%; coverage on call-back 95%; NICU/SCBU coverage 99%; sensitivity for **congenital** hearing impairment of ≥ 40 dB HL of at least 85%; maximum false-alarm rate of 10% for non-NICU/SCBU babies on first test and for NICU/SCBU babies 20%; false-alarm rate for non-NICU/SCBU babies after repeat (second screen) tests of 2–3% and for NICU/SCBU babies 5%
(ii) about 1% of all births should be audited at random to confirm that the testing was appropriately carried out, and that parents were satisfied with information given, consent sought and the procedures
(iii) surveillance procedures should be carried out as per recommendations of *Health for All Children* (Hall, 1996), with an annual appraisal of the procedures in place or when the management changes substantially
(iv) a targeted IDT should be implemented for all children who are not neonatally tested, who have moved into the District and for whom there is no record of a neonatal test (this will be a smaller burden when UNS is widely implemented) and for any child for whom there is parental or professional concern; this screen should be carried out in proper facilities by staff with appropriate audiological training and experience
(v) the ages of identification of PCHI and start of habilitation should be monitored, on a stratified basis of degree of hearing impairment and on key demographic variables to ensure on-going equity of the programme.

Structure and coordination

A single person or committee should have responsibility for planning and implementation of the hearing screening programme. The planning and coordination of the programme should be focused around:

(i) early detection
(ii) formulation of individual care plans in the first months of life.

Hence, it is important that all key professionals should be involved in the formulation of the policy. Implementation issues should be the responsibility of the programme manager and the local NHS Trust line management with due consideration for the impact this will have on other professionals. An alliance of the professionals in each District (with appropriate involvement of the voluntary sector) would lead to the formulation of a better integrated service, which is a first priority for clients and professionals alike. The framework of the *Children Act* and the *Children's Services Planning Order* to be implemented in 1997, should serve as a guideline for the provision of care and it should be realised that, for children with PCHI, the duty of care is on-going from the time of referral from screening.

Purchasing

There is a need for considerable integration of the services received by children with PCHI and their families. Therefore the framework of either joint purchasing or joint commissioning of services should be explored by the relevant purchasers and providers. In this way, the true benefits of early detection might be better achieved and audited. This is a complex procedure for which no model programmes exist and therefore gives considerable scope for innovation and service development; however, there may also be pitfalls because of boundary and other differences.

Management

It is preferable for the neonatal screening programme to be managed by an individual with appropriate training and expertise in audiology, test procedures and management of multi-disciplinary teams. The day-to-day management should be delegated to appropriate professionals (e.g. nursing, scientific) who have access and links to specialist advisors in audiology, medical physics, ENT, child health, and education. Consideration should be given to the information system requirements and the need for further specialist training.

Transition

It is vitally important that, in the transition between the services presently in place and any new services, adequate arrangements are made for the evolution of the services. An abrupt cessation of universal HVDT is neither possible, as mentioned above,

[3] We do not mean to be prescriptive here but to give an idea of what is required. Some Districts may need very different components because of demographics, ethnicity or geography. Thus a rural District with 40% of births outside the District general hospital may need to consider a different model programme using the acute, community or GP sectors, as appropriate.

nor desirable. The cessation of the HVDT does not mean that the health visitors do not have a role in case detection through surveillance and parental concern.

Detailed implementation issues

The number of at-risk neonatal screening programmes in the UK has grown in the last few years. In addition, there are two implemented UNS programmes in the UK and very many in the USA. The USA experience has led to the creation of NCHAM, one of whose aims is to assist services in implementing effective neonatal screening programmes. NCHAM has developed procedures and materials "...for training people to implement and operate successful newborn hearing screening programmes" (White & Maxon, 1995). There is, therefore, considerable expertise to be drawn upon in developing and planning a UNS programme, much of it is available in the published literature. In brief, the following issues need to be addressed.

1. **What is the case definition? That is, which group of children is the screen aiming to identify?** While many North American programmes aim to pick up all permanent hearing impairments, including mild and unilateral, those in the UK have tended to aim at (and audit, therefore) moderate and greater bilateral losses. That is not to say that mild or unilateral losses are unimportant, or that they should not receive appropriate support and intervention when found. However, the evidence for outcome benefits is less equivocal for moderate and greater bilateral losses, and treatment/support is therefore easier to provide and justify. We consider that a national neonatal hearing screening programme aimed at detecting bilateral impairments of ≥ 40 dB HL is extensively supported by the evidence summarised in chapter 8. Widening of the programme should only be considered if the evidence supports it.

2. **Which testing methods/device(s)?** At present the choice is between PARC, TEOAE, and ABR, or some combination of these three. The evidence indicates that a successful neonatal screen could be implemented with any of these (provided all other implementation issues were dealt with appropriately), although the PARC is likely to be inappropriate for at-risk special care screening and for outpatient postnatal screening. For UNS, a combination of TEOAE and ABR is the most researched and most likely to be optimal. The evidence is that automated ABR may be better for screening NICU/SCBU and early discharge babies (Mason *et al,* 1997; Lutman *et al,* 1997), while TEOAE may be better for the large numbers of healthy babies discharged after 12 hours in maternity units (see Appendix 3 for further discussion of testing devices).

   New technology in OAEs and ABR is currently undergoing trials. Simultaneous binaural ABR screening devices may reduce test time with no loss of sensitivity or specificity. Maximum length sequence OAEs (e.g. Picton *et al,* 1993; Thornton, 1993) may offer better specificity for early discharge babies.

3. **What screening system? That is, what combination of tests and retests constitute the total screen?** The possible combinations here are numerous, and include, for example, decisions on devices, test–retest options, inpatient and/or outpatient recall. Again, the evidence suggests that most combinations can be made to work given high quality implementation. ABR must be included at least at follow-up: the "...**local provision** [our emphasis] of a neonatal ABR [follow-up] service is considered an essential precursor to implementing a TEOAE screen" (Watkin, 1996), but automated ABR or ABR as a screen is probably to be recommended for NICU/SCBU babies.

   The most commonly emerging system is currently TEOAE as in-patient, followed by repeat TEOAE, if the first test is inconclusive. This is either as an in-patient or as a recalled out-patient. A referral on the second test would be followed immediately by screening ABR. Given non-attendance rates, and the evidence from outpatient or community-based programmes (Rowe, 1991; Kei *et al,* 1997) it is probably easier to achieve high coverage with inpatient neonatal testing, although a significant proportion of home births, small birthing units, or very early discharge of full-term babies may work against this. Comprehensive figures on neonatal discharge times in the UK are difficult to find but some current data (see *Figure 5)* suggest that 8% are discharged the same day and 25% the day following birth. About 1.4% are born at home (National Perinatal Epidemiology Unit: personal communication, 1997).

4. **What stimulus details and pass–fail criteria?** While detailed systematic trials have not been carried out to define the optimum stimulus

and pass–fail criteria, sufficient consensus is now emerging to guide new programmes. These choices will also depend upon earlier definitions, for example, of target cases, and on the NIH trial conducted by Professor Susan Norton.

5. **What are the staffing and training needs?** Both Kennedy and colleagues (1997) and Watkin (1996a) provide evidence that a new programme will take 9 months of familiarisation and improvement before stabilising at accepted levels of coverage and specificity. So it is important that staffing issues and training are carefully managed from the onset of the programme. Feasibility studies have indicated the time per test, number of babies screened per day or week, sickness and holiday cover requirements, and so on, for both at-risk screening and UNS (e.g. Stevens *et al,* 1991; Tucker & Bhattacharya, 1992; Watkin, 1996a; Kennedy *et al,* 1997). Data from Watkin (1996a) indicate the need for two full-time staff covering normal working hours for 7 days per week (including holidays and public holidays) to achieve acceptable coverage for his TEOAE universal screen (retest and ABR retest done at outpatient recall). His evidence suggested that one person could undertake 20 TEOAE tests per day, or 3–4 per hour. Hall and Garner (1988), using the ARC, suggested similar numbers of full-time employees for 95% universal coverage and 0.5 full-time employees for at-risk screening, allowing 30 minutes per test. The quoted test time of 2–3 minutes or so takes no account of time to, for example, input data, prepare the baby, talk to parents. Thus, Watkin's (1996a) test time for TEOAE was 3–4 minutes per (in-patient) child but the test rate was only 3–4 per hour.

   Daily birth-rate variability is another issue to be considered. Watkin (1996a) reports the local range in the daily rate for newborn babies as being 4–25, with 20% of days having more than 16 births and 10% having less than 8. In terms of efficient use of staff time this presents problems, and the question programme managers will face is whether to increase staff to cover all days, including the busiest, or accept a lower coverage and recall those missed. In Watkin's programme, 86% of babies were tested before discharge (mean age 32.7 hours, excluding NICU/SCBU babies) with the remainder called back (mean test age 7.4 weeks).

   Other staffing issues include the question of qualifications and training. Most workers argue for neonatal nurses to carry out at-risk screening although MTOs do play a large part at present, but less qualified screeners could be used for universal testing. Training of screeners is an important issue and there is a need for the development of an accredited national training programme. Packages such as that described by White & Maxon (1995) may be helpful models to build on. Drawing on the growing experience of established programmes, consortia need to be formed to support neonatal screening activity and to provide education and training. Apart from NCHAM in the USA, there is now the International OAE Data Centre (IODC) based in the UK. A subdivision of IODC, the UK Consortium on Hearing Assessment with OAEs, "...aims to foster communication between UK users of OAE technology, organising training and other events including a mutual self-help programme".

6. **What are the follow-up issues?** These can be divided into audiological/testing/timing issues, and questions concerning the management of true-positive cases.

   Given the anxiety for parents that may be engendered by cases that do not pass the screen, follow-up without undue delay is essential. Furthermore, full ABR testing without the need for sedation is easier the younger a baby is. However, there are neurological maturity considerations which argue for some delay, and most programmes suggest that 4–6 week follow-up testing may be optimal. Full 'diagnostic' ABR is essential (Watkin, 1996a), as is a well-found audiology service, in order to secure the fullest audiological profile of the child as quickly and efficiently as possible. Necessary procedures on follow-up will include oto-admittance testing and other age appropriate tests such as behavioural observation audiometry, visual reinforcement audiometry (including ear-by-ear insert phone testing) and further OAE and ABR testing. Some of the expected outcome benefits for children with PCHI depend upon early and appropriate hearing-aid fitting and services should be able to use the techniques and procedures appropriate for very young infants (e.g. Seewald, 1992; Westwood & Bamford, 1995). The estimation of frequency-specific thresholds in infants under 6 months of age is challenging and techniques such as notched-noise ABR (Stapells *et al,* 1995) need to be

considered. Cases will be confused by the presence of fluctuating OME, and procedures (e.g. bone conduction ABR) and personnel (e.g. specialist paediatric otologists) need to be available to remove uncertainty in this area.

The importance of the family at the centre of these procedures cannot be overemphasised (NDCS, 1996). This is a sensitive time for parents and families, and considerable damage can be done by service provision models which marginalise or de-skill parents – classic shortcomings of some traditional models of care. The understanding of what is meant by family-centred services is more highly developed in the USA than in the UK, and there is a need to enhance services in this respect in the UK.

With regard to the on-going management of true cases, it is emphasised in *Figure 1* (see chapter 1) that neonatal hearing screening and subsequent identification is not the end but the start of a lifetime of multi-agency provision, with child and family at the centre. Procedures for educational service involvement need to be immediate, preferably at the point of confirmation (NDCS, 1996). Information for parents about Social Services and the voluntary sector are a statutory obligation, once hearing impairment is confirmed. The voluntary sector has a key role to play in providing support and information for parents of hearing-impaired children, and in helping to bring about a family-centred service.

7. **What are the coordination issues?** Evidence from Focus Groups clearly showed that the top priority for parents and professionals alike was a seamless, well coordinated programme, with high levels of quality control and good, written parental information. Parents perceived current audiological services as fragmented and poorly coordinated. They argued that adding neonatal screening to existing fragmented services would be dangerous and anxiety-provoking **unless** steps were taken to ensure a well-coordinated screen and good coordination at follow-up, and good coordination between health and education services for true-positive cases. The evidence from the Focus Groups suggested that this was **the** major issue for parents and professionals alike.

Securing a transparent, seamless, well-coordinated screening and follow-up service requires quality control mechanisms, audit, a coordinator or coordinating team, and good quality information and tracking systems. A significant proportion of families with infants relocate and information systems need to be developed which allow case-tracking across geographical areas. Existing programmes and interested groups have designed appropriate case-management software which is available for use (e.g. the NCHAM HISCREEN® software, OZ systems software). Such software projects aim to assist in the management of neonatal screening programmes, and are essential for successful long-term implementation. However, such systems were developed mainly for use in the USA and are essentially stand-alone. There is a need to see how such systems might be integrated within the Child Health Information Systems that are currently used in the UK.[4]

NCHAM has produced a document which outlines some of the issues which need to be considered in selecting equipment. The document confirms the point that successful neonatal screening programmes are being run with different types of equipment and that any combination may be made to work effectively if the implementation issues are properly addressed. The NCHAM document is reproduced in Appendix 3 in its entirety (with permission); note that some of the figures quoted (e.g. on costs, on sensitivity) may differ somewhat from those presented in this review – where this is the case, readers are advised to defer to the review figures.

Finally, the formation of a national training strategy for hearing screening should be considered as a high priority. As well as short courses and workshops, a task force to help individual Districts plan for UNS should be considered, since the need for consistent practical and effective advice would overwhelm the resources of those who currently do this at the margins.

[4] As experience with service-based neonatal screening grows, so established programmes or consortia are producing advisory materials to help other services setting up new programmes. Thus, in 1996, Kimm (Wessex Neonatal Hearing Project) produced a practical handbook dealing with practical issues in ABR screening, OAE screening and baby handling. Copies are available from the project manager (WNHP, Princess Anne Hospital, Southampton, SO16 5YA) or through Otodynamics Ltd (36–38 Beaconsfield Road, Hatfield, AL10 8BB).

Summary

The options for neonatal hearing screening for PCHI have been evaluated. First, the extent and impact of PCHI are such that a systematic screening programme **is** warranted and should be a high priority in child health. Second, taking into account equity, responsiveness and efficiency, the preferred and most cost-effective option is to purchase UNS in place of the present heterogeneous system. In addition, a targeted IDT (or other arrangement) to screen children not tested neonatally or for whom there is concern should be purchased to complement the UNS system. Third, such a programme has been shown to be consonant with the principles of screening (Haggard, 1993). Considerable gains from the recommended screening programme may be made if training and quality control are implemented in a standard way across Districts, for example, regionally or (preferably) nationally.

Chapter 10

Research needs

Scope

This review has highlighted the need for further research in a number of areas and these have been mentioned in the text. Some of the research needs are basic, for example, understanding the development of language and communication in hearing-impaired and deaf children, while others are of a more applied nature, for example, what interventions lead to good outcomes. In this chapter, the research needs of an applied nature are synthesised; these flow from the recommendations and will lead to an improvement in screening and subsequent management of hearing-impaired and deaf children and their families in the medium and long term.

Introduction

Since screening is not to be seen in isolation from on-going care and appropriate outcomes *(Figure 1),* the research needs are linked but diverse. They include training, purchasing and information systems as well as screening technology. The ten most pressing needs have been organised here under four headings for convenience, although other schemes are clearly possible. We initially assigned a 3-point priority rating to each topic (the highest priority being indicated by 1). After consideration, all topics with ratings less than 2 have been taken out, in order to focus on the most relevant and pressing needs. The priority banding has been influenced by the extent to which the need is directly related to the issues of early identification of congenital PCHI by neonatal screening. The estimated scale of each project is also indicated (£–£££££, in roughly £75,000 bands). Finally, the details of how these issues might best be addressed have not been spelt out, nor have the questions been broken down into their further constituent questions – some of which will be obvious. This is more properly approached at the commissioning stage; for the moment, what follows is deliberately indicative rather than detailed and prescriptive. Many of the research questions overlap and the answers from each would feed into and inform the others; nevertheless, each may in some sense be regarded as discrete, and able to be prioritised independently of others.

Research priorities

Management of children identified by neonatal screening

Early identification of PCHI is only optimally beneficial if appropriate intervention follows. Under this heading there are two major health technology related research needs.

1. The first step in audiological follow-up is to make an accurate assessment of the child's hearing. There is a need to determine the most effective combination of tests and protocols (electro-physiological, electro-acoustic and behavioural) for the early and accurate determination of frequency-specific auditory thresholds and uncomfortable loudness levels in order to inform early audiological management.
   Priority: 1; Scale: £.
2. The audiological management of hearing-impaired children depends substantially on the degree of hearing impairment and the age at which the child is identified. Neonatal screening gives the greatest potential for such management (in combination with other services) to lead to good outcomes. However, there are several uncertainties that need urgent clarification so that the opportunity is not squandered. Management should be modelled around the interventions that have been shown to give (greatest) benefit. There are three priority areas that would benefit from research using RCTs of management strategies. Greater efficiency might be achieved if the first two areas, which are highly similar, were combined. The third area is different in nature.

   The first RCT covers children with moderate congenital hearing impairments (about 500 per year) who will be detected by the neonatal screens, and those who have a mild impairment (25–39 dB HL; at least 500 per year) who may be detected by the screen. There is no evidence as to whether very early

provision of hearing-aids is appropriate[1] for the large group of children with mild hearing impairments (i.e. about 25–39 dB HL). Indeed, it has been suggested that the distortion introduced by hearing-aids may even be detrimental to the maturing auditory system. In this case, where there is genuine doubt as to the most appropriate intervention, an RCT would be highly appropriate to guide cost-effective management in this difficult area and would, in addition, inform decisions about the case-definition used for neonatal screening. **Priority: 1; Scale: ££££.**

The second RCT covers the early management of those children who have a severe and profound impairment. In these groups the use of appropriate hearing-aids is not questioned as a major component of the management of these children. However, the appropriate first language of deaf children continues to be a controversial issue. There are no studies that look at the longer-term benefits of signing – or other focused early communication packages – because there have been a lack of outcome measures in this area. However, the development of outcome measures for use in the evaluation of paediatric cochlear implant studies (conducted by Summerfield, Davis, Bamford, Bloor and Fortnum) and in investigations of profoundly deaf children (conducted by Davis, Bamford and Gregory) means that a prospective RCT (or controlled trial) of the cost-effectiveness of early intervention packages for this group will be feasible within the next few years. **Priority: 1; Scale: ££££.**

The third RCT concerns the habilitative options for congenitally profoundly deaf children (about 180–200 per year). If neonatal screening is introduced then the majority of profoundly deaf children will be accurately assessed within 3 months and the lack of benefit from hearing-aids, in some cases, will be apparent much earlier than at present. There are two major controversies concerning the early use of cochlear implants, which would be clarified by an RCT.

