

PROJECT HIEDI

Hearing Impairment:
Early Detection and Intervention



RIGHT FROM THE START

I TE TIMATANGA

- In summary
- Why is early intervention important?
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In association with the National Foundation for the Deaf





New Zealand has a very poor record in the early identification of infants with permanent congenital hearing impairment. The average age of diagnosis of this type of hearing loss (moderate or greater in severity) was 46 months in 2003¹. As a result, interventions such as the provision of hearing aids, cochlear implants and the teaching of sign language are significantly delayed. These late average ages are in stark contrast to those in many countries, where, because of newborn hearing screening, diagnosis occurs before the child reaches three months of age and intervention begins before six months of age.

Nearly all of New Zealand's newborns are without hearing screening. Thus most children born with permanent hearing impairment are not identified at the optimal time. As a result, these children struggle to build the foundation skills required to succeed educationally and socially, and this in turn has significant long term effects on mental health, vocational choice and employment as they grow into adults².

For more than 10 years, objective screening technologies have been available to accurately screen babies for hearing loss during the birth hospitalisation. Currently over 80% of newborns in the United States are screened soon after birth. Project HIEDI is promoting the establishment of a nationally-coordinated universal newborn hearing screening and early intervention programme in New Zealand, so that infants can be identified much earlier, allowing intervention to take place at the optimal time. This approach has proven to be successful in many other countries.

The primary aim of a newborn hearing screening and early intervention (UNHSEI) programme is to detect and provide early intervention for children with Permanent Congenital Hearing Impairment (PCHI). PCHI is permanent sensorineural or conductive hearing loss present when the baby is born.

PCHI acts as a barrier, limiting the child's capacity to build foundation skills such as the ability to acquire language, read, write, and interact with others. The consequent developmental delays are in great part due to the lack of access to language during 'sensitive' acquisition periods³. In turn, these developmental delays have a significant impact on longer term outcomes such as employment and vocational choice.

In addition to placing stress on the hearing-impaired child, late diagnosis of a hearing loss also places pressure on families. Late intervention means the required intensity and duration of the habilitation process is much greater to attempt to help the child recover from the developmental deficits that have accrued. Unfortunately, as much of the intervention occurs after the optimal time, recovery is often not possible.

Minimising the negative effects of hearing impairment requires the early identification of PCHI, preferably soon after birth, followed by timely and appropriate intervention and support⁴. International research demonstrates that well-run UNHSEI programmes result in earlier identification and improved outcomes when compared to those achieved through the use of a risk factor approach, as currently operates in New Zealand.

Children who are enrolled early in intervention programmes outperform their late intervention counterparts on measures of expressive and receptive language. These improvements lead to downstream benefits in terms of access to more appropriate educational settings and better reading and educational performance⁵⁻⁷. With early intervention, even some children with the most severe hearing losses can develop normal language, removing a significant barrier to educational and social success. In addition, reaching these children before developmental deficits build can lead to a reduction in the amount of long term assistance needed.

Children with mild and unilateral hearing losses also benefit from early identification and intervention. In addition to moderate and greater bilateral hearing losses, mild and unilateral losses are also known to affect language development and educational performance. These hearing losses are notoriously difficult to detect and as a result are commonly diagnosed later than bilateral or more severe hearing losses, thus increasing their impact.



WHEN ARE CHILDREN WITH PCHI IDENTIFIED?

New Zealand has a very poor record for detecting and taking action on early childhood hearing impairment, with an average age of detection in 2003 of 46 months for losses that are moderate or greater. Maori children are over-represented in this group and are consistently identified later than other children. For example, in 2001, 80% of Maori children with PCHI were identified by 80 months of age, compared to 60 months of age on average for European children⁸.

The current method of identification of hearing loss in New Zealand infants uses risk factors and hearing surveillance by early childhood health providers. Risk criteria, including family history and maternal infection, identify children with a greater chance of PCHI. Children with risk factors, along with those suspected of having a hearing loss should then be referred to their local audiology department for diagnostic assessment. As around 60% of infants in New Zealand with PCHI have no known risk factor, a risk factor approach will never detect a large proportion of children with PCHI.

HOW MANY CHILDREN ARE AFFECTED?

