PROJECT HIEDI

Hearing Impairment:
Early Detection and Intervention

Improving outcomes for children with permanent congenital hearing impairment

The case for a national newborn hearing screening and early intervention programme for New Zealand.

November 2004

NFD

Tihei mauri ora
e ngā mana e ngā reo
e ngā matāwaka huri noa i te motu,
tēnā koutou, tēnā koutou,
tēnā tātou katoa

Ko te kaupapa huarahi, i tukuna tenei take mo te whakātūria i tenei panui mo ngā pēpi hou o Aotearoa. Ki te whakakaha ake i te mohio o te hunga mo te taringa whakarongo.

Ki te roopu e hapai i tenei kaupapa mo te awhi mo ngā taringa o ngā pēpi he mihinui tenei kia koutou katoa.

Ko te tumanako me awhi i ngā whaea me ngā whanau kia haere ki te whakāmatautau ngā taringa o ratou pēpi i waenganui o ngā kaimahi. Mo te whakatikatika, kia rongo ake kia tu tonu rātou i te hauoratanga o te iwi.

Ko rātou ngā pēpi, ia mātou taonga, e heke mai nei mo apopo.

Welcome to the people of Aotearoa (New Zealand). Greetings to one and all.

The purpose and objective of the document is to present a case demonstrating the need for a nationally co-ordinated universal newborn hearing screening and intervention programme in New Zealand.

The Project HIEDI Steering Team is grateful to all those people who assisted with the preparation of this document.

Our aim is to support mothers and whanau to ensure they can access early diagnosis and intervention to help them maximise future learning for their children and thus enable them to reach their potential.

Our babies are our future.

CONTENTS

2. PREFACE

This document has been produced by *Project HIEDI (Hearing Impairment: Early Detection and Intervention)* in consultation with a broad group of stakeholders (The Newborn Hearing Screening Consultative Group).

2.1 BACKGROUND

Over the last decade various efforts have been made to encourage successive governments to introduce a programme of universal newborn hearing screening and early intervention (UNHSEI). Although the need for screening has generally been acknowledged these efforts have not been successful and the age at which permanent congenital hearing impairment (PCHI) is diagnosed in New Zealand continues to increase.

To facilitate progress in the adoption of a universal newborn hearing screening and early intervention (UNHSEI) programme, the National Foundation for the Deaf funded a conference of interested stakeholders in Auckland in 2001. This conference provided a consensus view that a nationwide programme of UNHSEI should be introduced. A stakeholder group was formed (Newborn Hearing Screening Consultative Group, referred to as The Consultative Group within this document) and a mandate was given to members of that group to work on a case for action. A later conference, also supported by the National Foundation for the Deaf, was held in 2002. With the support and encouragement of the Consultative Group, Project HIEDI was formed in late 2002 and a Steering Team, was established. Funding from the NR Thomson Trust and The Todd Foundation enabled the appointment of a Project Manager and the start of work to assess and draw together the evidence for newborn screening and early intervention, to consult with the government, to support local universal screening programmes, and to raise awareness of the need for early identification of, and intervention for, children with permanent congenital hearing impairment (PCHI).

2.2 STEERING TEAM

The Steering Team comprises:

- **Dr Peter Thorne** (BSc, DipSc, PhD), **Project Leader:** Dr Thorne is an Associate Professor of Audiology in the Faculty of Medical and Health Sciences at the University of Auckland. He is the Head of the Section of Audiology, and Director of the Audiology Training Programme. He is the Chair of the Board of the National Foundation for the Deaf, and is a member of the Board of the Deafness Research Foundation and its Scientific Committee. Dr Thorne is a long-time advocate of universal newborn hearing screening and facilitated the establishment of the Consultative Group.

- **Margaret Cooper** (QSM): Margaret Cooper is a parent advocate who has a 32 year old profoundly deaf son. She has 26 years of experience with various parent groups and the deaf and special needs sector. In addition, Margaret is a past President and executive member and Honorary Vice-President of the New Zealand Federation for Deaf Children, an organisation she has served for 20 years. She is also an independent member of the Board of the National Foundation for the Deaf.

- **Dr Bill Keith** (QSO, MA Hons, PhD, MNZAS): Dr Keith was previously Principal Audiologist at the National Audiology Centre. He is a member of the Board of the Deafness Research Foundation and the Deafness Research Foundation Scientific Committee, and is a member of the Abilities Board. Dr Keith is currently Managing Director of Phonak New Zealand Ltd.

- **Dr Dianne Webster** (PhD, DHSM, FHGSA): Dr Webster is the Director of the National Testing Centre, which screens newborns for congenital metabolic disorders. She is also Chair of the Quality Assurance Committee of the International Society for Neonatal Screening and is Chair of the Human Genetics Society of Australasia and the Royal Australasian College of Physicians – Division of Paediatrics Joint Newborn Screening Committee.

- **Oriole Wilson** (MSc, DipAud): Oriole Wilson is an Audiologist and is Clinical Director of Auckland District Health Board Audiology Services and the National Audiology Centre. Oriole is a long-time advocate of universal newborn hearing screening and facilitated the establishment of the Consultative Group.

- **Bill Tangariki:** Bill Tangariki is an executive member of the Kaumatua/Kuia Te Roopu O Waipareira Trust West Auckland (Māori elders group). He is also the cultural advisor for the West Auckland Living Skills Mental Health Organisation (WALSH). Bill has been passionately involved in the health and well-being of Māori children for many years and has endorsed his support to be the kaumatua advisor for Project HIEDI.

- **Janet Digby, Project Manager:** Janet Digby was appointed as Project Manager of Project HIEDI in December 2002.

Prior to her appointment, Janet's experience centred on research and organisational change projects in a wide range of industries, within both the private and public sectors.

2.3   CONSULTATIVE GROUP

The Newborn Hearing Screening Consultative Group comprises representatives from a broad range of interested organisations, and was established in 2001 to act as a reference group for the establishment of a nationally coordinated UNHSEI programme.

Organisations (listed below) were invited to appoint a representative to join the Consultative Group. The Consultative Group endorsed the development of a Steering Team and Project Manager as a way of facilitating the introduction of a UNHSEI programme in New Zealand. (See also section 11.2.1 *New Zealand Consensus Statement.*)

Auckland Parents of Deaf Children	Joy Wells
Cochlear Implant Programme	Ellen Giles/ Phillipa Hunt/ Dr Colin Brown
Deafness Research Foundation	Dr Ron Goodey
Deaf Education Aotearoa New Zealand	Rachel Noble
Group Special Education, Advisers on Deaf Children	Glenys Yates and Valerie Smith
Hearing House	Anne Ackerman
Immunisation Advisory Centre (IMAC)	Dr Nikki Turner
Itinerant Teachers of the Deaf	Janet Wilson
Kelston Deaf Education Centre	David Foster
New Zealand College of Midwives	Norma Campbell
Ministry of Health, Chief Child Health Adviser	Dr Pat Tuohy
Ministry of Education	Sally Jackson (represented by Joanna Curzon)
National Audiology Centre	Oriole Wilson
National Ear Nurse Specialist Group of NZ	Margaret Couillault and Barbara Middleton
National Foundation for the Deaf	Marianne Schumacher
National Testing Centre	Dr Dianne Webster
Ngati Awa Society and Health Services	Sue Maloney and Myint Lin
New Zealand Society of Otolaryngology Head and Neck Surgery	Dr Bill Baber
New Zealand Federation for Deaf Children	Sabine Muller
The Pediatrlc Society of New Zealand	Dr Roland Broadbent
New Zealand Speech-Language Therapists Association	Judith Lemberg
New Zealand Audiological Society	Jo Mackie
Public Health Nurse	Leane Els
Royal New Zealand Plunket Society	Trish Jackson Potter
The University of Auckland, Audiology Department	Dr Peter Thorne
Van Asch Deaf Education Centre	Neil Heslop
Vision Hearing Technicians	Gay Mohi

2.4   ADDITIONAL ACKNOWLEDGEMENTS

Contributions to this document

Project HIEDI would like to acknowledge the following people for their individual contributions to this document: Phillipa Adams, Dr Nicola Austin, Dr Jim Bartley, Sue Barratt, Anne Fulcher, Kirsty Gardner-Berry, Dr Steve Hodgkinson, Emily Hunter, Bruce Kent, Jo Mackie, Sabine Muller, Letitia Nicolescu, Teresa Konieczny, Rachel Noble, Michelle Pokorny, Pat Pritchett, Dr Grant Searchfield, Leslie Searchfield, Doreen Singh, Michael Williams, Dr Donald Webster and Glenys Yates.

We would also like to thank advertising agency *DDB (Auckland)* and printers *Modern Print* for their contributions.

In addition to those individuals listed above, our Steering Team would like to thank the following people for reviewing or contributing to sections of the document:

Section 5: Hearing Loss in context
Professor Randall Morton[i] for reviewing a draft of section 5.1.

Section 6: Effects of permanent congenital hearing impairment
Liz Fairgray[ii], Judith Lemberg[iii] and Dr. Trecia Wouldes[iv] who reviewed various drafts and parts of this section.

Section 6.6: Impact on the child's family
The New Zealand Federation for Deaf Children for their contribution to the writing of this section. A number of parents contributed their own experiences to supplement references from international literature. In particular, we would like to thank Sabine Muller and Joy Wells.

Section 8: Technologies for identifying permanent congenital hearing impairment
Dr. Andrea Kelly[v] for reviewing various drafts and parts of this section.

Project Contributions

Project HIEDI would also like to thank the following people for their assistance with this project: Anne Ackerman, Dr Lara Harvey, Dr Melissa Wake, Kris MacDonald, Lorraine Fox, Lorraine Knutsen, Chris Blincoe, Christine Rhodes, Kathy Sandiford, Nicholas O'Flaherty, Monica Wilkinson and Richard Hipgrave.

Project HIEDI acknowledge the tireless and passionate effort of Sir Patrick Eisdell Moore to see the introduction of newborn screening for deafness in New Zealand. This vision and energy has been an inspiration.

Project Sponsors

And finally, Project HIEDI would like to acknowledge the financial support of The National Foundation for the Deaf, The NZ Guardian Trust as trustee of the N R & J H Thomson Charitable Trust, The Todd Foundation, Scanmedics (Australia), GN ReSound (New Zealand), The Oticon Foundation of New Zealand and the Auckland Medical Research Foundation.

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3. EXECUTIVE SUMMARY

3.1   SUMMARY

This document presents the evidence relevant to the need for a national universal newborn hearing screening and early intervention (UNHSEI) programme in New Zealand. It demonstrates that the introduction of such a programme complies with the National Health Committee's 'Criteria for Assessing Screening Programmes'. It would also make a considerable contribution toward the achievement of key Health, Education and Disability strategies.

Hearing loss and its effects: Permanent Congenital Hearing Impairment (PCHI) is an important population health issue both in terms of its prevalence and impact on the health and well-being of children. Hearing impairment can prevent or delay speech and language development and lead to poor communication ability. These effects can be ameliorated by early intervention[1]. However, because the auditory system becomes less receptive to intervention with time, a child born with PCHI has a limited window of opportunity within which exposure to language can optimise the chance of normal language acquisition[2-4]. Children born with PCHI that is diagnosed 'late' have limited exposure to sound during this critical window resulting in poorer development of the central auditory system and reduced ability to acquire language. A similar principle applies where intervention is non-auditory (ie sign language). Limited or absent exposure to sound through hearing impairment can have significant negative effects, not only on language acquisition but cognitive development, educational outcomes, mental health, social functioning and vocational choice[5-14].

