Structure and status of this report

The first part of this report concentrates on screening issues; the second on primary prevention and health promotion; and the third considers the wider implications and implementation issues. The appendices are devoted to screening topics not debated at the seminar; an account of systematic reviews; and a short note on the evolution of preventive child health in the UK, written primarily for the many overseas colleagues who have requested a copy of this report.

For each topic, I have either used the authors' abstract as presented for the seminar, or else I have summarised the content and key messages of the review. This section is followed by a commentary, which draws on observations made by an invited discussant at the seminar, and on comments from the floor; but it nevertheless reflects a personal opinion. Thus the commentary sections are my responsibility and do not necessarily reflect the views of the authors, the discussants, the National Screening Committee, or the Royal College of Paediatrics and Child Health.

The timescale for this report required that I either confined myself to factual summaries of the reviews themselves, or offered a personal opinion, since to consult widely and obtain a consensus would take many months. I believe that this latter task is one which would be more profitably dedicated to the revision of "Health for all children" (reference - see page …) and the eventual production of a fourth edition.

Terminology

In this report I have used the same terminology as was recommended in the third edition of "Health for all children". Thus screening refers to an activity or procedure which fulfils, or is intended to fulfil, the classic criteria set out by Wilson and Jungner. Child health surveillance refers to a wider set of procedures, of which screening is only one, whose aim is to identify conditions, disorders and problems at an early stage. The other methods of surveillance include observation, asking parents questions and inviting their comments, identifying abnormalities on an opportunistic basis, and making use of the observational skills of other individuals, for example play group leaders. Thus this use of the term "surveillance" can be related to the concept of secondary prevention, an approach recommended by Butler

See Butler J. *Child health surveillance in primary care*. London: HMSO, 1989).

Child health promotion in contrast is a wider philosophy whose aim is to promote good health and prevent ill health in the widest sense of the term. It includes surveillance, but also places emphasis on primary prevention wherever possible, and the creation of an environment in which individuals have the opportunity to optimise their health. Over the past decade, the three Joint Working Parties have progressively moved from a closely focused approach to screening issues, to a much greater emphasis on primary prevention.

Current concerns

Although it is gratifying to observe the impact of "Health for all Children", the pace of change has been rapid and it is time to take stock. Notwithstanding the overseas interest in what the UK has done, there is no doubt that we now have the leanest well-child health care programme in the Western world. This may be a matter for celebration and the shift of emphasis to primary prevention is the correct way to go.

Nevertheless, there is a danger that the pendulum will swing too fast. In the current climate, soft targets for disinvestment are attractive. Many of the services currently being debated were introduced for excellent reasons, and by the very nature of these reasons, and the aims of the services in question, their effectiveness is extremely difficult to assess. Health promotion does not take place in isolation but in the changing social context; many other factors also influence outcomes; and the outcomes desired are often many years in the future. Thus the maxim frequently quoted at the conference "no evidence of effect is not evidence of no effect" should be remembered when considering changes in our service network.

The political context

Economic pressures and health service re-organisation are potentially more powerful drivers of change at the present time than the publication of research or Systematic Reviews. From an academic point of view, one would often wish to defer making any decisions until better evidence is available. The reality is that most research programmes and Systematic Reviews answer only one or two questions, while creating four or five new ones. Not infrequently one has to make decisions on the basis of seriously inadequate evidence. Thus we have to operate on the "best buy" concept, knowing that in the future we may need to change our policy again.

General issues

A number of themes emerged at the meeting which were common to all the various programmes:

- For all screening programmes, it is important to be clear about the aims. For example, where hearing screening is concerned, it is not enough to specify that one is searching for children with hearing impairment, it is necessary to specify what type and severity, and by what age it should be found. The issue becomes much more complex in screening for speech and language impairment, and more difficult still when addressing the question of the school entrant examination, whose purpose is poorly defined and seems to vary widely between authors.

- Parents have a right to know about the screening programmes that are being offered. They should be told what is being provided and why; the limitations of the screening process; what is likely to happen if the screening test is positive; what the indicators might be of a child who had passed the screening test but nevertheless had the condition in question. Information of this nature could be provided more extensively in the personal child health record. However, currently this is usually provided only after the child is born and it might have more impact if given in pregnancy. Many parents will not absorb the information included therein, unless they are taken through it by a health professional or other knowledgeable person.

- Television and video are obvious ways to disseminate understanding more widely regarding the benefits and hazards of screening programmes, and how parents can contribute themselves to earlier identification of important conditions.

- The vital importance of adequate management time and commitment for any screening programme was stressed repeatedly. Many screening programmes involve several different trusts and professional groups. Commissioning and purchasing authorities need to specify in their requirements that one individual must be the named person responsible for quality control and monitoring. Surprisingly few health professionals have the requisite skills. The time commitment required to do this job properly is universally underestimated. Sometimes this is compounded by poor quality data or even by deliberately limiting access to data, due to misguided notions of commercial security.

Information systems

Most of the presenters at the meeting recognised the need for data on the coverage and impact of screening programmes. However, the issue was not discussed in detail. There are several obstacles to the use of routine community data for monitoring screening:

- there is still inadequate integration of community and hospital data systems.

- paradoxically at a time when the possibilities opened up by information technology are greater than ever before, the restrictions being placed on the use of this technology both by legislation and by ethics committees are becoming increasingly obstructive.

- children do not always stay in their district of birth, but move away, so that one does not always know about false negatives from screening programmes.

Questions to do with the confidentiality and ownership of data about individuals, held on large public health data bases, are still being debated (.e.g., the recently published Caldicott Report). At one

extreme, there is a view that all data is entirely the property of the patient or parent in question and that it should not be used in any way without the specific permission of the individual. Clearly this position virtually guarantees the end of most public health research and monitoring. At the other end of the scale, some believe that the monitoring of such data is not only desirable but essential and a public duty, that the confidentiality of individuals can be adequately protected by modern technology, and that individuals who have benefited from the evolution of health care research over the past century might reasonably be expected to contribute to the furtherance of knowledge when there is no cost or risk to themselves.

This debate involves not only the maintenance of routine health data bases, but also the development of registers for specific conditions. It will be apparent from the material that follows in this report that there is a particular need for such registers for children with congenital hearing loss, and for those with inherited metabolic disorders. Without national registers it will be virtually impossible to know which children have been missed by the screening programmes.

The rights of parents

Parents feel strongly about the early identification of important medical conditions. The fact that a particular disorder is not treatable in the strict medical sense of the term, or that the benefits of any treatment might be modest, does not alter their views on this subject. Parents feel profoundly let down by the health professional network if they discover that some chronic condition in their child could have been identified some years earlier.

These powerfully held views do not justify the establishment or maintenance of a screening programme that flies in the face of evidence - but the public funds our National Health Service and if professionals do not feel that the expense, or the medical hazards of a particular screening programme, can be justified, we owe it to parents firstly to explain precisely why this is so and to demonstrate the disadvantages of running unsatisfactory screening programmes; and secondly, we need to ensure that when parents do express concerns to their GP or other health adviser, these are taken seriously and there is prompt access to an appropriate specialist who can resolve the problem.

A Critical Review of the Role of Neonatal Hearing Screening in the Detection of Congenital Hearing Impairment

Adrian Davis, John Bamford

In 1995, the NHS R&D Health Technology Assessment Programme funded a critical review of the role of neonatal hearing screening in the U.K. The review is necessary because of increasing doubt about the ability of existing screening programmes (mainly the health visitor distraction test at 7-8 months) to deliver early identification of such children, and technological advances which have made newborn hearing screening an option.

Researchers at the MRC's Institute of Hearing Research and the University of Manchester's Centre for Audiology, Education of the Deaf and Speech Pathology carried out the review, and reported in 1997 to the National Co-ordinating Centre for Health Technology Assessment. The report is available as a long summary, as a full report, and as a book1. The review involved two major strands of work. First, a review of the available literature, published and unpublished; and second a comprehensive survey of current pre-school hearing screening provision in the U.K. coupled with a health economic study of hearing screening costs2, a number of focus groups and visits to key sites.

The report reviews and summarises the evidence in five areas: epidemiology of permanent childhood hearing impairment, evidence for improved outcomes with earlier identification, current U.K. practice, the effectiveness of existing screening programmes, and the evidence on costs of different programmes.

Conclusions

There are approximately 840 children a year born in the U.K. with significant permanent hearing impairment likely to affect their and their family's quality of life substantially. Present services do not identify about 400 of these children by 1½ years of age nor about 200 of these children by 3½ years of age. Such late identification of hearing impairment greatly reduces the responsiveness of the services for individual children. Hearing impaired children identified late are at risk of substantial delay in their acquisition of language and communication with consequent longer term risk to 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.

The survey of current practice indicated poor information systems, and a wide variety of differing practice. There are currently two programmes in which all newborns 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 health visitor distraction test. Intervention and habilitation for those screened neonatally is routinely well within six months, for those screened only by the health visitor distraction test begins on average at 18 months. The yield from the neonatal screening programmes is increasing, and the apparent yield from the health visitor distraction test is low, below 30%.

The cost comparisons between the different implementations of hearing screening in the first year of life were encouragingly uniform, with systematic differences observed between implementations such that universal neonatal screening appeared to have lower initial costs associated with it than the health visitor distraction test on a cost per child screened basis. The cost per case found would be several orders of magnitude lower with universal neonatal screening.

The published evidence on screen performance indicates poor sensitivity and relatively poor specificity for the health visitor distraction test, with relatively low yield. Median age of identification via the health visitor distraction test 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, and median identification ages for those screened in the order of 2 months.

Option Appraisal and Recommendations

The recommendations that stem from the evidence presented have been made in the light of the ongoing service context. If a hearing screening programme were being set-up ab initio a further large scale randomised control trial

might be required. However with neonatal hearing screening there is a very strong case for setting up the 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, there is an existing but inadequate screen in place and to withdraw it without replacement would be unacceptable, and third, the potential harm from treatment consequent on detecting cases is not a major factor in the case of screening for a hearing loss.

Nine screening options in 4 different categories (no screen, health visitor distraction test, at-risk neonatal screening and universal neonatal screening) were evaluated in terms of their running costs, incremental yield, efficiency, responsiveness and equity. The report makes a number of recommendations in three areas - service development, implementation and research, where the weight of the evidence strongly supports:

1. The introduction of a national screening programme for congenital hearing impairment based upon universal neonatal screening followed at 7 months by a targeted screen using an infant distraction test (primarily for those who have not had the neonatal screen).
2. The development of an information system strategy to facilitate the co-ordination of the services needed for screening and following up hearing impaired children. Such a system would involve the development of a local shared-list (or register) of hearing impaired children ultimately leading to regional and national lists and linked with the local child health record information systems.
3. The adoption of a model screening programme, including appropriate targets, around which the preferred option of universal newborn 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 (averaged in the mid-frequencies on the better ear). Responsibility for implementing and monitoring the programme should be explicit. Habilitation should be initiated early and be provided within a service context that is perceived as seamless by parents and their children. Service links with education are likely to be crucial and need to be well co-ordinated. The relevant groups in the voluntary sector, whose involvement is guaranteed by law, have a significant support and coordinating role.
4. Consideration of a number of research and development needs, in particular concerning;

   (i) habilitative management of children identified by neonatal screening,
   (ii) models for service co-ordination, including Joint Commissioning,
   (iii) further refinement of screening techniques, both neonatal and infant and
   (iv) prevalence and risk factors for late onset and progressive permanent childhood hearing impairment.

The recommendations have considerable policy implications and may result in changes of practice. However, this will not happen overnight. The National Screening Committee will consider the recommendations in early 1998. The outcome of their deliberations may be to convene a working group to define, among other things, (i) the quality standards that would define adequate performance, (ii) how to specify an information system that will allow performance to be measured, (iii) what should be done if screening falls below the required level of quality, and (iv) a recommended model for commissioners.

