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Preschool hearing, speech, language, and vision screening

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Preschool screening

Child health surveillance is part of a broad set of activities, the objective of which is to reduce childhood disability by identifying and managing a multiplicity of conditions at an early stage.[1] This includes several screening programmes which are focused on the detection of specific disorders.

The value of surveillance and monitoring of child health, growth, and development used to be regarded as self evident. The Hall reports emphasised the importance of applying rigorous criteria for screening programmes in community child health and helped to produce a more coordinated national programme.[2–4] However, there is still considerable variation both within and between health authorities in the content, timing, and delivery of child health surveillance.

This paper summarises the research evidence presented in a recent issue of the *Effective Health Care* bulletin, Vol 4, No 2; April, 1998 about hearing, speech and language, and vision screening and is based on recent systematic reviews commissioned by the National Health Service (NHS) Health Technology Assessment Programme. Details of the methods and the results are available in the full reports.[5–7]

Evaluation of screening

The objective of universal screening in childhood is to identify impairments which are not obvious or apparent, which will cause *considerable* disability or handicap and which are more effectively treated early. Screening does not include situations in which potential problems are noticed and are then referred for detailed evaluations. Because screening uses considerable resources and imposes tests on children who are not ill, and because it has been argued that some screening programmes could be potentially harmful due to the unnecessary worry, referrals, and procedures that may result, there is an ethical responsibility to ensure that screening is only carried out when there is confidence that it will result in more good than harm. "It is unethical to offer screening tests which cannot stand up to critical examination".[4] Several criteria are helpful when considering whether to carry out screening (box).[8 9]

Major criteria for assessing a screening programme

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

Hearing screening

EPIDEMIOLOGY AND NATURAL HISTORY OF CONGENITAL HEARING IMPAIRMENT

There are about 840 children born each year (1.12/1000 live births) in the United Kingdom who have congenitally impaired hearing with a permanent bilateral moderate, severe, or profound hearing impairment of $\geq$ 40 dB in the better ear.[10] Most permanent childhood hearing impairment is sensorineural in type due to lesions in the cochlea or auditory nerve and its central connections (unilateral or bilateral) which does not resolve.

Almost 85% of all permanent childhood hearing impairment will be present at birth with around 160 cases a year being acquired (often after meningitis). The impact of permanent hearing impairment on the children and their families can be considerable. Late identification may compound problems in communication, and language acquisition, and affect other areas of development.

SCREENING TESTS

The most common preschool hearing screening test used in the United Kingdom is the infant distraction test carried out by two health

Table 1 Key screening tests used to detect permanent childhood hearing impairment

Tests	*Comments*
Infant distraction tests (IDTs):	
Traditional health visitor distraction test (HVDT) universal in most districts	Test carried out at 6-9 months, usually in "protected" time. Cost about £25 per test including follow up
Targeted IDT	Proposed in tandem with universal neonatal screening on equity grounds
BeST test	New one person IDT, with calibrated sound source. Not yet available for trials.
Transient evoked otoacoustic emissions (TEOAE)	Quick test carried out within days of birth. Measures acoustic energy generated by the healthy cochlea in response to wide band clicks with a lightweight ear canal probe. Cost £14 per test. Presently most used for well babies. Need agreed criteria for pass or refer
MLS TEOAE	New very quick version of TEOAE that may have advantages in noisy situations. Not yet available for trials
Distortion product otoacoustic emissions (DPOAE)	Many implementations, need to monitor literature as to outcome
Auditory brainstem response (ABR)	Test carried out within days of birth. Wide band clicks are presented to one ear and the resulting electrical potentials of the early auditory pathways are measured with surface electrodes. Some ABR machines make pass or refer decisions, others need trained operators. High recurrent costs or long test times on some implementations. Presently most used in NICU/SCBU children
Portable auditory response cradle (PARC)	Automated, quick, behavioural test which presents a 70-80 dB SPL high pass noise to one or both of the baby's ears through an earphone or probe. The baby's response is measured by a cradle and associated computer software which compares head turns, and body movements in periods with the sound on and off. An automated decision algorithm is used to pass or refer. Probablygood for severe and profound impairments

MLC = maximum length sequence.

visitors (HVDT), or by a health visitor and a trained assistant (table 1). It is administered at about 6–9 months of age and it assesses the infant's ability to turn and localise a sound source. It is used as a universal hearing screen in about 98% of health districts and achieves coverage of about 90% of all infants but varies by socioeconomic status. There is also variability in the way it is carried out; the sound generators used, the number and level of training of the people doing the testing, and the adequacy of soundproofing of the room. This leads to concerns about the number of children with problems who are not identified during a screen under current arrangements.

