Universal screening of newborns for hearing impairment in New Zealand

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Introduction

Hearing impairment in children is an important public health problem.[1] Sensorineural deafness is known to have devastating effects on the development of a child's auditory, speech and language skills and ultimately a child's academic performance and employment opportunities in later life. The advent of new technologies enabling early detection and management of hearing impairment has led to the establishment of many Universal Newborn Hearing Screening (UNHS) programmes around the world over the last 10 years. This is due to a growing body of evidence suggesting that early exposure to language (either spoken or signed) correlates with better language development and that early intervention can reduce the need for special education and improve long-term outcomes.

Current strategies

The Ministry of Health currently has three main strategies for identifying hearing impairment:[2]

1. Neonatal paediatricians and obstetricians identify children with a high risk of hearing impairment. These children are then referred on for further audiological evaluation.
2. Questions regarding hearing are included at multiple points of the Wellchild Schedule. Plunket nurses usually administer this. General practitioners, paediatricians and audiologists also have significant input if concerns are present.
3. Children are also screened for middle ear disease and hearing loss at school entry as part of the National Vision and Hearing Screening Programme, which functions as a backstop for the early childhood programme. The National Audiology Centre coordinates this screening.

The National Audiology Centre has kept a database of deaf and hearing-impaired children since 1980, which on average receives 250 notifications per year. The majority of cases of permanent congenital hearing loss are first suspected by parents.[3] In 2002, the average age of identification for children with moderate or greater losses was 35.1 months.[3] The Ministry of Health's 1995 report *Preventing child hearing loss: Guidelines for public health services* stated goal was for all infants with hearing loss to be identified before three months of age and to receive intervention by six months of age.[1] Over the last seven years an average of 60% of hearing impaired infants in New Zealand have had no known risk factor,[3] limiting the effectiveness of the use of an 'at risk' register as a screening tool for deafness in newborns.

The current approach is not working. One issue is that the screening methods being used are of limited effectiveness. It is not easy to measure behavioural thresholds on very young children and the use of these methods for screening has not been effective in New Zealand or overseas. By the time hearing loss is suspected, the window of opportunity is already lost. To rectify these problems, UNHS is now being considered for New Zealand. The National Health Committee has published *Screening to improve health in New Zealand: Criteria to assess screening programmes* outlining criteria by which screening programmes can be evaluated.[4] A number of regional neonatal hearing screening programmes have operated over the years, many of which have ceased operation due to lack of sustainable funding. The exception is the Tairawhiti programme, which has been screening consistently since 1997.

Appropriate technologies

Screening using evoked Otoacoustic Emissions (OAEs) or Automated Auditory Brainstem Response (AABR) testing can be satisfactorily carried

out in the ward. OAEs measure the function of the inner ear by stimulation of specific frequencies.[5] An echo is produced by the mechanical action of the auditory sensory outer hair cells that can be recorded by a microphone in the outer ear canal. The AABR test detects brain activity in response to sound and uses small sensors that are placed on the baby's scalp. This test measures the integrity of the peripheral auditory system and auditory pathway up to the brainstem.[5]

The measurement of OAEs is a test based on acoustic output from the cochlea, and these emissions are affected to a greater extent by ambient noise and noise produced by the child than is the AABR. A limitation in using OAEs as a neonatal screening test is the finding that the occurrence of OAEs is low in the hours after birth and may not stabilise until approximately 48 hours afterwards. In addition, the accuracy of this test is reduced with the presence of vernix and amniotic fluid in the ear canal when performed soon after birth. Obtaining results from both these tests is easier with a settled or sleeping infant. However, OAEs are generally quicker and cheaper than AABR although higher false positive rates are common and they do not detect all cases of auditory neuropathy. The utility of both OAE and AABR as screening tools has been well documented in NZ and overseas.

Standard of care

Over the last decade the popularity of UNHS and early intervention programmes has soared, and it is now considered the standard of care in many parts of the world. Currently UNHS is mandated by law in 37 of the US States and is being implemented in the United Kingdom through a carefully controlled rollout. Some Australian states have implemented UNHS, along with three provinces in Canada (Ontario, Alberta and New Brunswick).

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Screening programmes

The sensitivity (proportion of hearing impaired children who test positive) and specificity (proportion of normal hearing children who test negative) of overseas newborn hearing screening programmes have continued to improve, both with experience, and as commonly used technologies have been refined. Hearing status is a continuous two dimensional variable: hearing threshold as a function of frequency. Sensitivities and specificities of screening programmes vary according to the degree of hearing loss and the screening protocol adopted. Recent programmes are attaining 98% specificity and a sensitivity of close to 100%, with false positive rates as low as 0.2%.[6,7] It is important to realise that a screening programme by its very nature is going to misclassify some children's hearing loss. In the well baby nursery the positive predictive value of a positive screening test varies according to the screening protocol used. False positive rates range from 0.2 to 4% in well run two tier or AABR based programmes (with the best results achieved where re-screening was routinely employed) to 35% in OAE based programmes with no routine rescreening. White et al. examined refer rates for varying protocols, finding that for every 1000 babies screened, a two stage OAE programme would likely refer 80 infants to a second screen, with eight of these being referred for full audiological assessment. An AABR based programme would refer 40 straight to diagnostic assessment, while an OAE/AABR programme would refer 20. All would find the three true cases of hearing loss.[8] As protocols have been modified, the positive predictive value of screening has also improved.