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

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

would continue to use support materials and training videos, such as those developed by McCormick and published by Nottingham District Health Authority (1987).

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1 Adrian Davis, John Bamford, Ian Wilson, Tina Ramkalawan, Mark Forshaw, Susan Wright.
A Critical Review of the Role of Neonatal Screening in the Detection of Congenital Hearing Impairment. Health Technology Assessment 1997: 1 (10).
2 Stevens J, Hall D, Davis A, Davies CM, Dixon S. The costs of Early Hearing Screening in England and Wales. 1998. Archives of Diseases in Childhood 78: 14-9.

Hearing screening: summary & commentary

The problem Severe hearing impairment is a devastating handicap which, if not detected, leads at worst to a frustrated, non-communicating adult with a high risk of mental illness. The benefits of intervention are correspondingly substantial. Management may involve the use of a hearing aid, a cochlear implant or a manual sign system. All of these rely on skilled educational support. Definitive evidence that neonatal diagnosis and early hearing aid fitting are better than diagnosis one or two years later is not yet available, but the evidence is persuasive and sufficient on which to act.

The current test (the health visitor distraction test - HVDT) has been in use for 40 years. It can work well if it is (1) done in protected time (this is the crucial difference in the costings offered by various speakers - it is a cheap test if done in baby clinics but in those circumstances it cannot produce good results) (2) it is closely and frequently supervised to maintain quality. However, many papers show that in reality it has produced disappointing results. With the advent of high-risk neonatal screening in many places, the number of cases left to be diagnosed by the HVDT has gone down. Even with an improved or cheaper HVDT, this still applies.

Poor performance of HVDT The child health surveillance programme recommended a development and health review at around 7-9 months of age, primarily because this is the optimal age for the HVDT. It soon became clear that the HVDT performs very poorly if it is entangled with a range of other health care issues, even though it is cheaper in professional time when this is done. Those few authorities who have reported good results have done so by insisting that the test is important enough to be done in protected quiet time, in proper surroundings and using agreed techniques.

If the routine universal HVDT were to be discontinued, it would be necessary to re-examine the issue of whether there are any other particular benefits in a review at 7 months. It may be a good time to re-check the hips and to address questions of diet, feeding and sleep problems and home safety, but it is possible that the hips could be reviewed at the third immunisation and that the other aims could be undertaken a few months later at age 12 - 15 months at the same time as the MMR immunisation. This would be done at the surgery or health centre, though whether there would be any significant savings or added value is less clear. The proposal needs to be examined in one or two districts.

New technology In view of the cost and poor performance of the HVDT, doing nothing is not an option. It must either be improved or stopped. Technology now exists which allows screening of all neonates for the identification of hearing loss in the first few weeks of life. This has been used in three sites in the UK. One is based on a behavioral method (auditory response cradle) which is not widely favoured, partly because of limited published data and experience, partly because of doubts over technical reliability in earlier trials, and also because of concerns about sensitivity. The other two use the newer objective methods of testing. One of the two sites is NHS funded, the other was supported by a research grant and has now finished. Neonatal screening is however widely and successfully used in the USA.

The logical approach now is to work towards discontinuing the HVDT and introducing universal neonatal screening. However, it is worth considering the costs involved in setting up neonatal screening. The cost per case detected, though less than for the HVDT, is inevitably going to be high. Purchasers and commissioners will doubtless observe that it may be double the cost per case of metabolic disease detected by TMS (see below).

Universal neonatal screening would nevertheless be an excellent investment if one were certain that early intervention made the difference between a functioning earning adult and permanent disability and dependency. The evidence is not sufficient to make such a claim

with absolute confidence, but it is enough to justify proceeding with neonatal screening since, as argued here, doing nothing is not an option.

Careful planning and evaluation are advisable in any national implementation plan because:

- Although there is considerable USA experience, there are only two established programmes of universal screening using NHS funding; experience must be gained in arranging funding, training , technical supervision, data handling, audiological and educational support. There is however extensive UK experience with targeted neonatal screening, which will form the basis for universal screening.

- The cost per case detected, though much less than for the HVDT, is inevitably going to be high. It may be double the cost per case of metabolic disease detected by TMS (see below). Nevertheless, stopping all screening for deafness, or doing nothing, are not options.

- Universal screening is cheaper than properly conducted HVDT. The necessary funds must be identified. "New" money may be forthcoming in some districts, since a good business case can be made for this programme. If districts decide to divert funds from the HV budget to neonatal screeners, this will need planning over two or three years. In the current financial climate this would be a sensitive issue. (see Naish J. Screening Infants' Hearing: what's it all about? Nursing Times 1998;)

- There is a potential conflict between the need (at present) to perform neonatal screening in hospital, and the wish of many mothers to have their babies at home; and also between hearing screening and other screening measures that may be introduced in the future.

- There may well be shortages of suitable staff to carry out these new tasks. Training unqualified staff to carry out neonatal screening is possible, but close monitoring is vital to maintain quality.

- It is possible that planning, introduction, supervision and management of universal screening may need to be done on a contract basis, with smaller trusts calling on large centres to provide the technical expertise, at least during the start-up phase.

- The LEA must be involved since the teachers of the hearing impaired will have to take on the teaching of very young infants and some may be ill-prepared for this task.

- Though parent response to universal screening seems to be generally positive, little is known about the psychological aspects of early diagnosis from the parents' perspective. Screening has its own hazards and can cause considerable anxiety to some parents; and changes in the communication process which follows the early diagnosis of deafness merit further study. Information and support to parents are crucial for a successful programme.

- There have been some concerns about the false positive rate of BSER; some parents might be told that their baby has a hearing loss, which later will turn out to be a "false positive". Provided that diagnostic testing is properly supervised by experienced people, this should not be a major problem.

- The distraction test will still have a role to play for those babies missing the neonatal screen and those whose parents become concerned about their hearing after the neonatal period.. There are virtually no data on how babies needing this service will be flagged on a register or how the delivery of this more "targeted" distraction test will be achieved

Recommendations

These are based on the view that (a) universal neonatal screening is now the "best buy" option and (b) that a controlled trial comparing differing approaches to hearing screening, although undoubtedly desirable, is not a practical proposition.

1. Trusts should require one person to take a lead in reviewing their staff and their current audiological service.

2. One person in each trust should then be identified to examine the potential for universal screening and should form a working group to consider how universal screening could be implemented.

3. There should be a national working group of the professionals involved to examine current and future staffing implications for screening and the spin-off effects for children's audiology services in general.

The options for improving the HVDT during the difficult transition phase should also be considered. A national multi-disciplinary implementation team would be the ideal solution.

4. The National Screening Committee should consider how best to encourage the introduction of this technology while seeking to maintain some quality control.

5. This might be done by inviting trusts to tender for the role of demonstration sites. However, the actual costs of setting up the programme should be met by the trust. There is no point in showing that the new method can work if it is centrally funded - the real challenge is to make it work by re-deploying existing resources.

6. The tender should include the preparation of detailed guidelines on implementation, including staffing and financial issues.

7. The Joint Working Party on Child Health Surveillance should re-examine the need for a general review of the baby's health and development at 8 months of age, if the hearing test is discontinued.

8. A nationally agreed specification for a database is needed for the audiological screening programme, linked with the population register for routine child health care and immunisation.

9. A National Register of hearing impaired children is needed to monitor the impact of the screening programme and of any changes that may be needed in the future. This was considered some ten years ago and it was clearly a difficult task, but with the support of the voluntary organisations it may now be possible.

SCREENING FOR
SPEECH AND LANGUAGE DELAY

A systematic review of the literature 1

ABSTRACT This review focuses on children with primary speech and language delays in the 0-7 years age range. Primary delays are those which cannot be attributed to other conditions such as hearing loss, developmental disabilities etc. Four domains integral to the screening process were identified, namely the prevalence of primary speech and language delay, its natural history,the effectiveness of intervention and the accuracy of the screening procedures themselves. Strict inclusion/exclusion criteria were applied to the data set.

The prevalence data suggest that the number of potential cases of primary speech and language delay, when determined by a cut off on a standardised test, is high with a median figure of 5.95% reported for delays in either speech or language. However, the reported range for apparently comparable conditions is wide. There has been little attempt to tie this evidence into prediction of subsequent case status, and there is little published evidence to support the perception either that the total number of children with language delays declines in real terms across the age range or that prevalence has been rising over recent years.`

The natural history data indicate that a substantial proportion of children identified on the basis of expressive delays alone are likely to have difficulties which resolve spontaneously in the pre-school period. However, the data do not, at this stage, make it possible to predict at the time of identification which of the children with expressive delays are likely to have persistent problems. The picture for older children is clouded by the lack of evidence from samples which have not received any additional educational or therapeutic support. Nonetheless it is clear from treated samples that children identified as having language difficulties in the first year of primary school are likely to have difficulties which persist through to secondary school. Risk factors for persistent problems include the initial severity, the extent to which the difficulties are generalised across speech and language domains (most notably the degree to which receptive as opposed to expressive language skills are implicated) and the extent to which other cognitive and developmental skills are also delayed. However, it is not clear from the data to what extent

1 This systematic review was commissioned by the NHS Centre for Reviews and Dissemination at the University of York on behalf of the Health Technology Assessment programme. Full copies of the report are available from the NHS Centre for Reviews and Dissemination, University of York, York YOI 5DD.3.

factors such as social class, family history, temperament and gender contribute to the prediction.

Randomised control trials (RCTs) (N=1O), quasi-experimental (N=12) and experimental single case designs (N=26) met the study inclusion criteria for the review of literature relating to primary language delay. The results from the RCTs and the quasi-experimental designs were combined using meta-analysis, where a standardised effect size of +1.0 is equivalent to progress from the 5th to the 25th percentile. Treatment outcomes in speech (i.e. articulation and phonology), expressive language, receptive language and auditory discrimination were analysed separately, with separate analyses for norm-referenced and criterion referenced measures. The results revealed positive and statistically significant effects of intervention relative to untreated controls in all areas. Comparable results for direct (clinician- administered) and indirect treatment were observed in the cases of expressive language, with average effect sizes ranging from +0.65 to +1.16. In contrast, direct intervention was more effective in the case of speech, where effect sizes for direct treatment ranged from +0.94 to +1.11, while those for indirect intervention ranged from -0.02 to +0.20. However indirect intervention was more effective in the case of receptive language, with an average effect size of +1.43 for indirect treatment compared with an average effect size of -0.02 for direct intervention. Data from the single case experimental designs were synthesised using the PND (percentage of non-overlapping data) statistic and provide confirmatory evidence for the positive effects of intervention. The data in particular provide evidence for the generalisation of treatment effects. However, the data reviewed do not provide information about the long-term outcomes of intervention, nor on the likelihood of intervention reducing prevalence in a given population. Similarly it is not possible to draw conclusions about the effects of subject variables such as socio-economic class or age upon the relative value of interventions.

The screening evidence indicates that, although a considerable number of assessments have been shown to perform adequately in terms of their productivity, few studies compare the performance of two or more screening tests when applied to one population nor do they compare single screening tests across different populations. The majority of studies examine single screens on single populations. It is, therefore, difficult to make judgements about the relative value of different procedures. In general, specificity is higher than sensitivity. Interpretation is further complicated by the considerable variation in the cut offs adopted on the range of reference "gold standard" measures, suggesting that there remains considerable disagreement as to what proportion of the population should be considered cases. There have been no explicit attempts to benchmark the target population in terms of prevalence estimates in terms of the prediction of case status or in terms of the impact of intervention.

The review concludes that it would be premature to introduce universal screening for speech and language delays on the grounds that, while screening tests can be shown to be reasonably accurate, they are not yet sufficiently predictive. The data do not allow conclusions to be drawn about withdrawing existing services.