The published evidence on test performance from clinic based retrospective studies and case note reviews indicates poor and variable sensitivity and specificity for the HVDT.[5] The cumulative yield is low, being about 50% by 18 months. The average age of confirmation of hearing impairment through the HVDT is between 12 and 20 months, with subsequent median age of hearing aid fitting after HVDT being about 18 months.

Alternatively, several neonatal screening tests that can be applied within the first few days after birth are available (table 1). These methods include the portable auditory response cradle (PARC), the auditory brainstem response (ABR), and the transient evoked otoacoustic emissions (TEOAE). The TEOAE is currently the preferred technique for well babies, and automated ABR for those in neonatal intensive care or special care baby units. Although the PARC had been extensively tested, its implementation has not been as well evaluated in multicentre studies as the TEOAE.

One controlled trial has been carried out which compared 21 000 babies given neonatal screening (TEOAE, with ABR for those failing the first test) with 29 000 babies who received only the HVDT at 6–8 months.[11 12] Interim results show that the neonatal screening test had a specificity of around 98% and gave a yield of 1.1/1000 births by 4 months, which corresponds to the expected prevalence, thus indicating a high sensitivity. The high specificity and sensitivity of the neonatal screen is confirmed by another United Kingdom study.[13–14] The cumulative yield in the HVDT only group was lower at 0.7/1000 by 18 months suggesting that false negatives will emerge later on. Only 0.1 hearing problems per 1000 births were actually detected by the HVDT as most were identified due to parental or professional concern, or passed the HVDT incorrectly. In the neonatal screening group 96% were identified under 9 months compared with around half in the HVDT only group.

Table 2 Key interventions for moderate to profound permanent childhood hearing impairment and ways they will be affected if universal neonatal screening is introduced

Intervention	*Effect of universal neonatal screening*
Family support, advice, and information	Needs to be effective from screen refer and onwards. Requires better multiagency cooperation
Provision of hearing aids	Better early diagnostic testing and aid fitting. Needs evaluations for mild impairments if screen to be extended to this group
Provision of communication support (spoken, or signed, or both)	Earlier support needed
Provision of preschool educational support	Earlier support needed. Different skill mix needed for children in first 18 months
Cochlear implants	Earlier implantation will be possible
Provision of other devices—eg radio aids, tactile aids, other assistive devices	No effect

INTERVENTIONS FOR CONGENITAL HEARING IMPAIRMENT

Interventions include amplification, cochlear implants, or helping the child to learn sign language (table 2). For children with a profound impairment a cochlear implant may enable the auditory neural pathway to be stimulated directly; this is currently being evaluated by a Medical Research Council (MRC) study.

Although there is a growing body of literature on the benefits of early intervention, few studies are of high quality. Three of the 18 studies identified provide reasonable evidence that early intervention is better for language acquisition than late. In a study of 69 children identified by a Colorado neonatal screening programme, those "habilitated" before 3

months of age scored 87% of normal for expressive language, compared with only 66% of those habilitated between 3 and 12 months.[15] Similarly, in the same study, 72 children whose hearing impairments were identified before the age of 6 months were found to have better vocabulary and expressive and receptive language than 78 children whose impairment was identified after 6 months (after having taken into account any differences in non-verbal cognitive skills).[16] In another study, subjective assessments by teachers of speech intelligibility of 153 children (matched for age, sex, age of onset of hearing loss, degree of deafness, and schooling) found that those fitted with hearing aids before 6 months achieved higher scores than any groups of children fitted with hearing aids later in life.[17]