Hearing impairment in New Zealand children: The prevalence of significant bilateral or unilateral PCHI in New Zealand is estimated to be 3.00 per thousand births, which is within the range reported internationally. The current system of detection is to identify those newborns at highest risk of hearing loss and refer them for audiological assessment. This system is clearly not working as the average age of detection of hearing-impaired infants in New Zealand with moderate or greater losses is currently 46 months[15]. This is much higher than the three month threshold by which diagnosis is recommended in many countries. The efficacy of the 'high-risk' approach in New Zealand is limited as 59% of children notified with a hearing impairment in the seven years to 2002 had no known risk factors for deafness[16-21]. Māori children are over-represented in the notification statistics, comprising 23.5% of the population under 19[22] yet they comprised 48% of all deafness notifications in 2001[20].

Newborn hearing screening: UNHSEI programmes are designed for the early detection of PCHI (that is a hearing impairment present at birth) enabling intervention to begin early. At-birth screening using objective, physiological techniques that measure auditory function is now possible and is commonly followed by diagnostic audiological tests in babies with a positive screening result. (See section *8.2.2: Objective tests*)

There is a strong international trend towards the early identification of hearing impairment in infants. Universal newborn hearing screening is now offered to more than 80% of newborns in the USA, and programmes are being or have been established in many other countries (eg UK, Australia, Canada, Sweden, Taiwan and Croatia). As a result UNHSEI programmes are rapidly becoming the standard of care internationally[23-28]. The processes involved in establishing and maintaining a quality programme are well understood as a result of 15 years overseas and local experience, and newborn hearing screening programmes are broadly acceptable to health professionals and key stakeholder groups.

Cost benefit research has been conducted overseas. Although comparison (and generalisation) between studies is difficult, the general view is positive when comparing universal newborn hearing screening to (1) risk register approaches, (2) other screening programmes or (3) to no screening at all. New Zealand specific analysis has not been conducted to date. (See section *10: UNHSEI Programmes: Costs, benefits and cost effectiveness.*)

Health professionals, parents and educators in New Zealand are unified in their support of the establishment of a national newborn hearing screening and early intervention programme. (See section *11:Local support and endorsements.*) A number of local programmes have been established, but most are no longer in operation due to a lack of sustainable funding.

Early detection and intervention: The very late age of identification of PCHI now current in New Zealand denies infants access to language during the optimal period for language acquisition[2, 4, 29-32]. Infants whose hearing loss is detected early and who receive appropriate habilitation demonstrate significantly better language and subsequent educational outcomes[5-12, 33]. Good language skills are also correlated with improved literacy, a basic skill required for educational success[34].

International research demonstrates that UNHSEI programmes result in significantly earlier detection of hearing losses[35-39]. This in turn results in earlier intervention for diagnosed children[40-43]. Evidence is accumulating that this earlier intervention leads to lasting improvements in outcomes. Examples of these improved outcomes include better receptive and expressive language for children experiencing earlier intervention. Further evidence of benefits will result from both larger sample sizes and prospective studies as more programmes are established and as data from existing programmes becomes available. (See sections *9.4: Do UNHSEI programmes result in earlier identification and intervention?* and *9.5 Does early intervention lead to improved outcomes?*)

Compliance with screening criteria: There is good evidence that the establishment of a UNHSEI programme in New Zealand would comply with the National Health Committee's Criteria to Assess Screening Programmes.

Compliance with key strategies: The establishment of a national UNHSEI programme would contribute significantly to the fulfilment of the New Zealand Disability Strategy, ensuring that hearing-impaired children have full access to appropriate educational opportunities and its many positive downstream effects in terms of well-being and quality of life. The establishment of such a programme would also make significant contributions to current Health, Education and Māori strategies. (See section *15: Appendices: Compliance with Disability, Health, Education, and Māori Health strategies.*)

Conclusions: With the establishment of a national UNHSEI programme we will have a unique opportunity to make significant improvements to the lives of many children and their families. Equal access to health and educational services will increase each child's ability to achieve his or her potential, both as a student and later, as a fully participating member of society.

Primary recommendation:

The authors of this report strongly recommend 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.

This proposal is well supported within the sector, with both professional and consumer groups unified around its value, across health and education, deaf and hearing-impaired, Māori and non-Māori.
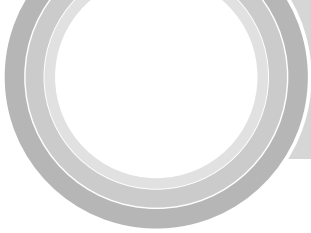
3.2   QUESTIONS OR COMMENTS

Questions or comments relating to this document should be directed to:

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4. COMPLIANCE WITH SCREENING CRITERIA

This section summarises the implications of universal newborn hearing screening and early intervention (UNHSEI) in relation to the National Health Committee's "Criteria for Assessing Screening Programmes"[44]. Evidence in support of each of these criteria is presented in more detail in the sections indicated

Criteria for assessing screening programmes	How UNHSEI meets these criteria
1. The condition is a suitable condition for screening	**Importance:** PCHI is an important health and disability issue due to its high incidence rate and the poor social, developmental and educational outcomes for those children not detected and treated early. (See section *6: Effects of permanent congenital hearing impairment,* and section *7.2: Prevalence.*) **Presence of a detectable disease marker:** PCHI at birth is revealed by the loss of inner ear or auditory nerve function. This loss can be effectively detected using objective physiological tests. (See section *8.2.2: Objective tests.*) **Natural history of the condition:** PCHI has significant effects on a child's language acquisition, social interaction, mental health, cognitive development, and educational performance. These effects can be ameliorated by early, appropriate intervention. (See section *9.5: Does early intervention lead to improved outcomes?*)
2. There is a suitable test ...	**That is safe** - Otoacoustic emissions (OAE) and automated auditory brainstem response (aABR) are the methods commonly used to screen newborns for PCHI. Both are non-invasive and safe for infants. (See section *8.2.2: Objective tests.*) **That is simple and reliable** - The commonly used tests are easy to perform, require minimal interpretation and are reliable. (See section *8.2.2: Objective tests.*) **That is accurate/valid** - Providing appropriate protocols are used, both aABR and OAE are a true measure of auditory function in the newborn. (See section *8.2.2: Objective tests.*) **That is highly sensitive** - The tests commonly used are highly sensitive. (See section *8.2.2: Objective tests.*) **That is highly specific** - The reported specificity of these tests is upwards of 95% with a number of programmes achieving referral rates of 2% or less. (See section *8.2.2: Objective tests.*)
3. There is an effective and accessible treatment or intervention for the condition identified	Many types of interventions are available, including cochlear implants (available through the North Island and Southern Cochlear Implant Programmes), hearing aids and FM (radio) aids (available through public audiology services), various types of speech language therapy, or teaching of manual language (eg NZ sign language), and other educational support. These interventions are provided currently by government funding with advisers on deaf children playing a key role in delivering and co-ordinating intervention services. There is good evidence that infants whose hearing loss is detected early and who receive appropriate habilitation have significantly better spoken language and subsequent educational outcomes than their late detected peers. This can be achieved by way of early language exposure via effective auditory stimulation using either hearing aids or cochlear implants. (See section *9.4: Do UNHSEI programmes result in earlier identification and intervention?* and section *9.5: Does early intervention lead to improved outcomes?*)

Criteria for assessing screening programmes	How UNHSEI meets these criteria
4. There is high quality evidence, ideally from randomised controlled trials, that a screening programme is effective in reducing mortality or morbidity	Evidence from randomised controlled trials (RCTs) is not available to determine the efficacy of UNHSEI. Results from RCTs may not become available as it may be considered prohibitively expensive, unethical and impractical to conduct randomised controlled trials on children with this relatively rare condition when an effective treatment is available[45-47].

However, there are a number of comparative studies which have provided detailed analysis of benefits resulting from early detection and intervention, including improvements in expressive and receptive language. These studies demonstrate that early intervention is associated with improved outcomes and evidence suggests such improvements in turn result in better longer term outcomes. (See section *9.5: Does early intervention lead to improved outcomes?*) |
| 5. The potential benefit from the screening programme should outweigh the potential physical and psychological harm (caused by the test, diagnostic procedures and treatment) | There is no physical harm caused to newborns screened with commonly used techniques. (See section *9: Universal newborn hearing screening and early intervention programmes: Issues and considerations.*)

There is potential psychological harm to parents due to increased anxiety (especially relating to false positives) resulting from the screening process. However, there is good evidence that the impact of screening on parents is not great and that it can be minimised through high quality protocols, the provision of accurate information and by requiring informed consent from parents before screening takes place. International experience shows that parental attitudes towards newborn hearing screening are on the whole positive, with very few parents refusing to have their infant screened. (See section *9: Universal newborn hearing screening and early intervention programmes: Issues and considerations.*)

The benefits of early identification of PCHI are well recognised, including by the Ministries of Health and Education. Outcome improvement data through to adulthood is not available due to the age of existing programmes and the high costs required to collect such data. However research indicates that earlier identification improves both language and educational outcomes and that many children born with PCHI who receive early intervention can function well in a mainstream environment, even achieving age appropriate speech and language. (See section *9.4: Do UNHSEI programmes result in earlier identification and intervention?* and section *9.5: Does early intervention lead to improved outcomes?*)

In addition, a national UNHSEI programme will result in a better understanding of the risk factors for progressive losses and potentially earlier identification of later developing losses as a result of increased awareness and parent education. (See section *9.4: Do UNHSEI programmes result in earlier identification and intervention?*)

Such a programme has the potential to reduce downstream costs, especially educational support costs. (See section *10: UNHSEI Programmes: Costs, benefits and cost effectiveness.*)

A programme of UNHSEI would reduce geographical inequalities that currently exist in service provision and therefore improve access to early and effective intervention. (See section *9.2.2: Potential benefits*). An effective universal programme would also reduce inequities due to ethnicity. For example, Māori have a higher prevalence of PCHI, the children are identified later and there is a longer delay before the hearing impairment is confirmed compared with non-Māori. UNHSEI would correct the inequities of later detection and intervention. (See section *7.7: Ethnic differences relating to the age of identification.*) |

Criteria for assessing screening programmes	How UNHSEI meets these criteria
6. The health system will be capable of supporting all necessary elements of the screening pathway, including diagnosis, and programme education	The current health and education systems have an appropriate support infrastructure for both diagnosis of and intervention for children with PCHI. The screening itself would require new personnel and systems. There is currently a shortage of qualified audiologists and specialist interventionists, but, with the likely staged roll-out of such a programme this could be managed. Further specialist training would also be required for the current workforce to improve skills in order to best serve the much younger children that will be diagnosed, although these children will be less delayed than those identified through the current system.
7. There is consideration of social and ethical issues	Universal newborn hearing screening programmes are well understood both overseas and in New Zealand and are acceptable to health professionals and key stakeholder groups. (See section *7.6.2: Existing regional UNHSEI programmes* and section *12: International support,* section *11: Local support and endorsements* and section *2.3: Consultative Group.*) Such a programme would allow equity of access to appropriate early health and education services for all babies born within New Zealand, regardless of ethnicity or geographic location. (See section *9.2.2: Potential benefits of UNHSEI and ethical considerations.*) Potential participants (parents and caregivers) in the screening programme would need to be provided with information allowing them to make an informed choice about their infant's participation. (See section *9.2: Potential benefits and harms of universal screening.*) Culturally appropriate, evidence-based information would be made available to assist with this decision-making process. This could be based on very well accepted information provided in relation to metabolic screening. There is a considerable international knowledge base relating to social and ethical issues which could be helpful in implementing a UNHSEI programme in New Zealand. (See section *9.2: Potential benefits and harms of universal screening.*)
8. There is consideration of cost benefit issues	This is not a condition which affects mortality. Furthermore, apart from the few cases where surgical or medical intervention is appropriate, early interventions are designed to manage the condition through medical, technological and behavioural interventions, supporting the family and the child through education. On the whole, the condition of hearing impairment remains. Within this context, new cost benefit information is being published on an ongoing basis from international UNHSEI programmes. The general consensus seems to be that these programmes are cost effective when compared to other approaches to identifying hearing impairment, other screening initiatives or to no screening at all. (See section *10: UNHSEI Programmes: Costs, benefits and cost effectiveness.*) Research is required to examine the cost benefit implications of introducing a UNHSEI programme in New Zealand.