(i) Would medium-term outcomes concerning the development of language and communication, and quality of life be substantially improved (towards those of normal hearing children) if implantation was at about 1 year of age rather than at 2–3 years of age (currently considered early)?

(ii) Would the benefits of cochlear implantation extend to children close to current eligibility criteria concerning the degree of impairment? **Priority: 2; Scale: £££££.**

Coordination of services

Parents of children with PCHI are highly critical of the coordination within health services and between health and education services. This lack of coordination leads to inefficient systems, that are not responsive to the individual needs of hearing-impaired children and their families, and is inequitable because only articulate or highly persistent parents can get the best out the system. The overall quality of service provision and outcomes for hearing-impaired children is likely to be severely compromised because of this lack of coordination. Three research priorities are identified in this area.

1. Pilot studies elsewhere in the NHS show that where continuing care is prejudiced by lack of coordination, there is the possibility of developing joint commissioning arrangements. In the case of PCHI, there might be the involvement of at least two NHS trusts, education services and Social Services in the provision of care from an early age. There is an urgent need to research the best management model to integrate with UNS, and to examine in more detail the feasibility of joint commissioning arrangements. Of particular interest is the cost of different options and the extent to which earlier appropriate management saves expenditure at a later stage. **Priority: 1; Scale: ££–££££.**
2. The recommendations concerning service development suggest the use of a shared-list (register) to help combat the lack of coordination. The case for a shared (health and education) register of children with PCHI is now very strong, for both service monitoring and longitudinal research purposes. Such a register would provide the means for effectively investigating a number of research questions, some of which have been highlighted here. There is an urgent need, therefore, for research into the minimum dataset for monitoring services, ways in which such information may be integrated into local health information systems

[1] We are not suggesting that early identification is not appropriate for this group, as there will be several benefits in terms of family knowledge and support. It is the form of management, following early identification, that is in question.

and the way in which research information can be added to the dataset at minimum cost. **Priority: 1; Scale: £–££.**

3. The extent to which services are 'family-centred' is likely to determine the effectiveness of early intervention for PCHI. There is a need to investigate the components of parent-perceived 'seamless' services and the ways in which these can be achieved, and the extent to which demographic variables such as race and social factors influence this perception. Such research would help to pinpoint the most effective ways of involving parents and families in the assessment and habilitative processes (e.g. use of hearing-aids, speech and language therapy, parent support services, audiological and otological assessment, clinical genetics and counselling). **Priority: 2; Scale: £–££.**

Screening techniques (including training and dissemination)

1. The effectiveness of neonatal screening and the targeted IDT is highly dependent upon quality of implementation. Protocols and procedures for cost-effective training of the personnel involved need to be developed and trials undertaken. **Priority: 1; Scale: £.**
2. As UNS develops, there is a need for the development of the methods, case definition and cost-effectiveness of later (6–12 months of age) screening systems of targeted IDT. **Priority: 1; Scale: ££.**
3. The efficiency and sensitivity of present neonatal screening techniques is lowest for two groups of children: (i) those who are born in hospital but who are discharged within 12 hours of birth and (ii) those children who are not born in hospital (or who are not screened) and who are called back for a test. It is would be highly desirable to develop fast, sensitive and specific techniques for testing these populations (which may be 10% of the birth cohort). Methods that work well currently in disadvantageous signal/noise ratios such as variations of TEOAEs (e.g. MLS, Quickscreen) and automated ABR devices show some promise in respect of the first group, and the former may prove to be useful for the second group. **Priority: 2; Scale: ££–£££.**

Epidemiology

1. There are doubts about the proportion of PCHI cases that are late-onset or progressive (see chapters 2, 3, 6 and 8). There is a requirement to determine accurately the proportion of such cases in the PCHI population, and to estimate the extent and priority of the needs of this sub-population. In addition, it would be very helpful to determine the most effective means of identifying such cases (e.g. to discover in what proportion there is a genetic marker). **Priority: 1; Scale: ££–£££.**
2. There is evidence (Fortnum *et al*, 1997) that inappropriate surgical intervention for OME in children with PCHI delays optimal intervention, particularly in those children identified by neonatal screening. The magnitude of this problem, how it occurs, and how it may best be avoided, needs further research. **Priority: 2; Scale: £.**

Summary

The research needs have been summarised, and an attempt made to rate the immediate priority and cost of the research. The research needs are prioritised so that they will form the basis for constructing better services giving better screening, habilitation for hearing-impaired children and support for their parents. Some of the research does depend on information systems for hearing screening being in place and on appropriate follow-up information being available. The key role that a national shared-list (or register) of hearing-impaired children would play in making some of the research feasible has been emphasised.

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Ian Wilson designed and implemented the questionnaire databases and their analyses, maintained and cross-referenced the bibliographic database, and took the lead in formatting, editing, proof-reading and then producing the final report and executive summary.

Dr Tina Ramkalawan contributed towards and advised on chapter 3, and was responsible for the final draft of that chapter.

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References

ACSHIP, 1981. Advisory committee on services for hearing impaired children: final report. London: DHSS.

Allen AD, Wallace AM, 1996. Audit of the distraction test of hearing at about 8 months of age: results of a survey of Scottish Health Boards. *Br J Audiol*;**30**:389–96.

Baart de la Faille L, 1991. Validity of large scale standardised behavioural screening. *Acta Otolaryngol Suppl (Stockh)*;**482**:92–101.

Batchava Y, 1993. Antecedents of self esteem in deaf people: a meta-analytic review. *Rehabil Psychol*;**38**:221–34.

Bench R, Bamford JM, 1979. The spoken language of hearing impaired children. London: London Academic Press.

Bennett MJ, 1979. Trials with the auditory response cradle I. Neonatal responses to auditory stimuli. *Br J Audiol*;**13**:125–34.

Bennett MJ, Lawrence R, 1980. Trials with the auditory response cradle II. *Br J Audiol*;**14**:1–6.

Bess FH, Paradise JL, 1994a. Universal screening for infant hearing impairment: not simple, not risk-free, not necessarily beneficial, and not presently justified. *Pediatrics*;**93**:330–4.

Bess FH, Paradise JL, 1994b. Universal screening for infant hearing impairment [Reply]. *Pediatrics*;**94**:959–63.

Bess FH, Tharpe AM, 1986. An introduction to unilateral sensorineural hearing loss in children. *Ear Hear*;**7**:3–13.

Bhattacharya J, Bennett MJ, Tucker S, 1984. Long term follow-up of newborns tested with the auditory response cradle. *Arch Dis Child*;**59**:504–11.

Black IB, 1995. Trophic interactions and brain plasticity. In: Gazzaniga MS, editor. The cognitive neurosciences. 9–16.

Blackman JA, Hein HA, 1985. Iowa's system for screening and tracking high-risk infants. *Am J Dis Child*;**139**:826–31.

Blakemore C, 1978. Maturation and modification in the developing visual system. In: Held R, Leibowitz HW, Teuber HL, editors. Handbook of sensory physiology, volume VIII. Berlin: Springer Verlag: 377–436.

Boothman R, Orr N, 1978. Value of screening for deafness in the first year of life. *Arch Dis Child*;**53**:570–3.

Brown J, Watson E, Alberman E, 1989. Screening infants for hearing loss. *Arch Dis Child*;**64**:1488–95.

Clark C, 1989. Screening for deafness in the health authority: outcomes and effectiveness. Manchester Health Authority.

Clark M, 1989. Language through living. London: Hodder and Stoughton.

Cochrane A, Holland W, 1971. Validation of screening procedures. *Br Med Bull*;**27**:3–8.

Conrad R, 1979. The deaf school child. London: Harper & Row.

Corballis MC, 1991. The lopside ape: evolution of the generative mind. Oxford: Oxford University Press.

Curnock DA, 1993. Identifying hearing impairment in infants and young children. *BMJ*;**307**:1225–6.

Das VK, 1988. Aetiology of bilateral sensori-neural deafness in children. *Scand Audiol Suppl*;**30**:8107–593.

Davidson J, Hyde ML, Alberti PW, 1989. Epidemiologic patterns in childhood hearing loss: a review. *Int J Pediatr Otorhinolaryngol*;**17**:239–66.

Davis A, 1984. Detecting hearing-impairment in neonates – the statistical decision criterion for the auditory response cradle. *Br J Audiol*;**18**:163–8.

Davis AC, 1993a. The prevalence of deafness. In: Ballantyne J, Martin A, Martin M, editors. Deafness. London: Whurr: 1–11.

Davis AC, 1993b. Public health perspective of childhood deafness. In: McCormick B, editor. Paediatric audiology 0–5 years. 2nd edition. London: Whurr: 1–41.

Davis AC, 1995a. Current thoughts on hearing screening. In: Spencer N, editor. Recent advances in community paediatrics. Edinburgh: Churchill-Livingstone.

Davis A, 1995b. The epidemiology of permanent hearing impairment in children: consideration of a cost model for alternative forms of early detection. *Audiens (BACDA Newsletter)*;(17):3.

Davis AC, Parving A, 1994. Towards appropriate epidemiological data on childhood hearing disability: a comparative European study of birth cohorts 1982–88. *J Audiol Med*;**3**:35–47.

Davis AC, Sancho J, 1988. Screening for hearing impairment in children: a review of current practice in the UK with special reference to the screening of babies from special care baby units for severe/profound impairments. In: International perspectives on communication disorders. Washington: Gaulledet University Press.

Davis A, Wood S, 1992. The epidemiology of childhood hearing impairment: factors relevant to planning of services. *Br J Audiol*;**26**:77–90.

Davis AC, Wharrad HJ, Sancho J, Marshall DH, 1991. Early detection of hearing impairment – what role is there for behavioral-methods in the neonatal-period. *Acta Otolaryngol Suppl (Stockh)*;**482**:103–10.

Davis A, Wood S, Healy R, Webb H, Rowe S, 1995. The public health implications of changes in the epidemiology of childhood hearing impairment in Great Britain over the last two decades. *J Am Acad Audiol*;**6**:365–70.

Dias OAM, 1990. Childhood deafness in Portugal: aetiological factors and diagnosis of hearing loss. *Int J Pediatr Otorhinolaryngol*;**18**:247–55.

Downs MP, 1994. The case for detection and intervention at birth. *Semin Hear*;**15**:76–83.

Downs MP, 1995. Universal newborn hearing screening – the Colorado story. *Int J Pediatr Otorhinolaryngol*;**32**:257–9.

Eggermont JJ, 1986. Defining and determining sensitive periods. *Acta Otolaryngol Suppl (Stockh)*;**429**:5–9.

Eilers RE, Oller DK, 1994. Infant vocalizations and the early diagnosis of severe hearing impairment. *J Pediatr*;**124**:199–203.

El-Refaie A, Parker D, Bamford JM, 1996. Otoacoustic emissions versus ABR screening: the effect of external and middle ear abnormalities in a group of SCBU neonates. *Br J Audiol*;**30**:3–8.

Ewing I, 1957. Screening tests and guidance clinics for babies and young children. In: Ewing A, editor. Educational guidance and the deaf child. Manchester: Manchester University Press.

Ewing IR, Ewing AWC, 1944. The ascertainment of deafness in infancy and early childhood. *J Laryngol Otol*; Sept:309–33.

Feinmesser M, Tell L, Levi H, 1982. Follow-up of 40,000 infants screened for hearing defect. *Audiology*;**21**:197–203.

Fisch L, 1981. Development of school screening audiometry. *Br J Audiol*;**15**:87–95.

Fonseca S, Forsyth H, Grigor J, *et al.*, 1996. Identification of hearing impairment in children: monitoring of audiology services. Audit report: unpublished.

Fortnum HM, Davis AC, 1993. Hearing impairment in children after bacterial meningitis: incidence and resource implications. *Br J Audiol*;**27**:43–52.

Fortnum H, Davis A, Butler A, Stevens J, 1997. Health service implications of changes in aetiology and referral patterns of hearing impaired children in Trent 1985–1993. Report to Trent Health. Nottingham/Sheffield: MRC Institute of Hearing Research and Trent Health.

Galambos R, Wilson MJ, Silva PD, 1994. Identifying hearing loss in the intensive care nursery: a 20-year summary. *J Am Acad Audiol*;**5**:151–62.

Galbraith W, 1976. The role of the teacher of the deaf within the NHS [Minority report]. In: ACSHIP. An interim report of the sub-committee appointed to consider services for hearing impaired children. London: DHSS.

Gallaway C, Nunes A, Johnston M, 1994. Spoken language development in hearing impaired children: a bibliography covering research from 1996 – present. Manchester: CAEDSP, University of Manchester.

Gerber SE, 1990. Prevention: the etiology of communicative disorders in children. New York: Prentice Hall.

Greenough WT, Black JE, Wallace CS, 1987. Experience and brain development. *Child Dev*;**58**:539–59.

Gregory S, 1995. Deaf children and their families. Cambridge: Cambridge University Press.

Gregory S, Mogford K, 1981. Early language development in deaf children. In: Wall B, Kyle J, Deucher M, editors. Perspectives on British sign language and deafness. London: Groom Helm.

Gregory S, Bishop J, Sheldon L, 1995. Deaf young people and their families. Cambridge, Cambridge University Press.

Haggard M, 1993. Research in the development of effective services for hearing-impaired people. London: Nuffield Provincial Hospitals Trust.

Haggard MP, Hughes E, 1991. Screening children's hearing. London, HMSO.

Haggard MP, McCormick B, Gannon MM, Spencer H, 1992. The pediatric otologic caseload resulting from improved screening in the 1st year of life. *Clin Otolaryngol*;**17**:34–43.

Hall DMB, editor, 1989. Health for all children: a programme for child health surveillance; the report of the Joint Working Party on Child Health Surveillance. Oxford: Oxford University Press.

Hall DMB, editor, 1992. Health for all children: a programme for child health surveillance; the report of the Joint Working Party on Child Health Surveillance. Oxford: Oxford University Press.

Hall DMB, editor, 1996. Health for all children. The report of the Joint Working Party on Child Health Surveillance. Oxford: Oxford University Press.

Hall DMB, Garner J, 1988. Feasibility of screening all neonates for hearing loss. *Arch Dis Child*;**63**:652–3.

Hall J, Lamb M, Freeman SD, Beeler J, 1996. Infant hearing screening with automated ABR in the NICU vs. WBN. In: Proceedings of XXIII International Congress of Audiology, Bari, Italy.

Hepper PG, Shahidullah BS, 1994. Development of fetal hearing. *Arch Dis Child*;**71**:81–7.

Herrman BS, Thornton AR, Joseph JM, 1995. Automated infant hearing screening using the ABR: development and validation. *Am J Audiol*;**4**:6–14.

Holland WW, Stewart S, 1990. Screening in health care: benefit or bane? London: Nuffield Provincial Hospitals Trust.

Hunter MF, Kimm L, Cafarelli Dees D, Kennedy CR, Thornton ARD, 1994. Feasibility of otoacoustic emission detection followed by ABR as a universal neonatal screening test for hearing impairment. *Br J Audiol*;**28**:47–51.

Hyde ML, Malizia K, Riko K, Alberti P, 1991. Audiometric estimation error with the ABR in high-risk infants. *Acta Otolaryngol (Stockh)*;**111**:212–19.

Hyson RL, Rudy JW, 1987. Ontogenetic change in the analysis of sound frequency in the infant rat. *Dev Psychobiol*;**20**:187–207.

Imai H, 1983. Early fitting of hearing aids and early education for the severely to profoundly hearing-impaired children. *Adv Otorhinolaryngol*;**29**:3065–71.

Irvine R, Rajan R, 1995. Plasticity in the mature auditory system. In: Manley G, Klump G, Koppl C, Fastl C, Oeckinghaus H, editors. Advances in hearing research. Singapore: World Scientific Publishers. In press.

JCIH, 1994. Joint Committee on Infant Hearing position statement. *ASHA*;**36**:38–41.

Johnson A, Ashurst H, 1990. Screening for sensorineural deafness by health visitors (The Steering Committee, Oxford Regional Child Development Project). *Arch Dis Child*;**65**:841–5.

Kei J, McPherson B, Smyth V, Latham S, Loscher J, 1997. Transient evoked otoacoustic emissions in infants: effects of gender, ear asymmetry and activity status. *Audiology*;**36**(2):61–71.

Kemp D, 1978. Stimulated acoustic emissions from within the human auditory system. *J Acoust Soc Am*;**64**:1386.

Kemp D, Ryan S, 1993. The use of transient evoked otoacoustic emissions in neonatal hearing screening programs. *Semin Hear*;**14**:30–45.

Kennedy C, 1996. Wessex trial of universal neonatal screening for hearing impairment. Interim report. Southampton: August 1996.

King AJ, Moore DR, 1991. Plasticity of auditory maps in the brain. *Trends Neurosci*;**14**:31–7.

Kitzinger J, 1995. Introducing focus groups. *BMJ*;**311**:299–302.

Kramer SJ, Vertes DR, Condon M, 1989. Auditory brainstem responses and clinical follow-up of high-risk infants. *Pediatrics*;**83**:385–92.

Kuhl PK, 1994. Learning and representation in speech and language. *Curr Opin Neurobiol*;**4**:812–22.

Kuhl PK, Williams KA, Lacerdo F, Stevens KN, Lindblom B, 1992. Linguistic experience alters phonetic perception in infants by 6 months of age. *Science*;**255**:606–8.

Kurtzberg D, Hilpert PL, Kreutzer JA, Vaughan HG, 1984. Differential maturation of cortical auditory evoked potentials to speech sounds in normal full term and very low birth weight infants. *Dev Med Child Neurol*;**26**:466–75.

Laurenzi C, Monteiro B, 1997. Mental health and deafness – the forgotten specialism? *ENT News*;**6**:22–4.

Lenneberg E, 1967. Biological foundations of language. New York: Academic Press.

Levitt H, McGarr N, Geffner D, 1987. Development of language and communication in hearing impaired children. *ASHA Monogr*;**26**:9–24.

Lippe WR, Rubel Ew, 1983. Development of the Place Principle: tonotopic organization. *Science*;**219**:514–16.

Lutman ME, Davis AC, Fortnum HM, Wood S, 1997. Field sensitivity of targeted neonatal hearing screening by transient-evoked otoacoustic emissions. *Ear Hear*;**18**:265–76.

Markides A, 1986. Age at fitting of hearing aids and speech intelligibility. *Br J Audiol*;**20**:165–7.

Marteau T, 1994. Screening for carriers of cystic fibrosis. Psychological consequences are unclear. *BMJ*;**309**:1429–30.

Marteau TM, Johnston M, Shaw RW, Michie S, 1989. The impact of prenatal screening and diagnostic testing upon the cognitions, emotions and behaviour of pregnant women. *J Psychosom Res*;**33**:7–16.