The benefits of early identification in hearing impaired children are supported by other studies which show earlier onset of babbling[18] or better communication skills[19 20] the earlier the children were fitted with hearing aids. One study however, found that the initial benefits of early intervention on receptive language did not persist; however, the number of children in this study who were identified in the first 6 months of life was likely to be few, if any.[21] Overall this research supports the view that these children (particularly those with more severe impairments) have poor outcomes at present compared with children with normal hearing. Earlier identification is associated with better language acquisition and communication. However, the extent to which even better outcomes may be achieved with very early identification is not yet clear although the early results from the research in Colorado point to this being the case.[16]

COST EFFECTIVENESS OF HEARING SCREENING

There is a significant difference in the cost of neonatal and HVDT screening. The cost (including follow up) for universal neonatal screening programmes is about £14 000/1000 births; and for HVDT is about £25 000/1000 children, when done in protected time or on a separate visit.[22] This translates into a "cost per child with a hearing problem identified" of around £17 000 for neonatal screening and £80 000 for HVDT screening. These figures do not take into account any of the benefits to the child of earlier detection and habilitation nor the extra costs of the earlier treatment and educational support which they will receive with neonatal screening. Conversely it does not take into account other health promotion activities which may be undertaken by health visitors at the same contact. However, in most English districts, hearing tests are carried out by health visitors in separate clinics or during protected time.

Speech and language delay

EPIDEMIOLOGY AND NATURAL HISTORY OF SPEECH AND LANGUAGE DELAY

Delay in speech and language is one of the most common neurodevelopmental difficulties in early childhood[23] with a prevalence of around 6% of children. The demand for services, particularly for children under 4 years of age is increasing.[24 25] As the age distribution at which normal children learn to speak is probably represented by a bell shaped curve of prevalence estimates are dependent to a great extent on the cut off used. Few data are available on bilingual or ethnically diverse groups and the association with social class is also unclear.

Spontaneous remission of speech and language delays identified in the preschool period can be high, particularly for children with specific expressive delays, in whom some 60% may resolve without treatment by 3 years of age.[6] The picture for older children is unclear due to a lack of research, but it is evident that if children go on to have difficulties in the first year of primary school they are at risk of experiencing problems throughout their schooling. Also, 41%–75% of children who present with early expressive language delay were found to have reading difficulties at the age of 8.

Risk factors for persistent problems include the initial severity of the delay, the extent to which the difficulties are generalised across speech and language, and the extent to which other cognitive and developmental skills are also delayed. There is reasonable evidence to suggest that speech and language development are affected by how well parents interact verbally with their children and by the general level of stimulation within the home environment. However, it is uncertain whether parental factors can actually create a clinical level of difficulty.

SCREENING TESTS

Several screening measures are used in the United Kingdom (table 3). No randomised controlled trials (RCTs) of screening programmes were identified by the review. Screening test performance varies considerably with sensitivity within the range 17%–100% and specificity in the range 43%–100%. Sensitivity

Table 3 Principal methods of screening for speech and language delay

Level of concern elicited from parent by professional (the parental evaluation of developmental status)
Parent provides information about speech and language milestones and the clinician interprets the results (the early language milestone scale, the clinical linguistic auditory milestone scale)
Parent reports on child's current level of speech and language functioning and the clinician interprets the results (the Minnesota child development inventory, the ward infant language screening test, the language development survey)
Clinician makes a judgement of child's performance based on mixed observation and reported data (The Denver developmental screening test)
Clinician tests child's speech and language performance by means of specific activities such as the: child's response to requests graded in terms of difficulty (eg the Hackney language screening test, the Mayo early language screening test, the Uppsala general language screening) child's capacity to imitate words and sentences (sentence repetition screening test) child's ability to retell stories

Table 4 Key intervention approaches for speech and language delay

Most interventions are primarily behavioural in nature and may be provided by speech and language therapists or specialist teachers; intensively within a specialist unit or less intensively but at regular intervals in a clinical setting, a school, or a daycare setting. The 3 main intervention types are:

Didactic intervention:

The child is given a model of a sound, a word, a communication behaviour, or a syntactic construction and an attempt made to elicit the child's production of that model with positive reinforcement. This approach is usually carried out by the therapist or teacher

Naturalistic intervention:

This approach recreates the environment which is known to optimise the child's language learning opportunities, not through explicit instruction, but by making the stimulus relevant to the child's focus of attention. This approach is aimed at promoting the acquisition and generalisation of functional language and often involves parents as active participants. It can be carried out directly by a therapist or teacher, or indirectly by others in the child's environment