Table 1: Compliance with National Health Committee Criteria for Assessing Screening Programmes

5. HEARING AND HEARING LOSS

This section provides a basic outline on the process of hearing, and describes the types of hearing loss, with a primary focus on children. The purpose of this chapter is to provide the reader with a background on hearing loss, in particular, Permanent Congenital Hearing Impairment (PCHI). PCHI is defined as a permanent hearing loss present at birth.

(A list of terms used within this document can be found in section *18: Glossary of terms.*)

5.1 HEARING LOSS IN CONTEXT

The World Health Organization (WHO) estimates there are some 250 million people in the world today with disabling hearing impairment[48]. Here in New Zealand, Greville estimates 400,000 (10.3%) people are affected by some kind of hearing loss[49]. (156,900 of this total were reported to require assistance for their hearing impairment, and 12,000 of the cases of hearing impairment were in children under 15 years of age[49]. The New Zealand Disabilities Survey Snapshot on sensory disabilities estimates that 18,300 children were 'deaf or had a hearing limitation that was currently not corrected'[50].) It is therefore not surprising that a significant proportion of the total disease burden in a given population results from hearing impairments. (The measure often used to calculate the burden of disease from hearing impairments is the Disability Adjusted Life Year [DALY]. This is calculated as the sum of the years of life lost due to either premature mortality and years lived with disability [YLD]. The YLD is in turn calculated by multiplying the number of cases of the condition by its average duration and then by the conditions specific 'disability weighting'.)

This burden is calculated considering the relative weighting of hearing impairment when compared to other conditions. *Table 2* shows disability weightings for various common conditions as described by a commonly cited Dutch study[51]. Deafness is highlighted in the 0.30-0.40 weighting category.

Condition	Weight
Gingivitis, caries	0.00-0.01
Mild depression, osteoarthritis (grade 2), epilepsy	0.10-0.15
Moderate depression, relapsing MS, severe asthma, chronic Hep B, **deafness**	0.30-0.40
Disseminated cancer, severe dementia, severe schizophrenia, quadriplegia	0.80-1.00

Table 2: Commonly used Years Lived with Disability (YLD) weightings

An example of the relative burden can be found in the Victorian Burden of Disease Study[52] which examined the burden of non-fatal disease in 1999 and projected the burden to 2016 in Victoria, and found mental disorders were likely to be the leading cause of disability, accounting for 26% of the non-fatal burden in Victoria. This was followed by nervous system and sense organ disorders, including hearing impairment (17%) and chronic respiratory diseases (9%).

A 2001 report examining the non fatal disease and injury burden in New Zealand listed hearing disorders ninth among 1996 YLD rankings[53].

The burden relating to permanent hearing impairment consists of hearing losses such as late onset, noise induced and congenital hearing impairment. These hearing impairments span all degrees of hearing loss.

This document focuses on permanent congenital hearing impairments (PCHI). The WHO acknowledge that this type of hearing loss retards children's development as it causes delays in language acquisition and impedes school progress[48]. In New Zealand, our best estimate of the size of this group among under 21 year olds is 2,800. This represents the number of children with permanent hearing loss who were accessing hearing aid funding[49]. Overall, approximately 11,000 – 16,000 New Zealanders are thought to have a permanent congenital hearing impairment. See section *7.4: Number of New Zealanders affected by PCHI* for further information.

In terms of relative disease burden, the authors of this report could not find weightings specific to each degree of PCHI. Rather, as seen in *Table 3*, the relative disease burden of PCHI seems to be calculated using the same weightings as adult onset deafness of the same degree. This raises a number of issues, as PCHI contributes significant developmental delays, in addition to the functional implications seen in adults with late onset or noise induced hearing impairment.

Condition	Weight
Overall weighting for hearing impairment	0.234
Congenital deafness	0.234
Early acquired severe, through otitis media	0.233
Mild permanent early acquired	0.110
Neonatal causes – severe hearing loss	0.370
Adult onset hearing losses	
Mild	0.020
Moderate	0.120
Severe	0.370

Table 3: Commonly cited hearing related YLD weightings[51]

In conclusion, hearing impairment contributes significantly to both the worldwide and New Zealand disease burdens. Although there may be issues in the calculation of the specific disease burden created by PCHI, all types of hearing impairment are known to have a significant impact on the Years Lost to Disability.

5.2   HOW WE HEAR AND TYPES OF HEARING LOSS

The auditory (hearing) system is a complex pathway that comprises the ear, neural pathways and auditory centres in the brain.

Sounds, such as speech, are detected by the ear and the complex characteristics of the sound are represented by activity in the auditory nerve to be interpreted by the brain.
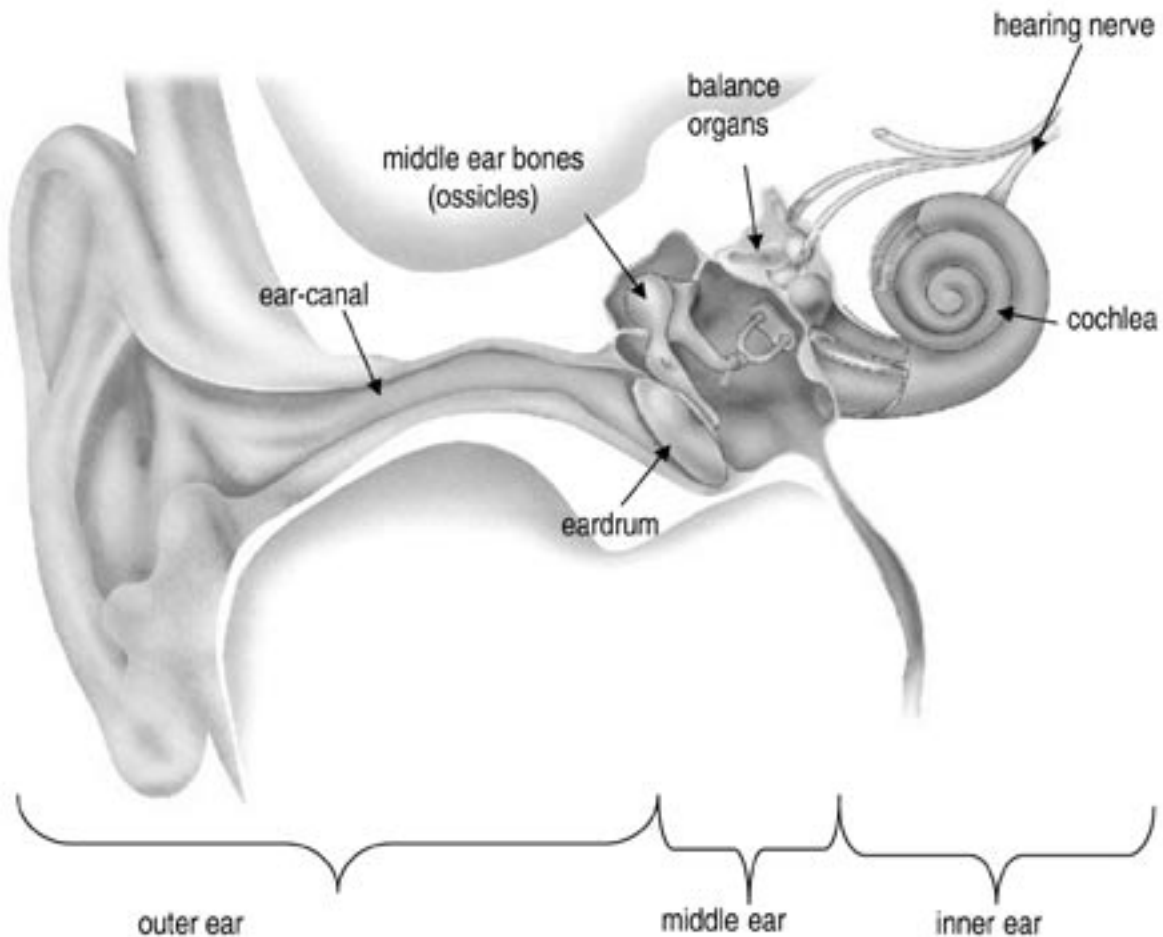
Figure 1: Anatomy of the ear (Adapted from an original diagram courtesy of Oticon)

As seen in *Figure 1*, the ear comprises three parts: the outer ear, middle ear and inner ear. The normal process of hearing requires correct function of all these parts and of the neural pathways to and within the brain.

- **The outer ear** consists of the external cartilaginous part of the ear and the ear canal. The eardrum is located at the end of the ear canal, and forms the boundary to the middle ear. The primary function of the outer ear (or pinna) is to collect and transmit sounds to the middle ear. Sound waves travel through the outer ear canal and strike the eardrum (tympanic membrane), which then vibrates.

- **The middle ear** is normally filled with air. The Eustachian tube, which connects the middle ear to the back of the throat, functions to keep the air pressure in the middle ear the same as external air pressure so that the eardrum is not under tension. There are three tiny bones (ossicles) in the middle ear cavity; the malleus (hammer), incus (anvil), and stapes (stirrup) that vibrate to the rhythm of the eardrum, amplifying the sound and passing the sound waves on to the inner ear.   Congenital abnormalities and acquired diseases of the outer and middle ear cause a conductive type of hearing loss because they reduce the conduction of sound to the inner ear.

- **The inner ear** is a series of fluid-filled cavities deep in the temporal bone of the skull. It contains two sets of sensory organs, the hearing organ or cochlea and the balance organ or vestibular labyrinth. The cochlea is shaped like a snail-shell (with two-and-a-half turns) and is connected to the balance organ, which is why hearing and balance disorders often occur together. The cochlea contains the hearing sensory organ lined with approximately 20,000 hearing sensory cells, or hair cells. These cells are attached to the auditory (hearing) nerve fibres. Vibrating sound waves cause ripples in the sensory organ, which in turn stimulates the sensory cells, sending impulses along the auditory nerve. Different parts of the cochlea respond to different frequencies (pitches) of sound, enabling us to hear different frequencies. Diseases or congenital abnormalities in the inner ear can cause a sensorineural type of hearing loss. Selective damage to different parts of the cochlea affects the ability to hear specific frequencies.

- **Auditory pathways and centres of the brain**: Approximately 30,000 nerve fibres are connected to the hair cells of the cochlea. These nerves make up the auditory or hearing nerve which passes from the ear to the lower part of the brain. Within the lower brain there are a number of parts that respond to sound and send response to the hearing centre in the auditory cortex. These auditory pathways and other parts of the brain are all involved in the complex interpretation of the sound messages from the ear to provide our sense of hearing. Abnormalities of the auditory brain pathways lead to complex problems of processing, interpreting and understanding sound. These are commonly referred to as auditory processing disorders.