Despite this conclusion, it is clear that early speech and language delays should be a cause of concern for those involved with child health surveillance, because of the problems they may pose for the individual child, because they may serve as a litmus test for other co- morbid conditions such as hearing loss, developmental and behavioural difficulties and because of the implications that early language delays may have for literacy and socialisation. The fact that there is not sufficient evidence to merit the introduction of universal screening does not imply that these children should not be identified.

The review suggests that more attention might be placed on sharing with parents the responsibility for identifying children with speech and language difficulties. Primary care workers (health visitors, GPs, school nurses and nursery staff) should be involved in eliciting parental concerns and in making appropriate observation of children's communication behaviours. This would require formal training for such professionals in current knowledge relating to delayed speech and language development and risk factors pertaining to it. Appropriate information would also have to be available for parents to allow them to play an active role in judging need.

Given the reported value of indirect approaches there is a case for widening the range of professionals able to promote good interactive practice in parents of young children. Speech and language therapists are in a good position to play an active role in disseminating this

information. Children who do not respond to such primary prevention could then be given access to speech and language therapy services and appropriately structured nursery input.

There are many gaps in the literature. The review identified a number of research priorities. There is a need for further work on screening measures which have good predictive validity. It is suggested that natural history data should be combined with report and observation as part of such a screening measure. There is an urgent need for randomised control trials to examine the medium and long term effects of appropriately described models of intervention. These should include an appropriate range of outcome measures including where possible, economic analysis.

Speech and language: commentary

This was a valuable review. It showed modest benefits for speech therapy but also showed that involving parents and carers could be as effective and possibly more effective than therapy given just by a professional. The report did not find support for the wholesale adoption of first-year preventive schemes such as "Wilstar".

It is important to remember the limitations of the review - these are not criticisms but reflect points raised by the authors themselves, and the subsequent discussion. The primary research on which it is based has its own problems:

- Most studies were carried out in the clinic setting by therapists and it is difficult to ensure that selection and allocation are truly unbiased and that evaluation of treatment effects is blinded.

- The selection of cases and the exclusion of many categories of speech and language problem may tend to polarise the findings and bias them in the direction of greater treatment effect.

- Children with global learning problems, autistic spectrum disorder or hearing impairment may present with language delay. We know very little about the efficiency of the speech and language therapy service in sorting out these categories.

- Few of the studies distinguished between "innate" and "environmental" causes of language delay, and hardly any addressed the issue of children from ethnic minorities whose first language is not English.

- There is an educational literature on the general theme of "enrichment" for children in deprived circumstances. This educational approach seeks to prevent the effects of deprivation across all domains of development of which language is just one.

Recommendations

1. This review is to be discussed at two dissemination seminars shortly and it would be wise to await professional response to this controversial report.

2. The early intervention programme known as Wilstar should not be introduced at present except as a properly funded formal research study.

3. Primary prevention through "enrichment" programmes must be considered in the context of other initiatives, especially those in the education sector. It should be examined in collaboration with educational academics who have expertise in pre-reading work. The DfEE should be involved.

4. The review did not address the problem of distinguishing children with primary language delay from those with learning problems or autism (or deafness) and a professional response, perhaps from a working group on this issue, should be sought - it is rather artificial to just review primary speech and language delay, since children do not present conveniently labeled.

5. The time has come to examine service delivery - see next section.

Implications of Systematic Reviews on hearing and on speech therapy for the surveillance of the two to four year old age group: what next?

Secretory otitis media is a fluctuating condition for which a screening programme would be inappropriate. Nevertheless, there are occasional children who do have a prolonged and significant disability as a result of the hearing loss arising from glue ear. The benefits of speech and language therapy are measurable but modest, and there is much scope for intervention through parents and carers. The older the child, the easier it is to identify those children with delayed language development who may

have significant language impairment, whether isolated, or secondary to other problems.

A new approach? The time has come to re-think how we provide for this age group within the health services.

- Health visitor update on language Health visitors should monitor the child with slow language development and provide supportive advice themselves, rather than referring immediately to the speech and language therapy service. By delaying referral for a few months, the numbers of children needing referral might be much reduced.

- Triage therapist: Those who do need an assessment should be seen by an experienced therapist who can sort out those who need detailed assessment or follow up from those who do not. This "triage" approach is already widespread.

- The one-stop shop for language delayed children Currently, the parent of a child with language delay will be referred simultaneously to the speech and language therapist and the audiologist; they may also be referred to a paediatrician, a clinical psychologist, and an educational psychologist.

- Speech therapists all receive training in linguistics, audiology and acoustics, and have the skills to secure the co-operation of young children. An experienced speech and language therapist could learn to test hearing, measure non-verbal IQ, and carry out a basic checklist for autistic features. On an experimental basis, they could then offer a "one-stop shop" for children where there is concern about their language development. Many therapists would welcome this idea.

- The impending manpower crisis in paediatric audiology This proposal, if implemented together with universal screening, would help to address the predicted shortage of community doctors trained in paediatric audiology within the next decade.

- Language development schemes need to be tested We need an investigation of the various schemes designed to promote language development, in close collaboration with educational experts and academics, and local authority collaborative schemes.

Pre-school vision screening.

Sara Snowdon and Sara Stewart Brown.

Summary of the main issues identified and reviewed:

A systematic review of research on the effectiveness of pre-school vision screening was undertaken. The primary aim of pre-school vision screening is to identify amblyopia, refractive errors and the non cosmetic obvious squint which cannot be detected without screening. We have called these target conditions. The research questions, formulated using the criteria for evaluating screening programmes, were concerned with the Natural history and prevalence of the target conditions, the disability associated with them, and the detection and treatment through screening children's vision at the age of three for five years.

Findings and recommendations:

The search involved 17 electronic databases, reference lists of retrieved papers, a limited amount of hand searching and making contact with relevant professionals. This yielded 05,000 references and 85 studies were included in the main analysis. While no studies were found with the primary aim of establishing the prevalence of the target conditions in pre-school children, data from studies of screening programmes report on a range of yield for all the target conditions of 2.4-6.1 per cent, which is sufficiently common to justify screening. No studies designed with the intention of documents in the Natural history of these conditions were found. Without this information, it is impossible to estimate the effect of treatment from studies which have no control group. Few experimental studies of treatment were found and no studies which included a "no treatment" control group. The literature investigating disability in relation to the target conditions is insufficient to draw any firm conclusions about the impact on quality of life. Evidence from retrospective for observation of Studies suggests that primary orthoptic screening programmes can be provided in the UK with acceptable uptake and referral rates, and that these programmes are more effective at identifying children with straight to my amblyopia and refractive errors than screening by health visitors or general

practitioners. However, the one prospective controlled study we identified does not support the belief that pre-school vision screening reduces the prevalence of the amblyopia at seven years of age.

Given the lack of good research into the Natural history of the target conditions, the disabilities associated with them, and the efficacy of available treatments, it may be appropriate to continue screening only in the context of a controlled trial of treatment. Providers currently offering screening programmes should be considered discontinuing them and new programmes should not be implemented. If policy makers to not feel able to take this step, the reasonable compromise could be that they insisted that screening and treatment be offered only following a clear statement of the limitations of medical knowledge about the conditions, the treatment and its side effects, to enable parents to make informed choices.

Major research areas requiring investment:

- The extent of disability attributable to the target conditions.
- The prevalence of blindness or partial sight attributable to amblyopia in the UK.
- The prognosis for vision in the amblyopia I falling loss of vision in the better I.
- The impact of orthoptic treatment on family life and the psychological well-being of the child.
- The effectiveness of orthoptic treatment for amblyopia on vision and quality of life. This should be a randomised controlled trial in which the control group is not treated, using health outcome measures defined in studies of disability. This would also provide data on the Natural history of amblyopia. Trials undertaken in groups of children aged three to four and five to seven would determine whether screening in the pre-school years confers in the benefit of the screening at school enrty
- The effectiveness of treatment of non cosmetic obvious squint and refractive errors in this age group.

Commentary by discussant, Professor Fielder.

We know very little about the natural history and treatment of amblyopia. Nobel prize winners demonstrated around 30 years ago that there is a period of visual development when the visual system is especially susceptible to insult and visual experience plays a role; this period of sensitivity ends around six to seven years. Interruption to normal development during the sensitive period by strabismus or a blurred retinal image (e.g., refractive error) results in the visual deficit known as amblyopia. This research showed that amblyopia should be identified and treated as early as possible. It is difficult to overestimate the world wide wave of enthusiasm the studies generated, leading to the establishment of a UK national pre-school vision screening. So it is no surprise that in the systematic review no studies of the natural history of amblyopia or rigorous treatment trials were identified: they would have been precluded on ethical grounds as during the recent past studies involving a no- treatment group would have been considered unethical.

This systematic review did not consider alternatives to whole population screening such as the screening of high risk groups. These include children with a family history of amblyopia or strabismus, children with developmental problems and those who were born pre-term. This review has highlighted that the natural history and management of amblyopia, the, eye condition of childhood, have not been adequately studied.

See: Rahi JS, Dezateux C. The future of pre-school vision screening services in Britain. British Medical Journal 1997; 315: 1247-8.

Fielder A et al. Compliance in amblyopia therapy: objective monitoring of occulsion. British Journal of Ophthalmology 1995; 79: 5859.

Parker J et al. Vision screening of children selected by family history-how many children would be missed? Invest Ophthalmol Vis Sci 1997; 38: S 844

Vision screening: commentary

This presentation by Dr Stewart-Brown and colleagues was the subject of a very lively debate. The first part of the discussion centered around the issue of whether the evidence she had reviewed was sufficient justification for the recommendations made. The two key messages of her review were:

- amblyopia is probably not very disabling; while the possibility of becoming visually disable by loss of the one good eye, by accident or disease, is important, numerically it is probably not enough in itself to justify screening

- the treatment (patching) is of dubious effectiveness and may even be harmful from a psychological point of view

There are however several reasons why we should not abandon vision screening for children at the present time as suggested by the review:

- The review did not consider the question of screening at school entry (typically age 5 though progressively children start school earlier than this). However, if the main target disorder at age 5 is also amblyopia, then the same arguments would apply.

- It assumed that unilateral amblyopia is the only target condition. It could be argued that bilateral amblyopia and refractive error are also important.

- There is a political or parental dimension. It is counter-intuitive to suggest that vision screening is not useful. Many parents are angry and upset at the discovery of amblyopia at age 7 or even older in some cases.

- A further review should consider the role of screening vision in school age children. It does not seem wise to abandon pre-school or school entry screening, without simultaneously considering what role (if any) is played by screening the older age groups. Screening for colour vision defects is part of vision screening in most districts and should be included..

- If vision screening were to be withdrawn from a national programme some parents would almost certainly take their child to an optometrist for a vision check. This would have two undesirable outcomes - it would increase inequity between social groups and it would probably cost as much to test some children in the optometrists' premises as all children in school.

Recommendations

1. Further discussion in a working group or seminar of ophthalmologists, orthoptists, optometrists, paediatricians, and public health doctors is needed to consider the implications of this report. It may be wise to sample public opinion as well.

2. No new programmes should be introduced unless they have a built-in research component; but it would be inappropriate at present to discontinue old ones. "Today's best buy" may be to continue with a vision screening programme with the aim of ensuring that every child has had a vision test by the age of five. This is best done by an orthoptist but if this is not possible then the personnel who do the tests must be trained by orthoptists and there must be some form of quality control by random checks.

3. The review only addressed pre-school vision screening but the implications are probably the same for school entry screening.

4. If one is to screen at all, professionals trained to do the task properly (usually orthoptists) should do it rather than nurses or doctors.