Hybrid intervention:

This approach combines elements of both didactic and naturalistic interventions. It recognises that children with delayed speech and language development may learn language in different ways from one another and from their normal peers and may need to be exposed to a range of different types of environmental modifications

Other intervention approaches include non-directive therapy, auditory training comprehension monitoring, and cognitive therapy

was generally lower than specificity, particularly in the better quality studies, suggesting that it may be easier to indicate those children who are not cases than to be clear about those who are. Few studies have attempted to compare the application of two or more screening tests to a single population or to compare a single screening measure across different populations. It is, therefore, difficult to make a judgement about the relative value of different procedures or to single out any one measure as outperforming the others. In general, however, screens that used parents as informants were as accurate as those that used formal testing procedures.

Most of the screening procedures currently available are applicable after the age of 2 years when the reported accuracy of screening is greater, although work currently in progress is exploring a method of identifying those at risk of subsequent difficulties based on auditory skills at 9 months.[26] Given the variability in the natural history of speech and language delay, and the high level of subsequent spontaneous improvement, particularly in the very early years, the use of a single measure at this stage in a child's development is unlikely to be valuable. Tests which can identify those children who will fail to progress without treatment need to be developed.

INTERVENTIONS FOR SPEECH AND LANGUAGE DELAY

Several types of interventions have been used for helping children with speech and language delays (table 4). Ten RCTs and 12 controlled studies were identified which evaluated treatment, mostly for problems of articulation or phonology and expressive language.[27–45] These studies show that interventions are effective in enhancing speech, expressive language, receptive language, and auditory discrimination relative to untreated controls. The size of the benefits represented progress from the 5th to the 25th percentile on a norm-referenced test. This corresponds to an overall standardised effect size of around 1.0—that is, an increase in the average performance equivalent to 1SD of the distribution of performance scores. These results are supported by data from 26 single case experimental designs which were synthesised separately.[6] No studies specifically compared the effects of different timing of interventions on social and educational outcomes and there are few reliable data with which to identify the best choice for any area of delay.

One of the interesting issues is who most effectively provides the interventions—professionals (speech and language therapists or specialist teachers) or parents and others in the child's environment. Studies have shown comparable results for both in the case of expressive language (effect size of norm-refereced measures was + 0.65 for professionals and +1.08 for parents, and the effect size for criterian-referenced measures was +1.11 for professionals and 1.16 for parents).In speech delay, professionals (effect sizes +0.95 for norm-referenced measures and +1.11 for criterion referenced measures) were more effective than parents (-0.02 and +0.20). (The sores of norm-referenced measures and standardised on a population, whereas criterion referenced measures are effectively sills achieved.) For receptive language the reverse was found—indirect treatment by family and friends was more effective, with an average effect size of 1.43 compared with an average effect size of 0.02 for direct intervention (only seven studies had receptive language outcomes). There is some evidence from the United States which suggests that home based intervention may be more cost effective.[46]

Preschool vision screening

EPIDEMIOLOGY AND NATURAL HISTORY OF ASYMPTOMATIC PROBLEMS OF VISION

The aim of vision screening at the age of 3–4 years is the prevention or reduction of disability due to one or more of the following target conditions: *amblyopia* (reduced visual acuity usually in one eye in the absence of organic disease which cannot be improved by spectacles), *refractive errors*, and the types of *squints* which are unlikely to be detected without screening (phorias and microsquints) and so are cosmetically not obvious.

No studies were found which had the primary aim of establishing the prevalence of visual defects at 3–4 years of age. However, data from studies of primary orthoptic screening programmes for this age group reported a range of yields for the target conditions of 2.4%–6.1%.[47–55]

No studies were found that aimed to document the natural history of these conditions in untreated preschool children. A few studies however, give some information on what would be expected to happen to the vision of children at this age with amblyopia,[56] squints,[57 58] and refractive errors[59] in the absence of intervention. These suggest that

Table 5 Common contents of preschool vision screening

Checking the appearance of the eyes
Cover and uncover test for squint
Ocular movements
Convergence
Prism test (eg 20 dioptre base out prism)
Test of stereoacuity (eg Fisby or Lang stereotest)
Single optotype or linear visual activity test (eg Sheridan Gardiner or Snellen)

mild amblyopia (due to non-cosmetically obvious squints or mild refractive error at 3–4 years) in some children at least may resolve without treatment. However, there are many important gaps in the data.