In summary, hearing loss is often categorised by the part of the auditory system that is affected. There are three basic types of hearing loss: conductive hearing loss, sensorineural hearing loss and auditory processing disorders. In addition, some individuals will have 'mixed' hearing loss, where there is both a conductive and sensorineural component to the hearing loss. Conductive hearing loss is due to abnormalities in the outer and/or middle ear, sensorineural due to disease of the inner ear or cochlea and auditory processing disorder refers to a problem in the auditory pathways (leading from the cochlea to the auditory cortex of the brain) or with associated regions of the brain itself.

PCHI can be either sensorineural or conductive in nature. The primary aim of a universal newborn hearing screening and early intervention (UNHSEI) programme is to provide intervention as early as possible for children with PCHI.

5.2.1   Describing hearing loss

In addition to the type of loss, hearing losses are also described by configuration and degree.

Configuration: The 'configuration' (shape) of the hearing loss refers to the extent of hearing loss at each frequency and the overall picture of hearing that is created. 'Hearing thresholds' are typically measured using an audiometer and plotted
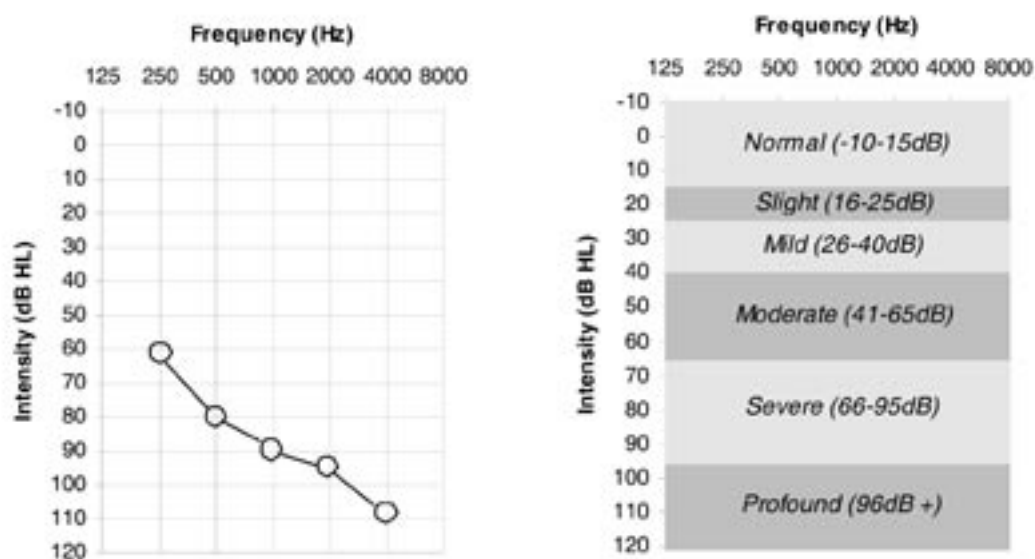

Figure 2: Sample audiogram showing a severe-profound hearing loss (left), and an audiogram showing the categories of hearing loss used in the Deafness Notification Database (right)

on an audiogram, which is a graph of an individual's hearing thresholds in dBHL (decibel hearing level) as a function of frequency *(Figure 2)*. A normally hearing person would have hearing thresholds between -10 and +15dBHL, based on the average thresholds of a group of young people with no history of ear disease. The frequencies tested span the range 125-8000 Hz, which includes the range of frequencies used in speech. Hearing loss may occur in many different configurations from losses at only the high, middle or low frequencies to one which is similar across all frequencies or various combinations in between.

Degree: The extent of hearing loss is often categorised by degree as mild, moderate, severe or profound. These categories are defined by the average of the thresholds across these frequencies in the better hearing ear *(Figure 2)*.

Although an individual's hearing loss may not fit within a particular band of severity across all frequencies, the overall degree of hearing loss tends to reflect the level of hearing disability a person will experience. The degree of loss is therefore determined by the average loss across frequencies spanning 500-4000Hz. The following descriptions aim to demonstrate how, in general, the severity of a hearing loss is likely to affect an individual. These descriptions have been adapted from Australian Hearing's *Choices* Book[54].

Mild Hearing Loss (26-40dBHL)	Persons with mild hearing loss have some difficulties hearing soft speech and conversations but can often manage in quiet situations with clear voices. Persons may sound as though they are mumbling to the person with a mild hearing loss. Speech and language usually develop normally if a child is fitted early with hearing aids.
Moderate Hearing Loss (41-65dBHL)	Persons with moderate hearing loss will have difficulty understanding conversational speech particularly in the presence of background noise. Volume on the television and radio would have to be turned up to be heard. Speech and language development are generally affected if a hearing aid is not provided early. Hearing aids assist most hearing difficulties if speech discrimination is good and the listening environment is not too noisy.
Severe Hearing Loss (66-95dBHL)	For those with losses in the most severe side of this category, normal conversational speech is inaudible. Only raised voices at close distance can be understood by those with less severe losses in this group. Speech and language will not develop spontaneously in children with this degree of hearing impairment. Hearing aids will amplify many speech sounds and will greatly assist a child with severe hearing loss to develop speech, although speech quality is likely to be affected. Some children with severe hearing loss will obtain benefit from a cochlear implant.
Profound Hearing Loss (96+dBHL)	Learning to speak without significant habilitation and support is very difficult for children born with a profound hearing loss but there are differences between individuals. There is greater inconsistency in the benefit derived from hearing aids for this group. Some profoundly deaf people can understand clear speech in quiet conditions when they are wearing hearing aids. Others derive little benefit. Children with a profound hearing loss will be considered for cochlear implants and will obtain benefit from this procedure, especially if implanted young.

5.3   NORMAL HEARING DEVELOPMENT

The development of hearing is complex and is only briefly described here to provide a context for subsequent discussions. For those interested in obtaining more detail, excellent reviews of the development of the auditory system and hearing are provided by Downs and Northern[55], Werner and Marean[56] and Rubel and colleagues[57].

The ear, particularly the inner ear, is well formed by the end of the first trimester. There is little change to the structure of the inner ear after birth but changes do occur to the outer and middle ear during childhood, especially within the first six months. The central auditory pathways also continue to develop structurally after birth (eg increased myelination and size of nerve cells in the auditory centre in the brain). The foetus seems to respond to high intensity sounds from about 22 weeks gestation, and this is confirmed by observations of premature babies born at 25 weeks who are shown to be able to detect and respond to sound[58]. Following birth, there are continuing improvements in many indices of hearing function, for example, absolute sensitivity, frequency discrimination and sound localisation. Many indices of hearing continue to improve into the mid to late teens. The relationship between hearing development and auditory and non-auditory factors is not well understood, but it is clear that many of these processes require auditory stimulation.

6. EFFECTS OF PERMANENT CONGENITAL HEARING IMPAIRMENT

Permanent Congenital Hearing Impairment (PCHI) is an important public health issue both in terms of prevalence (See section *7.2: Prevalence.*) and prognosis. This section discusses PCHI and its far-reaching effects on a child's ability to acquire language, develop cognitively, achieve in education, function socially, and establish good self-esteem. This section also discusses the effects of hearing loss on mental health, employment, vocational choice, and family dynamics. It is important to note that development in areas discussed within this section are interconnected, so developmental delays in one area will negatively influence development in other areas, compounding the direct effects of the hearing impairment on each area.

Four key variables are known to increase the impact of the hearing impairment

- Age of onset

- Severity of the loss

- Intervention delay

- The presence of other disabilities

To complicate the picture, less severe hearing losses can have potentially greater impact as they are often detected later, allowing time for deficits in speech and language to develop.

Notes on terminology

People with a hearing impairment may refer to their hearing status or cultural affiliation through use of many terms, including 'deaf', 'Deaf', or 'hearing-impaired'. These terms can have different meanings when used in different contexts, or when used within different groups. The term 'Deaf' is usually retained for those persons who align themselves with 'Deaf Culture' and generally, use sign language. The terms deaf and hearing-impaired usually relate to the broad spectrum of hearing loss from mild to profound. Because of the differing usage, and to improve readability of this document, the authors use the term 'hearing-impaired' to describe all persons with hearing impairment, unless reporting research findings, in which case the original author's terminology is retained or when specifically describing the Deaf community.

6.1   LANGUAGE ACQUISITION

One of the key effects of PCHI is language delay. This effect is well studied and language delays have effects on other areas such as educational performance, and cognitive and social development, which are discussed later in this section. This section examines normal language acquisition and then compares this to acquisition of language in children with PCHI. Current understanding of so-called 'critical' or 'sensitive' periods for language acquisition is also discussed.

6.1.1   Normal language acquisition

Language and theories of language acquisition

Language has been called the symbolisation of thought. It is a learned code, or system of rules that enables us to communicate ideas and express wants and needs. Language falls into two main divisions: receptive language (understanding what is signed, written or spoken); and expressive language (speaking, writing or signing)[59].

Contributions towards our understanding of language have come from a wide range of disciplines, combining efforts of psychologists, linguists, educators, neuroscientists and communication scientists. Language development can be divided into first and second language learning with the literature on first language learning being most relevant to child development while second language learning pertains to all ages, including adults.

Theories of language acquisition can be grouped broadly into those based in empiricism (focusing on the dominance of environmental influences, eg Vygotsky[60, 61], Skinner[62], Piaget[63, 64]), nativism (focused on the dominance of genetic factors, eg Fodor[65, 66], Universal Grammar Theory[67, 68]) or more recent interactionist theories (focusing on the interaction between environment and genetics[69]).

Language development

Development of signed or spoken language requires the learning of phonology (learning to understand and communicate the speech sounds or signs in the language), syntax and morphology (the grammatical rules), and vocabulary and semantics (meaning of words or combinations of words)[70-74]. The learning of this framework proceeds in a similar way and along similar timelines for both spoken and signed languages[75, 76]. Acquisition of spoken language is also related to the acquisition of listening, speaking, reading, and writing processes[77, 78].

Linguists classify speech sounds contained within a language into a number of categories called phonemes, which are contrasting units in a given language. Each of these phonemes has its own frequency pattern, which distinguishes it from other phonemes. Phonemes are the basic building blocks of all spoken languages. *Figure 3* shows the key frequencies or phonemes of English speech, superimposed on a 'pure tone' audiogram showing the loudness and approximate frequency of each speech sound. For example, the vowel sounds are dominated by low frequencies whereas the consonant sounds predominately contain higher acoustic frequencies. The sounds are enclosed by the 'speech banana', which is used by audiologists to illustrate the range of hearing required to perceive speech sounds.

There is evidence that some aspects of language in humans are innate, indicating that centres in the brain associated with language are already developed to some extent at birth, providing a foundation for language acquisition[79, 80]. Even before birth, normally hearing babies listen to sound, and begin to form a phonemic map specific to their language. This process continues after birth, and normally around one year of age the baby will put together the first set of phonemes to say his or her first word. As the baby practices these speech sounds he or she will lose the ability to say or accurately hear speech sounds that do not occur in the language to which they are exposed[81].