Because newborns are not routinely screened for hearing loss in New Zealand, it is difficult to determine the exact number of New Zealand babies with PCHI. However, we can estimate the number of children in New Zealand born with PCHI by examining the number of significant bilateral and unilateral notifications to the Deafness Notification Database. The resultant estimate of 3.00 cases per thousand (or approximately 170 children per year) equates to the number of children that would likely be detected by a UNHSEI programme⁸⁻¹⁰.

Incidence of PCHI is higher than that of any of the medical conditions currently screened for at birth through metabolic screening. Late detection of hearing loss, and its consequences, place a significant burden on families and on our education system, which at any one time has to deal with hundreds of late diagnosed children requiring considerable resources to assist them through school.

LOCAL SCREENING PROGRAMMES

There have been several attempts to establish regional UNHSEI programmes in New Zealand, with varying success. The longest running of these programmes is funded by the Tairāwhiti District Health Board and has been operating continuously since the mid-1990's. Other regional programmes (Invercargill and Whakatane) have not been able to continue screening due to a lack of sustainable funding, or staffing issues. A limited programme operates through Christchurch Women's Hospital. Historically, the regional programmes have been small, and driven by a single individual, often an audiologist, who has had to set standards and implement a programme largely without assistance or sustainable funding.

In February 2004 the Waikato District Health Board established a new hospital-based UNHSEI programme. This programme will eventually screen 3500 babies annually making it the largest programme of its type ever established in New Zealand. The Waikato programme has the potential to become a pilot for a nationally-coordinated programme.

SCREENING NEWBORNS FOR HEARING IMPAIRMENT

Two simple and inexpensive tests are commonly used to accurately screen infants for hearing loss in the first days of life, often while the baby is asleep. They pose no risks for babies and, if used and interpreted correctly, are highly accurate and reliable. This technology is already used here in the regional programmes mentioned and also to assess hearing in infants and children suspected of hearing loss.

The tests used are:

- **OTOACOUSTIC EMISSIONS (OAE)**
For this test a miniature earphone and microphone are placed in the ear to produce a sound and measure a response from the ear. If a baby hears normally, an echo is reflected back into the ear canal and this is picked up by the microphone. The lack of an echo on the OAE test is indicative of a possible hearing loss and the child is referred on for further testing.
- **AUTOMATED AUDITORY BRAINSTEM RESPONSE (AABR)**
For this test, sounds are played to the baby's ears. Small sensors that are placed on the baby's head detect brainwaves in response to the sound, thus providing a measure of the sound-induced activity in the hearing pathways of the brain. If there is no response or the response is outside the normal range then the child is referred on for further testing.

The two test methods may be used individually or in combination. In some hospitals, a two-tier approach is used, with babies being screened first using OAEs with those that do not pass then being tested using AABR.

If a problem is suspected as a result of the hearing screen, the child is sent for a full audiological assessment to confirm whether a hearing loss exists. Intervention is the crucial next step to ensure the child has the best possible start to life. Intervention may include the fitting of hearing aids or a cochlear implant, learning of sign language, speech therapy or the provision of other services to support the child and family in learning communication skills. In New Zealand, much of the support and intervention is coordinated or provided by an Adviser on Deaf Children, who contacts the family after diagnosis.

Overseas programmes demonstrate very high rates of coverage, often screening more than 95% of total births within the target population. The hearing screening is carried out as soon as possible after birth, with the informed consent of the parents.



INTERNATIONAL SUPPORT

Due to technological advances, there has been rapid growth in the number of universal newborn hearing screening programmes throughout the world, leading to its description as an *international standard of care*.

Currently, universal newborn hearing screening (UNHS) is mandated by law in 37 states plus the District of Columbia in the United States of America and is being implemented in the United Kingdom following a series of carefully controlled pilot studies. Some Australian states have implemented UNHS, along with three provinces in Canada (Ontario, Alberta and New Brunswick). National programmes exist or are being implemented in Sweden, Scotland, Croatia and Denmark. Regional or hospital-based screening exists in countries such as Russia, Lithuania, Romania, Hungary, Brazil, Poland, Italy, Belgium and Austria.

International cost benefit research indicates significant cost savings can be achieved through the establishment of UNHSEI programmes. Such programmes reduce the need for educational support and improve educational outcomes which in turn lead to wider vocational choice and projected reductions in the levels of unemployment for individuals with PCHI.