5. The trend to school entry at age 4 may have several implications: (a) the urgency of answering the question - "is screening at age 3 ½ better than at age 5?" - will diminish (b) there will increasingly be a captive audience at a younger age so that the whole population could be tested before the age of 5 (c) the younger the child, the more challenging the testing and therefore the greater the argument for use of a specialist to screen (d) we need to know whether school nurses, appropriately trained ,could obtain results as good, or almost as good, as those obtained by an orthoptist.

6. There are a number of research issues arising from the review. In particular, further research is needed on the efficacy of treatment for amblyopia and the extent of any disability caused by the condition. However, these questions will be difficult and expensive to answer. Policy decisions have to be made now before all the desirable evidence is available.

7. In the future, it may be possible to identify at risk children much earlier and prevent amblyopia but this is some years away.

8. The Personal Child Health Record should be updated to include information about vision screening and parents should also be told about amblyopia - briefly in the Record, but in more detail when they attend eye clinics. Ophthalmologists may not accept the finding of the review, that patching is of little benefit - but they should at least ensure that parents understand what we do and do not know about amblyopia and its treatment.

9. The review does not apply to screening after school entry because the target disorders are different - i.e., primarily refractive error, not amblyopia or squint. A separate examination of the evidence is needed to consider whether there are any benefits in testing vision (including colour vision) in children age 7 and upwards.

10. The evidence would probably not support such a programme. Routine school vision tests after the age of five probably could be stopped. The yield of important new findings must be very low. Parents and children could be informed about the free access to eye testing by optometrists. Children starting secondary school could be reminded of the need to get an eye check if they had any difficulty with school work or sport. Those who have career aspirations needing good vision or normal colour vision should go to the optometrist for advice. Of course, any child with difficulties in school should have a vision check as a routine. These proposals need to be tested in the field.

Neonatal Screening for Inborn Errors of Metabolism: Cost, Yield and Outcome

R. J. Pollitt (Sheffield Children's Hospital), A. Green, N. K. Virdi (Birmingham Children's Hospital), J. V. Leonard (Institute of Child Health, London), C.J. McCabe, N. J. Cooper, P. Nicolson, J. R. Tunaley, A. Booth (School for Health and Related Research, Sheffield)

The Commission

A systematic review of the literature is required to establish:

- the burden of inborn errors of intermediary metabolism.
- the proportion identifiable by existing and emerging tests.
- sensitivity and specificity of the tests. the natural history of untreated cases.
- effectiveness of existing interventions for these conditions.
- the number of cases presenting before 10 days (the point at which about 75% of Guthrie cards have been tested).
- The review will need to gather and use information on: costs (including direct and indirect costs); effectiveness and psychological and social impact, including acceptability to patients.

The review The multidisciplinary nature of the team lead to a "productive clash of cultures" and particularly to an examination of some of the underlying (Wilson and Jungner based) assumptions of the commissioning brief. The conclusion was that any serious inherited disorder presenting in infancy or early childhood is a candidate for neonatal screening. Whether it is acceptable will depend on:

- the performance of the screening test
- the balance between costs and resulting health benefits

The review concentrated on disorders for which there is already significant experience of large-scale screening and for diseases which may be detected by the emerging technique of tandem mass-spectrometry. The disease-specific section of the review used a causal pathway approach based on that from the NHS Centre for Reviews and Dissemination (1996) and assessed:

(a) prevalence of the condition
(b) the clinical significance of the biochemical abnormality concerned.
(c) accuracy of the screening test
(d) effectiveness of early treatment

The number of cases presenting before 10 days was judged to be less important in view of the difficulty of distinguishing many of the disorders covered from more common causes of neonatal morbidity and the substantial literature on late and missed diagnoses. Additionally we reviewed the literature on the psychological impact of neonatal metabolic screening programmes on parents.

The "quality" of the literature and the quantitative or qualitative information it contained were assessed separately. In many instances the "quality" of the clinical literature was low in that there is a tendency for interventions producing very clear beneficial effects to be accepted without formal trial.

The literature on economic evaluation of neonatal screening was also reviewed. An economic model of tandem mass-spectrometry based screening was constructed, taking into account additional treatment costs as well as the marginal costs of changing screening technique.

Overall conclusions

The current national screening programmes for phenylketonuria and congenital hypothyroidism:

- have largely fulfilled their objectives in that mental retardation due to these conditions is now very rare
- are cost-effective
- would benefit from a broader overview at national level

Cystic fibrosis: Screening should be encouraged

Galactosaemia: Dedicated screening should be abandoned

Duchenne muscular dystrophy: The Welsh screening study should be continued until more data on family well-being and reproductive behaviour have been collected

Congenital adrenal hyperplasia: Literature inconclusive

Biotinidase deficiency: There would be a good case for screening except for the low incidence (high cost/case detected)

Replacing current methods of screening for phenylketonuria by tandem mass spectrometry would be both practicable and cost effective:

- some 17 additional diseases would be detected
- approximately 1 additional diagnosis per 5,000 babies screened
- high specificity, though for a few diseases sensitivity is low
- additional cost per baby screened ranging from £0.60 to £1.40 depending on service configuration, giving for medium-chain acyl-CoA dehydrogenase deficiency a cost per QALY in the rage £179 to £415

HOWEVER - Technical and organisational change on this scale is best tested by a large-scale pilot study to:

- provide a technical appraisal of large-scale screening by tandem mass-spectrometry, building on the work currently being performed in the UK on a research basis.
- agree minimum criteria for follow-up and definitive diagnosis for each disease.
- produce generic information pamphlets for parents and professionals, and immediate advice pamphlets for use with newly-diagnosed patients and assess their effectiveness.
- establish registration of screening-diagnosed and screening-missed cases in order to determine sensitivity and effect on outcome.
- report on general practicability, including organisational and staffing requirements, and costs

A clear lead on this a matter of some urgency or the débacle surrounding the introduction of screening for phenylketonuria is likely to repeat itself.

Neonatal screening for inborn errors of metabolism: a systematic review.

Topic of Review: Neonatal screening for inborn errors of metabolism: a systematic review.

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Funded by: Research & Development Directorate, NHS Executive, Department of Health

Start and Finish Dates: October 1995 - May 1997

Objectives To establish a database of literature and other evidence on neonatal screening programmes and technologies for inborn errors of metabolism: To undertake a systematic review of the data as a basis for evaluation of newborn screening for inborn errors of metabolism: To prepare an objective summary of the evidence on the appropriateness and need

for various existing and possible neonatal screening programmes for inborn errors of metabolism in relation to the natural history of these diseases. To identify gaps in existing knowledge and make recommendations for required primary research. To make recommendations for the future development and organisation of neonatal screening for inborn errors of metabolism in the UK.

How the research was conducted

There were three parts to the research:

A systematic review of the literature on inborn errors of metabolism, neonatal screening programmes, new technologies for screening and economic factors. Inclusion and exclusion criteria were applied, and a working database of relevant papers established. All selected papers were read by two or three experts and were critically appraised using a standard format. Seven criteria for a screening programme, based on the principles formulated by Wilson and Jungner (World Health Organisation, 1968) were used to summarise the evidence. These were:

- Clinically and biochemically well defined disorder
- Known incidence in populations relevant to the UK
- Disorder associated with significant morbidity or mortality
- Effective treatment available
- Period before onset during which intervention improves outcome
- Ethical, safe, simple and robust screening test
- Cost effectiveness of screening

Methods: A questionnaire which was sent to all newborn screening laboratories in the UK. Site visits to assess new methodologies for newborn screening. The classical definition of an inborn error of metabolism was used (i.e. a monogenic disease resulting in deficient activity in a single enzyme in a pathway of intermediary metabolism).

Research findings

- Inborn errors of metabolism
- Phenylketonuria (PKU) (incidence 1:12,000) fulfilled all the screening criteria and could be used as the 'gold standard' against which to review other disorders despite significant variation in methodologies, sample collection and timing of screening and inadequacies in the infrastructure for notification and continued care of identified patients.
- Of the many disorders of organic acid and fatty acid metabolism, a case can only be made for the introduction of newborn screening for glutaric aciduria type 1 (estimated incidence 1:40,000) and medium-chain acyl CoA dehydrogenase (MCAD) deficiency (estimated incidence 1:8,000 - 1:15,000). Therapeutic advances for glutaric aciduria type 1 offer prevention of neurological damage but further investigation is required into the costs and benefits of screening for this disorder. MCAD deficiency is simply and cheaply treatable, preventing possible early death and neurological handicap. Neonatal screening for these diseases is dependent upon the introduction of tandem mass spectrometry. This screening could however also simultaneously detect some other commonly-encountered disorders of organic acid metabolism with a collective incidence of 1:15,000.
- Neonatal screening for congenital adrenal hyperplasia (CAH) due to 21-hydroxylase deficiency (incidence 1:17,000) has been shown to be beneficial in other countries and similar benefits should accrue in the UK. A national programme of neonatal screening for CAH would be justified, with re-assessment after an agreed period.
- Biotinidase deficiency is of low incidence in the UK (estimated 1:100,000), but this may be outweighed by the simplicity of the screening methodology and the benefits in prevention of serious neurological disease in patients with profound biotinidase deficiency. This question requires further investigation

and a national neonatal screening programme would be justified, with reassessment after an agreed period.

- Neonatal screening for galactosaemia (incidence 1:44,000) has been based upon prevention of neonatal mortality. However, evidence suggests that, despite early treatment, long-term outcome is poor with neurological dysfunction and a high incidence of ovarian failure in females. The accepted criteria are not currently met by galactosaemia and newborn screening is not justified.

- The accepted criteria for a neonatal screening programme are not currently met by non-PKU amino acidopathies (including tyrosinaemia type 1, homocystinuria and maple syrup urine disease), familial hypercholesterolaemia (FH), peroxisomal disorders, urea cycle defects, trace metal disorders, purine or pyrimidine disorders, or lysosomal disorders.

Screening technologies

- Automation of all or parts of the screening process is technically possible but some current methodologies are not amenable to automation. Fully automated neonatal screening utilising time-resolved fluorescence is currently being developed.

- Current molecular (DNA) techniques do not permit the simultaneous screening of large numbers of mutations and can be very expensive. At present there is no indication for newborn screening for inborn errors of metabolism using these techniques.

- Tandem mass spectrometry (tandem MS) can be considered as the most important of the new technologies for newborn screening for inborn errors of metabolism. It has the potential for simultaneous multi-disease screening for selected disorders of amino acid and organic acid metabolism using a single analytical technique and is complementary to immunoassay-based methods for congenital hypothyroidism (CH) and CAH screening. The technology has been demonstrated to be robust (accurate, sensitive, lack of false positives) and suitable for the reliable detection of PKU and some other inborn errors of metabolism. However, introduction of new technologies for neonatal screening must be determined by the perception and evidence for the need for screening for each disorder or group of related disorders and by the need for the new technology in existing programmes. Of those disorders detectable by tandem MS in addition to PKU, evidence has identified only glutaric aciduria type 1 and MCAD deficiency as disorders for which a case for newborn screening can be made. Further, evidence for the utility of tandem MS in prospective neonatal screening for inborn errors of metabolism has come from only one source, based on relatively small numbers screened. Thus this technology requires further evaluation through primary research in the UK with prospective screening of more than 1,000,000 neonates for PKU, glutaric aciduria type 1, MCAD deficiency (and possibly other selected disorders) in order to fully validate the utility of tandem MS for newborn screening for inborn errors of metabolism.

Economic evidence

- PKU screening provides a positive net monetary benefit to society and justifies the collection of blood samples from neonates. There is insufficient economic evidence to support a change from current methodology to tandem MS-based screening solely for PKU. More information is needed on the cost-effectiveness of extending screening to other disorders. There is insufficient evidence to assess the economic value of screening for any other inborn errors of metabolism.