The ability to hear the key speech frequencies is therefore crucial to ensuring early vocal development[82, 83] as babble informs the infant how to produce different sounds required for spoken language[55]. This can be seen in the strong positive relationship between the frequency of input and expressive language abilities among children, with those exposed to a greater number of utterances showing higher levels of expressive language[84].
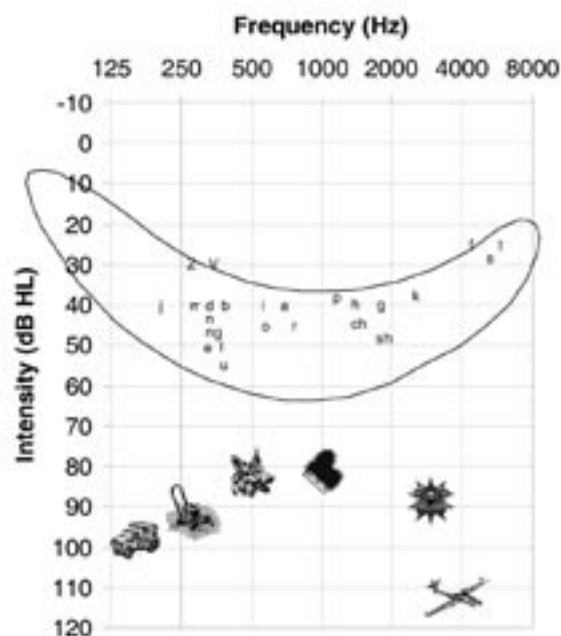
Figure 3: Audiogram illustrating key frequencies in speech sounds. Adapted from Downs and Northern (1999)[55]

Language development relies on social interaction, and children's comprehension grows as they discover what another person is thinking through language[85]. Adults assist a child's language development in a number of ways including paraphrasing, expansion, discussing the child's interests, listening to the child and asking specific questions. Reading aloud can provide important opportunities for such learning, and it has been shown that children who are frequently read to often develop language earlier than other children[85].

By the time a child is three he or she can usually use 900 to 1,000 different words, understanding about 20,000 words by the age of 6 [85]. This happens as the child moves from short simple sentences, to using ones with multiple clauses, plurals, possessives, declaratives, conjunctions and prepositions.

6.1.2   Language acquisition in deaf and hearing-impaired children

Hearing-impaired children may acquire either spoken language (through access to key speech frequencies) or signed language. As previously discussed, acquisition of both these types of language requires the learning of phonology,

morphology, syntax and semantics, and proceeds along similar timelines[75, 76]. Prelingually hearing-impaired children who have not had early exposure to language (signed or spoken) display significant delays in language acquisition[86-90].

Critical and sensitive periods

A critical or sensitive period is a period in which 'a specific stimulus (such as sound) is required for normal development of the system (such as the auditory part of the brain) and during which the organism is vulnerable to environmental manipulation'[91]. Critical periods by nature begin and end abruptly. A sensitive period on the other hand begins and ends more gradually and this is considered to be a time during development in which the organism is maximally sensitive to environmental factors. As there is no definitive conclusion in the debate on whether critical or sensitive periods are involved in language acquisition, these terms we will use interchangeably throughout this document.

The concept of 'critical' and 'sensitive' periods relies on plasticity. Plasticity is the term given to the ability of the brain to alter structurally and functionally in response to experience and stimulation. Brain plasticity occurs throughout life but it is often classified into different types reflecting various stages of life or response to injury. For example, during development, rapid changes (*developmental plasticity*) in brain structure and function occur when the immature brain begins to process sensory information (eg auditory information). Damage to the body (for example damage to the ear) can cause changes within the brain (*injury-related plasticity*) because of an alteration in the balance of sensory input to the brain. Finally, *learning or memory-related plasticity* underlies the changes in behaviour that occur as we receive new sensory information. Adjustments in the strength of connections between brain cells and an increase or decrease in the connections seems to underpin all forms of plasticity. The ability of the brain to adapt to new stimuli and experience is the essential basis of learning and is based substantially on the reception of sensory information, such as sound.

Critical or sensitive periods and language acquisition

Evidence for the existence of critical or sensitive periods has been found in a number of sensory systems (eg visual and somatosensory) and across many species[9, 70, 86, 92]. Evidence indicates that such periods also apply to the auditory system[93].

The theory that a child's ability to learn language is determined by exposure to spoken language during early brain development has existed since early last century[70]. Evidence contributing to the present view on the plasticity of the auditory system is derived from a number of sources including: studies on second language acquisition, 'wild-child' research, clinical audiology experience, physiological measurements, behavioural modification experiments, cross-species research and more recently, research examining the differential effectiveness of hearing devices such as cochlear implants[8, 94-97].

There is strong evidence that learning-related plasticity in the auditory cortex deteriorates with age[2-4, 98]. For example, young congenitally deafened animals who are provided with cochlear implants develop normal sound-evoked cortical potentials (sound-induced electrical activity in the cortex) compared with abnormal potentials in older implanted animals[2]. This demonstrates a higher level of auditory plasticity in younger animals.

Lennenberg[70] asserted the existence of a critical period for language acquisition, citing three key observations as evidence. Firstly, that deaf children hearing speech early in life seemed to learn language more easily. Secondly, that acquisition of language in deaf and intellectually handicapped children stopped at puberty. And finally, that children suffering from brain injury recovered linguistic ability if other parts of the brain remained intact, whereas the loss of language was irreversible in adolescents and adults[70].

The study of Genie and Victor[85] (so-called 'wild-children') who were found after years of social and auditory deprivation, and were unable to speak, provides an interesting observation of the impact of critical periods on language development. Although studies of these children provide some evidence that later first language development is limited, alone they are not sufficient to prove the existence of critical periods, as it is impossible to separate the effect of the many confounding factors involved. In the case of Genie, who was found in 1970 at age 13 after years of mistreatment, social isolation and

abuse probably retarded her socially, emotionally and cognitively[85]. Together, and with a lack of auditory stimulation, these effects probably contributed to her subsequent inability to acquire normal language skills, despite significant remedial assistance.

The limitations of such studies have led to the study of deaf children who are without early language but are raised in otherwise normal environments[85, 99]. These include a study of deaf individuals in Nicaragua, in which some of the confounding factors existing in studies of 'wild children' are not present. In this case, until around 1985 there was no deaf education or sign language available. As assistance for this group was provided, educators found around 300 deaf individuals, many of whom had been raised in otherwise supportive environments, who were incapable of learning effective language skills[99].

Other studies, such as those of Newport[10] and Neville[100], have examined native learners of sign language and compared their language proficiency to that of early learners (exposed to sign between four and six years of age) and late learners (exposed after the age of 12). Children exposed later to sign language showed a lower proficiency than their early identified counterparts[10, 100].

Similarly, a study of children with cochlear implants which measured cortical auditory evoked potential (sound evoked electrical activity in the brain as an indicator of central auditory maturation) showed children with the longest period of auditory deprivation prior to cochlear implant had abnormal cortical latencies in response to speech. In contrast, children implanted before three and a half years, showed age appropriate cortical responses six months after implantation.[101]

Most studies on *second* language learning indicate grammatical outcomes worsen considerably when exposure to the language is after the age of eight[74, 86, 102, 103].

Similarly, studies of congenitally deaf children who receive cochlear implants have established that maturation of the auditory system is limited by the duration of the deafness prior to implantation[104] and that there is a sensitive period in the first few years during which the auditory system shows maximal plasticity[105, 106]. Children implanted after about 3.5 years of age show poorer auditory cortical function[107]. Correspondingly, the loss of hearing function in adults leads to rapid and profound changes within the auditory centres of the brain[93].

Timing of sensitive or critical periods

As outlined earlier, development of language requires the learning of phonology (learning to hear and speak the speech sounds in a language), its syntax and morphology (the grammatical rules), and vocabulary and semantics[70-73]. Of particular interest for newborn hearing screening is the timing and length of the sensitive period for language acquisition.

Current evidence suggests that the acquisition of syntax relies on learning during 'sensitive periods', mainly within the first three to four years although acquisition of syntax can occur up to the age of eight years[108]. The ability to acquire phonology appears to be more pronounced in the first six months after birth and decreases rapidly after that time until the child reaches four years of age[31, 32]. The sensitive period for acquisition of semantics is thought to be longer and may last until the end of the 15th or 16th year of life. Vocabulary is acquired throughout life, and research suggests adults have an advantage in increasing their vocabulary. There appears to be a lack of a critical period for learning of vocabulary as demonstrated by second language learning among adults and the fact that children exposed to language late in life are able to acquire new words[86]. Mayberry and colleagues demonstrated that normal grammatical acquisition (signed or spoken) occurs only when subjects are exposed to language (signed or spoken) in early life[5]. There is also some evidence suggesting that the rate of language development is set within the first year of life and is resistant to change after that time[47].

Kuhl and colleagues examined normally hearing 6 month old infants from Sweden and the United States, and found that exposure to a particular language in the first half year of life alters phonetic perception, providing evidence that auditory stimulation in the first six months is critical to the development of speech and language[29].

Although some success can be seen in prelingually deaf children who are fitted with a cochlear implant after a long period of deafness[109], there seems to be a general consensus that the greatest benefit for auditory intervention with deaf and hearing-

impaired children lies within the first three years of life[30, 110-116]. Children born with permanent hearing impairment can acquire spoken language normally if they are exposed to sound through amplification in early development[5-12], although this ability diminishes with age.

Where exposure to sound is absent during these critical/sensitive periods of development it becomes very difficult to fully redress the deficit incurred by an absence of normal social and linguistic experience. This is supported by evidence of improved outcomes in children who are detected early and receive appropriate amplification. This is discussed in section *9.5:Does early intervention lead to improved outcomes?*

6.1.3   The effects of hearing loss on spoken language development

In practical terms, it is difficult to learn the sounds associated with speech without good access to sound in the relevant speech frequencies. As a result of limited access to speech, hearing-impaired children display fewer and shorter spoken utterances, have less developed articulation, and have speech with abnormal spectral characteristics[89, 90, 117].

Hearing-impaired children's spoken language development differs from that seen in normally hearing children with a lack of canonical babbling observed[82, 118, 119].

Lack of access to early audition has significant effects on language development as the child matures. For example, one US study examining 89 prelingually deaf children found that on average, without amplification, these children could only be expected to achieve 5 months of expressive language growth per year in the early years of language development[120]. Thus, the gap between the chronological and language age of the hearing-impaired child widens as the child ages[90, 121].

In her book *Language Disorders From Infancy* Paul describes how hearing-impaired children fall behind their peers demonstrating: poor conversational initiation, poor ability to respond to the initiations of others, and difficulty in using the rules of entering and continuing conversation, particularly in the classroom[122].

Further information on the key milestones for language development in children can be found in *Appendix 16.3: Milestones in development of speech and language.*

6.1.4   Conclusions

Overall, there is a high degree of plasticity within the central auditory and language areas of the brain during early development. Without early auditory input spoken language development is delayed[105]. The exact length of the sensitive or critical periods for learning foundation language skills are still not known, but there is evidence that exposure to language (signed or spoken) within the first three to four years of life is essential to allow optimal language development. Normal language development is also reliant on normal social-emotional interactions and this is discussed in section *6.4: Social functioning, mental health and self-esteem.*

6.2   COGNITIVE DEVELOPMENT

Although lesser degrees of hearing impairment do not impair basic cognitive development, the more severe degrees are known to effect later stages of cognitive, language and socio-cognitive development[123, 124].

In children with more severe hearing impairments, 'cognitive development proceeds but is limited to the results of direct experience without language'[123]. Although a mild or moderate hearing loss may not directly affect cognitive development, distorted auditory perception along with less than optimal parental attachment are believed to lead to sub-optimal cognitive development[125]. Normal cognitive development is also reliant on normal social-emotional interactions. This is also discussed in *section 6.4 Social functioning, mental health and self-esteem.*

Once cognitive concepts become more abstract, language is required for interpretation and understanding, for example, in the use of concepts such as before and after. By the time a child reaches the age of three many concepts to be learned are of this more

abstract type, and language is needed to relate to concepts outside direct experience and to give meaning to experiences[76, 126].