Twenty one studies were found which aimed to investigate whether various disabilities were associated with any of the three target conditions. Most studies either compared the performance of children with visual defects in tasks such as reading with that of their peers with normal vision, or compared the vision of children with and without disabilities such as dyslexia. The only strong and consistent relation to emerge is that children with myopia perform better than their peers on reading tests.[60–63] Studies that investigated the relation between squints and reading ability produced inconsistent findings.[64–67] However, children with squints have been found to perform less well than their peers without squints in neurodevelopmental tests.[68–70]

Amblyopia in one eye can disrupt depth perception, but the effects that this might have are poorly understood and are currently the subject of debate.[71–73] The only study found which investigated the perceptual difficulties associated with amblyopia in adulthood suggested that amblyopia in one eye had little impact on perception of space or contrast and was unlikely to affect everyday life, although this study was methodologically flawed.[74] No studies have been carried out with a design that is appropriate for establishing a causal link.

Physiological data from animal studies showing that blurred vision at a critical stage of neurological development could result in permanent impairment of the relevant brain functions gave rise to the enthusiasm for early detection of amblyopia. However, the quality of the publications on visual defects and disability, and on the natural history of these conditions in humans is insufficient to know with any certainty what might be expected to happen in an individual child with amblyopia, a non-cosmetically obvious squint, or a refractive error if they were left untreated. One large RCT in Avon comparing vision screening programmes in children under 3 years old should provide useful information on associated disability in older children.[75] There is a very strong professional view however, that amblyopia is disabling and should be treated.

Table 6 Treatments for visual problems identified at preschool screen

Amblyopia:
Intermittent occlusion of the amblyopic eye with a patch
Intermittent squints:
Followed up and may be treated with surgery
Latent squints with hypermetropia (long sighted):
Often spectacle correction only
Microsquints and small latent divergent squints:
Not treated, but small latent convergent squints are often associated with hypermetropia for which spectacle correction is prescribed
Refractive errors:
Left untreated or corrected by spectacles

SCREENING TESTS

The principal tests used in preschool vision screening are visual activity tests which can identify children with amblyopia and significant refractive errors. Tests are also carried out to identify non-cosmetically obvious squints because these may be a cause of amblyopia and also sometimes with the aim of treating them in their own right (table 5). No randomised controlled trials of screening programmes for the 3–4 year age group were identified. One prospective controlled study was found, which compared visual outcomes at the age of 7 years in children who were screened at 3 years of age by orthoptists, general practitioners, or health visitors in Newcastle.[56] Children with straight eyed amblyopia and refractive errors were identified significantly earlier in the orthoptic screening cohort, but there was no difference in the time of identification of squint. Despite the fact that many more children with amblyopia were identified and treated in the orthoptic screening cohort, the prevalence of amblyopia at 7 years of age was the same in all three cohorts.[56] However, this study has certain methodological weaknesses.

Sixteen other studies which aimed to establish the effectiveness of preschool vision screening were either observational or audits (Nolan J, 1996, personal communication, and James J, 1996, personal communication).[47–50 52–55 76–81] Uptake rates for primary orthoptic screening ranged from around 44% to 80%. Vision screening by health visitors, general practitioners, or clinical medical officers, undertaken as part of a routine surveillance contact, had a mean uptake rate of 76.2%. Rates of referral from primary orthoptic screening programmes ranged from 4.1% to 10.6% of the screened population, and from 1.6% to 15.2% in other professional groups.

In five studies of orthoptic screening programmes, the positive predictive value (the % of those referred who are true positives) varied from 47% to 66%.[47 51–53 55] Positive predictive values >90% were achieved when the definition of a positive case was broader.[49 54] In programmes run by health visitors or clinical medical officers the positive predictive value was much more variable, ranging from 14% to 62%.[47 49 51 80] In other words, orthoptists are generally better at identifying problems than doctors or health visitors. A significant number of areas use orthoptists (usually on a separate occasion) to test for visual problems at around the ages of 3–4 years.