Conclusions and recommendations

- Universal neonatal screening for PKU is worthwhile and should be continued. Cost-benefit analyses show that

screening for PKU by itself justifies the collection and testing of neonatal blood spots.

- If the neonatal screening programme is to be expanded a clinical and supportive infrastructure for paediatric metabolism urgently needs to be established to provide adequate treatment and care for identified patients and their families.

- Programmes for the screening of profound biotinidase deficiency and CAH should be introduced with structured, coordinated, on-going evaluation to ensure that these programmes are cost-effective, with review after 5 years.

- Screening for MCAD deficiency should be seriously considered for inclusion in newborn screening programmes. Similarly, a case can be made for the introduction of newborn screening for glutaric aciduria type 1. The clinical effectiveness and cost-effectiveness of such screening would need to be carefully monitored, with review after 10 years. Such screening is dependent upon the introduction of tandem MS technology into newborn screening programmes. Tandem MS could simultaneously detect other selected disorders.

- There is however insufficient evidence at present for the widespread introduction of tandem MS technology into newborn screening programmes in the UK. Tandem MS for newborn screening for PKU, MCAD deficiency and glutaric aciduria type 1 should be further evaluated by primary research conducted over 5 years with a defined timetable and external and independent statistical, health economic and scientific monitoring and evaluation of the technology and programmes. This research should be conducted at four selected centres that have been identified to have the required infrastructure and appropriate expertise. During this primary research, and until reports are presented and decisions made, there should be an embargo on the introduction of tandem MS technology into newborn screening laboratories in the UK.

- There is no evidence to support a newborn screening programme for galactosaemia and any current newborn screening for galactosaemia should be discontinued.

- Screening for other inborn errors of metabolism is not warranted at this time.

- Technologies for fully automated immunoassay-based screening are not yet sufficiently developed. The benefits from a fully automated neonatal screening system remain to be demonstrated. These benefits will probably only be achieved if the range of tests is expanded from CH (and PKU) alone and this will in turn depend upon decisions about other diseases to which newborn screening should be extended.

- At present there is no indication for newborn screening using molecular techniques.

Major research areas requiring investment:

1. Pilot studies on tandem MS in neonatal screening for inborn errors of metabolism, specifically PKU, MCAD deficiency, glutaric aciduria type 1 and possibly some other disorders of organic acid and fatty acid metabolism, together with the infrastructure for the essential external and independent statistical, health economic and scientific monitoring and evaluation of the technology and programmes. Minimum of 3 year funding for the pilot studies and 3-5 years of funding for the monitoring and evaluation team.

2. Initiation of nationwide screening for CAH and biotinidase deficiency, together with infrastructure for external and independent monitoring and review. Initiation funding plus five years of funding for the monitoring and evaluation team.

Details of the review and/or published papers:

Thomason MJ, Lord J, Chalmers RA et al. Neonatal screening for inborn errors of metabolism: a systematic review. Health Technol Assessment 1997; 1 (11) [ISSN 1336-5278]

[Available from: The National Coordinating Centre for Health Technology

Assessment, Boldrewood, University of Southampton, Highfield, Southampton, SO16 7PX]
Executive Summary: http://www.soton.ac.uk/~hta/pubs.htm Crown Copyright

Secondary analysis of economic evaluations: A case study using neonatal screening for PKU. J.Lord, M.J.Thomason, P.A.Littlejohns, R.A.Chalmers, M.D.Bain, G.M.Addison, A.H.Wilcox and C.A.Seymour. Proc. 2nd International Conference on the Scientific Basis of Health Services, Amsterdam, October 5-8, 1997.

Systematic review of neonatal screening for inborn errors of metabolism: Evidence on the cost-effectiveness of existing programmes and new technologies. J. Lord, M.J. Thomason, G.M. Addison, M.D. Bain, R.A. Chalmers, F.Cockburn, P.Littlejohns, C.A. Seymour, A.H.Wilcox, Proc. European Public Health Association Annual Meeting, London, December 12-14, 1966

A systematic review of neonatal screening for inborn errors of metabolism. M.J. Thomason, G.M. Addison, M.D. Bain, R.A. Chalmers, F. Cockburn, P. Littlejohns, J.Lord, C.A. Seymour, A.H. Wilcox, Proc. 3rd Meeting of the International Society for Neonatal Screening, Boston, October 21-24, 1966, P175

Neonatal screening programme: an audit of phenylketonuria and hypothyroidism screening in England and Wales.

Streetly A, Corbett V.

Main issues identified and assessed:

Development of standards

- Identifying Organisation of the programme in England and Wales
- how a identifying and recorded coverage in a sample of the areas
- Investigation of the initial management of the screen positive cases of congenital hypothyroidism
- findings:
- Standards were broadly acceptable
- Organisational arrangements are in need of review with responsibility and accountability for the programme often unclear
- Coverage can be monitored locally using child health population registers
- Management of screen positive cases is very variable.

Recommendations:

- Develop and disseminate up to date national policy
- Developing a quality assurance programme which should include formalising co-ordination, arrangements for monitoring and reporting on the operation of the programme and maintaining and developing adequate systems to meet data requirements.
- Review of treatment services to support screening programmes including its future development in sure that they are adequate.
- Identify funding to ensure that recommendations proposed for registers, quality assurance research are achieved.

Main research areas requiring investment:

- Overview of the programme incorporating the findings of all recently commissioned health technology assessments to determine priorities for development of the programme and for further funding of research questions
- Clarification on of the research questions to be answered by existing registers what screen positive cases of congenital thyroid and P and new registers proposed for new programmes.

Published papers: Corbett V. Bedford H, Gatford A, Addison M, Streetly A. Heel prick screening of neonates: the role of the health visitor. Health Visitor 1997; 17: 428-30.

Report: Streetly A, Corbett V. The national newborn screening programme: an audit of PKU and hypothyroidism screening in England and Wales. London: Faculty of Public Health Medicine, 1998.

Copies are available from: Dr A. Streetly, Department of public health medicine, Bexleyheath & Greenwich Health Authority, 221 Erith Rd., Bexleyheath, Kent DA7 6HZ.

Other steering group members: Addison M, Bedford H, Bickler G, Chapple J, Davies S, Garvie D, Gatford A, Macfaul R, McCormick C, Nicoll A, Pollitt R, Ross E, Taylor B.

Biochemical screening: commentary

Screening programmes for phenylketonuria and hypothyroidism have been success stories. New screening procedures considered in these two reviews have great promise. The two reviewers came to differing conclusions regarding the target disorders, largely because they differed in their interpretation of the classic screening criteria. Both groups supported TMS screening for glutaric aciduria type 1 and medium-chain acyl CoA dehydrogenase deficiency (MCAD) and both agreed that galactosaemia screening is not justified. TMS could identify a number of other conditions but the groups differed as to how many should be included in the TMS programme.

The St George's Group supported a trial of screening for congenital adrenal hyperplasia (a project also supported by the British Society for Endocrinology and Diabetes). Both groups noted the simplicity of screening for biotinidase deficiency but both were concerned about the high cost per case detected. The issue of screening for cystic fibrosis was discussed and subsequently two reviews have appeared - this is a controversial topic and a national policy needs much further debate.

(See: Desai M, Weller P. Impact and future of screening for cystic fibrosis. Current Paediatrics 1997; 7: 184-6: Wald NJ, Morris JK. Neonatal screening for cystic fibrosis. Brit Med J 1998; 316: 404-5).

The new method of TMS is an exciting technology, with considerable potential. Other biochemical screening methods would be needed for the detection of some conditions - for example, congenital adrenal hyperplasia and cystic fibrosis. Decisions about screening for these should be separated from the issue of TMS.

Opportunities, challenges and hazards

Too many sites? A strategic overview is important so that decisions can be made about which laboratories should undertake TMS screening. We need 3 to 5 trial sites to gain experience of the method and the benefits. There may be important economies of scale if the service is planned carefully and centralised, so that the potential cost savings are not wasted by the development of too many different services. This will be a sensitive issue because some labs clearly would be losers if a highly centralised TMS service becomes the norm.

One way of controlling the process is by inviting tenders to be pilot sites for an evaluation and supplying funds, and to embargo screening by TMS outside those sites; but this control mechanism may be difficult to sustain.

Need for computerised data handling Re-organisation under the new White Paper may present hazards to computerised data storage and monitoring of screening programmes. It is essential that a minimum data-set is agreed and collected for biochemical screening and that centralised monitoring and quality control continue and improve.

Ethical issues Implementation of TMS for conditions where screening has already been shown to be worthwhile, or where the main constraint has been cost, is a service development and should not require Ethics Committee review. However, trials of TMS screening for conditions where screening has not yet become established (for example, biliary atresia or peroxisomal disorders) will be regarded by some ethics committees as research rather than service development. The NSC might set out certain criteria by which a TMS programme could be judged to be safe and ethical - these would include a whole range of quality control procedures. National fora would be the ideal solution but if these prove unacceptable, then Local and Multi-centre research ethics committees might be provided with information which could be useful when applications are made for new TMS schemes.

Need for testing both by standard methods and by TMS initially? During the change-over phase it may be necessary to analyse samples both by the existing method and by TMS but this is unlikely to present major financial or practical problems.

Recommendations

The main issue in TMS screening is how proliferation of this technology can be controlled so that adequate evaluation can be undertaken and planning can ensure the optimal siting of the service for the maximum efficiency and economy. This requires a centrally controlled implementation strategy. There are also issues about which conditions should be included.

- Voluntary groups with an interest in metabolic diseases should be invited to help in planning the information for parents about these new procedures.

- The relevant professional societies should have the opportunity to comment on the reports and debate the issues raised in the reports, since TMS will affect their members' pattern of work

- A central group reporting to the NSC is needed both to oversee the implementation of TMS technology and to encourage and respond to professional debate about screening for the other conditions mentioned - in particular, cystic fibrosis and congenital adrenal hyperplasia.

- A professional group (perhaps including the relevant Royal Colleges and professional societies) should consider what changes and developments in supporting services may be needed.. There must be broad professional and parental ownership of any new screening programme, to ensure that all clinicians and laboratory staff, and the parents of babies identified in the screening process, are committed to the success of the scheme and to gaining the maximum research and development opportunities from the programme.

- Mechanisms for quality control, monitoring of coverage and timeliness etc. will be needed and must be in place soon. Experience already gained in biochemical screening is clearly relevant here.

The need for surveillance, monitoring and a Register. There is also a need to monitor the changes in the epidemiology of inherited metabolic disease which may result from screening, and to develop a register Complete ascertainment of the inherited metabolic diseases for which screening is offered is a desirable goal. This needs surveillance and monitoring schemes, so that it is possible to trace screening failures and ascertain why the case was missed. There is an urgent need to establish a baseline register of children with metabolic disorders. The Research Trust for Metabolic Diseases for Children (RTMDC) and the National Reye's Syndrome Foundation of the UK would encourage such an initiative.

REVIEWS OF WORK IN PROGRESS

Screening for congenital dislocation of the hip: a cost-effectiveness analysis

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Start and finish dates 1997 - 1998

Summary A national policy of universal neonatal screening for congenital dislocation of the hip (CDH) was formally introduced in 1969 and the current recommendations, revised in 1986, advise that all newborns be screened by a clinical examination to detect hip instability on three occasions during the first 6 weeks of life, and that infants with recognised risk factors for CDH be identified and followed clinically until walking normally. This policy was not evaluated formally before its introduction.

The effectiveness of the current programme is being increasingly questioned but a national picture was lacking as outcome is not routinely monitored. Evaluation is complicated because of problems in case definition, uncertainty regarding the natural history of CDH, lack of a 'gold standard' with which to judge the performance of screening tests, as well as uncertainty about the effectiveness of non-surgical treatment.