6.3 EDUCATIONAL OUTCOMES

The following discussion on educational outcomes is based on a review of research on hearing children for whom intervention was provided at various ages. As such, many of the educational outcomes discussed do not reflect the result of early detection and intervention. These studies are not limited to children with PCHI but may include those who have acquired hearing losses.

6.3.1 Learning foundations

Hearing-impaired children are more often without the age-appropriate language and communication skills when compared to their hearing counterparts. These deficits in communication mean it is more difficult for children with PCHI to succeed in acquiring early learning foundations. This in turn makes it more difficult for these children to perform in an education system that requires direct communication with teachers and peers[77, 78].

Interestingly, along with vision and touch, hearing may also contribute to development of coordinated physical movement which contributes to positive behaviour at school and is essential for 'school readiness'[127, 128]. Because a hearing-impaired child has a lower level of 'readiness', he or she is unable to take advantage of the many opportunities available at school to learn and practice skills that are required for a healthy lifestyle[129]. In addition, there is evidence to suggest that there is a positive relationship between academic performance and psychological adjustment[130]. It could therefore be assumed that poor academic performance in hearing-impaired children could contribute to poor psychological adjustment.

Approximately 90% of a young child's knowledge can be attributed to incidental reception of language around them[131]. Without good audition, hearing-impaired children are limited in their ability to learn incidentally, and their knowledge base inevitably falls behind that of their peers as a result[131].

For students with disabilities, psychological adjustment and quality of life are not directly related to their given health condition, but are the product of the interaction of the student with his or her social and physical environment[132]. However, the negative effects of the disability can be ameliorated or removed by appropriate and timely intervention.

Without appropriate intervention, hearing impairment also has an ongoing impact on communication which in turn limits subsequent social access and participation. This increases the likelihood that children will suffer from isolation and disrupted social relationships as they mature[34]. Similarly, it is very difficult for hearing-impaired children without spoken language to become strong readers and writers, another key for performance in our educational system, and a key to independence later in life[34].

6.3.2 Achievement levels of deaf and hearing-impaired people

Deaf and hearing-impaired people, including those with mild losses, have significantly lower educational achievement levels than their normally hearing counterparts[55, 128, 133-138].

Reading and mathematical performance have been the focus of most of the research into the educational performance of deaf and hearing-impaired students. Reading requires familiarity with a language and an understanding of the mapping between that language and the printed word. Children who are profoundly deaf are disadvantaged on both counts making reading difficult[34]. The frequency with which children are read to, and the methods used, are known to affect how soon children read and their reading proficiency[85]. In a study undertaken in the UK in 1979, deaf school leavers were shown to have an average reading age of 9 years[140].

In a large study of over 1,000 pupils (including 540 deaf pupils) in the UK, Wood and colleagues reported an average maths age of 15.5 years for the hearing pupils and 12.3 years for the deaf pupils[141]. There does not appear to have been a more recent study that defines the current situation in the UK.

Three large scale studies in the United States have demonstrated that hearing-impaired students have lower reading and mathematics achievement levels. The Center for Demographic Studies and Assessment (CADS) at Gallaudet University examined the academic achievement levels of deaf students in the US in 1974, 1982, and 1990 using the Stanford Achievement Test - Hearing-Impaired Edition (SAT-HI). The average achievement levels in reading comprehension and mathematics of deaf students in special education at age 17 were comparable to hearing students at 8 or 9 years of age[142]. Other US studies support this assertion, finding that the reading comprehension of many deaf and hearing-impaired children reaches a plateau at about the third or fourth grade level[13, 142, 143].

As a group, those deaf adults using manual communication methods alone have lower educational levels, lower family income, are more likely to be unemployed, and have a particularly poor assessment of well-being compared with hearing individuals[144].

Powers studied moderately, severely and profoundly deaf pupils in all types of mainstream schooling in England and showed significant differences between the exam successes of deaf pupils compared to their hearing counterparts. For example, in 1995, only 14% of deaf students achieved 5 or more 'A' to 'C' grade passes in national examinations compared with a 44% average for hearing students[145].

A recent review of the performance of deaf and hearing-impaired children in Australia reports that the mathematical achievements of hearing-impaired children are significantly lower than those of their hearing peers[14]. This assertion is supported by a recent study examining long term outcomes of hearing loss in a cohort of 86 seven and eight year old children born in Victoria, Australia, who were fitted with hearing aids before 4.5 years of age and who had no additional disabilities. Although non-verbal IQ in the hearing-impaired group was in the normal range, the sample scored 1.3-1.7 standard deviations below the normative population on language tests.

Unfortunately, academic performance of hearing-impaired persons in New Zealand has not been studied extensively. A small random sample study commissioned in 2000 by Special Education Services[146] demonstrated that the more severe the hearing loss the further behind children were from their peers. It also showed that Māori and Pacific children in the study were among those with the most significant educational disparities. The educational gap between the hearing and hearing-impaired children included in this study increased with age, in line with international research. This study also underscores the critical nature of identity development, particularly during adolescence, for deaf and hearing-impaired children[146].

Again in New Zealand, Pritchett studied the comprehension skills of severe to profoundly deaf 9-19 year olds, finding that 47% were at the low end of comprehension rankings compared to just 4% of their hearing counterparts[147].

6.3.3 Effect of degree of hearing loss on educational performance

The US Preventative Services Taskforce recognises hearing losses of greater than 35-40dB as having educationally significant and potentially devastating impacts on children's speech, communication and general development[148]. However, there is a growing body of literature that indicates even slight and mild losses impact on a child's ability to speak, learn and, therefore, perform well educationally[1, 128, 149-151]. Increasing numbers of children with milder hearing losses are receiving treatment for losses previously thought of as 'insignificant', both in New Zealand and overseas[152, 153]. Similarly there is evidence that children with unilateral sensorineural hearing loss regularly experience complications in communication, behaviour and education[88, 137, 153].

The degree of hearing loss has a significant effect on speech outcomes[154]. Academic achievement is not, however, simply related to degree of impairment, since children suffering from milder forms of hearing loss still perform well below the levels of their hearing peers[141, 155-157]. It is possible that the effect of degree of loss has not been observed due to confounding factors[154] that cannot be separated, such as intelligence; peer, teacher and family support; and quality of amplification devices[154].

6.4   SOCIAL FUNCTIONING, MENTAL HEALTH AND SELF-ESTEEM

Healthy social-emotional development is vital, not only for future emotional health, but also for normal cognitive functioning and language development[85].

Mothers and babies contribute to the strength of their attachments through their personalities and the way they behave toward each other[85]. Mothers of securely attached children are likely to be sensitive and responsive and the more secure the attachment between a child and his or her parent, the more likely it is that the child will become independent of that parent and develop good relationships with others as they mature[85]. An example of the social impact of hearing impairment is the effect on infant-parent attachment. Mechanisms by which secure attachment is formed between an infant and parent can be impeded through lack of access to sound, or through altered signals as a result of undiagnosed hearing loss. Opportunities for emotional intimacy are less when a child cannot hear or is hearing-impaired[85]. As a result, hearing loss may make it more difficult for the hearing-impaired child to become independent and develop other healthy relationships.

As a group, it has been demonstrated that hearing-impaired children rate poorly in social functioning, mental health and self-esteem compared with hearing children. As an example, the US Preventative Services Taskforce reported that 31% of US children with sensory disability (loss of hearing or vision) were sad, unhappy, or depressed, compared with only 17% of children without such disabilities[148]. In part, this was attributed to frustration with environmental barriers that limit the child's ability to fully engage in home, school, community or social activities[148].

Research into the effects of hearing impairments also demonstrate increased behavioural difficulties, delayed social development, increased social needs, and a higher need for mental health services among the hearing-impaired population. The Gallaudet Research Institute's 1997-1998 annual survey of deaf and hard of hearing children and youth in the United States found that half of the students surveyed had communication difficulties, and that almost half reported at least one cognitive, behavioural, or social limitation[158].

Zingeser demonstrated that communication disorders such as hearing impairment not only impact on speech, language and the development of social skills, but that they may also affect social and emotional development[159]. It has also been demonstrated that hearing impairments, especially those undiagnosed or diagnosed late can lead not only to poor language skills and academic performance but also to lack of motivation, feelings of frustration and alienation, academic problems, and inadequate social skills through poor communication competence[160-162]. These effects may in turn increase the risk of children dropping out of school, gang involvement, juvenile delinquency, and eventual adult criminal behaviour[159]. Recent studies support this assertion. For example, there is a much higher prevalence of hearing loss in prison populations in the US, than that found in the general population[163].

Even *mild* PCHI is thought to contribute to problems in the areas of social and emotional function[164] in addition to reducing educational performance, although the effects are often more difficult to attribute to hearing loss due to it's mild nature[164].

This area is not well researched in New Zealand, although there have been three small studies.

A small random sample study of deaf people was conducted in 2000, a study of 222 randomly selected hearing-impaired children with varying levels of need, across differing ethnic groups and regions concluded that the academic performance of this group was lower than that of hearing peers, and that more than half the students had significant social and personal development needs, were socially isolated, and displayed anti-social behaviour or low self-esteem[146]. These problems appeared to be worse in older children[146]. The report summarises the views of the only counsellor employed specifically for Deaf and hearing-impaired children in New Zealand, concluding that '[Hearing-impaired children]…often feel left out and not important because they cannot easily access the information around them….Children need all avenues explored to give these children language[146].'

Kent examined identity issues of hearing-impaired young people aged 11, 13 and 15 and compared these to a group of hearing students of matched age, gender and ethnicity[165]. This study found the hearing-impaired group were significantly more likely to experience a sense of loneliness. Those hearing-impaired in the sample who were willing to self-identify were also significantly more likely to experience bullying[165].

Finally, a national survey of 200 randomly selected deaf adults in 2000, examined mental health among deaf people and showed that the deaf community has a higher risk of mental health problems and higher need for mental health services[166].

In conclusion, hearing impairment can have significant negative implications for social functioning, mental health and self-esteem. Without healthy social-emotional development, cognitive function and language development of hearing-impaired children suffer.

6.5   VOCATIONAL CHOICE AND EMPLOYMENT STATUS

Not only does hearing impairment restrict vocational choice, even among very well educated hearing-impaired adults[134, 167, 168] but hearing-impaired people are more likely to be unemployed than hearing people[133, 169]. Crammatte conducted two landmark studies in this area within the United States. The first in 1968 showed that only 6.6% of employed deaf respondents were in professional, technical or kindred occupations compared with 10.6% of the general population[170]. In a follow-up study in 1987, Crammatte determined that although deaf people were employed in a wider range of occupations than in the previous survey, they still worked in a narrow range of occupations, with 63% working in education, compared with only 10% in the general population[171]. The definition of 'deaf' within this research was that none of the sample could use the telephone unaided and therefore required visually oriented communication[171].

These findings were supported by Schein and Delk who discussed the channelling of deaf people into a narrow band of occupations. For example 60% of deaf white males were working as linotype operators at this time[134]. In addition, a study by Fritz in 1986 demonstrated that deaf and hearing-impaired adults in the US were less likely to be promoted to managerial positions[168]. More recently, Parving and Christensen examined the long term outcomes of early and later acquired hearing losses and found that those with early acquired or congenital losses were more likely to work in manual occupations and less likely to be tertiary educated than their hearing counterparts[169].