Marteau TM, Michie S, Johnston M, Kidd J, 1990. The impact of prenatal screening and diagnostic testing upon the cognitions, emotions and behaviour of pregnant women [Reply]. *J Psychosom Res*;**34**:340–1.

Marteau TM, Slack J, Kidd J, Shaw RW, 1992. Presenting a routine screening test in antenatal care: practice observed. *Public Health*;**106**:131–41.

Marteau TM, Kidd J, Michie S, Cook R, Johnston M, Shaw RW, 1993. Anxiety, knowledge and satisfaction in women receiving false positive results. *J Psychosom Obstet Gynaecol*;**14**:185–96.

Martin JAM, Hennebert D, Bentzen O, *et al.,* 1979. Childhood deafness in the European Community. Brussels: Commission of the European Communities.

Martin JA, Bentzen O, Colley JR, *et al.,* 1981. Childhood deafness in the European Community. *Scand Audiol*;**10**:165–74.

Mason SM, 1984. On-line computer scoring of the auditory brainstem response for estimation of hearing threshold. *Audiology*;**23**:277–96.

Mason SM, Davis AC, McCormick B, 1987. Screening hearing in neonates using an automated electric-response audiometry system. *Int J Neurosci*;**34**:3–4.

Mason S, Davis A, Wood S, Farnsworth A, 1997. Field sensitivity of targeted neonatal hearing screening using the Nottingham ABR screener. *Ear Hear*; in press.

Mauk GW, Behrens TR, 1993. Historical, political, and technological context associated with early identification of hearing loss. *Semin Hear*;**14**:1–17.

Maxon AB, White KR, Vohr BR, Behrens TR, 1993. Using transient evoked otoacoustic emissions for neonatal hearing screening. *Br J Audiology*;**27**:149–53.

McClelland RJ, Watson DR, Lawless V, Houston HG, Adams D, 1992. Reliability and effectiveness of screening for hearing-loss in high-risk neonates. *BMJ*;**304**:806–9.

McCormick B, 1983. Hearing screening by health visitors: a critical appraisal of the distraction test. *Health Visit*;**56**:449–51.

McCormick B, 1988a. Behavioural hearing tests 6 months to 5 years. In: McCormick B, editor. Paediatric audiology 0–5 years. London: Taylor & Francis: 97–115.

McCormick B, 1988b. Screening for hearing impairment in young children. London: Croom Helm.

McCormick B, 1990. Commentary on Scanlon and Bamford. *Arch Dis Child*;**65**:484–5.

McCormick B, 1993. Behavioural hearing tests 6 months to 3.6 years. In: McCormick B, editor. Paediatric audiology 0–5 years. 2nd edition. London: Whurr: 102–23.

McCormick B, Curnock DA, Spavins F, 1984a. Auditory screening of special care neonates using the Auditory Response Cradle. *Arch Dis Child*;**59**:1168–72.

McCormick B, Wood SA, Cope Y, Spavins F, 1984b. Analysis of records from an open-access audiology service. *Br J Audiol*;**18**:127–32.

McFarland WH, Simmons FB, Jones FR, 1980. An automated hearing screening technique for newborns. *J Speech Hear Disord*;**45**:495–503.

Meadow-Orlans KP, 1987. An analysis of the effectiveness of early intervention programs for hearing-impaired children. In: Guralnick MJ, Bennett FC, editors. The effectiveness of early intervention for at-risk and handicapped children. Orlando, Florida: Academic Press: 325–62.

Midgley J, 1957. Screening tests of hearing in primary schools. In: Ewing A, editor. Educational guidance and the deaf child. Manchester: Manchester University Press: 107–27.

Moeller M, 1996. Family matters: making sense of complex choices. In: Proceedings of the 4th International Symposium on Childhood Deafness, Kiawah Island, South Carolina.

Moeller M, Osberger M, Eccarius M, 1986. Receptive language skills. In: Osberger M, editor. Language and learning skills. 23.

Mott A, Emond A, 1994. What is the role of the distraction test in hearing. *Arch Dis Child*;**70**:10–13.

Musselman CR, Kircaali-Iftar G, 1996. The development of spoken language in deaf children: explaning the unexplained variance. *J Deaf Stud Deaf Educ*;**1**:108–21.

Musselman CR, Wilson AK, Lindsay PH, 1988. Effects of early intervention on hearing impaired children. *Except Child*;**55**:222–8.

NDCS, 1983. Discovering deafness. A report for National Deaf Children's Week. London: National Deaf Children's Society.

NDCS, 1994. Quality standards in paediatric audiology. Vol I: guidelines for the early identification of hearing impairment. London: National Deaf Children's Society.

NDCS, 1995. Quality standards in paediatric audiology. Occasional papers in the field of early identification of hearing impairment in children. London: National Deaf Children's Society.

NDCS, 1996. Quality standards in paediatric audiology. Vol II: the audiological management of the child with permanent hearing loss. London: National Deaf Children's Society.

Neville HJ, 1991. Neurobiology of cognitive and language processing: effects of early experience. In: Gibson KR, Peterson AC, editors. Brain maturation and cognitive development: comparative and cross-cultural perspectives. Aladine de Gruyter Press: 355–80.

Newport E, 1990. Maturational constraints on language learning. *Cognitive Sci*;(14):11–28.

Newton VE, 1985. Aetiology of bilateral sensorineural hearing loss in young children. *J Laryngol Otol Suppl*;**10**:1–57.

NHS, 1994. Health technology assessment programme: commissioning document. London: NHS Executive.

NHS, 1995. Responsibilities for meeting continuing health care needs. London: NHS Executive, HSG (95)8, LAC (95)5.

NHS, 1996. Priorities and planning guidance for the NHS. London: NHS Executive.

NIH, 1993a. Early identification of hearing impairment in infants and young children. Bethesda, Maryland: National Institutes of Health.

NIH, 1993b. NIH recommends universal screening of infants for hearing impairment. *Am Fam Physician*;**3**:521–2.

Norton SJ, 1994. Emerging role of evoked otoacoustic emissions in neonatal hearing screening. *Am J Otol*;**15** suppl 1:4–12.

O'Hare AE, Grigor J, Cowan D, 1993. Screening and assessment of childhood deafness – experience from a centralised multi-disciplinary service. *Child Care Health Dev*;**19**:239–49.

Oller D, 1991. Effects of hearing impairment on the development of speech. In: Early identification of hearing impairment in infants and young children. Proceedings of NIH Consensus Development Conference. Bethesda, Maryland: NIH.

OPCS, 1996. Office of Public Census and Surveys report 1996. London: OPCS.

Parving A, 1984. Early detection and identification of congenital early acquired hearing disability – who takes the initiative? *Int J Pediatr Otorhinolaryngol*;**7**:107–17.

Parving A, 1992. Intervention and the hearing-impaired child – an evaluation of outcome. *Int J Pediatr Otorhinolaryngol*;**23**:151–9.

Parving A, 1996. Study group in the epidemiology of genetic hearing impairment. *HEAR Infoletter;* (2) November:18–22.

Parving A, Christensen B, 1993. Training and employment in hearing-impaired subjects at 20–35 years of age. *Scand Audiol*;**22**:133–9.

Peckham CS, 1986. Hearing impairment in childhood. *Br Med Bull*;**42**:145–9.

Picton TW, Kellett AJC, Vezsenyi M, Rabinovitch DE, 1993. Otoacoustic emissions recorded at rapid stimulus rates. *Ear Hear*;**14**:299–314.

Plant A, Pick G, 1995. The screening and surveillance of infant hearing in North Staffordshire: clinical audit report. Unpublished audit report.

Pollitt RJ, Green A, McCabe CJ, *et al.*, 1997. Neonatal screening for inborn errors of metabolism: cost, yield and outcome. A review. *Health Technol Assessment*;**1**(7).

Powers S, 1996. Deaf pupils' achievements in ordinary subjects. *J Br Assoc Teachers Deaf*;**20**:111–23.

Prieve BA, Gorga MP, Schmidt A, *et al.*, 1993. Analysis of transient-evoked otoacoustic emissions in normal-hearing and hearing-impaired ears. *J Acoust Soc Am*;**93**:3308–19.

Rajan R, Irvine DRF, Wise LZ, Heil P, 1993. Effect of unilateral partial cochlear lesions in adult cats on the representation of lesioned and unlesioned cochleas in primary auditory-cortex. *J Comp Neurol*;**338**:17–49.

Ramkalawan TW, 1997. Factors that influence the language and communication of hearing impaired children [unpublished PhD thesis]. Nottingham: Institute of Hearing Research, University of Nottingham.

Ramkalawan TW, Davis AC, 1992. The effects of hearing loss and age of intervention on some language metrics in young hearing-impaired children. *Br J Audiol*;**26**:97–107.

Recanzone G, Schreiner C, Merzenich MM, 1993. Plasticity in the frequency representation of primary auditory-cortex following discrimination-training in adult owl monkeys. *J Neurosci*;**13**:87–103.

Robertson C, Aldridge S, Jarman F. Saunders K, Poulakis Z, Oberklaid F, 1995. Late diagnosis of congenital sensorineural hearing impairment: why are detection methods failing? *Arch Dis Child*;**72**:11–15.

Robertson D, Irvine DRF, 1989. Plasticity of frequency organisation in auditory cortex of guinea pigs with partial unilateral deafness. *J Comp Neurol*;**282**:456–71.

Robinette MS, 1994. Universal screening for infant hearing impairment [letter]. *Pediatrics*;**94**:952–4.

Robinshaw HM, 1995. Early intervention for hearing impairment: differences in the timing of communicative and linguistic development. *Br J Audiol*;**29**:315–34.

Robinshaw H, 1996a. The pattern of development from non-communicative behaviour to language by hearing-impaired hearing infants. *Br J Audiol*;**30**:177–98.

Robinshaw H, 1996b. Acquisition of speech pre- and post cochlear implantation: longitudinal case studies of a congenitally deaf infant. *Eur J Disord Commun*;**31**:433–51.

Robinshaw H, Evans R, 1995. Assessing the acquisition of the auditory,communicative and linguistic skills of a congenitally deaf infant pre- and post-cochlear implant-ation. London: British Association of Teachers of the Deaf.

Robinson K, 1983. The scandal of late diagnosis of deafness in children. *Health Visit*;**56**:452–3.

Rowe SJ, 1991. An evaluation of ABR audiometry for the screening and detection of hearing loss in ex-SCBU infants. *Br J Audiol*;**25**:259–74.

Rubel EW, 1978. Ontogeny of structure and function in the vertebrate auditory system. In: Jacobsen M, editor. Handbook of sensory physiology (Development of sensory systems, volume 9). New York: Springer Verlag: 135–237.

Rubel EW, 1985. Strategies and problems for future studies of auditory development. *Acta Otolaryngol Suppl Stockh*;**421**:114–28.

Rubel EW, Lippe WR, Ryals BM, 1984. Development of the Place Principle. *Ann Otol Rhinol Laryngol*;**93**:603–15.

Ruben RJ, Levine R, Baldinger E, *et al.*, 1982. Moderate to severe sensorineural hearing impaired child: analysis of etiology, intervention and outcome. *Laryngoscope*;**92**:38–46.

Rubsamen R, 1992. Postnatal development of auditory frequency maps. *J Comp Physiol*;**170A**:129–43.

Ryals J, Rubel EW, Lippe W, 1992. Issues in neural plasticity as related to cochlear implants in children. *Am J Otol*;**12**:22–7.

Scanlon PE, Bamford JM, 1990. Early identification of hearing loss: screening and surveillance methods. *Arch Dis Child*;**65**:479–84.

Seewald RC, 1992. The desired sensation level method for fitting children: version 3.0. *Hear J*;**45**(4):36–41.

Shepard NT, 1983. Newborn hearing screening using the Linco-Bennett auditory response cradle: a pilot study. *Ear Hear*;**4**:5–10.

Shiu J, Purvis M, Sutton G, 1996. Detection of childhood hearing impairment in the Oxford Region. Report of the Regional audit project. Oxford: Oxfordshire RHA.

Smurzynski J, Jung MD, Lafreniere D, *et al.*, 1993. Distortion-product and click-evoked otoacoustic emissions of preterm and full-term infants. *Ear Hear*;**14**:258–74.

Staples DR, Gravel JS, Martin BA, 1995. Thresholds for auditory brainstem responses to tones in notched noise from infants and young children with normal hearing sensorineural hearing loss. *Ear Hear*;**16**:361–71.

Steel KP, 1995. Inherited hearing defects in mice. *Annu Rev Genet*;**29**:675–701.

Stevens JC, Webb HD, Hutchinson J, Connell J, Smith MF, Buffin JT, 1991. Evaluation of click-evoked oto-acoustic emissions in the newborn. *Br J Audiol*;**25**:11–14.

Stevens JC, Hall DMB, Davis A, Davies CM, Dixon S, 1997. A survey of the costs of hearing screening in the first year of life in England and Wales. *Arch Dis Childhood*; in press.

Stewart-Brown S, Haslum MN, 1987. Screening for hearing loss in childhood: a study of national practice. *BMJ*;**294**:1386–8.

Stewart-Brown S, Haslum MN, Howlett BC, Lyons PJ, Matthews C, 1986a. Screening for hearing loss in the preschool period. Report to DHSS of a survey of health districts in England and Wales in 1985; Pt I. Bristol: University of Bristol, Department of Child Health.

Stewart-Brown S, Haslum MN, Howlett BC, Lyons PJ, Matthews C, 1986b. Screening for hearing loss in schoolchildren in England and Wales in the 1980s. Report to DHSS of a survey of health districts in England and Wales in 1984, Pt II. Bristol: University of Bristol, Department of Child Health.

Summerfield Q, Marshall D, 1995. Cochlear implantation in the UK 1990–94. London: HMSO.

Sutton G, Rowe S, 1997. Risk factors for childhood deafness in the Oxford Region. *Br J Audiol*;**31**:39–54.

Sutton G, Scanlon P, 1996. Health visitor sceening versus vigilance for childhood hearing impairment in West Berkshire – a 10-year review. Unpublished.

Sutton GJ, Stokes J, 1994. Benefit of early fitting of hearing-aids. *J Pediatr*;**125**:844.

Sutton G, Gleadle P, Rowe SJ, 1996. Tympanometry and ocotacoustic emissions in a cohort of special care neonates. *Br J Audiol*;**30**:9–17.

Sweetow RW, Barrager D, 1990. Quality of comprehensive audiological care: a survey of parents of hearing impaired children. *ASHA*;**32**:844–7.

Tait M, 1987. Making and monitoring progress in the preschool years. *J Br Assoc Teachers Deaf*;**11**:143–53.

Thorton ARD, 1993. High rate otoacoustic emissions. *J Soc Audiol*;**94**:132–6.

Thornton ARD, Kimm L, Kennedy CR, Cafarelli Dees D, 1993. External-ear and middle-ear factors affecting evoked otoacoustic emissions in neonates. *Br J Audiol*;**27**:319–27.

Torgerson D, Donaldson C, 1994. Compliance in screening programmes. *BMJ*;**308**:978.

Tsukuhara N, 1981. Synaptic plasticity in the mammalian central nervous system. *Ann Rev Neurosci*;**4**:351–79.

Tucker SM, 1987. Auditory screening of normal and preterm infants using the auditory response cradle. *Audiol Pract*;**2**(4):5–7.

Tucker SM, Bhattacharya J, 1992. Screening of hearing impairment in the newborn using the auditory response cradle. *Arch Dis Child*;**67**:911–19.

Tye-Murray N, Spencer L, Woodworth GG, 1995. Acquisition of speech by children who have prolonged cochlear implant experience. *J Speech Hear Res*;**38**:327–37.

van Zanten BGA, Kok MR, Brocaar MP, Sauer PJJ, 1995. The click-evoked otoacoustic emission, c-oae, in preterm born infants in the post conceptual age range between 30 and 68 weeks. *Int J Pediatr Otorhinolaryngol*;**32** suppl:S187–97.

Varghese CM, 1997. An audit of 8 month hearing screening test in Bolton. *Bolton Med J*;**9**:30–7.

Vernon M, LaFalce-Landers E, 1993. A longitudinal study of intellectually gifted deaf and hard-of-hearing people – educational, psychological, and career outcomes. *Am Ann Deaf*;**138**:427–34.

Wanjohi A, 1996. Background noise in rooms used for infant hearing screening [unpublished MSc thesis]. Manchester: Manchester University.

Watkin PM, 1991. The age of identification of childhood deafness – improvements since the 1970s. *Public Health*;**105**:303–12.

Watkin PM, 1996a. Neonatal otoacoustic emission screening and the identification of deafness. *Arch Dis Child*;**74**:F16–25.

Watkin PM, 1996b. Optimising the specificity of a neonatal otoacoustic emission screen. Unpublished report.

Watkin PM, 1996c. Outcomes of neonatal screening for hearing loss by otoacoustic emissions. *Arch Dis Child*;**75**:F158–68.

Watkin PM, Baldwin M, Laoide S, 1990. Parental suspicion and identification of hearing impairment. *Arch Dis Child*;**65**:846–50.

Watkin PM, Baldwin M, McEnery G, 1991. Neonatal at risk screening and the identification of deafness. *Arch Dis Child*;**66**:1130–5.

Watkin PM, Beckman A, Baldwin M, 1995. The views of parents of hearing-impaired children on the need for neonatal hearing screening. *Br J Audiol*;**29**:259–62.

Watkin P, Baldwin M, Dixon R, 1997. Parental anxiety and attitudes to universal neonatal screening. In press.

Webster DB, 1983. Auditory neuronal sizes after a unilateral conductive hearing loss. *Exp Neurol*;**79**:130–40.

Weinberger NM, 1993. Learning-induced changes of auditory receptive fields. *Curr Opin Neurobiol*;**3**:570–7.

Weisel TN, Hubel H, 1963. Single cell response in striate cortex of kittens deprived of vision in one eye. *J Neurophysiol*;**26**:1003–17.

Westwood GFS, Bamford JM, 1995. Probe-tube microphone measures with very young infants – real ear to coupler differences and longitudinal changes in real ear unaided response. *Ear Hear*;**16**:263–73.

Whetnall E, 1955. Discussion on the management of deafness in the young child. *Proc R Soc Med*;**49**:455.

White KR, Maxon AB, 1995. Universal screening for infant hearing impairment – simple, beneficial and presently justified. *Int J Pediatr Otorhinolaryngol*;**32**:201–11.

Wilson JMG, Jungner G, 1968. Principles and practice of screening for disease. Geneva: World Health Organization.

Wood S, Farnsworth A, Davis A, 1995. The identification and referral of babies with a family history of congenital hearing loss for hearing screening. *J Audiol Med*;**4**:25–33.