Work leading up to the current project

A multidisciplinary MRC Working Party is reviewing current and alternative screening policies on behalf of the Department of Health. As part of this review, research has been undertaken to identify current screening and management practices nationally, to obtain

nationally representative estimates of the prevalence of non surgical and surgical treatment for CDH in the UK and to identify the most cost-effective screening strategy through reviewing existing data and, if appropriate, a research agenda which will assist policy decisions.

Work undertaken to date for this review has shown that:

- Ultrasound imaging of newborn hips is practised in more than two thirds of maternity units nationally, representing a five fold increase over the preceding decade.
- Ultrasound is not however used as a primary screening test but to assess infants with clinically detected hip instability or those with risk factors for CDH.
- There is wide variation nationally in screening and management practices, reflecting clinical uncertainty.
- In the UK, the prevalence of a first surgical procedure for CDH in children aged < 5 remains high despite screening and is comparable to that reported before screening was introduced.
- Two thirds of the children requiring surgery were not detected by screening.
- Approximately twice as many infants per 1000 live born are treated non-surgically for hip instability in the UK than would be expected from the reported rate of CDH before screening was introduced. This suggests overtreatment.

Universal imaging of the newborn hip has been introduced as a primary screening test for CDH in some European countries, but its effectiveness in reducing false negative diagnoses is uncertain. Furthermore, as this policy is associated with much higher rates of non-surgical treatment than currently reported for clinical screening (4% vs 0.2% of all live births respectively) and with high rates of follow up for initially ambiguous ultrasound appearances (13% of all live births), the public health implications of such a policy require careful review.

Formal evaluation of current and alternative primary screening strategies is required but the best approach remains unclear. This reflects concerns about the specificity of universal ultrasound screening, the most appropriate outcome measure to assess, the cost-effectiveness implications, and the investment in a trial which would be needed in order to detect a clinically important difference in outcome. The Standing Group on Health Technology Assessment have identified primary screening for CDH as a priority area and a cost-effectiveness analysis has been commissioned to inform the priority for and, if appropriate, the design of, a randomized trial of primary screening methods.

Overview of current project

The cost-effectiveness analysis is being conducted using decision trees representing the different screening strategies for CDH. The current policy of universal clinical screening and risk factor assessment is being compared with alternative strategies of ultrasound used selectively, universal ultrasound screening, and 'no screening'. Data on the pathway probabilities and associated costs are being obtained from a systematic review of published and unpublished data sources, supplemented where required by information from collaborating experts. The analysis is being conducted from a health service perspective and the analytic horizon will extend to postpubertal children to reflect the objectives of screening which are to ensure a functionally and developmentally normal hip joint at the end of the period of growth in childhood and adolescence.

Relevant published papers Dezateux C , Godward S. Screening for congenital dislocation of the hip: in Recent Advances in Paediatrics ed David TJ; London: Churchill Livingstone, 1997: 41-57.
Dezateux C, Godward S. A national survey of screening practices for congenital dislocation of the hip. Archives of Diseases in Childhood 1996;74:445-448
Dezateux C, Godward S. Evaluating the national screening programme for congenital dislocation of the hip. Journal of Medical Screening 1995:2:200-206.

Commentary Work on congenital dislocation of the hip (CDH) continues and for the moment the Health For All Children recommendations (based on those of the expert working party) still stand. The clinical examination is still the only practical means of screening; better training is vital; parents must be told that the examination is not foolproof; alertness must be maintained; and the role of ultrasound is still uncertain. There is currently NO place for primary screening by ultrasound. Screening by ultrasound of cases which have suspect signs or are at risk is a sensible approach at present for

those units not participating in a randomised trial of the issue.

Population Growth Analysis using the Child Health Computing System: a Method of Assessing the Value of Child Growth Surveillance

Hulse J.A., Schilg S., Blount J., Beattie A.M, Cole T.J., Williams S., Hall D.M.B.

Departments of Audit and Paediatrics, Maidstone Hospital & All Saints Hospital Chatham, Kent; Dunn Nutrition Laboratory, Cambridge; Department of Paediatrics, University of Sheffield.

There is increasing agreement that children whose heights are below the 0.4 centile or above the 99.6 centile should be evaluated for the possible presence of a health or growth disorder but there has been less agreement about the value of assessing children whose height velocity might be abnormal (the "channel crossers"), especially when their height is within the normal range.

In an attempt to answer this question, routine growth records from over 4500 children born during 1988-91 in Kent and held on the Child Health Computer System were examined to assess the feasibility of using the CHCS as a method of identifying undiagnosed children in the community with growth disorders and as a possible growth research tool.

After 2% of measurements were excluded as unlikely, comparison with research studies suggested that measurement errors varied from around 1.7 cm at 2-3.5 yrs to 0.5 cm at 5-6 yrs.

Examination of the health records of 59 children with a height recorded as <-2.67 SDs (0.4th centile) revealed that 52% were data errors or probable measurement errors. 34% were having their growth monitored and 6 (11%) had organic diagnoses including 2 with growth hormone deficiency. 16% were of low birth weight compared with 5.6% of those with height >+2.67 SDs (99.6th centile).

The Kent Child Health Growth Surveillance Project

Age Interval (yrs)	Sample Size	Observed correlation	Research correlation	Measurement errors (cms)
2-3.5	686	0.805	0.914	1.7
2-5	215	0.822	0.860	1.1
3.5-5	3443	0.897	0.955	1.3
5-6	155	0.974	0.981	0.5

Conclusions
At present growth data on the CHCS contains too many errors to make it of value for screening. Errors could be reduced by simple checking procedures, especially by plotting on charts at the time of measurement and there is a need to develop systems for quality control of data at all levels. Despite these problems, there is still potential for the use of the CHCS for growth monitoring and research. The data so far supports the value of assessing children with heights below the 0.4th centile.

Commentary Hulse et al found that many children had several growth charts in their community records and incorrect plotting and interpretation were common The problem of growth monitoring is being actively studied and debated in several quarters and the Child Growth Foundation will host a further research seminar in June 1998. No changes are advised at present. Parents have strong views on early detection. Nevertheless, the evidence that growth monitoring works offers value for money or adequate benefits in relation to the number of false positive cases generated is unconvincing. Decisions about this are intertwined with the future role of the school nurse.

If primary care teams seek to be the main providers of medical care for children (in parallel to the primary care paediatrician in Europe and the USA) they must surely accept the responsibility to keep the child's growth chart up to date by measuring the child in the health centre, and not "delegate" this task to the school nurse. Growth monitoring (other than identifying children below the 0.4 centile or above the 99.6) is not likely ever to fulfil the criteria for screening - but measuring a child's height is as much part of clinical assessment as measuring the temperature of an acutely ill child.

INSERT HERE ABSTRACT / NEWSLETTER OF SANDERSON ON PURCHASING OF CHILD HEALTH SURVEILLANCE

This work is not yet complete. In due course it will offer further insights into the costs of routine child health surveillance, though it may

be hard to disentangle these from the other tasks often carried out at the same contact.

Data will shortly be available from Sheffield on the costs of community child health services and the ways in which health visitors use their time.

(See Bowns I, et al. Hitting the Target - a study on the allocation of health visitor resources. Report to the NHSE, 1998: Cotton L, et al. Purchasing of community child health services. Report to the NHSE 1998. Summaries will be available from Prof. Hall - address at front of report).

The school entry medical examination

Jane Barlow, Sarah Stewart-Brown, John Fletcher.

Summary of main issues identified and reviewed.

- Is the doctor component of school entry medical examinations effective and efficient?

- Are selective school entry medical examinations in which children are seen by the school doctor only when there is concern about the child's health, as efficient as routine school entry medicals in which all children are seen by the school doctor?

Findings and recommendations:

A total of 64 studies were identified but only 16 studies met all the inclusion criteria. These comprised: 1 RCT (unpublished); 2 comparative studies (unpublished); 13 prospective and retrospective observational studies (published). There were significant differences in the identification and referral of new and ongoing problems not only between the routine and selective SEMs but also within the two types of SEMs and little overlap of the 95% confidence intervals. There were also large differences in the numbers of children selected for SEMs. None of the studies defined the methods or criteria used to identify screen positive children. None provided follow-up data after referral to estimate the positive predictive value or yield, of follow-up to identify false negatives.

Conclusion: data on the effectiveness and efficiency of routine and selective SEMs are not available at present. The SEM is based on fragile evidence.

Research areas needing investment: a concerted attempt to define the conditions being sought at SEM, to address questions concerning prevalence, natural history and disability; more robust studies on the efficiency of SEMs to identify children with problems.

Commentary The school health service has been conducting school entrant medical examinations since 1908, but we still know little about the yield of new, significant, treatable disorders. This review highlighted the shortcomings of available data. Virtually nothing is known about the benefits of the school nurse review which in many places has displaced examination by a doctor.

The role of the school entrant medical should be reviewed in the context of a re- examination of child health surveillance and this is being planned in conjunction with the Royal College of Paediatrics and Child Health. There are several practical problems:

- The transfer of pre-school child health surveillance to the primary care team is not quite complete, since in some districts much of this work has stayed with the community doctors, usually on the grounds that the standard of general practice locally is said to be low. However, there is not much hard evidence about this.

- So long as there is some form of school entrant screening and so long as school nurses seek to "monitor" children with short stature or medical problems in school, there will be a split of responsibility between the primary care team and the community doctors and nurses. Sooner or later the responsibility to identify problems must be placed unequivocally in the primary care team (excepting of course those for which there is a district wide programme such as biochemical screening and neonatal hearing testing). Similarly it is unequivocally the job of consultants to ensure that the medical advice they give is conveyed to schools and acted upon, for any child where there are implications of the illness or condition for the school staff.

- Most of the identification of problems can be (and often is) done by health visitors and school nurses. If these nurses are integrated

into a primary care team, or a local network of primary care teams, as seems likely under the White Paper proposals, they should be able to fulfil these functions perfectly well. They would however need the support of community based paediatricians and they should have the right to get a medical opinion themselves in cases where the GP is not on the CHS list.

- There are exceptional problems for a small number of GPs working in very difficult circumstances. Perhaps they should be able to contract for extra help to be provided if they cannot provide the general health surveillance required - but the responsibility should remain with them to ensure that the service is provided.

INSERT HERE ABSTRACTS OF ROBINSON ON HEALTH VISITING

Tomorrow's world: complete emotional and social well-being

Sarah Stewart-Brown

The current configuration of child health surveillance and school health services with its emphasis on screening for minor disorders cannot impact on the main health problems of childhood and adolescence and has limited potential for improving children's health. The most important causes of death and disability in childhood are:- behaviour problems in children, mental health problems in adolescents, injuries and social inequalities in health.

The epidemiological picture of both behaviour problems and mental health problems is one that begs a population approach to prevention rather than a high-risk approach.
Parenting programmes appear to have great potential for health gain and a service is currently being provided in the UK by the voluntary sector. The research on these open access parenting programmes provided in the community is observational and qualitative. There have been no controlled trials in the UK.

In contrast there is an adequate evidence base for parenting programmes for clinical populations provided in the context of child psychiatry services and for parenting programmes for high risk populations. School based emotional literacy programmes present a different and potentially complementary approach to the prevention of behaviour and mental health problems. Controlled trials in the USA suggest that some of these programmes are successful, but as yet no systematic reviews have been conducted and no trials have been run in the UK.

There is sufficient research evidence to propose that a child health service based around the promotion of emotional well-being in universal parenting groups and school classrooms with easy access to health information is likely to have a major impact on health and would be affordable. Such a model has the potential to increase the number of young people adopting health enhancing behaviours and to reduce social inequalities in health and childhood injuries.

Major research areas requiring investment: this model needs evaluating in large community intervention trials now.