In a study of deaf people in Finland, Jarvelin and colleagues observed that 14% of the deaf group were unemployed at 25 years of age compared with only 7% of randomly selected controls[133]. This finding was supported by Parving and Christensen who found that hearing-impaired people were less likely to be employed, with 31% of those with early acquired or congenital losses being unemployed compared with only 12% in an age matched background population[169]. These differences can also be found when comparing gifted deaf and hearing-impaired individuals to their hearing counterparts. Vernon and LaFalce-Landers demonstrated that 30% of gifted (IQ of 130 or more) hearing-impaired adults were unemployed with 40% requiring some kind of mental health support[172], demonstrating that these employment differences remain when intelligence levels are taken into consideration.

Danermark and colleagues, report that in addition to educational issues, hearing-impaired students in Sweden have much more difficulty than their hearing peers coping with shifting educational intentions and goals during later school years[173]. They found that these difficulties often mean that normally intelligent hearing-impaired people are channelled into secondary education paths focused narrowly on particular vocational choices, and must be encouraged to participate in paths oriented towards post-secondary education[173].

Hearing-impaired adolescents also seem to be more strongly influenced by the opinions of significant others, such as parents and careers advisers, than their hearing peers[174]. This further restricts vocational choice. Decaro and colleagues found that this advice often severely limited the occupations considered by deaf people[174].

In New Zealand, the National Audiology Centre examined differences between hearing and hearing-impaired adults in 1992 finding that those with a hearing impairment were less likely to have a higher income and more likely to have a community services card than those in the hearing population[175].

6.6   IMPACT ON THE CHILD'S FAMILY

The diagnosis of a hearing impairment has significant impact on many people, including parents, grandparents, siblings, other whanau, teachers and family friends[176]. This is a complex topic, which should not be oversimplified. The following subsection attempts to provide an overview of the kinds of effects that are commonly experienced. It is worth noting that the reaction of hearing-impaired parents to the diagnosis of their child as hearing-impaired is likely to be different from the reaction of hearing parents who have a deaf child. Positive impacts, especially those in the long-term, have also been described, although this section will focus on the negative effects, mainly within the immediate family, to give the reader an idea of some of the feelings and difficulties associated with the diagnosis of hearing impairment[176]. Positive outcomes are more likely with effective early intervention.

Parents and caregivers

Parents or grandparents are most likely to be the first to suspect a hearing impairment in their child. The fact that approximately 90% of parents of hearing-impaired children are hearing[177-179] means that few parents have any direct experience with congenital hearing loss. In addition, many parents report difficulties and stress associated with the process of obtaining a diagnosis and having to convince professionals to take action as a result of their suspicion.

Much has been written of the grief experienced by parents of a child newly diagnosed with PCHI. This grief is associated with the loss of expectations that parents often experience, as their expectations of the family's future life are profoundly changed[180, 181]. The degree of the hearing loss does not necessarily determine the significance of the impact on the family, with the impact of a more mild or unilateral diagnosis often being similar to that of more severe losses[153, 180]. Depression is often reported by parents of hearing-impaired children as they come to terms with their child's lack of hearing and all this implies for their families' future[181-183].

Feelings commonly experienced include inadequacy, anger, isolation, denial, guilt, vulnerability, confusion, depression and sorrow[180, 183-186]. Parents of hearing-impaired children often describe how they continue to experience some or all of these feelings after many years have passed. The confusion parents report often centres on the many choices they are asked to make regarding options for intervention, with conflicting information coming from the proponents of various approaches.

The parent(s) present at diagnosis and intervention appointments often report feelings of stress resulting from being seen as an instant expert, being asked questions by their spouse and other interested parties. At the beginning of the process this can be very difficult, as much of the learning about hearing impairment has not begun and this is occurring during a period when the parents are dealing with their own feelings around the diagnosis of their child[180, 184, 187].

Isolation from families with hearing children is often reported by parents of a hearing-impaired child. However, there is also isolation from other families with hearing-impaired children, especially in the provinces and isolation from resources needed to assist the child (eg sign language classes). Parents often speak of the importance of talking with other parents of hearing-impaired children and that these encounters are an important source of emotional support[184].

The logistics associated with managing a child's hearing impairment are substantial, and the diagnosis and intervention can significantly affect the functioning of the family unit. Intervention regularly involves transportation, sometimes over significant distances, to audiological appointments, specialist surgeons and special education services. The goal of these interventions is to ensure the child has access to language, and to provide habilitation to prevent them from falling further behind their peers, especially in speech and/or language.
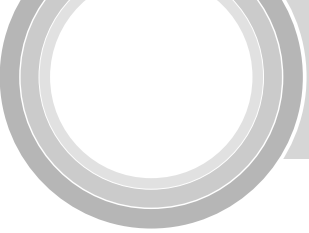
Diagnosis often means that parents or caregivers need to spend many hours of one-on-one time habilitating the hearing-impaired child. Late detection requires more intensive and longer duration habilitation. It may also mean the parents learn sign language to enable them to communicate with their child. Many parents also embark on a significant education process learning about their child's hearing impairment and enabling them to make informed choices about management of their child's disability. Many parents find this decision-making process stressful and difficult. All these activities must happen around normal family life and can place significant financial, logistical and emotional pressure on families[182].

Parents, especially the primary caregiver, are at risk of focusing on the hearing-impaired child to the exclusion of the other family members, including themselves. They may also focus on the impairment to the exclusion of the child. Often the primary caregiver becomes an intermediary between other family members and the hearing-impaired child[176].

Siblings

Parents of hearing-impaired children often describe feelings of guilt when they cannot pay enough attention to, or do not have enough emotional energy for, their hearing child(ren). As a result, hearing siblings often report feelings of neglect in addition to the belief they had to behave particularly well as they understood the strain their parents were under in dealing with the hearing-impaired child. In some cases these siblings themselves felt anxious or upset for the difficulties experienced by their parents or deaf sibling.

Hearing siblings also report feeling guilty for having normal hearing and anxiety about whether their own children will be born

deaf. They may experience loss of childhood as they are asked to carry additional responsibility. In addition, they may also feel ashamed at having a hearing-impaired brother or sister, feel pressured by an expectation to perform, or experience bullying by peers because of their hearing-impaired family member[176].

6.7   CONCLUSION

A lack of access to sound during periods of maximal auditory plasticity significantly affects a child's ability to learn language. PCHI thus limits a child's ability to build the foundation language skills required to succeed educationally. As a result, many hearing-impaired children do not reach their academic potential. In addition, where language is unavailable, later stages of cognitive development in early childhood are at risk.

Diagnosis of a hearing loss places pressure on families, with significant remedial effort often being required to assist late detected hearing-impaired children and prevent them from falling further behind their peers. Stress on the hearing-impaired child can also be significant, with social isolation and mental health issues more common among this group. Together, these difficulties often result in reduced employment opportunities and limited vocational choice.

Language development, cognitive development and social emotional development are all interrelated. This increases the effects of hearing impairment on development.

Minimising the far reaching consequences of hearing impairment requires the early identification of hearing impairment, preferably at birth, followed by timely, appropriate intervention and support.

7. PERMANENT CONGENITAL HEARING IMPAIRMENT IN NEW ZEALAND

This section presents the available data relating to permanent congenital hearing impairment (PCHI) in New Zealand. It also describes the support available for hearing-impaired children, and briefly outlines the history of regional screening programmes.

7.1   NEW ZEALAND DATA ON PCHI

There is an annual reporting system in New Zealand (Deafness Notification Database), which since 1982 has collected data on the number and age of children diagnosed with *permanent* hearing loss. This database is contracted by the Ministry of Health to The National Audiology Centre. The database provides the only source of information from which the prevalence of PCHI and the characteristics of children with PCHI can be assessed.

Estimating prevalence from this database has limitations. Firstly, the Deafness Notification Database is designed to provide broad information on childhood hearing impairment. It does not focus solely on congenital hearing loss. The determination of congenital hearing loss is made through the exclusion of acquired hearing losses. This is made especially difficult due to the late age of identification, which allows a relatively long window of time during which progressive hearing losses can develop. As a result it is difficult to determine which children notified have hearing losses that are congenital as opposed to those which are acquired later in life. Secondly, there is no assurance that all hearing-impaired children are included within the database, although it is likely to include the great majority of cases. Thirdly, longitudinal comparisons of the data are difficult due to changes that have occurred in the classification system over time. This relates particularly to the data on the age of identification. Although this database provides valuable information, the data on PCHI must be viewed with these limitations in mind. As a result, the data present an indicative picture of the extent of PCHI and of trends over time, although small changes within those data may not be reliable.

Data in this report reflects notifications until the end of 2002, although where data for 2003 was available they have been included. See Section *16.4* for information on the classification system used within the database.

7.2   PREVALENCE OF PCHI

As universal newborn hearing screening is not carried out in New Zealand, the exact incidence of PCHI is not known. However, an estimate of the total prevalence of PCHI in New Zealand can be made by examining the number of children in each category notified to the Deafness Notification Database.
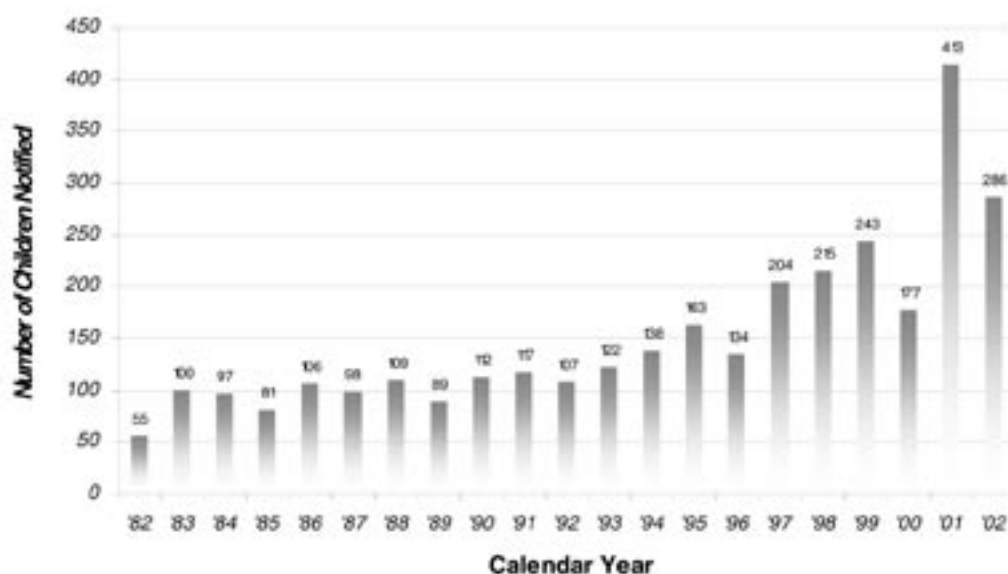

Figure 4: Number of notifications per year: (Deafness Notification Data 1982-2003, National Audiology Centre)[188]

Figure 4 illustrates the increasing number of total notifications to the Deafness Notification Database since record keeping began in 1982. This increase has been more substantial since 1996 and is much greater than would be predicted solely by the population increase over those years. It is assumed that this trend reflects an improvement in reporting systems and an increase in reporting of cases of milder hearing loss rather than an increase in the prevalence of PCHI. For example, there was a dramatic increase in the number of notifications in 2001 which was a result of a drive to ensure retrospective notification forms were registered and so does not indicate a real increase in the number of children detected in that year.