Wood S, Davis A, McCormick B, 1997. Changing yield of the health visitor distraction test (HVDT) when targeted neonatal screening is introduced into a health district. *Br J Audiol*; in press.

Yoshinaga-Itano C, Sedey A, Apuzzo M, Carey A, Day D, Coulter D, 1996. The effect of early identification on the development of deaf and hard-of-hearing infants and toddlers. In press.

Appendix 1

Methodologies: literature review and Focus Groups

Literature review

There is a noticeable lack of literature/guidelines on systematic reviews of studies other than RCTs. Although the literature on neonatal hearing screening tends not contain many reports of RCTs (such studies have rarely been performed for ethical reasons), it is still possible to review the literature on the basis of a scheme which is as explicit, bias-free and structured as possible, thus facilitating critical peer review.

To begin, the expert members of the review team generated a list of potential keywords. This was added to later as new keywords occurred to the members of the team and also when the researcher responsible for the literature search began the trawl and was able to 'snowball' out into the literature from the reference lists of key papers.

Using these keywords, a large number of bibliographic references were from derived from a coverage-overlapping set of on-line databases (BIDS, Embase, ISSI and ISI, Eric, PsychLit, Medline, CINAHL, the British Library catalogue and thesis listings) and a search of Internet/World-Wide Web URLs. These were added to from other sources, such as bibliographies on paediatric audiology which were well known in the field and readily available from their creators.

A first pass through the file was undertaken by the researcher responsible for the literature review to eliminate obviously irrelevant articles which had been selected by the computer searches because they contained a chosen keyword. For instance, a Boolean search for the keywords 'hearing' and 'aids' will produce papers on the subject of hearing problems reported by sufferers of AIDS.

The next step was for two of the senior expert members of the research team to look through the titles and abstracts of the remaining papers, indicating which were to be kept for review. Both experts reviewed the same papers, and the criterion for inclusion at this stage was if **either** had requested a paper to be retained; hence, only those papers deemed irrelevant by **both** experts were excluded.

This left a set of required references and a set to be discarded. However, to minimise the possibility of discarding relevant studies, one of the senior members of the team made an additional pass through the items to be discarded to see if anything had been missed. This pass **did** reveal some papers which might not otherwise have been included in the review.

A 3-point rating system was devised so that papers could be further assessed in terms of their relevance to the review. The scale was thus: 1, essential reading, highly relevant; 2, not essential but relevant – include if time permits; 3, do not read, not relevant. People with expertise in particular fields, such as the epidemiology of deafness or otoacoustic emissions, were identified and approached to act as assessors in this respect. (Some of this was done by senior members of the research team, since they are acknowledged experts in a number of the appropriate areas.) They were briefed on the nature of the project, given details of the 3-point scale and asked to rate each title/abstract according to the scale. It should be noted that at this stage no judgement was made as to the quality of papers, only their relevance to the review of infant hearing screening. Raters were informed that ratings should be made strictly on salience, ignoring quality completely.

The final set of papers derived from this step-wise procedure was added to at intervals in a number of ways; on-going literature searches to keep up with papers being published as the study progressed; information and pre-publication papers sent to us by interested professionals; single items which had been missed by the search but which were found once the review process proper had begun. Literature trawling is, and should be, a recursive process, since the results of searches are constantly used to direct further searches. For this review, this continued throughout the time available for the trawl.

The next step was for the papers to be read. An attempt was made to make this process as systematic and explicit as possible. A checklist was created in the same mould as those sent by academic journals to reviewers. This checklist was then completed for each paper read, and a summary of the paper

and its conclusions was appended to this. This summary formed the basis for the annotations in our annotated bibliography.

It must be stated that this process suffered from the unavoidable flaw that senior members of the team were already familiar with a great many of the papers and so were forced to make ratings which could have been biased because of prior experience. However, all such studies suffer in the same way. It is impossible to expect an entirely disinterested person to read a hundred papers on, for example, the ABR because (i) they would not have the relevant knowledge-base to interpret the papers and (ii) if they are **dis**interested they are likely to be **un**interested and so unavailable.

Focus Groups

There is a great deal of information that cannot be derived from the literature on any subject, simply because there are opinions and attitudes that are not expressed formally, ways and means of achieving ends in practical settings which have never been formally published, and also data which are either not suitable for publication or fail to be published because of either financial or political constraints. In order to sample these, both a survey of current practice and a set of national Focus Group meetings were organised.

Methodology of Focus Groups

The purpose of the Focus Group meetings was to bring together professionals involved with children's hearing screening services (both providers and purchasers), together with parents of hearing-impaired children and relevant voluntary organisations to obtain their views on a range of issues relating to existing services for hearing-impaired children and their ideas for the future development of these services. These were planned with particular reference to the role of neonatal screening in the early detection of children with congenital hearing impairments. The issues to be discussed in the groups were:

(i) levels of satisfaction with existing children's hearing screening services
(ii) the importance of early identification of hearing-impaired children
(iii) views on neonatal screening including UNS
(iv) the importance of the education services and other support services in early identification of hearing-impaired children
(v) the value and priority given to the different screens
(vi) thoughts on the future development of these services over the next 5–10 years.

Participants were also asked to comment on the contribution of the NDCS publication, *Quality Standards in Paediatric Audiology,* volume 1, to the development of services and give their views on the value of a Register for hearing-impaired children.

The Groups and their composition

Five Focus Groups were organised in different cities around the country from June to December 1995. The intention was to bring together people from the different geographical areas with experience of very disparate services, for example, from inner city London to rural areas in South-West England and parts of Scotland. Meetings were organised in Nottingham, Glasgow, Manchester, London and Bath. A central venue was booked in each city for 1 day from 0930–1600 hours to cater for between ten and 30 people.

The research team drew up a list of names of people involved in children's hearing screening services and possible contacts for obtaining names of parents of hearing-impaired children and voluntary organisations for four of the five Focus Groups. The Glasgow list was produced by a Senior Registrar of the Grampian Health Board. The team attempted to invite a wide range of professionals involved in children's hearing screening services although no attempt was made to invite people as representative samples of professionals or parents from the country as a whole. Each Group had a different composition of professionals, parents and voluntary group representatives.

A total of 171 people were invited to the five Groups; the numbers and composition of those who attended each Focus Group are given in *Table 25,* details of the people that were invited are given in *Table 26,* and overall participation is summarised in *Table 27.* Fewer educationalists, speech therapists, parents, voluntary organisation representatives and purchasers were invited than practitioners of audiology and medicine. Attempts were made to rectify this; for the London Focus Group, the research team worked closely with the NDCS in London to increase the numbers of parents of hearing-impaired children. Similarly, for the last Focus Group in Bath, names of purchasers in the South-West were taken from the database of names used for part 1 of the survey and invited to this meeting. Purchasers were less likely to attend than any other major group; in particular, there was a statistically significant difference between the purchasers and the audiologists,

TABLE 25 *Summary of Focus Group participants, 1995*

Position	Nottingham	Glasgow	Manchester	London	Bath
Health Visitor Manager		1			
Health Visitor		2			
Specialist Health Visitor, Audiology			1		
Community Nurse Manager			1		
Clinical Nurse Specialist for Child Protection/Child Health					1
TOTAL for sub-group = 6					
SCMO	2	2	2		
SCMO, Audiology		2	1	1	2
SCMO Audiological Medicine			1		
CMO		1		1	2
Clinical Scientist	1				
Associate Specialist	1				
Medical Officer, Audiology				1	
TOTAL for sub-group = 20					
Community Paediatrician		1		2	
Consultant Paediatrician	1				
Consultant Community Paediatrician	1	1	3		1
Paediatrician, Audiology			1		
Senior Registrar		1			
Senior Registrar, Community Child Health		1			
Senior Registrar to Audiological Physician	1				
TOTAL for sub-group = 14					
Head of Audiology				1	
Chief Audiologist		1			
Senior Chief Audiologist		1	1		
District Audiological Services Manager					1
Area Audiological Services Manager		1			
Audiological Scientist	1	2	3		1
Paediatric Audiological Scientist				1	
Consultant Audiological Scientist	1		1	2	
Consultant Audiologist				2	
Consultant Audiological Physician				2	
Consultant in Audiological Medicine				1	
Medical Technical Officer	1				
Chief Technician			1		
TOTAL for sub-group = 25					
Consultant ENT Surgeon/ENT Surgeon		2	2		
TOTAL for sub-group = 4					
Speech Therapist				1	
Speech and Language Therapist				1	
TOTAL for sub-group = 2					
Senior Peripatetic ToD				1	
Teacher of the Hearing Impaired	1				
Head of Educational Services for the Hearing Impaired	1		3	1	
Deputy Head Teacher			1		
Senior Educational Audiologist	1				
Educational Audiologist		3			2
TOTAL for sub-group = 14					

continued

TABLE 25 contd *Summary of Focus Group participants, 1995*

Position	Nottingham	Glasgow	Manchester	London	Bath
Hearing Screening Tester				1	
TOTAL for sub-group = 1					
Nuffield Support Services Coordinator				1	
TOTAL for sub-group = 1					
NDCS, Chief Executive				1	
DCS Regional Representative			1*	3*	1*
NDCS, Management Committee Member				1*	
TOTAL for sub-group = 7					
Parents			9 and (1*)	1 and (4*)	(1*)
TOTAL for sub-group = 10 + (6*)					
Director of Public Health				1	
Healthcare Programme Manager					1
Consultant in Public Health Medicine		2			
Registrar in Public Health			1		
TOTAL for sub-group = 5					
Focus Group totals	13	23	33	27	12
Total number of people attending =108					

* *NDCS Regional Representatives were also there as parents*

medical professions, parents and educationalists (the sessions were held on a weekday, and needed at least 4–5 hours attendance).

Organisation of Focus Groups

The organisation and format were very similar for each Focus Group. The day was organised primarily to facilitate discussion among participants. However, two short presentations were planned at the start of the meeting to give everyone some background information to the project and to outline the rationale for these groups. Two workshop sessions were organised in which participants would not get together in smaller groups to share their views, opinions and experiences on a range of issues relating to children's hearing screening services. It was emphasised in both the first letter of invitation and in the information pack sent to participants that the day had been organised specifically to facilitate discussion.

Each person on the invitation list was sent a letter inviting them to attend the Focus Group together with further information on the purpose of the Focus Group (see *Table 28*), the project, a draft agenda and a list of people who had been invited to that Focus Group meeting. If they were unable to attend they were asked to nominate someone in their place. If they did confirm their attendance they were sent more information at least 2 weeks prior to the meeting. In addition to administrative material, this included an agenda, a list of participants, further information about the format of the day and the questions they would be asked to consider in the small workshop groups, together with background reading material (see *Tables 29* and *30* for details).

Two workshop sessions were organised, each followed by a plenary session where all groups would come back together for the report back and general discussion.

Before each meeting, the names of those attending were organised into two groups for the workshop sessions, each with a similar composition of participants. The Manchester Focus Group had three groups because of the larger number of people attending this meeting. One of these groups was organised specifically for parents and voluntary organisation representatives. It was thought, given the large numbers of parents and representatives attending this meeting, they would feel more at ease talking about their experiences and sharing their views about children's hearing screening services without professionals for these services being present.

On the day all participants were given name badges and a summary of the questions listed in the information pack, the agenda, the list of names for the two or three groups for the workshop sessions and brief notes about the rationale of the Focus Group meetings. Following a general introduction

and welcome, each participant was asked to briefly introduce themselves. A member of the research team gave a brief review of the project and a second an overview of the purpose of the meeting and the aims of the workshop sessions. After the presentations, participants joined their smaller workshop groups, each of which had a member of the team as facilitator and another as notetaker. A participant from each group was asked to record the main points of the discussion on a flipchart and to report these back to the full group during the plenary session. Facilitators worked through the questions listed on the handout and encouraged all participants to contribute. The day ended with a general discussion and summary from a member of the research team.

The record of the Focus Group discussion

Detailed notes were made of the discussions in both workshop and plenary sessions. A summary of the discussion was produced for each Focus Group and a record was kept of the main points made by each group. These were used to produce an overall summary of the main points of discussion for all five Focus Groups. This summary was sent to all the people who participated in the groups and is discussed in chapter 6.

TABLE 26 *Summary of those invited to Focus Group meetings, 1995*

Position	Nottingham	Glasgow	Manchester	London	Bath
Health Visitor Manager		1			1
Health Visitor		2			
Specialist Health Visitor, Audiology			1		
Community Nurse Manager			1		
Lead School Nurse					1
Acting Community Services Manager					1
Senior Nurse Manager					1
Senior Nurse/Health Visitor					1
Community Directorate Manager					1
Nursing Officer					1
TOTAL for sub-group = 12					
SCMO	4	2	3		1
SCMO, Audiology		2	2	1	3
SCMO Audiological Medicine			1		
CMO		1		1	
Clinical Scientist	1				
Associate Specialist	1				
Medical Officer, Audiology				1	
GP			2		
TOTAL for sub-group = 26					
Community Paediatrician		1		2	
Consultant Paediatrician	3				1
Consultant CommunityPaediatrician	1	1	4		1
Senior Registrar		1			
Senior Registrar in Community Child Health		1			
Consultant Paediatrician and Neonatalogist			1		
Consultant Paediatric Neurologist					1
TOTAL for sub-group = 18					
Scientist, Institute of Hearing Research					1
TOTAL for sub-group = 1					
Research Fellow					1
TOTAL for sub-group = 1					
Head of Audiology	2			2	
Chief Audiologist		1			
Senior Chief Audiologist		1			
District Audiological Services Manager					1
Area Audiological Services Manager		1			
Audiological Scientist	1	1	3		1
Paediatric Audiological Scientist				1	

continued

TABLE 26 contd *Summary of those invited to Focus Group meetings, 1995*

Position	Nottingham	Glasgow	Manchester	London	Bath
contd					
Consultant Audiological Scientist	1		1	2	
Consultant Audiologist				3	
Consultant Audiological Physician	1			2	
Consultant in Audiological Medicine				1	
Medical Technical Officer	1				
Chief Technician			1		
Principal Audiological Scientist	1			1	
Paediatric Audiologist			1		
Senior Audiological Scientist					1
TOTAL for sub-group = 32					
Consultant ENT Surgeon/ENT Surgeon	3	2	2		1
TOTAL for sub-group = 8					
Hearing Screening Tester				1	
TOTAL for sub-group = 1					
Speech Therapist				1	
Speech and Language Therapist				1	
TOTAL for sub-group = 2					
Senior Peripetetic ToD					1
Teacher of the Hearing Impaired	1				
ToD, Cochlear Implant Programme	1				
Deputy Head of Service			1	1	
Senior Educational Audiologist	1				
Educational Audiologist	1	3			3
Team Leader, Sensory Impaired Services	1				
Head of Hearing Impaired Services/for Children	1		4	2	3
Peripetetic ToD/Educational Audiologist			1		
Hearing Services Manager			1		
Head Teacher					1
TOTAL for sub-group = 27					
Coordinator, Nottingham Paediatric Cochlear Implant Programme	1				
TOTAL for sub-group = 1					
Social Worker for Hearing-Impaired Children				1	
TOTAL for sub-group = 1					
Nuffield Support Services Coordinator				1	
TOTAL for sub-group = 1					
NDCS, Chief Executive				1	
DCS Regional Representative			1*	7*	1*
NDCS, Management Committee Member				1*	
NDCS, Vice-Chairperson			1*		
TOTAL for sub-group = 12					
Parents			6 and (2*)	2 and (8*)	(1*)
TOTAL for sub-group = 9 + (11*)					
Director of Public Health				1	8
Consultant in Public Health/Medicine		2	1		2
Registrar in Public Health/Medicine		1	1		
Senior Registrar in Public Health Medicine	1		1		
Director of Borough Focusing Commission				1	
TOTAL for sub-group = 19					
Focus Group totals	28	24	41	38	40
Total number of people invited = 171					

* *DCS Regional Representatives were also invited as parents*

TABLE 27 *Summary of the numbers of people invited to, and attending, all Focus Groups by position*

Position	Invited	Attended
Health Visitor/Nurse	12	6
Medical Professions	44	34
Audiologists	32	25
ENT Surgeons	8	4
Speech Therapists	2	2
Educationalists	27	14
Social Workers	1	0
NDCS/Parents	20	16
Purchasers	19	5
Related Groups	5	2

TABLE 28 *Purpose and role of Focus Groups and workgroups*

Why have discussion/Focus Group activity in this project?

- To make research and development more relevant to health service provision.
- To ask people their views on the project.
- To talk directly with 'consumers' of health care.

Role of workgroups

1. To explore:
- views and opinions
- beliefs
- values
- priorities

in relation to paediatric audiology services and, in particular, whether the role of neonatal screening should be expanded.

2. To discuss how the results of research should be disseminated to decision-makers, professionals and consumers.

TABLE 29 *Questions for consideration in workshop groups*

How satisfied are you with your existing screening services for permanent childhood hearing impairment in your District and for the country as a whole?
Satisfaction **0** (very dissatisfied) – **10** (very satisfied):
- with service in own District
- with service in the country as a whole
- why?

Views on neonatal screening
What views do you have on universal neonatal screening?
- What are your views on the NIH consensus statement?
- Do your future plans include the possibility of introducing universal neonatal screening?

What importance and priority would you give to early identification – given your experience and knowledge? How can 'importance' be explored further?
- What are the benefits of early detection?
- How much would you pay – overall – per child?
- How could such a service be funded? For example, what could be cut to fund such a programme?

What importance do you attach to identifying the following 'types' of hearing-impaired children in the first year of life?
- Unilateral (all severities)
- Mild bilateral (< 40 dB)
- Moderate bilateral (40–69 dB)
- Severe bilateral
- Profound bilateral

Should unilateral losses, or those for children with other disabilities, be treated differently?

How important is educational provision following early identification?
- What sort of educational provision is needed?
- What sort of provision is presently provided?
- How is it presently funded?

What improvements would you make to current paediatric audiology services and with what priority?
- What are the most important aspects of the service that you would like to change?
- What support would you need to make these changes?
- How do we go about changing the service?

Value and priority for screens
- universal neonatal screening
- targeted neonatal screening
- health visitor distraction testing
- school-entry screening

- Which needs to be improved most?
- How well coordinated are they?
- Would you abandon any of these?
- Can they be improved?
- What research priorities do you have?

Foresight – plans for the future
In 5 years and in 10 years time:
- what changes do you foresee in screening services?
- in your District?
- in the UK?

Is there a need for a register?
- Information system to underpin the raft of screening and surveillance?
- How do we implement it?
  – by District?
  – by Region?
  – by country?

What do you think of the NDCS guidelines, *Quality standards in paediatric audiology*, vol. 1?
- Are the targets attainable?
- Have they influenced your service in any way?

TABLE 30 *Supporting information for Focus Group meetings*

The following journal articles, booklets and other papers related to the subject of children's hearing screening services were sent to those attending Focus Groups before the meeting.