Additional Reviews

A review of reviews of health promotion in schools:

Dr John Fletcher: MRC Health Services Research Fellow, Health Services Research Unit, University of Oxford. Dr Sarah Stewart-Brown: Director, Health Services Research Unit, University of Oxford. Dr Jane Barlow: MRC Health Services Research Fellow, Health Services Research Unit, University of Oxford

Funded by: NHS R&D Programme (Anglia and Oxford)

Start date September 1995
Finish Date September 1997

Summary of the main issues identified and reviewed School health promotion programmes which have been demonstrated to have an impact on health related behaviour include "innovative approaches" as well as acquisition of knowledge. These innovative approaches cover the development of skills in assertiveness, conflict resolution, relationships and emotional literacy. These approaches seem to have much in common with approaches that have been used in school mental health promotion programmes. The most promising model for health promotion in schools appears to be a core curriculum of emotional literacy and skill development run

throughout the school years in PSE classes combined with information about substance misuse, sex and contraception, healthy eating, exercise, injury prevention and other aspects of health, delivered as part of the national curriculum.

Systematic review of the effectiveness of parent training programmes in improving behaviour problems in children aged 3-10 years.

Dr Jane Barlow MRC Health Services Research Fellow, Health Services Research Unit, University of Oxford.

Funded by: MRC

Start date September 1996
Finish date April 1997

Summary of the main issues identified and reviewed Parent training programmes are effective in reducing conduct disorder in children aged 3-10 years: Group based programmes are more effective than individual programmes and those run in the community appear more effective than those run in clinics. Parent education programmes have been devised from a range of theoretical models Those which have been subject to the most robust evaluation are based on behaviour modification. More trials are needed of programmes based on other models and on programmes which have primary prevention as their aim, but these approaches seem promising.

Commentary The review on health visiting is not yet complete, but it seems likely that it will support the findings of previous reviews. Thus, health visiting probably has benefits on social support, postnatal depression, accident prevention and the primary prevention of child abuse, as well as contributing to the child health surveillance programme. There is an increasing public health role in community development and support for creation of better social networks involving other agencies, both statutory and in the voluntary sector .

The promotion of mental health and emotional well-being is a very ambitious goal but it begins to look attainable and offers potentially great rewards. The current medical pre-occupation with identification of defects should be replaced by a parent supporting and "nurturing" approach, emphasising environmental enrichment and more positive child rearing methods. It is not only parents designated as poor, deprived or inadequate who could benefit from this, though they may need a higher intensity of support.

Input must not be restricted to the pre-school years. Although long term follow-up after early environmental enrichment programmes shows that some improvements persist into adult life, parents who needed help with the rearing of their toddler because they lacked understanding of children's needs or had major problems in their own lives may not find it easy to sustain enhanced educational and emotional support for their child throughout the school years.

Recent research strongly suggests that merely telling children not to smoke, use drugs or engage in casual or unsafe sex will not work - what is needed is an approach which helps children to value themselves and each other, and gives them hope for the future. School-based programmes of mental health promotion ("emotional literacy") are being investigated in the USA and may contribute to the solution of the apparently intractable problems now facing schools and parents, particularly in areas of high unemployment where a sense of hopelessness pervades the current youth culture.

No-one imagines that this on its own will be enough, but perhaps such an approach might help - and it may be a better way of using the skills and experience of our community nursing staff. One implication of this work will be the need to invest in training and support, which in turn implies a need to define the tasks more clearly and to make sure there are people available with the experience and knowledge to provide what is needed.

INTER-RELATIONSHIPS OF PRIMARY PREVENTION AND HEALTH PROMOTION - NEED FOR A NATIONAL CO-ORDINATING GROUP?

Screening programmes in childhood cannot be considered in isolation. It is right that each individual screening test and procedure should be subjected to critical scrutiny, but on its own

this is not enough. Even if a procedure is accepted as useful and valid, it is vital to consider how it would integrate with other aspects of the programme.

There have been several examples during the course of these reviews. For example, discontinuing the health visitor distraction test removes the primary reason why eight months was selected as a sensible age to review a baby's progress. Discontinuing school entrant medical exams altogether would place a greater onus of responsibility on the primary health care team in general and the GP in particular. Stopping vision screening and growth monitoring in schools would radically alter the role of the school nurse.

Although the evidence base varies in its quality for each of the activities and procedures set out in this report, there are sufficient data on which to base a programme of child health promotion and surveillance. The clinical programme must be tied in closely with public health initiatives designed to improve community facilities and support for parents. In turn, this whole exercise must be related to an information system which collects data that are really useful, both to the clinicians for monitoring their own performance and goals, and to those who have a responsibility for the public health perspective.

Those who have particular expertise in the evaluation of screening programmes have a vital contribution to make whenever a new or existing programme comes under scrutiny. However, the separate task of bringing all this material together and advising on the implications for policy requires a much broader church.

There is a need for a group with a composition similar to that of the Joint Working Parties on Child Health Surveillance, who would be able to perform this task. A case could be made for such a group becoming a standing committee, rather than a working party, since the pace of change and the accumulation of new evidence is so rapid that it may no longer be sufficient to re-convene every three or four years.

The funding, support, relationships and loyalties of such a committee would need much further thought and debate. There is however support for it at a number of levels and the proposal in principle has been welcomed by the Executive Committee of the Royal College of Paediatrics and Child Health.

There is little doubt that one of the implications of the kind of approach being proposed is a need for individuals at district level who can take responsibility for delivering these proposals. This in turn implies a knowledge of paediatrics, nursing, social sciences, and public health skills. This is a tall order, but it may well be achievable by closer collaboration between the professional bodies concerned, both in training a new generation of people with the requisite combinations of skills, and in the shorter term, ensuring that there is better and closer collaboration.

Proposal A national forum is needed to monitor the evidence as it emerges and ensure that when one change is proposed the effects on other areas of child health are taken into account before recommending changes. This might take the form of a national Committee on the Public Health of Childhood. It would need representation from the key professional and voluntary organisations, and a clearly defined relationship to the Department of Health and NHSE.

APPENDIX 1:

These abstracts refer either to work in progress or to material not discussed at the meetings in London and are included for those readers who are interested in other areas of screening relevant to child health.

COST-EFFECTIVENESS OF ANTENATAL HIV SCREENING PROGRAMMES

Gibb DM1, Sculpher MJ2, Peckham CS1, Ades AE1

1Department of Epidemiology and Biostatistics, Institute of Child Health; 2Health Economic Research Group, Brunel University

The case for antenatal testing for HIV is well-established as anti-retroviral treatment and counseling against breast-feeding are of proven efficacy in limiting mother-to-child transmission. In the UK, where most infected women breastfeed, screening would be expected to reduce the probability of transmission from over 25% to 5%. This two year project funded by the Department of Health is intended to establish the most cost-effective strategy for delivering an antenatal screening service. The final report is now being completed, but several components have already been published.

The project includes two data collection exercises: a telephone survey of London Maternity units,1 and a study of the uptake of antenatal HIV screening in 6 inner London units all claiming to offer HIV testing to all women.2 These studies have highlighted the inadequacies in staff training and in the information available to women on the advantages of being tested for HIV. Most strikingly, they have shown that even in maternity units with the highest prevalence of undetected HIV, as determined by anonymous testing, uptake of screening was between 3% and 50%.

An outline of the economic analysis has been published.3 It focuses on the relative cost-effectiveness of a range of selective screening strategies based on groups considered at risk and of universal screening, carried out in regions of the UK that have different prevalences of unrecognized HIV among the antenatal population, and different risk group compositions. An epidemiological model of British regions, defining the size of potential target groups for screening and the prevalence of HIV within them is being designed. This is used to calculate the costs and effects of each screening strategy implemented with given uptake rates in any region. A second component of the analysis is a 'risk reduction' model,4 which estimates the effect on mother-to-child transmission rates of different combinations of bottle-feeding, caesarean section, and anti-retroviral treatment. The uptake of each of these interventions, and the uptake of TOP among infected women aware of their infection status was estimated from obstetric and paediatric surveillance data.5 Finally, in order to assess the costs of paediatric infection and the benefits of averting it, it is necessary to distinguish between children followed from birth and those presenting later with HIV-related symptoms or when another family member is diagnosed. In order to take account of rapid changes in anti-retroviral treatments, a modeling approach was adopted6

The screening strategy, epidemiological, risk reduction and paediatric components are being drawn together in an overall analysis that will assess the incremental cost-effectiveness of different screening strategies per additional life-year gained. The economic and health gain implications of screening for the mother herself will also be taken into account.

1. MacDonagh S, Helps BA, Masters J, et al. Descriptive Survey of antenatal HIV testing in London. BMJ 1996; 313: 523-3.
2. Gibb DM, MacDonagh S, Gupta R, et al. Factor affecting uptake of HIV testing in London: results of a multi-centre study. BMJ in press.
3. Briggs A, Gibb DM, Sculpher M, et al. Antenatal screening for HIV: developing a model for economic evaluation. 1996. HERG Discussion paper 14, Brunel University.
4. Ratcliffe J, Ades AE, Sculpher MJ, et al. Prevention of mother-to-child transmission of HIV-1 infection: alternative strategies and their cost-effectiveness. Submitted.
5. Gibb DM, MacDonagh S, Tookey P, et al. Uptake of interventions to reduce mother-to-child transmision of HIV in UK and Eire. AIDS 1997; 11:F53-F58.
6. Sculpher M, Gibb DM, Ratcliffe J, et al. Modelling the costs of paediatric HIV infection and AIDS: comparison of infected children born to screened and unscreened mothers. HERG Research Report 23, Brunel University

PREVENTION OF TRANSMISSION OF HIV INFECTION FROM MOTHER TO CHILD

Graham Davies

(Opinions expressed those of the author)

There is now clear evidence that transmission of HIV infection from an infected mother to her child can be greatly reduced by the use of antiretroviral therapy and by the avoidance of breast feeding. The key to providing these preventative measures lies in being able to diagnose infection in the mother before or during pregnancy. Currently in the UK over three-quarters of HIV infected pregnant women remain undiagnosed at the time of delivery. Diagnosis of HIV infection in most affected families only occurs when the child develops AIDS or other symptoms of infection.

This situation is unacceptable and urgent action is required. The following relevant issues need to be considered:

- The provision of appropriate information on F{[V and its transmission from mother to child to all pregnant women and the making of HIV testing easily accessible to them.
- The need to offer and recommend antenatal HIV testing to all women in areas of high seroprevalence such as Greater London (where currently I in 520 infants are born to HIV positive mothers).
- The normalisation of the HIV test so that it becomes integrated as part of the routine testing for other conditions in pregnancy.
- The need to ensure that testing for any condition in pregnancy is done with the woman's knowledge and consent.
- The provision of support, counselling and specialist medical care for mother and baby when HIV infection is diagnosed in pregnancy.
- The monitoring of rates of HIV diagnosis in pregnancy as a marker of good practice in antenatal care.

An Intercollegiate Working Party has been addressing these issues and will report early in 1998.

SCREENING FOR HAEMOGLOBINOPATHIES IN THE UK: SYSTEMATIC REVIEW AND ECONOMIC ANALYSIS.

Zeuner D1, Ades, AE1, Karnon K2, Brown J2, Dezateux C1, Anionwu E1.

1Department of Epidemiology and Biostatistics, Institute of Child Health; and 2Health Economic Research Group, Brunel University.

This report is currently being reviewed by the NHS Executive HTA Programme.

Antenatal haemoglobinopathy screening is conducted in order to offer reproductive choice to women with affected fetuses, while neonatal screening is carried out to identify and provide early prophylactic care to infants with sickle cell disease. The objective of the study was to determine whether these screening programmes should be offered to all (universal) or offered only to non-North European women and their children (selective). Universal screening would be indicated if either the fetal prevalence of sickle cell disease among North European women was in itself sufficiently high to warrant screening them, or if the process of selection lead to a high proportion of affected non-North Europeans being missed.