INTERVENTIONS FOR VISION PROBLEMS

The treatments for amblyopia include patching the non-amblyopic eye and spectacle correction of associated refractive error possibly combined with surgery to correct squints (table 6). Five prospective RCTs of treatment and six non-randomised controlled trials were found. None

were specifically relevant to this age group and in no study was the treatment compared with an untreated control group and thus the absolute effects of treatment are not known.

Three of the RCTs compared the effect of the CAM (a vision stimulator grating) with conventional orthoptic treatment and showed no significant advantage.[82–84] One small RCT showed that adding the drug levodopa/carbidopa to orthoptic treatment for amblyopia improved visual acuity and contrast sensitivity, but that at 1 month after treatment the intervention group had regressed slightly and the control group had not maintained improvement.[85] This latter finding is supported by a controlled study comparing different occlusion regimes, in which 33% of those with improved acuity after treatment showed some deterioration after 3 months.[86] Drugs and CAM are now rarely used in the United Kingdom.

Five controlled trials compared different approaches to amblyopia treatment.[86–90] All of these have methodological flaws which limit the value of their findings. Overall, although there is evidence that the vision of children with amblyopia improves with treatment,[82–90] these improvements may not be sustained.[82 84–86]

Seven studies evaluating screening programmes reported improvements in visual acuity of two or more Snellen lines in 50%–80% of children who were treated for amblyopia after screening.[47 48 50 53 55 56 91] However, as none of these have a comparison group of untreated children it is difficult to assess the degree to which these changes are attributable to treatment. None of the studies assessed long term outcomes of treatment or evaluated treatment in terms of disability or other patient perceived outcomes. Also, none of the studies assessed the potential negative impact of orthoptic treatment (such as patching) on children or their families which has been suggested by recent qualitative work.[92]

An RCT[93] and a non-randomised controlled trial[94] showed that the use of preoperative prism correction improved the outcome of squint surgery. However, these trials only included patients with obvious squints; no controlled studies of treatment for latent or microsquints were found. Spectacles are highly effective in correcting the disability casued by major refractive errors but the level at which the different types of refractive error cause mjor diability is uncertain and likely to vary with age. The treatment of refractive error in the absence of amblyopia or manifest squint is of unproved benefit and may even cause harm by inhibiting the normal refractive development of the eye (emmetropisation).[95]

Implications

HEARING SCREENING

On the grounds of equity, responsiveness, and cost effectiveness, the transition from universal HVDT to universal neonatal hearing screening in combination with targeted infant distraction tests is considered the best value for money. Some health authorities will be able to free some resources as well as improving the service in moving from HVDT to universal neonatal hearing screening.

SPEECH AND LANGUAGE DELAY

There are insufficient data available to recommend the introduction of *population screening* for early speech and language delay because there is not yet adequate agreement as to which children will not progress *unless* they are given intervention and on the grounds that the screening measures themselves have yet to be shown to have adequate predictive validity.

None the less, early primary speech and language delay should remain a cause for concern because of the problems it may pose for the individual child, the concern it causes parents; the fact that it may serve as a litmus test for other problems which commonly accompany it such as cognitive impairment, behaviour, and conduct disorders, and because of the implications that it may have for literacy and socialisation in school.

PRESCHOOL VISION SCREENING

Amblyopia can cause a considerable reduction in visual acuity, as measured by the Snellen test, but this may not be the best outcome measure. Equally, the physical, psychological, and social implications of reduced visual acuity in one eye are not well understood. Thus it is not clear that amblyopia should be seen as the cause of considerable disability or handicap. No study has adequately considered the possible negative aspects of treatment for amblyopia. Further research is needed to ascertain both the importance of this condition and the most effective and acceptable treatment.

Preschool screening for refractive errors and non-obvious squint, without associated amblyopia, does not seem to be justified as these conditions do not appear problematic by themselves and their treatment at an asymptomatic stage has not been shown to confer benefit. Research is needed to establish whether preschool screening is of benefit. Given the current uncertainty over the potential benefits and harms of testing and some corrective measures it is particularly important that professionals give adequate and accurate information to parents.

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