1. Davis A. Current thoughts on hearing screening services. In: Spencer NJ, editor. Progress in community child health 1. London: Churchill Livingstone, 1995 (see, in particular, *'Recommendations...',* pages 17 and 18).
2. Curnock DA. Identifying hearing impairment in infants and younr children. *BMJ* 1993;**307**:1225–6.
3. Bess FH, Paradise JL. Universal screening for infant hearing impairment: not simple, not risk free, not necessarily beneficial and not presently justified. *Pediatrics* 1994;**93**:330–4.
4. White KR and Maxon AB. Universal screening for infant hearing impairment: simple, beneficial, and presently justified. *Int J Pediatr Otorhinolaryngol* 1995;**32**:1–11.
5. National Institutes of Health. Early identification of hearing impairment in infants and young children. NIH consensus statement, vol. 11 (no. 1, March 1–3). Bethesda, Maryland: NIH, 1993.
6. National Deaf Children's Society. Quality standards in paediatric audiology, vol. 1. London: NDCS, 1993: 1–2. (Only pages 1–2 included; page 2 covers the NDCS targets.)
7. National Deaf Children's Society. Quality standards in paediatric audiology, occasional papers in the field of early identification of hearing impairment in children. London: NDCS, 1995.

Appendix 2

Survey of current practice

Contents

Directors of Public Health

Letter

Questionnaire

A Critical Review of the Role of Neonatal Screening in the Detection of Congenital Hearing Impairment

Dr A Davis
Dr H Fortnum
Dr S Wright
Mrs R Dobbins
(Nottingham)

Prof J Bamford
Dr M J Forshaw
(Manchester)

MRC
Medical Research Council
Institute of Hearing Research
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THE UNIVERSITY of MANCHESTER
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<date>

<name>
<address1>
<address2>
<address3>
<address4>
<address5>

Dear <name>,

Critical review of the role of neonatal screening in the early detection of congenital hearing impairments.

Questionnaires to Co-ordinators of Children's Hearing Screening Services.

I am working with Dr Adrian Davis and Professor John Bamford on the above project. As part of the project we are undertaking a survey of District Health Authorities to find out about their current children's hearing screening services. A sheet briefly describing the project is enclosed.

As part of this study, I am sending out questionnaires to coordinators of a) the neonatal screening services and b) the Health Visitor Distraction Test and the School Entry Screening Service in each District. I would be grateful if you would send out for me these questionnaires to the appropriate people in your District. I have enclosed 4 neonatal screening questionnaires and 4 HVDT and School Entry Screening questionnaires.

Please contact me on 0115 967 8743 if you have any queries or if you require more questionnaires.

Thank you very much for your help.

Yours sincerely,

Dr Sue Wright.

Critical Review of the Role of Neonatal Screening in the Early Detection of Congenital Hearing Impairments

CONTACT QUESTIONNAIRE

Name:_____ **District Health Authority:** _____

Demographic Information

What was the number of live births in your District for 1994?

Please write in:

What percentage of live births in 1994 were at Hospital Maternity Units?

Please write in:

How many Maternity Units are there in the District?

Please write in:

How many Special Care Baby Units (SCBU's) are there in the District?

Please write in:

How many children were discharged from SCBU's in your District in 1994?

Please write in:

Hearing Screening Services in your District

Within your District which of the following hearing screening services are currently in place?

(Please tick all those which apply)

Neonatal - universal (i.e. carried out on all neonates) ☐

Neonatal - targeted (i.e. on "at-risk" neonates) ☐

Neonatal - other ☐

Health Visitor distraction test, 6 - 9 months ☐

Health Visitor surveillance, 0 - 12 months ☐

Intermediate screens, 1 - 5 years ☐

Surveillance, 1 - 5 years ☐

School screen ☐

Do you have an Audiology Working Party (or equivalent) in your District?

Yes ☐ **No** ☐

If YES, please give a contact name, address and telephone number.

Name: _____

Address: _____

_____

_____

Telephone: _____

Do you purchase any hearing screening services from providers outside your District?

Yes ☐ **No** ☐

If YES, please give brief details of what you purchase and from where.

_____

_____

_____

Names and Addresses of Coordinators for the Services

Who is the coordinator (or equivalent) for each of the hearing screening services listed below whom we could contact to get further information?

Please write details below where appropriate:

Neonatal	**Name:** _____ **Position:** _____ **Address:** _____ _____ _____ _____ **Postcode:** _____ **Telephone:** _____
Health Visitor distraction test/surveillance 0 - 12 months	**Name:** _____ **Position:** _____ **Address:** _____ _____ _____ _____ **Postcode:** _____ **Telephone:** _____
Intermediate screens/ surveillance 1 - 5 years	**Name:** _____ **Position:** _____ **Address:** _____ _____ _____ _____ **Postcode:** _____ **Telephone:** _____
School screen	**Name:** _____ **Position:** _____ **Address:** _____ _____ _____ _____ **Postcode:** _____ **Telephone:** _____

Audit Reports and Costs

We are interested in whether you have an audit report on any of the following hearing screening services and if these could be made available to us. We would also like to know if you have costed these screening services.

(Please tick all which apply)	Is there an audit report available?	Could the report be made available to us?	Do you have the contracted cost of the service?
Neonatal - universal	☐	☐	☐
Neonatal - targeted	☐	☐	☐
Neonatal - other	☐	☐	☐
Health Visitor distraction test 6 - 9 months	☐	☐	☐
Health Visitor surveillance 0 - 12 months	☐	☐	☐
Intermediate screens 1 - 5 years	☐	☐	☐
Surveillance 1 - 5 years	☐	☐	☐
School screen	☐	☐	☐

Your Comments

If you have any further comments please continue on another sheet.

THANK YOU FOR COMPLETING THE QUESTIONNAIRE
PLEASE RETURN IT IN THE REPLY-PAID ENVELOPE

Coordinators of Neonatal Screening Services

Letter

Questionnaire

Tabulated data from the Questionnaire

A Critical Review of the Role of Neonatal Screening in the Detection of Congenital Hearing Impairment

Dr A Davis
Dr H Fortnum
Dr S Wright
Mrs R Dobbins
(Nottingham)

Prof J Bamford
Dr M J Forshaw
(Manchester)

MRC
Medical Research Council
Institute of Hearing Research
University Park
Nottingham NG7 2RD
Telephone (direct): 0115 967 8743/7905
Telephone (IHR): 0115 922 3431
Fax: 0115 942 3710
Email: sue.wright@ihr.mrc.ac.uk

THE UNIVERSITY of MANCHESTER
School of Education
Centre for Audiology, Education of the Deaf & Speech Pathology
The University of Manchester
Oxford Road
Manchester M13 9PL
Telephone: 0161 275 3373
Fax: 0161 275 3519
Email: mark.forshaw@man.ac.uk

<date>

Ref. No.: «DScode» / «Pcode»

«Ptitle» «Pname1» «Pname2»
«Pposition»
«Paddress1»
«Paddress2»
«Paddress3»
«Paddress4»
«Paddress5»
«Ppostcode»

Dear «Ptitle» «Pname1» «Pname2»,

Critical review of the role of neonatal screening in the early detection of congenital hearing impairments.

Neonatal Screening Questionnaire

As part of the above review a questionnaire has been circulated to Directors of Public Health to find out which childhood hearing screening services are currently being provided in each of the Districts (or equivalent) in the UK. A sheet that briefly describes the aims of the project is enclosed.

This questionnaire has now been returned and your name has been given to us by the Director of Public Health as the appropriate person to contact for information about the neonatal screening services in the District. We would be very pleased if you could spend the time to complete the enclosed questionnaire which will provide the vital information we need to give an accurate account to the Department of Health about childhood hearing screening services in the UK, and the possibilities for developing these services.

If you do not screen "at-risk" children but conduct a full assessment (e.g. Brainstem Response Audiometry) we would still like you to complete the questionnaire and treat the word "screen" as "assessment", annotating the questionnaire accordingly. If you are not the appropriate person please pass the form on to someone who could complete it. If you have any difficulties and/or queries please contact Dr Sue Wright on 0115 967 8743.

We have to submit our report to the Department of Health in early 1996. So in order for us to analyse the replies and discuss our conclusions we need the questionnaires returned as soon as is possible in the reply-paid envelope.

The time and effort that you and your staff put into completing this questionnaire is greatly appreciated.

Yours sincerely,

Dr Adrian Davis and Dr Sue Wright.

Encl.

Critical Review of the Role of Neonatal Screening in the Early Detection of Congenital Hearing Impairments

NEONATAL SCREENING QUESTIONNAIRE
Preliminary Statistics
(all figures are out of a total of 128)

This questionnaire is part of the above review funded by the Department of Health and undertaken by a team based at the MRC Institute of Hearing Research, Nottingham and the Centre for Audiology, Education of the Deaf and Speech Pathology, University of Manchester. The questionnaire is designed to gather information about current childhood hearing screening services in District Health Authorities (or equivalent) in the UK. Your name has been given to us by the Director of Public Health as the person who co-ordinates the neonatal hearing screening services in the District.

Questions in Part A are concerned with the structure of the neonatal hearing screening service. Part B covers the management of the service. Part C is concerned with the performance of the screen(s). Questions in Part D are designed to gather information about the follow-up services.

THANK YOU FOR YOUR HELP

Name: _____

Position: _____

Trust: _____
(if applicable)

District Health Authority/Health Board or equivalent:

_____

Address: _____

_____

_____

_____

_____

Postcode: _____

PART A: STRUCTURE AND ORGANISATION OF NEONATAL HEARING SCREENING SERVICES

NEONATAL SCREENING SERVICE

Which population of neonates do you routinely screen in the District and in what year did the service start?

(Please tick all those which apply and write in where applicable)		Year started
Universal screening - all neonates	2	85, 92,
Targeted - neonates in SCBUs and/or NICUs	84 (65)	see table 1
Targeted - neonates with a family history of hearing impairment	86 (64)	see table 2
Targeted - neonates with cranio-facial abnormalities	83 (63)	see table 3
Non-routine screening e.g. children referred by other professionals	69 (55)	see table 4
Other	19 (18)	see table 5

If other, please specify

_____

_____

_____

Are you the person responsible for co-ordinating the neonatal screening service?

Yes 106 No 20

If no, please could you give the name and tel. no. of the person who is? _____

_____

SCREENING TECHNIQUES AND EQUIPMENT

What techniques and equipment do you use for the neonatal screening service?

(Please tick all those which apply)		If ticked, please give details of equipment
Otoacoustic Emissions - Transient Evoked (TEOAE)	68 (51)	see table 6
Otoacoustic Emissions - Distortion Product (DPOAE)	1	_____
Behavioural Methods eg. Auditory Response Cradle (ARC)	7 (7)	see table 7
Brainstem Response Audiometry (BRA)	79 (66)	see table 8

If using BRA, what screening level is used see table 9

Other	4	_____

If other, please specify _____

_____

LOCATION OF NEONATAL SCREENING SERVICES

Where is the neonatal screening service routinely located in the District (or equivalent) before and after discharge?

(Please tick all those which apply)	Before discharge	After discharge
Hospital Maternity Unit	23	6
Hospital Outpatients Department	25	67
Neonatal Intensive Care Unit	54	16
Other e.g. at home	5	25

If other, please specify 56 had only 1 place, 23 had 2

Does the test take place:
(Please tick all those which apply)

At the bedside	24 (24)	In a separate sound proofed room	41 (37)
In a separate room	44 (36)	In a separate room adapted specially for screening	15 (15)
Other	8 (8)		

If other, please specify _____

PREMATURE BABIES

Is there a policy on when to test premature babies? Yes 85 (68) No 23 (22) (20 blank)

If yes, please specify _____

PARENTAL INVOLVEMENT

(Please tick all those which apply)	Yes	No	
<u>Prior to the screen</u>			
Is parental consent sought?	76 (63)	27 (25)	(25 blank)
Are parents given any written information about the screening procedures and the service?	60 (53)	42 (37)	(26 blank)
<u>On the occasion of the screen</u>			
Are parents routinely encouraged to be present whilst their child is being tested?	89 (70)	15 (15)	(24 blank)
<u>After the screen</u>			
Are parents given any materials e.g. "Hints for Parents"?	60 (47)	39 (37)	(29 blank)

PART A continued

RECORDS

How are the results of the neonatal screening service recorded?

(Please tick all those which apply)

On a parent held record	**42 (35)**	
On paper records in the Hospital or Clinic	**98 (75)**	
On the Child Health Computer Database	**9 (9)**	
On another computerised database	**46 (38)**	**Please specify** _____

NEONATAL SCREENING STAFF

For each member of staff who carries out the neonatal screening please give details of the type of staff e.g. Nursery Nurse, Neonatal Nurse, other Nurse, Audiologist, Technician etc., their grade and their whole-time equivalent.

Type of Staff		Grade	WTE
Includes:			
ENT	1		
Audiologists	71		
Scientists	26		
Nurses	14		
ATO / MTO	13		
Consultant / CMO / Paed	14		
Other	35		

STAFF TRAINING

Please describe the training relevant to screening received by staff who are involved in the neonatal screening service (please include, if applicable, any professional training, specialised training and/or refresher courses).

Professional Training

75 give details

Specialised Training

58 give details

Refresher Courses (please indicate how often)

60 give details

PART A continued

WRITTEN PROTOCOLS FOR STAFF

Do staff have written protocols to follow when screening neonates?

Yes 59 No 42 (28 blank)

If yes, and if easily available, please enclose.

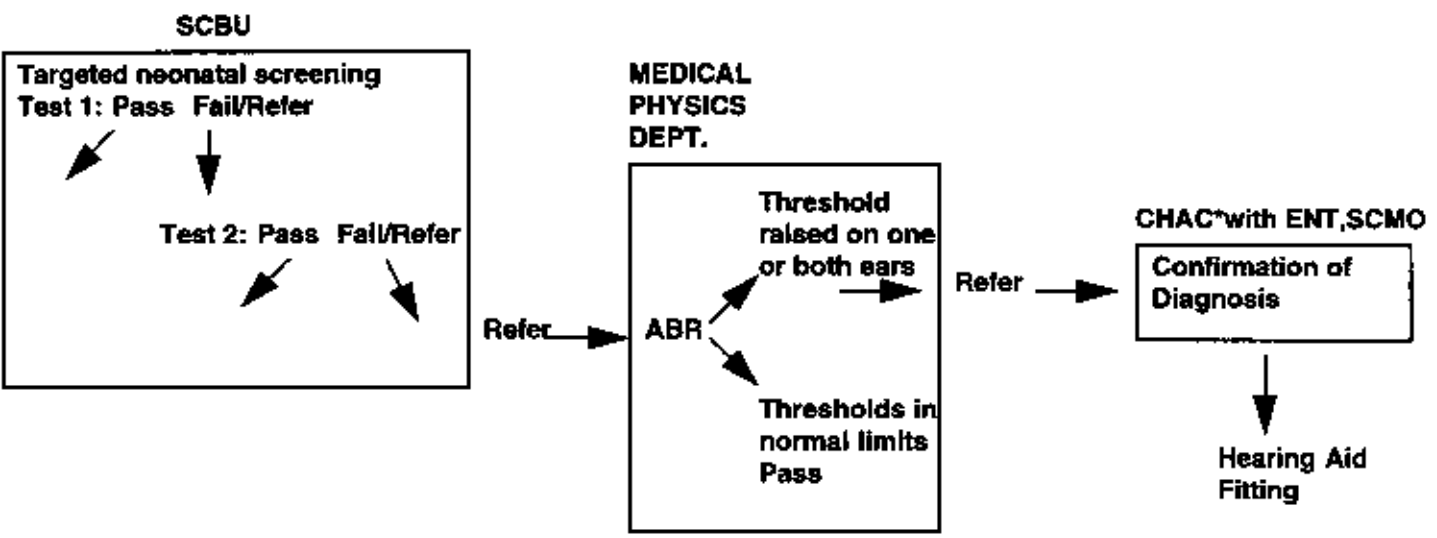
DIAGRAM OF NEONATAL SCREENING STRUCTURE

Could you provide a diagram of the structure of the neonatal screening service?

Yes 93 No 9 (26 blank)

If yes, please outline below or attach as a separate sheet. To indicate the relevant level of detail, an example of a neonatal screening structure is given below.

EXAMPLE



*Children's Hearing Assessment Centre

Space for your diagram

This is no pattern in the diagrams given.
Each is fairly unique !

PART B: MANAGEMENT OF THE NEONATAL SCREENING SERVICE

CONTRACTS WITH PURCHASERS

Who has the contract to provide the neonatal screening service? Please indicate if such screening is a specific part of the contract or part of the block contract by ticking the box in the appropraite column below.

(Please tick all those which apply)	Provided by	Specific part of contract	Part of block contract
Hospital Maternity Unit	3	0	6
SCBU and/or NICU	8	2	8
ENT Department	27	3	25
Child Health	12	0	15
Other	24	3	14

If other, please specify That is 70 overall, representing 59 districts.

SERVICE FUNDING

How is the neonatal hearing screening service funded?

(Please tick all those which apply)

	Salaries	Equipment	Cover for tester(s)	Training
Health Authority Purchasers	72 (56)	51	46	53
From research monies	4	5	0	2
From charitable donations	1	28	0	7
Other	8	6	8	5

If other, please specify

AIMS AND PLANS

Are there written or agreed aims for the neonatal screening service?

Yes 50 (44) No 42 (37) (36 blank)

If yes, please give details below or use a separate sheet if necessary

What are the development plans for the service for the next 5 years (if any)? Please describe briefly

24 (23) to universal screening
21 (20) to targetted screening

PART B continued

QUALITY CONTROL

What procedures are in place to monitor the quality of the neonatal screening service?
Please describe

Audit/Monitoring	38
check	9
Discuss	3
Routine follow-up	5
Train	1
Others	31

AUDIT

(Please tick those which apply)	Yes	No	
Has an audit been carried out on the neonatal screening service?	35 (32)	63 (50)	(30 blank)
Can the report be made available to us?	27	23	(79 blank)

If yes, please circle* where applicable: Report enclosed* / Report to be forwarded* / Enquiries being made*

COSTS

Have you been able to cost the service in any way? Yes 27 (24) No 63 (55) (38 blank)

If yes, please write in the cost and details:_____

_____

PART C: PERFORMANCE OF THE NEONATAL SCREENING SERVICE

NUMBER OF CHILDREN TESTED

How many neonates from SCBUs and NICUs did you screen in 1993 and 1994?