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Benefits of early identification

There is a large body of research examining the effects of hearing impairment, including milder losses, on language acquisition, learning and school performance. The importance of early language experience (both signed and spoken) in language acquisition is well recognised and has been linked to normal cognitive development as well as later educational success. With the growing number of universal newborn hearing screening programmes, interventions enabling earlier access to spoken language have been possible. As an example, Yoshinaga-Itano et al. tested 150 children between the ages of 12 and 36 months with varying degrees of hearing loss. The data reveals that the average total language quotient for infants receiving intervention prior to six months of age is significantly better than late identified children, surprisingly, across all degrees of hearing loss. These benefits cross communication modes, ethnicity, socioeconomic status and multiple disability status.[9]

Potential harm of screening

The manner in which parents are notified of the initial test results is a key consideration for all UNHS programmes. All screening has the potential to result in psychological

harm and unnecessary parental anxiety. In particular, anxiety can occur when a child does not pass the first screening test and where it takes time for parents to be provided with a final report on their child's status. False positive cases must be managed carefully to minimise parental stress and anxiety. Neonatal screening appears to be well accepted by parents with refusal rates from 0.4% to 2.4% quoted in the literature and parents indicating they would want hearing screening for their future children. The other potential issue is with false negatives: children who have a hearing loss and who are not detected through screening. False negatives need to be subject to audit as part of the programmes evaluation, although they do appear to be rare with universal newborn hearing screening. There are other reasons (see under 'Infants who may not be identified') as to why children may later present with hearing loss after a normal screening test. General practitioners, paediatricians, otolaryngologists and audiologists still need to be suspicious if there are ongoing concerns about a child's hearing even if screening programmes have passed them as having normal hearing. There is still much to learn to ensure that any potential New Zealand programme achieves adequate coverage, low false positive and false negative rates with minimal parental anxiety.

Follow-up and coverage

A potential weakness of any screening programme is the difficulty in adequately following up individuals who screen positive. Challenges to successful UNHS programmes include maintaining continuity in screening personnel, accurate timely diagnosis, overcoming a lack of knowledge about services for follow-up,[10] ensuring timely enrolment in early intervention programmes, and the establishment of screening protocols and continual monitoring of outcomes.[11] A process for handling home births, early discharges, private births and transient populations must also be established in order to provide high coverage and a consistent service to all parents.[12] Maori as a special group would need to be involved and consulted fully to ensure participation and equity of access. Many programmes set out to screen more than 95% of the target population. A United States survey completed in 1998 of 120 screening programmes reported that the coverage of the 64 OAE based programmes averaged 94.9% and the coverage of the 56 AABR based programmes averaged 96.2%. The percentage of children passing the first screen was 91.6% at discharge for OAE and 96.0% at discharge for ABR.[13] Those very people who slip through the net are often those very people whom the screening programme is attempting to reach. Coverage and follow-up rates are therefore a key measure of a programmes success.

Programme monitoring

The long-term benefits of most UNHS programmes have not been fully established. This is in part due to the difficulties in conducting randomised controlled trials in this area and the age of existing programmes. More evidence of the effectiveness of UNHS is needed. In time, more population based data will be collected on how programmes impact on the educational and social success of the children they reach. Late diagnoses (false negatives) and wrong diagnoses need to be routinely examined to determine the reasons and to improve the programme. Today's children with hearing impairment, almost all of whom are detected 'late', go on to require a considerable investment of health and educational resources.[14] Calculations on the potential cost savings in these areas, resulting from a New Zealand UNHS programme will be useful in decision making.

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Infants who may not be identified

The aim of universal newborn hearing screening programmes is to detect infants with permanent congenital hearing losses. As no screening programme will detect all cases of the screened condition, infants developing hearing loss after birth, such as those with progressive familial sensorineural hearing loss, and some infants with special neural dysfunction will not be identified. Two groups that would not always be detected are infants with cytomegalovirus and those with auditory neuropathy. Congenital cytomegalovirus is the most common foetal viral infection encountered, occurring in two to 22% of all live births. Of infants who are infected, only 10% are born with symptomatic cytomegalovirus. In a study of 388 children who had had a congenital cytomegalovirus infection the incidence of sensorineural hearing loss was 15.4%. This hearing loss can develop after birth and may be present in children who have had no symptoms or other signs of congenital cytomegalovirus infection.[15]

Auditory neuropathies are defined as hearing loss in the presence of preserved cochlear outer hair cell function (normal OAEs) but abnormal auditory nerve activity (abnormal AABR).[16] The incidence of auditory neuropathy in the infant population has not been established, although some research indicates as many as 10% of children with permanent congenital

hearing impairment may be affected. Clinically, the patients' speech recognition is poorer than would be expected from their audiograms and the expected benefits from amplification are also poorer. To test for Auditory Neuropathy cochlear function must be measured. The gold standard is AABR with presence of a cochlear microphonic, ideally with presence of OAEs.

Conclusions

UNHS programmes are becoming the standard of care around the world. New Zealand's poor record in the early detection of permanent congenital hearing impairment means that there is little doubt that with the passage of time a newborn hearing screening programme will be initiated; the potential benefits are great. The challenge is not just the introduction of such a programme but ensuring that quality standards are met, nationwide, with adequate coverage, and ongoing data collection, and independent evaluation in addition to timely and appropriate intervention. The overall success of UNHS programmes will ultimately be judged on population detection rates and long-term outcomes.[17]

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A more fully referenced paper is available on request