A computer model of the population to be screened and the screening process was designed. Given an antenatal population with any specified ethnic composition, the model first calculates the expected fetal prevalence of haemoglobinopathies from assumptions about ethnic group specific haemoglobinopathy carrier frequencies and inter-ethnic mixing. The model then calculates the expected numbers of women with affected fetuses who are able to exercise choice over the outcome of pregnancy, the number of affected live births, and the costs of the screening program, under a range of conditions. Parameter values relating to coverage of screening, uptake of partner testing, uptake of prenatal diagnosis, uptake of TOP, inter alia, were determined from literature review supported by expert opinion.

The main outcome in the analysis of antenatal screening was the number of women with affected fetuses offered choice, and for neonatal screening it was the number of late diagnoses of sickle cell disease avoided by the combined antenatal and neonatal programmes. An incremental cost-effectiveness analysis was performed, which considered the additional costs incurred per additional choice offered or late diagnosis prevented, on switching from a selective to a universal policy. Costs and effects of other antenatal and neonatal screening programmes, litigation costs in the UK arising from failure to offer choice, and life-years gained, were used to derive a range of amounts of money it would be worth paying to offer one

additional choice or prevent one late diagnosis. The analysis was applied to each HA in Great Britain, and was used to determine in which HAs universal antenatal and/or neonatal screening should be carried out, given the stated economic criteria.

Selective antenatal screening is cost effective in any district compared to no screening, whether on the basis of choices offered to women with affected fetuses, or affected live births prevented. Similarly, selective neonatal screening based on Guthrie cards is cost-effective in comparison to no neonatal screening. On current evidence, the fetal prevalence of SCD in North Europeans is too low, in itself, to justify universal antenatal or neonatal screening. Instead, the difference between universal and selective screening programmes in the rate at which they fail to screen non-North Europeans is the critical factor. The higher the fetal prevalence of haemoglobinopathies, the higher will be the uptake of screening in ethnic minorities that would be required of a selective strategy, in order to avoid having to adopt universal screening.

See also: Modell B et al. Audit of prenatal diagnosis for haemoglobin disorders in the UK: the first 20 years. Brit Med J 1997;315:779-84.

Neuenschwander H, Modell B. Audit of process of antenatal screening for sickle cell disorders at a north London hospital. Brit Med J 1997;315:784-5.

Antenatal Screening for Syphilis

Preamble Serological screening for syphilis is undertaken in the UK (1997) among pregnant women (routine), blood donors (mandatory), other donors of tissue body fluids (should be mandatory) and STD clinic attenders (routine). Currently most people being screened are unaware that they are being tested.

The Disease Infectious syphilis and congenital syphilis are important health problems. Infectious syphilis is difficult to diagnose because of its multiple presentations. Its importance stems from its severe lifetime sequelae for the infected individual. Because it is highly infectious it is also a threat to sex partners and the unborn foetus. Congenital syphilis threatens the viability of the foetus which maybe stillborn or emerge congenitally damaged. The natural history of the conditions are well known. Primary prevention for infectious syphilis is relatively unsuccessful as has been shown be recent local outbreak of syphilis in Bristol (41 persons, including cases among pregnant women).

Epidemiology and Prevalence Syphilis is endemic in most developing countries. An epidemic of syphilis occurred in the USA in the late 1980s and early 1990s. Epidemics are occurring currently in a number of states of the former USSR. In 1995 there were 96 cases of infectious syphilis in women diagnosed at STD clinics in England. In a three year British Co-operative Clinical Group survey (1994-7) of pregnant women treated by STD clinicians in the UK there were 31 new cases of infectious or probably syphilis identified. An incidence of 1.38 per 100,000 pregnancies. Twenty seven of the cases would seemingly only have been detected through antenatal syphilis. These are minimum estimates as cases may also be treated by obstetricians without referral. Risk factors in the 31 cases included being born abroad, living in London and being of an ethnic minority group. However, cases also occurred among white women outside of London born in the UK. A parallel British Paediatric Surveillance Unit survey found only nine presumptive and no definitive cases of congenital syphilis being seen by paediatricians suggesting that antenatal screening was successful.

Serologic Testing There is a simple inexpensive serologic screening test with high sensitivity. However, false positives are common due to non-specific cross reactions (biologic false positives) and all positives patients require to be referred for further evaluation, preferably by STD specialists. All positive specimens should be referred to the 6 PHLS syphilis reference laboratories (Birmingham, Bristol, Manchester, Newcastle, St George's (London) and Sheffield). Surveillance through these serological referrals shows some infections acquired in association with time spent in
Russia and Eastern Europe.

Treatment of an infected mother or child is highly effective but uncomfortable as it involves parental injections of penicillin. An agreed proforma for management of maternal syphilis cases identified in pregnancy has been developed for North Thames.

Controlled Trials None have been carried out and they probably would be considered unethical.

Cost Benefit Studies Three studies have been carried out in the UK. These have all concluded that antenatal screening was cost beneficial. The monies saved by stopping testing would be small.

Screening Policy and Practice At present there is a paucity of written policy. A policy options paper is being prepared by the PHLS for the national screening committee. A national survey of policy and practice is being undertaken by the Institute of Child Health (London), the Scottish Centre for Infection and Environmental Health and the PHLS.

Screening Brief prepared by HIV/STD Division, PHLS Communicable Disease Surveillance Centre
61 Colindale Avenue, London NW9 5EQ

For further details and references contact Dr Angus Nicoll, 0181 200 6868 ext:4695, anicoll@phls.co.uk

Fragile -X syndrome.

See: Barnicoat A. Screening for fragile-X syndrome: a model for genetic disorders. Brit Med J 1997;315: 1174-5.

Murray J, Cuckle H, Taylor G, Hewison J. Screening for fragile - X syndrome. Health Technology Assessment 1997; 1 (4): executive summary.
(also available on http://www.soton.ac.uk/~hta/summ104.htm)

Appendix 2
The history of preventive paediatric care in the UK

Interest in the early identification of children with developmental impairments and disabilities dates back to the 1950s, when parents became increasingly dissatisfied with the difficulties they experienced in obtaining help and advice for children who were not developing normally. The skills that were acquired by leading paediatricians in this country and the United States were gradually translated into a range of developmental screening procedures. During the '60s and '70s, accumulating scientific evidence from a range of disciplines strengthened the prevailing idea that early identification of disabling conditions would substantially improve outcome.

In the Western world it became the norm for all children to be offered a series of developmental and physical checks, beginning with the neonate and continuing through into secondary school age. The number of such checks, the conditions which were sought and the methods used, varied widely between and within countries. Nevertheless remarkably few data were available to determine the value of this exercise.

In 1966, Wilson and Jungner published their classic work on the criteria for screening programmes. These had little impact on child health screening however. At the beginning of the 1980s there were still very few studies available on the benefits of screening.

By the beginning of the 1980s, the climate was changing. All screening programmes, along with other health care activities, were coming under more intense scrutiny. It was realised that screening activities which could not meet the Wilson and Jungner criteria were not merely a waste of resources but might even be unethical, as well as presenting increasing legal hazards.

In 1982, the benefits of the available vision screening methods for young children were questioned and three years later the advances in our understanding of developmental paediatrics were reviewed in the context of the child health screening programme.

Since 1986, the pace of change has quickened. More research has been published than ever before. There have been three Joint Working Parties on Child Health Surveillance - I had the privilege of chairing the first and third of these, while the second was chaired by Dr. Colin Waine. Simultaneously with the publication of the report of the first Working Party, Professor John Butler published his own independent review of child health surveillance which still stands as a model of clarity and unbiased scholarship.

The Butler Report together with the three Joint Working Parties have undoubtedly been responsible for a major change in the approach to the routine care and reviewing of apparently healthy children. The approach adopted in the reports ("Health for all children") has been duplicated in Australia, the Netherlands, and Norway; and similar reviews are underway in Italy and Portugal. The Report also attracted some interest in the United States, which has recently launched its own new well child care programme, "Bright Futures", which specifies some 29 routine contacts with each child from birth until the age of 21.

Surveying the literature for the first edition of "Health for all children" was a relatively straightforward task, but the volume of material available has grown exponentially and it has become increasingly difficult to maintain an overview of all the issues, even with the help of a large number of expert advisers. Furthermore, the size of the task made it increasingly difficult to be certain that all points of view were being taken into account. Although the composition of the Working Parties guaranteed lively debate on all the issues, it is virtually impossible to retain total objectivity in such a situation.

The emerging concept of the Systematic Review (SR) has therefore been welcomed. The Systematic Review aims to offer a genuinely objective, unbiased and comprehensive review of the literature on a particular topic. This approach has been particularly powerful when reviewing the impact of a single treatment on a specified condition, a situation which is susceptible to study by the randomised controlled trial. The application of this methodology to more complex issues is still in its infancy, particularly with regard to statistical techniques.

For an overview of the UK programme, its background and a literature review, see:

Hall, DMB (ed.). Health For All Children. Report of the Joint Working Party on Child Health Surveillance. Oxford:OUP, 1996 (3rd edition).

For “Bright Futures” see:
http://www.brightfutures.org.

Appendix 3:
Systematic Reviews - summary of the paper presented by Dr. Stuart Logan, Institute of Child Health, Guilford Street, London WC1.

Reviews of medical literature have been commonplace for many years. They are however subject to bias in a number of ways. A Systematic Review represents an attempt to take a more objective look at available evidence, using a range of techniques to minimise bias.

Systematic Reviews are usually supported by a steering group or committee, who provide background evidence and expertise, and review successive drafts of the report. In most cases however, the review is primarily the work of those commissioned to undertake it and it is they who take responsibility for the findings.

The first stage in undertaking an SR is to define the question precisely. This can be very difficult in some cases. It is important to identify a question that is capable of being answered. Many childhood screening issues are complex and have to be broken down into several stages.

The next step is to set out the criteria by which papers on the subject will or will not be accepted for inclusion. These might for example deal with matters such as sample size, specification of the subjects involved, type of tests used, etc.

The third important element is to search the literature thoroughly and extensively including journals that are not normally regarded as mainstream. There is evidence that much research material remains unpublished. Papers that fail to demonstrate an effect or a difference between two groups are less likely to be published than those that do, thus creating a bias in favour of intervention. Papers that demonstrate only modest effects or no differences are less likely to be published in the English language and are less likely to be presented in mainstream journals.

The next element is the extraction of the relevant material from each of the papers selected. Finally, this is brought together and where appropriate subjected to a statistical analysis, so that the final conclusion draws on evidence from a range of different studies.

The interpretation of these findings is unavoidably more subject to the bias of the reviewer. It is important to set out the reasoning and judgements being made before any recommendations are included.

The Systematic Review should be treated like any other research product. Its conclusions should not be taken as gospel. The methodology should be examined carefully and the justification for the conclusions must be considered.

There are inevitably limitations in the method. Often useful items of information appear in papers that were not selected for the review. To control the size of the task, a number of closely related issues have to be ignored - for example, in the review on speech and language problems, the related questions of children with autism or learning problems were ignored and the educational literature on enrichment of the child's language environment was not examined.

Similar remarks could be made about the inclusion of health economics methods in the reviews. Often the data on which economics studies are based are incomplete or unavailable. It is difficult to put a value on aspects such as how much parents value a programme or the hazards of litigation. It is easy to over-estimate the benefits of a screening programme conducted as a research study - many screening programmes deteriorate in quality when they pass into the service domain. This may lead to over-estimation of the cost benefit ratio.

In summary, the Systematic Review, although a powerful tool which should increase our ability objectively to assess research, is not a complete answer. Ultimately policy decisions have to be made by integrating all the available evidence, incomplete though it may be, and assessing the relative value of different health opportunities, so that in the last analysis the decision is political rather than medical or scientific.