(Please write in and circle where applicable)*

	Number screened from SCBU/NICU		Number screened from SCBU/NICU
1993**	6303 (46)	1994**	9883 (64)
	32 actual figure*		41 actual figure*
	14 approximate figure*		23 approximate figure*
	- don't know*		- don't know*

** We are interested in the number of children tested within specific time periods. In this and subsequent questions it does not matter whether these time periods relate to a calendar or financial year. If you don't have figures for 1994 we would alternatively be interested in the figures for 1992 and 1993 (please write in).

PART C continued

FAILURE RATES

Please give the bilateral failure rates (if possible) for the first and second neonatal screens for 1993 and 1994.

(Please write in and circle* where applicable)

	Failure rate for first neonatal screening test		Failure rate for second neonatal screening test, if appropriate	
1993	11.7 (avg) %	see table 10	6.4 (avg) %	see table 12
	actual figure* 18		actual figure* 12	
	approximate figure* 17		approximate figure* 11	
	don't know* 18		don't know* 20	
	(119 blank)		(129 blank)	
1994	10.5 (avg) %	see table 11	4.7 (avg) %	see table 13
	actual figure* 27		actual figure* 17	
	approximate figure* 21		approximate figure* 13	
	don't know* 14		don't know* 16	
	(110 blank)		(126 blank)	

FALSE POSITIVE RATES

What are the false positive rates (children who failed the screen and had normal hearing on initial assessment) for 1993 and 1994 (if known)?

(Please write in and circle where applicable)*

1993	8.5 (avg) %	see table 14	1994	5.6 (avg) %	see table 15
	actual figure* 13			actual figure* 16	
	approximate figure* 11			approximate figure* 13	
	don't know* 25			don't know* 26	
	(122 blank)			(116 blank)	

YIELD AND SENSITIVITY

Please give the numbers of children who have been identified (by the neonatal screen) with a permanent hearing impairment (bilateral) of ≥ 50 dB HL better ear average over 0.5,1,2,4 kHz (or equivalent) who were born in the following years:

(Please write in and circle where applicable)*

(number of sites involved are in parentheses)

	1990	1991	1992	1993	1994
Numbers of children identified with a permanent hearing impairment	37 (17)	35 (16)	56 (20)	63 (27)	100 (39)
				51 T 12 U	89 T 11 U
	table 16	table 17	table 18	table 19	table 20

	1990	1991	1992	1993	1994
Please give the numbers of children who were passed by the screen and later found to have a permanent hearing impairment	5 (3)	8 (4)	4 (1)	3 (1)	4 (1)
	table 21	table 22	table 23	table 24	table 25

PART C continued

INTERPRETATION AND COMMUNICATION OF SCREEN RESULTS

How do you decide whether the result of the screen constitutes a "pass" or a "fail"?

(Please tick all those which apply)

Machine gives an automated readout of "pass" or "fail"	17	Personal interpretation of display by Screener	79
Other	16	If other, please specify	

If necessary can an expert opinion on the result of the screen be sought on the day of the test?

Yes 80 No 8

Who is responsible for telling parents the result of the screen?

Please write in: 3 doctors, 19 consultants, 27 audiologists, 12 testers, 7 nurses

PART D: DETAILS OF FOLLOW-UP SERVICES

ASSESSMENT

We are interested in gathering information about the assessments which children receive following a referral from the neonatal screen.

Please give details for the following.

a) What tests are used for assessment (e.g. ABR, VRA)?

ABR 83, OAE 10, Behaviour/obs 17, VRA 11, out of 99.

b) Where are the assessments carried out (e.g. Medical Physics Dept., Children's Hearing Assessment Clinic)?

Audiology 42, CHAC 15, Child Health 23, Neurophys 2, Medical physics 8

c) When is the follow-up assessment carried out (include immediately if appropriate)?

Immediate/ASAP 45, about 1 month 25, about 2 months 11

HEARING AID FITTING AND AGE OF FITTING

When young children (up to 3 years of age) are fitted with hearing aids in your District which of the following procedures (if any) are used in the verification of appropriateness of the aids?

(Please tick)

Probe-tube microphone facilities	36	Aided threshold testing	94	Other	20

If other, please describe

If hearing aids are fitted to a hearing-impaired child (up to 3 years of age) found by the neonatal screen, at what age is this routinely carried out?

Please write in: ASAP 46, 2-4 mnths 13, 4-6 12, within a year 3, depends on age 8

HEARING AID FITTING continued

Typically, how often do moderately, severely, or profoundly hearing-impaired young children (up to 3 years of age) have a hearing aid review using aided threshold or probe-tube microphone measures?

(Please tick)

Once a month or more	17	Once every six months	38
Once every three months	56	Once a year or less	10

EDUCATION SERVICES

Typically, when are the Education Services (for the hearing-impaired) informed of newly-identified, permanently hearing-impaired children?

(Please tick)

Within 24 hours of confirmation of permanent hearing impairment	54
Within 48 hours of confirmation of permanent hearing impairment	16
Within 7 days of confirmation of permanent hearing impairment	27
Other	9

If other, please specify

_____

_____

_____

On average, how often do peripatetic education staff (for the hearing-impaired) visit the family of a young severely hearing-impaired child i.e. up to 3 years of age?

(Please tick)

Twice a week	13	Once a month	9
Once a week	68	Less than once a month	3
Twice a month	12		

Please indicate if this is a year-round service or only provided during term time

(Please tick)

Year-round	27	Term time only	57

VOLUNTARY ORGANISATIONS

Are parents of newly-diagnosed permanently, hearing-impaired children given any information about voluntary organisations which could provide further advice and support?

Yes 92 (71) No 5 (75 blank)

If yes, please describe briefly

includes NDCS, RNID, Elizabeth ?

PART D continued

OTHER FAMILY SUPPORT

Are there any other services and/or support available to parents of newly-diagnosed permanently, hearing-impaired children?

Yes 90 (67) No 9 (29 blank)

If yes, please indicate who organises it and give a brief description

	(Please tick)	Brief description of service
Health Services	67	
Education Services	69	
Social Services	59	
Voluntary Organisations	61	

35 (30) had all 4, 24 had 3, 14 had 2, 16 had 1.

NATIONAL DEAF CHILDREN'S SOCIETY QUALITY STANDARDS

Are you aware of the NDCS Quality Standards in Paediatric Audiology, Volume 1?

Yes 97 (67) No 6 (68 blank)

If yes, please describe how they have influenced the service in any way, if at all

None	13
Minimal	5
Focus	9
Planning	41
Employment	7

OTHER REPORTS ETC.

Have any other documents, reports and/or articles published since about 1990 influenced the neonatal screening service in any way e.g. at the policy level or implementation stage?

Yes 68 No 18 (85 blank)

If yes, please describe

includes: Nottingham Workshop, NIH, BJA, OAE lit, BAAP, Hall report, BACDA, Archives, Peter Watkin

Critical Review of the Role of Neonatal Screening in the Early Detection of Congenital Hearing Impairments

NEONATAL SCREENING QUESTIONNAIRE Summary Statistics

TABLE 1

Year	Number
81	1
84	1
86	3
87	5
88	2
89	4
90	2
91	6
92	12
93	11
94	18
95	11
96	1

TABLE 2

Year	Number
81	1
84	1
86	4
87	5
88	2
89	4
90	3
91	8
92	11
93	10
94	16
95	12
96	1

TABLE 3

Year	Number
81	1
84	1
86	4
87	4
88	2
89	4
90	1
91	8
92	11
93	8
94	16
95	11
96	2

TABLE 4

Year	Number
84	1
85	3
86	3
87	3
88	3
89	3
90	1
91	7
92	6
93	7
94	10
95	7
96	1

TABLE 5

Year	Number
86	1
87	1
89	2
91	1
92	4
93	1
94	4
95	2

TABLE 6

Name	Number
ILO	44
Biologic Navigator	2
POEMS	4

TABLE 7

Name	Number
Linco-Bennett 1986	2
Modified DT for 4months of age	1
Portable ARC	2

TABLE 8

Name	Number
Algo Tek	2
Amplaid	3
Bio-logic systems	12
CED, medelec, Nimbus	1
Medelec	19
Navigator SLE	1
Nicolet	8
Nicolet, medelec	4
Nottingham ABR screener	8
Own system, Nicolet , medelec	1
SLE ,Nicolet, Medelec	1
Various	1

TABLE 9

dB nHL	Number
20	3
25	1
30	8
35	4
40	20
45	1
50	16
70	1
'threshold'	15

TABLE 10

% 1st fail	Number
blank	136
0	2
2	1
2.9	1
3	1
3.7	1
4	2
4.04	1
5	3
6	2
6.2	1
6.25	1
8	2
9	1
9.3	1
10	3
11	1
11.1	1
13	1
13.8	1
14	1
16	1
17	1
20	2
30	1
33	1
45	1

TABLE 11

% 1st fail	Number
blank	122
0	1
1	1
2	2
2.2	1
2.5	2
3.6	1
4	1
4.1	1
4.3	1
5	4
6	1
7.8	1
8	3
9.7	1
10	1
10.5	1
10.7	1
10.97	1
11	2
12	1
13	3
13.8	1
15	2
16.6	1
18	2
18.3	1
19	1
20	1
21.7	1
25	2
25.7	1
26	1
30	2
31	1
42	1

TABLE 12

% 2nd fail	Number
blank	148
0	4
1	1
1.01	1
1.5	1
1.9	1
2	3
3.6	1
3.7	1
4	1
4.9	1
5	2
6	1
6.25	1
8	1
9	1
12	1
33	1

TABLE 13

% 2nd fail	Number
blank	140
0	2
1	5
1.6	1
1.9	1
2	3
2.1	1
2.2	1
2.5	2
2.67	1
3	1
4.87	1
5	5
8	2
8.6	1
10	2
28	1
60	1

TABLE 14

false +ve	Number
0	11
1	1
1.3	1
2.2	1
2.5	1
3	2
3.6	1
4.5	1
7.8	1
8	1
10	1
11	1
20	1
25	2
50	1

TABLE 15

false +ve	Number
0	11
0.5	1
1	1
1.3	1
2	1
2.2	1
3	2
5	1
5.5	1
6	1
7	1
7.2	1
7.8	1
10	3
20	1
31	1
44.5	1
45	1

TABLE 16

identified	Number
0	4
1	3
2	4
3	3
4	1
6	1
7	1

TABLE 17

identified	Number
0	5
1	9
3	4
4	2
6	1

TABLE 18

identified	Number
0	6
1	5
2	6
3	4
5	3
6	2

TABLE 19

Identified	Number
0	12
1	7
2	11
3	6
4	2
8	1

TABLE 20

identified	Number
0	13
1	12
2	12
3	7
4	2
5	2
6	3
7	1

TABLE 21

identified	Number
0	13
1	2
3	1

TABLE 22

identified	Number
0	15
1	1
2	2
3	1

TABLE 23

identified	Number
0	19
1	4

TABLE 24

identified	Number
0	33
1	3

TABLE 25

identified	Number
0	38
1	4

Coordinators of HVDT/SES

Letter

Questionnaire

Tabulated data from
the Questionnaire

A Critical Review of the Role of Neonatal Screening in the Detection of Congenital Hearing Impairment

Dr A Davis
Dr H Fortnum
Dr S Wright
Mrs R Dobbins
(Nottingham)

Prof J Bamford
Dr M J Forshaw
(Manchester)

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Institute of Hearing Research
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Nottingham NG7 2RD
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Telephone (IHR): 0115 922 3431
Fax 0115 942 3710
Email: sue.wright@ihr.mrc.ac.uk

THE UNIVERSITY of MANCHESTER
School of Education
Centre for Audiology, Education of the Deaf & Speech Pathology
The University of Manchester
Oxford Road
Manchester M13 9PL
Telephone: 0161 275 3373
Fax: 0161 275 3519
Email: mark.forshaw@man.ac.uk

<date>

Ref. No.:«DScode» / «Pcode»

«Ptitle» «Pname1» «Pname2»
«Pposition»
«Paddress1»
«Paddress2»
«Paddress3»
«Paddress4»
«Paddress5»
«Ppostcode»

Dear «Ptitle» «Pname1» «Pname2»,

Critical review of the role of neonatal screening in the early detection of congenital hearing impairments.

Questionnaire: Health Visitor Distraction Test, 6 - 9 months/ Surveillance, 0 - 12 months and School Entry Screen

As part of the above review a questionnaire has been circulated to Directors of Public Health to find out which childhood hearing screening services are currently being provided in each of the Districts (or equivalent) in the UK. A sheet that briefly describes the aims of the project is enclosed.

This questionnaire has now been returned and your name has been given to us by the Director of Public Health as the appropriate person to contact for information about either the Health Visitor Distraction Test, 6-9 months screening service, the surveillance 0-12 months screening service and/or the School Entry screening service in the District. The enclosed questionnaire covers all three of these services. We would be very pleased if you could spend the time to complete the sections of the enclosed questionnaire which are relevant to your service(s). This will provide the vital information we need to give an accurate account to the Department of Health about childhood hearing screening services in the UK, and the possibilities for developing these services.

If you are not the appropriate person please pass the form on to someone who could complete it. If you have any difficulties and/or queries please contact Dr Sue Wright on 0115 967 8743.

We have to submit our report to the Department of Health in early 1996. So in order for us to analyse the replies and discuss our conclusions we need the questionnaires returned as soon as is possible in the reply-paid envelope.

The time and effort that you and your staff put into completing this questionnaire is greatly appreciated.

Yours sincerely,

Dr Adrian Davis and Dr Sue Wright.

Encl.

Critical Review of the Role of Neonatal Screening in the Early Detection of Congenital Hearing Impairments

QUESTIONNAIRE

HEALTH VISITOR DISTRACTION TEST/SURVEILLANCE, 0 - 12 MONTHS AND SCHOOL ENTRY SCREEN
Preliminary Statistics
(all figures are out of a total of 164)

This questionnaire is part of the above review funded by the Department of Health and undertaken by a team based at the MRC Institute of Hearing Research, Nottingham and the Centre for Audiology, Education of the Deaf and Speech Pathology, University of Manchester. The questionnaire is designed to gather information about current childhood hearing screening services in District Health Authorities (or equivalent) in the UK.

This questionnaire is concerned with the following children's hearing screening and surveillance services:

Health Visitor Distraction Test, 6-9 months
Surveillance, 0-12 months*
School Entry Screen

***Surveillance refers to both the routine tasks and observations Health Visitors carry out to monitor children's hearing and to the specific use of questionnaires.**

Questions in Part A are concerned with the structure of the hearing screening/surveillance service, Part B covers the management of the service and questions in Part C are concerned with the performance of the screen(s) or surveillance procedure. Questions in Part D are designed to gather information about the follow-up services and the possible influence of reports and documents on the service(s).

THANK YOU FOR YOUR HELP

Name: _____ **Address:** _____

Position: _____ _____

Trust: _____ _____
(if applicable)

_____

District Health Authority/Health Board or equivalent: _____

_____ **Postcode:** _____

PART A: STRUCTURE AND ORGANISATION

SCREENING AND SURVEILLANCE SERVICES

Please indicate below which childhood hearing screening services you coordinate in your District and whether the service is provided to all children (i.e. universal) or to a group (i.e. targeted).

(Please tick all those which apply)

	The service you coordinate	Universal	Targeted
Health Visitor Distraction Test, 6-9 months	112 (87)	148 (104)	3
Health Visitor Surveillance, 0-12 months	68 (55)	94 (70)	7
School Entry Screen	123 (92)	144 (102)	7

AGE OF TESTING

At about what age are children tested by the screening services below?

(Please write in the average age of testing)

Health Visitor Distraction Test, 6-9 months [ ] **range 6-9.5 months 153 providers (108)**

School Entry Screen [ ] **range 3.5-6.5 months 151 providers (105)**

SCREENING TECHNIQUES AND EQUIPMENT

Please specify which tests and which test stimuli are used for the different screening services.

(Please write in)

	Test technique	Test stimuli
Health Visitor Distraction Test, 6-9 months:		**table 1** **high freq (2k or more): table 2** **low freq (1k or less): table 3**
	What screening level is used?	**_____ dBnHL**
School Entry Screen:	**table 4**	
	What screening level is used?	**_____ dBnHL**

Part A continued

LOCATION OF SCREENING SERVICES

Please indicate where the screening takes place in the District and where appropriate, the approximate proportions in these locations e.g. 80%, 20% etc..

(Please tick all those which apply and write in proportions where known)

	Health Visitor Distraction Test, 6-9 months	Approximate proportions	School Entry Screen	Approximate proportions
Child's Home	103		7	
School	1		142	table 5
School Clinic	0		18	table 6
Clinic	144		18	table 7
GP Practice	121		4	table 8
Other	9		0	

If other, please specify

_____

_____

SURVEILLANCE TECHNIQUES AND AGE OF CHILD WHEN USED

Please indicate which procedures are used (if any) e.g. a questionnaire for surveillance of young children between 0 and 12 months of age and, where appropriate, at what age(s) these are used.

(Please tick all those which apply and write in the average age)

		Average age(s) of children when used
Hints for Parents Sheet e.g. by B. McCormick, CHAC	115	table 9
Parental Questionnaire	46	table 10
Other	43	table 11

If other, please specify

_____

_____

_____

_____

_____

If you think it would be helpful to the project, please attach any surveillance questionnaires or leaflets.

PART A continued

STAFF DETAILS

Please indicate which staff carry out the screening tests/surveillance procedures for i.) the Health Visitor Distraction Test, 6-9 months, ii.) the surveillance service, 0-12 months and iii.) the School Entry Screen.

(Please tick all those which apply)	Health Visitor DistractionTest, 6-9 months	Surveillance Service, 0-12 months	School Entry Screen
Trained Health Visitors	152	106	13
Trainee Health Visitors	37	33	3
Clinical Medical Officers	20	46	12
General Practitioners	21	45	2
Nurses	18	0	8
School Nurses	8	1	99
Nursery Nurses	28	8	5
Trained Assistants	74	11	25
Other	10	2	48

If other, please specify staff title

Please indicate below the actual numbers of staff involved in the service(s), full time (F/T) and part time (P/T), and give an estimate of their total time spent on this testing per year in person weeks (please include traveling time if possible).