LOCAL SUPPORT

Over the past decade various efforts have been made in New Zealand to encourage successive governments to screen all babies for hearing impairment at birth and provide early intervention for those diagnosed with a hearing impairment. Although the need for earlier identification and intervention has generally been acknowledged, these efforts have not been successful, and the majority of New Zealand babies are not screened for hearing impairment.

Project HIEDI (Hearing Impairment: Early Detection and Intervention) was formed in late 2002 to build on previous efforts. The project utilises the expertise and support of many individuals and organisations skilled in this area through the Newborn Hearing Screening Consultative Group. This group includes all major stakeholder organisations, including parents of deaf and hearing-impaired children, healthcare professionals and educational organisations involved in working with deaf and hearing-impaired children and their families.

Many statements of support have been collected, from both Consultative Group organisations and other health and education organisations. Organisations that have endorsed the Consensus Statement on universal newborn hearing screening include: New Zealand Federation for Deaf Children, NZ

Speech-Language Therapists Association, Advisers on Deaf Children, Deafness Research Foundation, National Foundation for the Deaf, The Paediatric Society of New Zealand, New Zealand Audiological Society, Royal New Zealand Plunket Society and Royal New Zealand College of General Practitioners. (For a full list of organisations endorsing the statement please contact Project HIEDI.)

Project HIEDI has also been working to further raise awareness of this important issue within government and among the public and stakeholder groups. Much of Project HIEDI's effort has been concentrated on compiling a review of the evidence for the need to establish a universal newborn hearing screening and early intervention programme in New Zealand. The review, "Improving outcomes for children with permanent congenital hearing impairment – *The case for a national newborn hearing screening and early intervention programme for New Zealand*" has been prepared and will be presented to the Ministries of Health and Education. This booklet provides a brief outline of the rationale for screening and early intervention. Copies of the review can be obtained by contacting Project HIEDI.

In addition to broad sector support, government agencies have acknowledged the importance of early identification and intervention for children with PCHI. The establishment of a Newborn Hearing Screening and Early Intervention Programme provides an excellent contribution to many government policies and strategies. In particular, such a programme would assist with government Disability, Education and Maori strategies by reducing existing inequities and allowing young deaf and hearing-impaired children to maximise their educational opportunities and fulfil their potential.

In 2003, the National Health Committee published "Criteria for Assessing Screening Programmes", to establish a framework for assessing current and potential future screening programmes. A well considered nationally-coordinated universal hearing screening and early intervention programme would meet these criteria, with accurate testing and diagnosis available, effective early intervention programmes, and acceptable ways to minimise stress for parents. Current health and education systems already deliver diagnostic and intervention services for children though systems would need to be refined to better deal with earlier identified infants.



Because of New Zealand's very late average age of detection, there is a unique opportunity to make significant improvements to the lives of many children and their families through the implementation of a national UNHSEI programme.

Implementation of such a programme would not increase the overall number of children diagnosed with PCHI, rather it would allow intervention to begin at the optimal time. This would result in improved educational, cognitive and social outcomes while providing equity of access and reducing downstream educational costs.

It is recommended that the Ministries of Health and Education urgently consider approaches to improve outcomes in children with permanent congenital hearing impairment; in particular, that they consider the strong evidence for superior outcomes that can be obtained by a universal newborn hearing screening and early intervention programme.

Subsequent to any decision to implement a national programme, the following should be considered:

- Establishment of a small number of pilot programmes which would be evaluated in addition to existing regional programmes, informing the design of a national programme.
- Utilisation of local and international expertise to assist in the design of a national programme.
- Formalisation of the pilot programme status of existing programmes and modification of these programmes as necessary to fit the agreed design.
- Development of policies to ensure consistent application of protocols and standards throughout New Zealand.
- Development of policies to ensure effective collaboration between screening, diagnostic and intervention services.
- Review of workforce and facilities for diagnosis and intervention and urgent action to address any identified deficits with due consideration of lead time for training.

The establishment of such a programme will not remove the need for later hearing surveillance to identify progressive or acquired hearing losses that may develop, rather it would be the first step to ensuring hearing losses present at birth are detected at the earliest possible time, enabling children with PCHI to fulfill their potential through timely intervention.

If you would like more information about Project HIEDI, please contact hiedi@nfd.org.nz, visit our website www.nfd.org.nz/nfdnews/projecthiedi, or phone The National Foundation for the Deaf on (09) 307 2922.

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