(Please write in where applicable)	DistractionTest, 6-9 months			Surveillance, 0 -12 months			School Entry Screen		
	Nos. F/T	Nos. P/T	Estimate of time	Nos. F/T	Nos. P/T	Estimate of time	Nos. F/T	Nos. P/T	Estimate of time
Trained Health Visitors	42(90)	25(64)	40 (45)	36(44)	26(32)	57(22)	7(6)	7(4)	71(4)
Trainee Health Visitors	3(13)	6(1)	2(5)	3(6)	4(2)	1(2)	0	0	0
Clinical Medical Officers	4(16)	4(10)	10(4)	8(11)	10(8)	26(4)	6(7)	4(5)	5(4)
General Practitioners	46(5)	65(2)	0	111(3)	0	0	0	0	0
Nurses	3(5)	4(6)	6(3)	0	3(2)	1(1)	3(1)	9(5)	12(2)
Nursery Nurses	5(6)	5(12)	24(4)	3(3)	8(3)	1(1)	2(1)	2(5)	5(1)
School Nurses	13(3)	3(3)	5(1)	1(1)	8(1)	0	14(39)	14(41)	31(24)
Trained Assistants	15(9)	8(21)	29(6)	1(1)	6(3)	0	4(7)	4(12)	38(9)
Other	0	19(3)	19(3)	0	15(1)	1(2)	2(21)	2(19)	45(18)
Overall			47						40

values indicate means and (number of sites)

PART A continued

STAFF TRAINING

Please describe the training relevant to screening or surveillance received by staff who are involved in the different parts of the children's hearing screening and surveillance services.

(Please write in)

Staff Training relevant to the Heath Visitor Distraction Test, 6-9 months:
Pre-registration Training:

table 12

Specialised Training (following professional registration):

table 13

Refresher Courses (please indicate how often):

table 14

Staff Training relevant to the Surveillance Service, 0 - 12months
Pre-registration Training:

table 15

Specialised Training (following professional registration):

table 16

Refresher Courses (please indicate how often):

table 17

Staff Training relevant to the School Entry Screening Service
Pre-registration Training:

table 18

Specialised Training (following professional registration) :

table 19

Refresher Courses (please indicate how often):

table 20

PART A continued

WRITTEN PROTOCOLS FOR STAFF

Do staff have written protocols to follow for the hearing screening tests and/or surveillance procedures?

(Please tick all those which apply)	Yes	No
Health Visitor Distraction Test, 6-9 months	129	14
Health Visitor Surveillance, 0-12 months	61	36
School Entry Screen	123	21

If yes, and if easily available, please enclose.

RECORDS

How are the results (test results and details of the numbers of procedures) from the different hearing screening services recorded?

(Please tick all those which apply)

	On parent held records	As paper records	On the Child Health Computer	Other computerised database
Health Visitor Distraction Test, 6-9 months	129	79	112	15
Surveillance Service, 0-12 months	74	41	43	7
School Entry Screen	22	116	77	7

If other computerised database, please describe

DIAGRAM OF REFERRAL ROUTES

Could you provide a diagram of the referral routes for the hearing screening and/or surveillance service(s) you coordinate?

(Please tick all those which apply)

	Yes	No
Health Visitor Distraction Test , 6-9 months	139	4
Surveillance Service, 0-12 months	76	10
School Entry Screen	132	6

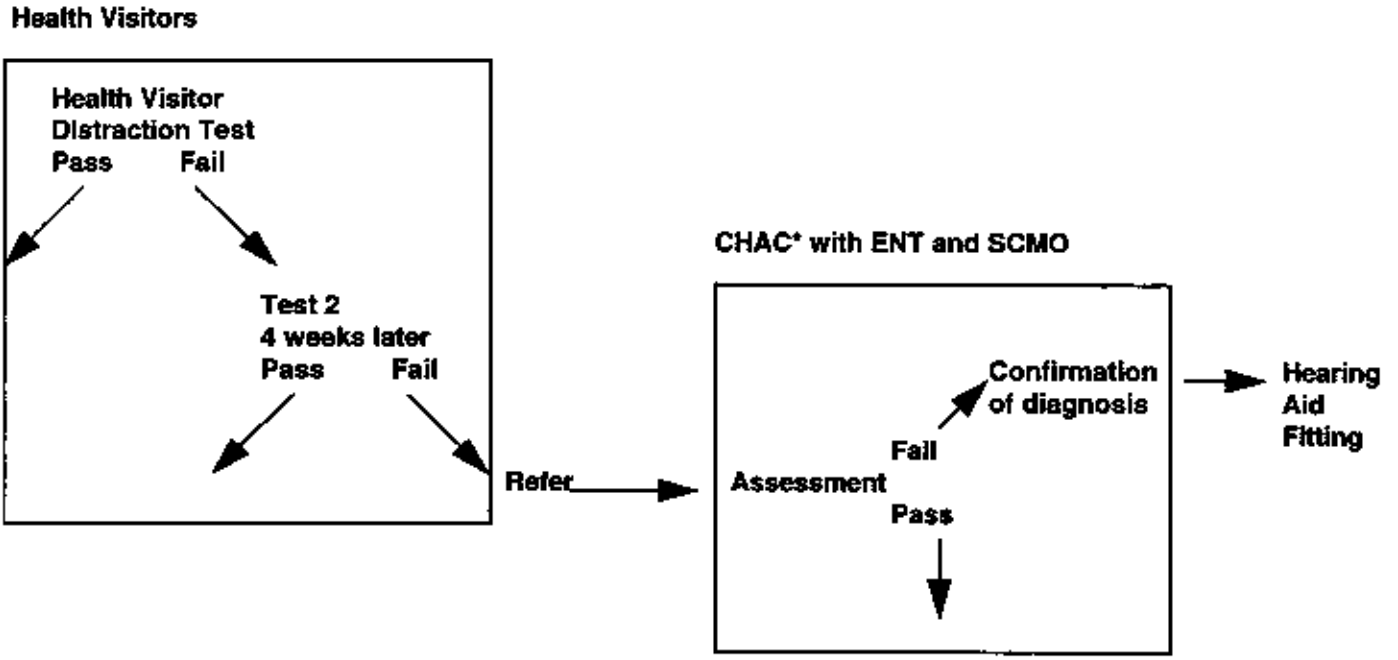
If yes, please outline opposite or attach as a separate sheet. To indicate the relevant level of detail, an example is given opposite of the referral routes for a Health Visitor Distraction Test screening service.

Part A continued

DIAGRAM OF REFERRAL ROUTES

To indicate the relevant level of detail an example of referral routes for a Health Visitor Distraction Test, 6 - 9 months, screening service is given below.

EXAMPLE



*Children's Hearing Assessment Centre

Space for your diagram(s)

Again, this was very variable

Please use the back page or attach a separate sheet if necessary.

PART B: MANAGEMENT OF THE SCREENING AND SURVEILLANCE SERVICES

AIMS AND PLANS

Please indicate if there are written or agreed aims and/or development plans (for the next 5 years) for the different parts of the children's hearing screening services listed below.

(Please tick all those which apply)	Written aims	Development plans for the next 5 years	
		Yes*	No
Health Visitor Distraction Test, 6-9 months	54 (47 districts)	42 (41)	67
Health Visitor Surveillance, 0-12 months	35 (33 districts)	20 (20)	51
School Entry Screen	57 (51 distrcits)	37 (37)	75

***If yes to development plans, please describe briefly what these are for the service(s) below (or append):**

a) Health Visitor Distraction Test, 6-9 months

24 - Audit, NS, BeST test, training

b) Surveillance Service, 0-12 months

27

c) School Entry Screen

30 - School health review, to continue as before

Is there a discussion group that meets regularly to consider the aims of the screening/surveillance services?

Yes 79 No 59

QUALITY CONTROL

What procedures (e.g. update courses, supervised training sessions, calibration procedures etc.) are in place (if any) to monitor the quality of the screening and surveillance services?

Please describe briefly stating the service referred to.

PART B continued

AUDIT

Has an audit been carried out on any of the following hearing screening/surveillance services and could the audit report be made available to us?

(Please tick all those which apply)	Has an audit been carried out?	Could the report be made available to us?	If available, please circle * where applicable
Health Visitor Distraction Test, 6-9 months	67	14	15 Report enclosed* 10 Report to be forwarded* 21 Enquiries being made*
Surveillance Service, 0-12 months	16	2	1 Report enclosed* 1 Report to be forwarded* 10 Enquiries being made*
School Entry Screen	51	9	8 Report enclosed* 3 Report to be forwarded* 21 Enquiries being made*

COSTS

Have you been able to cost the service(s) in any way? If yes, please write in the cost and give details.

Health Visitor Distraction Test, 6-9 months _____

Surveillance Service, 0-12 months _____

School Entry Screen _____

PART C: PERFORMANCE OF THE SCREENING AND SURVEILLANCE SERVICES

COVERAGE

What percentage of the children <u>that you aim to test</u> were actually tested or covered by the screening and/or surveillance service(s) listed below in the 2 years, 1993 and 1994?

*(Please write in and circle * where applicable)*

values are avergae % (number of sites)

	1993** table 21	1994**
a) Health Visitor Distraction Test, 6-9 months	90.1% (89) %	90.5% (99) %
	actual figure* 26	actual figure* 27
	approximate fig.* 57	approximate fig.* 66
	don't know* 19	don't know* 12
b) Surveillance service, 0-12 months	92% (41) %	92.7% (44) %
	actual figure* 8	actual figure* 5
	approximate fig.* 30	approximate fig.* 34
	don't know* 28	don't know* 25
c) School Entry Screen	94.3% (90) %	93.0% (91) %
	actual figure* 15	actual figure* 19
	approximate fig.* 66	approximate fig.* 64
	don't know* 30	don't know* 29

** We are interested in the percentage of children tested within specific time periods. In this and subsequent questions it does not matter whether these time periods relate to a calendar or financial year. If you don't have figures for 1994 we would alternatively be interested in the figures for 1992 and 1993 (please write in).

REFERRAL RATE

What percentage of children tested in the years 1993, 1994 was referred by the screening tests and/or surveillance service for further hearing tests?

*(Please write in and circle * where applicable)*

	1993	1994
a) Health Visitor Distraction Test, 6-9 months	10.4% (61) %	9.3% (63) %
	actual figure* 30	actual figure* 28
	approximate fig.* 28	approximate fig.* 31
	don't know* 34	don't know* 36
b) Surveillance service, 0-12 months	6% (16) %	8.5% (17) %
	actual figure*	actual figure*
	approximate fig.*	approximate fig.*
	don't know*	don't know*
c) School Entry Screen	9.3% (55) %	10.4% (68) %
	actual figure* 17	actual figure* 27
	approximate fig.* 35	approximate fig.* 40
	don't know* 54	don't know* 45

YIELD

Please give the numbers of children who have been identified by the hearing screening and/or surveillance service(s) with a permanent, bilateral hearing impairment of ≥ 50 dB HL better ear average over 0.5,1,2,4 kHz (or equivalent) tested in the following years:

*(Please write in and circle * where applicable)*

	1990	1991	1992	1993	1994
a) Health Visitor Distraction Test, 6-9 months				table 22	table 23
				actual fig.* 30	actual fig.* 33
				approx. fig.* 15	approx. fig.* 14
				don't know* 31	don't know* 31
	1990	1991	1992	1993 35 (26)	1994 39 (28)
b) Surveillance service, 0-12 months				table 24	table 25
				actual fig.* 18	actual fig.* 18
				approx. fig.* 5	approx. fig.* 6
				don't know* 31	don't know* 32

Please give the numbers of children who have been identified by the School Entry Screen with a permanent, bilateral (Bi) or unilateral (Uni) hearing impairment of ≥ 50 dB HL better ear average over 0.5,1,2,4 kHz (or equivalent) tested in the following years:

1990	1991	1992	1993		1994	
			Bi 28 (30)	Uni 44 (24)	Bi 27 (37)	Uni 68 (31)
			tables 26	27	tables 28	29
			actual fig.* 23	actual fig.* 15	actual fig.* 27	actual fig.* 19
			approx. fig.* 7	approx. fig.* 8	approx. fig.* 9	approx. fig.* 10
			don't know* 52	don't know* 59	don't know* 49	don't know* 56

FAILURE ON FIRST SCREEN

If a child fails on the first Health Visitor Distraction Test and/or the first School Entry Screen how many more subsequent tests are carried out before they are referred on for assessment?

(Please write in where applicable)	number of tests - number of replies
Health Visitor Distraction Test, 6-9 months	blank - 17, 0 - 5, 1 - 133, 2 - 9
School Entry Screen	blank - 27, 0 - 26, 1 - 101, 2 - 9, 3 - 1

PART D: FOLLOW-UP SERVICES AND REPORTS

ASSESSMENT

We are interested in gathering information about the assessment services which children receive following a referral from the Health Visitor Distraction Test, from the first year Surveillance service and/or the School Entry screening service.

(Please write in where applicable)

a) What tests are used for assessment following referral from the screening and/or surveillance service(s) listed below?

Health Visitor Distraction Test, 6-9 months

Surveillance Service, 0-12 months

School Entry Screen

b) Where are the assessments carried out (e.g. Community Clinic, Children's Hearing Screening Service) following referral from the screening and/or surveillance service(s) listed below?

Health Visitor Distraction Test, 6-9 months

Surveillance Service, 0-12 months

School Entry Screen

ASSESSMENT continued

c) When is the follow-up assessment carried out following a test or surveillance procedure?

(Please write in)

Health Visitor Distraction Test, 6-9 months: _____

Surveillance Service, 0-12 months: _____

School Entry Screen: _____

HEARING AID FITTING AND AGE OF FITTING

When young children (up to 5 years of age) are fitted with hearing aids in your District which of the following procedures (if any) are used in the verification of appropriateness of the aids?

(Please tick those which apply)

Probe-tube microphone facilities	43	Aided threshold testing	106
Other	25		

If other, please describe

_____

If hearing aids are fitted to a hearing-impaired child (up to 5 years of age) found by the Health Visitor Distraction Test or the surveillance procedures, at what age is this fitting routinely carried out?

Please write in: _____

Typically, how often do moderately, severely, or profoundly hearing-impaired young children (up to 5 years of age) have a hearing aid review using aided threshold or probe-tube microphone measures?

(Please tick those which apply)

Once a month or more	25	Once every six months	65
Once every three months	61	Once a year or less	23

Typically, when are the Education Services (for the Hearing Impaired) informed of newly-identified, permanently hearing-impaired children?

(Please tick all those which apply)

Within 24 hours of confirmation of permanent hearing-impairment	70
Within 48 hours of confirmation of permanent hearing-impairment	17
Within 7 days of confirmation of permanent hearing-impairment	40
Other	18

If other, please specify

_____

_____

EDUCATION SERVICES

On average, how often do peripatetic education staff (for the hearing-impaired) visit the families of permanently hearing-impaired children (up to 5 years of age)?

(Please tick all those which apply)

Twice a week	30	Once a month	23
Once a week	93	Less than once a month	18
Twice a month	28		

Please indicate if this is a year-round service or only provided during term time.

(Please tick)

Year-round	27	Term time only	86

OTHER FAMILY SUPPORT SERVICES

Are there any other services and/or support available to parents of newly-diagnosed permanently, hearing-impaired children?

Yes 127 No 4

If yes, please indicate who organises this and give a brief description

	(Please tick)	Brief description of service
Health Services	93	
Education Services	87	
Social Services	87	
Voluntary Organisations	66	

VOLUNTARY ORGANISATIONS

Are parents of newly-diagnosed permanently, hearing-impaired children given any information about voluntary organisations which could provide further advice and support?

Yes 124 No 5

If yes, please describe briefly

PART D continued

NATIONAL DEAF CHILDREN'S SOCIETY QUALITY STANDARDS

Are you aware of the NDCS Quality Standards in Paediatric Audiology, Volume 1?

Yes 119 No 22

If yes, please describe how, if at all, they have influenced the service in any way.

OTHER REPORTS ETC.

Have any other documents, reports and/or articles published since about 1990 influenced the neonatal screening service in any way e.g. at the policy level or implementation stage?

Yes 62 No 21

If yes, please describe

YOUR COMMENTS

If you would like to make any additional comments we would like you to write these below and please use a separate sheet if necessary.

We would particularly welcome your comments on the possible future role of neonatal hearing screening.

If you have any further comments please continue on the opposite page.

HEALTH VISITOR DISTRACTION/ SCHOOL ENTRY SCREEN QUESTIONNAIRE
Summary Statistics

Table 1

value	count
1	1
1,3	2
1,3,6	1
2	3
2,3	3
2,6	1
3	39
3,	1
3,1	2
3,1,5	1
3,2	8
3,2,5	2
3,4	1
3,4,	1
3,5	10
3,5,6	1
3,6	1
3,7,1	1
4,2,	1
4,3	2
5	18
5,1,3,2	1
5,2,3	1
5,3	6
5,6	2
6	1

key: 1 rattle, 2 hum,
3 voice, 4 chime, 5 warble
6 sand, 7 bell

Table 2

value	count
1	87
1, 5	1
1,2,3	1
1,2,3,5	1
1,3	8
1,3,4	2
1,5	6
1,6	1
3	2
4,3,6	1
5	9
5,1	3
5,1,2,3	1
5,1,3	1
5,1,6	1
6,1	1

key: 1 rattle, 2 hum,
3 voice, 4 chime, 5 warble
6 sand, 7 bell

Table 3

value	count
1	3
1,2,3	1
2	6
2,6	1
3	6
3,2,5	1
3,4	1
3,5	1
3,6	1
5	20
5,2,3	2
5,3	1
5,6	1
6	3

key: 1 rattle, 2 hum,
3 voice, 4 chime, 5 warble
6 sand, 7 bell

Table 4

value	count
Co-operative	1
Cooperation	1
Imped &pure tone	1
McC T/test, high freq sweeps	1
McCormick	2
McCormick Toy Test/Rattle	1
MWL, PTA	1
PTA/Tympanogram	1
pure tone Audiom.	138
sweep audiometry	1

Table 5

value	count
70	1
80	3
85	1
90	11
95	7
98	2
99	4
100	104

Table 6

value	count
0	1
2	1
5	3
10	7
20	1
30	1
100	1

Table 7

value	count
1	3
2	1
5	2
10	3
15	1
20	1
45	1
90	2
95	1

Table 8

value	count
45	1
98	1
99	1

Table 9

value	count
0	2
1	2
1-7 month	1
1.5 months	2
1st Birth Visit+	66
2 weeks	1
2 weeks old	1
3 years and above	1
4/12 months	1
6 weeks	6
6 weeks onwards	1
6/8 weeks	1
7month	1
8 months	2
9 months	1
At Birth	21
At Birth-4yrs	1
Used during 1st 2 yrs	1

Table 10

value	count
0	2
1st Birth Visit+	15
3 years &above	1
4.5	1
5 year	4
5.5	1
6 months	1
6 weeks	3
7 months	1
7.75	1
8	3
8 months	4
8 weeks	2
At Birth	2
School entry	1

Table 11

value	count
0	2
1-2 years	1
1.5	1
18/36 months	1
1st Birth Visit+	7
36	2
6 weeks	4
6-9 month	1
6wks/7wks	1
8 months	1
8 weeks	2
9 months	1
At Birth	2
HV visits	1
on-going surveillance	1
Varirous	1

Table 12

value	count
1/2 day in house	1
2 day course	1
3 days with ENT Sur and ToD	1
Course with audio staff	1
Local Training	4
Local Training + CPT	1
Local training and aud input	1
nil	2
None	3
Not known	1
Observation	2
Provided in inversity	1
Theory	2
Theory+Practice	15
Trained by audiometrician on DT	1
Trained by CPT	1
Training	1
Training at CHAC	1
Training once a year	1
Within HV Training	71

Table 13

value	count
1/2 day in house	1
2 day course on employment	1
3 1/2 hours	1
Assessed by TOD	1
Attend secondary clinic	1
Course	1
Course by Nuffield Trust	1
Course+Tests/ACC	1
Courses run by SCMO	1
In Service	26
In service with Aud	41
In service with Educational Aud	1
In-service by SCMO	1
Local training	1
None	5
Post registration	1
Post-reg training	1
Practice	9
Regular training	1
Theory+Practice	16
Trained by audiometrician on DT	1
Training by Aud staff	1
Training by TOD, Edu Auth	1
Updates	1
Updating training	1
WIth Ed audiologist	1

Table 14

value	count
4 years	1
Every 2 years	1
Five Years	2
Nil at present	1
None	1
None at present	1
Only one in 8yrs	1
Three Years	29
Three Years and 5 years	1
Three Years, theory yearly	1
Twice yearly	1
Two Years	54
Updates (no time)	16
Yearly	27

Table 15

value	count
1/2 day in house	1
3 days w ENT Sur and ToD	1
None	2
None specifically	1
Not known	1
Observation	3
Practice	1
Theory + Practice	5
Theory:	1
TOD	1
Trained by CPT	1
Training by Aud.Staff	1
Training by CMO/CPT	3
Training by CPT	1
Within HV Training	48
Within pre-reg training	1

Table 16

value	count
1 hour	1
1/2 day in house	1
Awareness	1
Child Health Surveill. course	1
Course	1
Course with Tests/ACC	1
Courses run by SCMO	1
In Service	12
In service (Aud.staff)	24
In-service by SCMO	1
Local training	1
None	1
None specifically	1
Not yet in place	1
On a needs basis	1
Practice	2
Theory	2
Theory+Practice	9
Trained by audiomet. on DT	1
Training with aud	1
Updates	1
Upto individual HVs	1

Table 17

value	count
3 yearly, theory is yearly	1
6 monthly	1
Every 2 years	1
Five Years	2
None	3
None specifically	1
Policy set by SCMO	1
Three Years	8
Twice a year	1
Two Years	20
Updates (no time)	20
Yearly	13
Yearly/2 years	1

Table 18

value	count
1/2 day in house	1
3-4 months	1
Audiom cert/1 unqualified	1
BAAT Parts I and II	2
BAAT/ATO	1
BSA course	1
BTEC Physiolo Measures	1
BTEC/BAAT	1
In Service Training	5
In service with MTO's	1
Instruction by SCMO/CMO	1
Nil	1
No special training pre-ref	1
None	7
Not known	1
Observation	1
Observation only	1
PTA Training	1
Theory	2
Theory+Practice	6
Theory, BBAT/BSA for Aud	1
Trained	1
Trained Audiologists	11
Training by CMO/CPT	2
Training in community	1
Within HV training	3
Within Med Training	3
Within pre-Reg Training	28
Within teacher training	1

Table 19

value	count
1 day supervision	1
1/2 day in house	1
2 days on pure tone	1
As needed	1
As part of MTO training	1
Attend courses	1
Attended courses	1
Audiometricians have certs	1
BAAT Pts I and II	1
BAAT training	1
BAAT/BTEC	1
BSA Course Pt I and II	1
Clinics with more exp staff	1
Core training	1
Courses as available	1
Courses SCMO/Aud courses	1
Has been, failure uncertain	1
In service	20
In service by Aud Staff	46
In service by Educ. Aud Staff	1
In service by SCMO	1
In-service with CMO	2
None	2
ONC medical physics	1
Part of Further Training	2
Practice	2
Theory	1
Theory+Practice	9
Varies - 3 day course	1

Table 20

value	count
6 monthly	1
Annually	1
Approx 1/12months	1
Courses as available	1
In-service by Aud staff	1
Nil	1
No regular courses at present	1
No requirement	1
None	7
None at present	2
None, start Jan 96	1
Not held regularly	1
Not in place yet	1
Not known	1
On request	1
Three Years	9
Two Years	23
Under discussion	1
Under review	1
Updates (no time)	34
Yearly	30

Table 21

value	count
60	1
62	1
65	1
74	1
75	1
76	1
77	1
78	2
80	3
81	4
83	2
84	1
85	7
86	1
87	2
89	1
90	11
91	5
92	3
93	1
94	1
95	10
96	3
97	2
98	6
99	6
99.9	1
100	10

Table 22

value	count
0	18
1	11
2	7
3	4
4	2
5	3
10	1
22	1

Table 23

value	count
0	14
1	23
2	8
3	4
8	1
12	1

Table 24

value	count
0	12
1	3
2	6
3	2
4	1
5	2

Table 25

value	count
0	12
1	6
2	1
3	7
4	1
6	1

Table 26

value	count
0	21
1	4
2	1
3	1
5	2
9	1

Table 27

value	count
0	10
1	3
2	4
3	2
5	3
6	2

Table 28

value	count
0	26
1	5
2	3
5	2
6	1

Table 29

value	count
0	13
1	3
2	5
3	1
4	3
5	1
6	3
8	1
9	1

Appendix 3

NCHAM advisory document

Since the National Institutes of Health (NIH) Consensus Development Conference on Early Identification of Hearing Loss in Infants and Young Children recommended that all newborns be screened for hearing loss before being discharged from the hospital, there has been a dramatic increase in the USA in the number of hospitals doing newborn hearing screening. In fact, the number of hospitals with universal newborn hearing screening programs has more than quintupled since March 1993.

The rapid expansion of universal newborn hearing screening programs has brought into focus questions about the most appropriate technique for newborn hearing screening. Through the 1980s, the approach recommended by most people (including the Joint Committee on Infant Hearing (JCIH), the American Speech-Language-Hearing Association (ASHA), and the American Academy of Audiology (AAA)) was to identify children who were at risk for hearing loss (this comprised approximately 10% of the population) and to use conventional auditory brainstem response (ABR) to determine whether those children had hearing losses. However, data from multiple studies showing that only about half of all children with congenital hearing loss exhibited any of the risk factors, coupled with the emergence of new techniques for screening, have caused most people to abandon the risk factor approach to newborn hearing screening. Instead, the vast majority of newborn hearing screening programs are now using automated auditory brainstem response (AABR), distortion product otoacoustic emissions (DPOAE), or transient evoked otoacoustic emissions (TEOAE). Equipment in each of these categories is currently being used in successful newborn hearing screening programs. But which technique is best?

Although this is probably the most frequently asked question by people considering the implementation of a newborn hearing screening program, the fact that there are so many different programs being conducted successfully with equipment in each of these categories suggests that the answer to the above question is not simple or straightforward. Indeed, because the characteristics of these techniques are so heterogeneous, there is probably not a definitive answer about which type of equipment is best. Individual hospitals will have to continue making their own decision, and there may well be situations where one type of equipment is best for the situation of one hospital, while a different type of equipment is best for the situation of another hospital.

The purpose of this brief document is to outline some of the issues that should be considered in selecting equipment. It would be good if there were definitive information for each of those issues. Unfortunately, such definitive information does not exist for many of these issues. What we have are results of a few studies and a lot of opinions based on clinical experience. The table below summarizes the research evidence and the clinical experience in an effort to help people select equipment to use in their own newborn hearing screening program.

Issue	Automated ABR	DPOAE	TEOAE
1. Cost of equipment (These figures are based on suggested retail prices by the manufacturer and include all necessary equipment to conduct newborn hearing screening. In those cases where the screening equipment is sold separately, but requires a computer to operate, we have included the cost of a moderately priced computer. The cost for a printer is not included.)	Natus Algo 2 $15,500 Algo 1E $9,000 Intelligent Hearing Systems Smart Screener* $11,850	GSI 60 $9,500 Bio-logic $12,750 Virtual $10,000 Mimosa $8,500	Otodynamics ILO88 $8,000
2. Cost of Supplies (Included here is the cost of all necessary supplies and reoccurring expenses (e.g., calibration) for doing screening. It does not include supplies for communicating the results of screening with parents of pediatricians, printing educational materials, or other ancillary materials. Costs are estimated per baby based on reported usage by typical programs with 1000–4000 births per year.)	$8.50–10.00 per baby includes the costs of disposable earphones and electrodes	$.50–1.50 per baby includes the costs of disposable tips for the probe assembly, calibration, and probe replacement	$1.00 per baby includes the costs of disposable tips and for the probe assembly and replacing the probe assembly every 750 babies

* *For completeness, the price of the HIS Smart Screener and the Algo 1E are listed here, even though the rest of the commentary only applies to the Algo 2 since it is the most frequently used automated ABR screener.*

continued

Issue	Automated ABR	DPOAE	TEOAE
3. Initial Training of Screening Technicians (Although it is possible to start any program by reading the literature which comes with the equipment and teaching yourself, most programs find that hands-on, competency-based training by someone who is already experienced with that particular equipment and has used it successfully is the best way to begin a program. Estimated times are based on the experience of operational programs and includes only the initial training screening technicians. Regular supervision with additional upgrading of skills should be included in addition to this initial training.)	2 hours	4 hours	4 hours
4. Time to do Screening per Baby (This is often misunderstood because the term "screening time" is used by people to refer to different aspects of the screening process. As used here, it is the total amount of time devoted screening babies and includes getting the baby ready for screening, talking to the parents if necessary, setting up the equipment, conducting the screening, recording information about the baby so results can be retrieved later, etc. "Screening time" is best computed by taking the total number of hours worked by screening technicians and dividing that time by the number of babies screened during that period. Numbers for each device are based on reports of well-established programs.)	15–40 minutes per baby	10–30 minutes per baby	10–30 minutes per baby
5. What is Being Measured? (None of the devices is a direct measure of hearing. Instead, each one measures slightly different physiological mechanisms which are related to hearing. Issues related to this are discussed below.)			
5a. What Degree of Hearing Loss is Likely to be Detected?	As used in most programs, the Algo 2 uses a 35 dB nHL click and, consequently, would probably miss children with very mild sensory hearing losses (25 or 30 dB). An alternative mode for the Algo 2 measures at 40 dB and 70 dB.	Although there is not unanimous agreement, some researchers believe that DPOAEs will only be detected when hearing is better than 40 dB nHL. Others believe that with the proper parameters, hearing loss as low as 25 dB nHL can be detected.	There is substantial agreement that TEOAEs will be detected if hearing threshold is 25 dB nHL or better.
5b. Is Frequency Specific Information Available? (In addition to indicating whether or not a child has a hearing impairment, some people are interested in knowing at what frequencies that hearing impairment is likely to occur. Others argue that the purpose of a screening test is not to provide detailed information about the nature of the loss, but to identify those children who need further diagnostic tests, during which information about frequency and severity of hearing loss can be determined.)	The Algo 2 is a dedicated screening device. Screening is a selection procedure for diagnostics where hearing loss is confirmed and its characteristics defined. No frequency specific information is obtained by click evoked auditory potentials screening, but is available through completion of diagnostic ABR follow-up where it is used to make treatment decisions.	DPOAEs have the best potential for providing frequency specific information, and some argue that DPOAEs can be used as a diagnostic tool. However, this has not been sufficiently demonstrated. There is general agreement that DPOAEs provide more information about the higher frequencies (6–10 kHz) than do TEOAEs, but most would agree that the improved information in these higher frequency areas is not very critical for hearing screening.	TEOAEs provide information about the frequencies at which emissions are detected between 1 and 5 kHz. However, the absence of an emission at a particular frequency does not always correspond to a hearing loss at that frequency.
5c. What is Being Measured?	The AABR provides information about the auditory pathway up to the brainstem (including the middle ear, the inner ear, and the VIII nerve).	DPOAEs provide information only up to and including the cochlea. Hence, infants with central auditory processing problems would not be discovered. Although definitive prevalence data are not available, most experts agree that this represents less than 1% of all children with hearing loss, or less than 3 children per 100,000 in the general population.	TEOAEs provide information only up to and including the cochlea. Hence, infants with central auditory processing problems would not be discovered. Although definitive prevalence data are not available, most experts agree that this represents less than 1% of all children with hearing loss, or less than 3 children per 100,000 in the general population.

continued

Issue	Automated ABR	DPOAE	TEOAE
6. Scoring Criteria and Ease of Interpretation (Because DPOAEs and TEOAEs produce a waveform for each infant, users must decide what constitutes a pass or a refer. Because the widespread use of these techniques is fairly recent, there is not universal agreement on what criteria should be used. In practice, however, this lack of agreement affects a very small number of infants, since in most cases emissions are clearly present or clearly absent, and it's only the relatively small number of infants around the cut point where disagreement occurs.)	The Algo 2 matches the ABR to a template derived from the waveforms of normally hearing neonates to 35 dB nHL click stimuli. The algorithm employs binomial sampling and a statistical test to determine that data collected sufficiently discriminates between the presence of a response + noise vs. pure noise at > 99% level of confidence. There is no operator interpretation needed. Studies which have compared the results of the Algo 2 with expert scoring of conventional ABR have found agreement ranging from 83% to 98%.	DPOAEs are the most recent of the techniques and, not surprisingly, there is a lot of disagreement about what constitutes a pass or a refer. Most people have tended to use fairly conservative pass criteria until more data are available. The numerical criteria are easy to interpret, and most programs use technicians to make this determination in a few seconds per baby.	Although they have been used extensively since the early 1990s, there are still many different pass/refer criteria being used in TEOAE-based newborn hearing screening programs. The most frequently used criteria recommended by NCHAM is a very conservative criteria. Using this numerical criteria, interpretation is straightforward and is done in most programs by technicians in a few seconds per baby.
7. Flexibility of Administration	Because it was intended to be a completely automated system, the Algo 2 is designed to have very little flexibility. This is viewed by most people as an advantage. It is possible to screen at either 35 dB or at 40 dB and 70 dB, and it is possible to screen both ears simultaneously or each ear separately.	There is much flexibility in how the test is administered. Unfortunately, there is little agreement about what parameters are best for screening, particularly true with respect to the different primaries to be used for f_1 and f_2 and the intensity of the stimulus. There is also little agreement on how many data points per octave are required for an adequate test.	Although there is a great deal of flexibility with regard to collecting TEOAE information, parameters used in screening programs are usually those recommended by NCHAM (e.g., QuickScreen, low frequency filter, 50 low noise samples, peak stimulus between 78 and 83 dB SPL).
8. Flexibility of Use	The Algo 2 is a dedicated screener designed for use only with infants. Consequently, it can only be used for screening newborns.	In addition to being used for infant screening, DPOAE equipment is used with children and adults for monitoring the effects of surgery and drug administration and various diagnostic applications.	In addition to being used for infant screening, TEOAE equipment is used to screen hearing with children and adults for monitoring the effects of drug administration and various diagnostic applications.
9. Referral Rates (Screening is designed to identify a small group of at-risk infants who will require further diagnostic testing. As in all screening programs, it is expected that some children who have normal hearing will be referred for further diagnostic testing, but the lower this number is, the better.)	Reported referral rates at the time the infant leaves the hospital for programs using the Algo 2 equipment range from 1% to 10%, with an average of about 4%.	Reported referral rates at the time the infant leaves the hospital for DPOAE programs range from 4% to 15%, with an average of about 8%. Since most DPOAE programs do a two-stage screening process where those who do not pass before discharge from the hospital are rescreened before referring them for diagnostic testing, the percentage referred for diagnostic testing is about 1%.	Reported referral rates for infants at the time they are discharged from the hospital range from 3% to 12%, with an average of about 7%. Since most TEOAE programs are a two-stage screening program, with infants who are referred at the time of discharge from the hospital being screened a second time before being referred for diagnostic assessment, the percentage of infants referred for diagnostic assessment ranges from $^1/_2$% to 1%.

continued

Issue	Automated ABR	DPOAE	TEOAE
10. Screening in Noisy Situations (Noise which interferes with screening can come from the external environment or from the baby. Because newborn nurseries can be quite noisy, many people have questions about the effects of noise on newborn hearing screening procedures (this is especially true for intensive care nurseries.))	The Algo 2 manual recommends choosing a baby in a favorable state, "sleeping, having been fed recently", for most efficient screening. An artifact reject system automatically interrupts data collection when ambient noise > 50 dB SPL at 2000 Hz and automatically resumes when conditions meet criteria again. Thus, the Algo 2 screens in noisy settings, but noise may slow data collection.	The key to screening in noisy situations is achieving good probe fit. Not all DP equipment provides feedback regarding adequacy of probe fit. Most DP units have artifact reject systems which exclude noisy data from averaging. Thus, the equipment can be used in noisy settings, but data collection is slower. Because DPOAEs measure one frequency at a time, they are moe susceptible than TEOAEs to a response at that frequency being obscured by noise. Babies no not need to be asleep, but a noisy baby will slow data collection substantially.	The key to screening in noisy situations is achieving good probe fit. The ILO88 provides excellent real-time information to monitor probe fit and has an artifact reject system which excludes noisy data from averaging. Thus, the equipment can be used in noisy settings, but data collection is slower. Babies do not need to be asleep, but a noisy baby will slow data collection substantially.
11. How Many Children with Hearing Loss will Pass the Screen? (These children are often referred to as false negatives and reported as a measure of the test's sensitivity. It is important to minimize the number of infants in this category. While no screening test is perfect, ideally, as few children as possible should be in this group. This does not refer to children who have late onset losses, but instead is only concerned with those children who have impaired hearing at the time of the test and still pass the screen.)	Infants with very mild losses (25 to 30 dB) will likely pass the screening, as will infants with high frequency losses, reverse slope losses, or precipitous losses.	Depending on the parameters used, children with hearing losses less than 40 dB, as well as children with reverse slope losses and neural or central auditory pathology, may pass.	Children with neural or central auditory pathology or children having reverse slope losses may pass.
12. Cost per Infant Screened (Although there have numerous reports in the literature and anecdotal reports about the cost per baby screened in newborn hearing screening programs, most of these analyses are based on gross estimates of time devoted to different tasks or have been incomplete (e.g., have ignored fringe benefit costs for personnel, indirect costs, supervisory costs, or costs associated with supplied and equipment). How the program is organized can also have a big impact on the cost per baby. Because of such factors, people trying to interpret reported costs should be very cautious and remember that cost per baby is primarily a function of how long it takes to do the tasks, coupled with the hourly rate of people doing the work and the cost of supplies, equipment, and facilities.)	Reported costs range from $15 to $75 per baby.	Costs per baby are not available for DPOAE programs, but they should be similar or a little bit higher than those reported for TEOAE.	Reported costs range from $8 to $30 per baby.

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