The notifications within the database are grouped into five categories. (See *Table 4: Deafness Notifications 2000-2002.)* The number of children in each category is shown for the period 2000-2002. This period has been chosen as it reflects the current situation and data within this period are more easily compared. (This total differs from the statistic published in the DND report for 2000.)

Type of case	Number of cases 2002	Number of cases 2001	Number of cases 2000
1. Children with bilateral hearing loss greater than 26dB in the better ear	113	202 (includes 44 retrospective notifications)	92
2. Children with slight hearing losses	70	56	53
3. Children with unilateral hearing losses	38	54	14
4. Children born overseas	25	34	5
5. Children with losses thought to be acquired.	40	67	6
TOTAL NOTIFICATIONS	**286**	**413**	**170**

Table 4: Deafness Notifications 2000-2002[19-21]

The number of PCHI notifications is calculated by subtracting suspected acquired losses from the total number of notifications (Group 5 in *Table 4*). Some acquired losses may have been misclassified and so this is only an estimate of this group.

From this information, an estimate of the prevalence of PCHI can be obtained by dividing the number of notifications to the database by the number of births in the relevant years. Due to the currency and reliability of the data, the notifications for the period 2000-2002 have been used to estimate prevalence. The average age of identification was approximately three years during this period, and so prevalence has been calculated by dividing the number of notifications in the corresponding years by the number of births in the six year period prior and then averaging this figure[189]. The estimated prevalence of *PCHI* in the three years to 2002 is on average 4.40 per thousand births[19, 188]. Once cases of children born overseas are removed this drops to 4.05 per thousand births.

The estimate of prevalence that is most comparable to overseas data includes only those cases where the hearing loss is significant bilateral or unilateral (26dB or greater). This estimate for New Zealand is 3.00 per thousand, dropping to 2.38 per thousand when unilateral losses are removed. It is therefore estimated that the number of cases that could be identified through a UNHS programme in New Zealand is between 2.38 and 3.00 per thousand births depending upon the thresholds for inclusion. This equates to approximately 135-170 cases per year at current birth rates.

International prevalence rates, based on data from universal hearing screening programmes overseas, for PCHI vary widely from 0.9 to 5.95 per thousand infants screened *(See Table 5).* This is in part due to differences in the criteria for inclusion, and so direct comparison is difficult. The New Zealand estimate is in line with international rates and it is unlikely that our true incidence of PCHI is markedly different from rates reported overseas.

A detailed list of prevalence data can be found in section *16.1: Prevalence* .

Region	Prevalence	Programme/Study
United States	0.9-5.95 in 1000	USA[190], Rhode Island[36], Atlanta[191], New York State[41], Hawaii[192], New York State[193], Washington[194], Utah State[195].
UK	1.2-3.5 in 1000	England[196,] Trent region[197], Wessex[43], UK[198], Nottingham[199].
Europe	1.0-3.25 in 1000	France[200], Denmark[201], Denmark[202], Denmark[203], Northern Finland[204], Austria[205], Norway[206].

Table 5 : Permanent congenital hearing impairment – International prevalence rates per thousand births

7.3   SEVERITY OF PCHI

The severity of hearing loss reported in the Deafness Notification Database is categorised as mild, moderate, severe, or profound as determined by the average hearing level over 4 audiometric frequencies (500, 1000, 2000 and 4000 Hz) in the better ear. The criteria used to define the level of hearing loss are as follows:

Category of Loss	Hearing Level
Mild	26-40dB
Moderate	41-65dB
Severe	66 -95dB
Profound	Greater than 95dB

These categories reflect the criteria used in clinical practice in New Zealand but differ from those used prior to 1996. As a consequence, it is not possible to accurately compare the age of identification between various categories of hearing loss over time.

Mild losses are most common, followed by moderate losses with severe and profound losses forming a smaller proportion (14%, see *Figure 5*). The number of children notified with mild losses has increased over the previous 10 years. This is likely to be due to improved reporting of these losses, which were previously thought to be insignificant, rather than to increasing incidence rates. (In a small number of children, hearing status may change over time, meaning their categorisation may be different from the severity at birth due to the late age of identification. These changes are not tracked through the Deafness Notification Database.)

These proportions are slightly different from those reported overseas, with a greater proportion of children with moderate hearing losses and fewer profound and severe losses than reported in England and Australia[21, 188, 205]. As an example, cases notified to Australian Hearing in 2001 were; mild 48%, moderate 26%, severe 12% and profound 12%[21]. However, Australian Hearing's categories of loss are defined slightly differently and their data includes acquired hearing losses. The categories of loss used in 2001 were; mild - less than 40dB, moderate 40-60dB, severe 61-90dB, profound 91+dB[207].
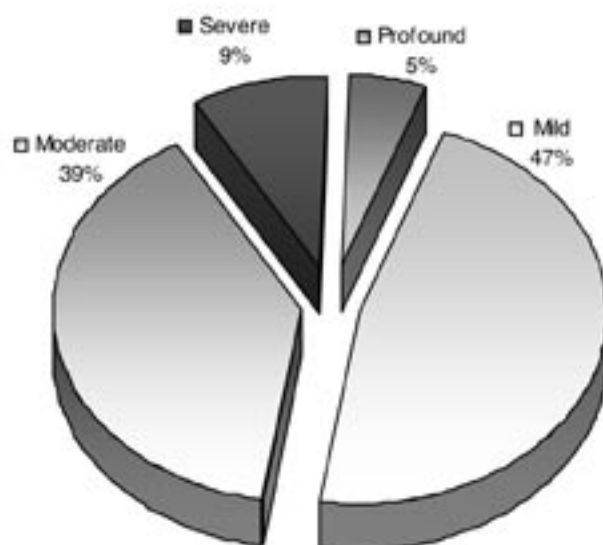
Figure 5: Proportion of hearing loss by category (Deafness Notification Data 2002, National Audiology Centre)[188]

7.4   NUMBER OF NEW ZEALANDERS AFFECTED BY PCHI

Using the estimates of prevalence that range from 2.38 to 4.40 per thousand (depending on the criteria for inclusion as discussed in section *7.2 Prevalence of PCHI*) and with a population of 3,942,100[208] it is estimated that the total number of people (children and adults) living in New Zealand who were born with various degrees of PCHI is from 9,382 (with significant bilateral PCHI) and 17,345 (with all degrees of PCHI).

Another way of estimating the total number of people in New Zealand who were born with PCHI is to use the data on the number of children receiving hearing aid funding in New Zealand. In 2002, 2,800 children 0-21yrs were receiving funding for their hearing aids from the National Audiology Centre Hearing Aid Fund[49]. (As acquired and congenital losses cannot be separated, and as some children with hearing loss go without intervention, these data can only be regarded as estimates.) Extrapolating this to the total population gives an estimate of 9,014 people in New Zealand born with PCHI. Probably, because many of the slight and mild hearing losses do not receive any interventions paid for from the fund, this figure is similar to the lower estimate in the range above, which excludes mild losses.

The data accuracy in this area could be improved to inform estimates of the prevalence and nature of hearing loss in New Zealand, which in turn would be valuable in making health policy and planning decisions.

7.5   CAUSES OF PCHI

At the time of diagnosis, PCHI could only be attributed to a known 'cause' in 41% of cases notified between 1997 and 2002[16-21, 209]. Thus, in 59% of cases, no cause of the hearing loss was identified. *(See Figure 6.)* This proportion is higher than observed overseas where between 40% and 54% of cases are reported to be of unknown cause[210-215]. For example, Australian statistics for 2001 show 46.8% of cases of hearing loss were of unknown cause at birth. In the UK, the Trent Region statistics show 41% of unknown cause at birth[197] and in Spain, 50% of cases were associated with risk factors present at birth[216]. See section *8.1.1: Risk factors* for more information.

The most commonly stated 'cause' of PCHI in the New Zealand Deafness Notification Database between 1997 and 2002 (22%) was listed as 'genetic', based on reports of a family history of 'deafness'. There is no standardisation within this reporting and so these data provide only an indication as to whether an individual loss *may* be genetic in cause. Similarly, those that are categorised as a non-genetic cause may actually be genetic as there was no family history identified or the impact of a family history is misconstrued. There is no routine genetic testing for deafness conducted currently in New Zealand and research into the genetic basis for PCHI in New Zealand is required.

It is interesting that these data are similar to the 23.3% of children found to have a positive family history of deafness in the UK[217] and to an epidemiological study of congenital hearing loss in Victoria (Australia) which cited genetic causes in 24% of the 134 cases reported. Present research suggests up to 50% of all hearing losses present in children are due to genetic cause[218].
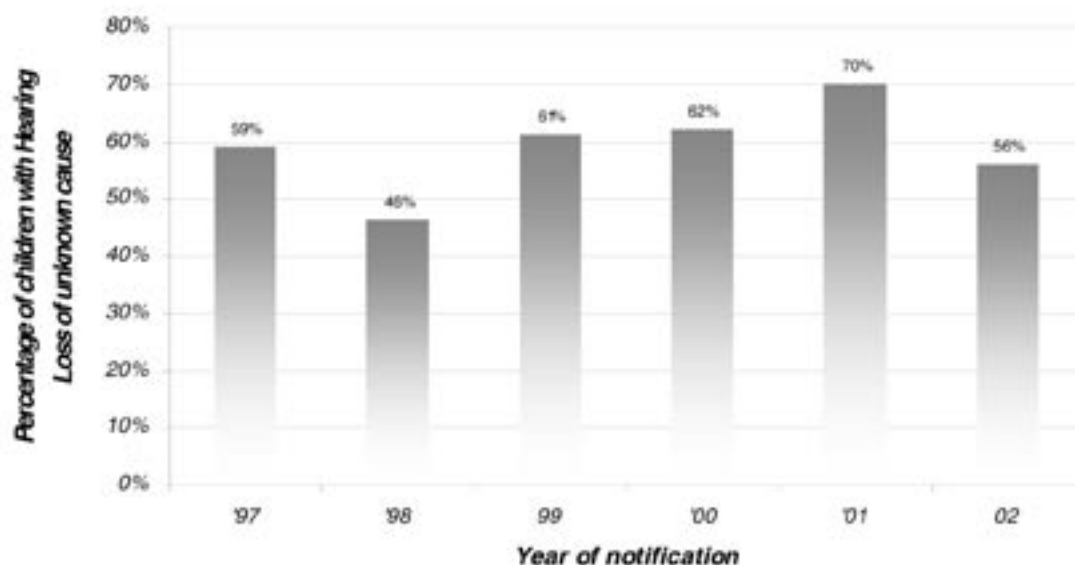
Figure 6: Proportion of children by year of notification with PCHI of unknown cause 1997-2002. Deafness Notification Data 1997-2002, National Audiology Centre[188]

7.6   IDENTIFICATION OF PCHI IN NEW ZEALAND

7.6.1   Approach to identification – the risk factor approach

The risk factor approach identifies infants who are at a greater risk of PCHI. Children categorised as 'high-risk' are then referred to their local audiology department for diagnostic assessment. This approach has been used in New Zealand as a targeted hearing 'screening' technique since the 1980's (See section *8.1: Targeted approaches* for more discussion on the risk factor approach to identification).

The high-risk criteria used in New Zealand are:

1.   Family history of hearing loss

2.   Jaundice requiring exchange transfusion

3.   Craniofacial abnormalities

4.   Ototoxic drugs

5.   Mechanical ventilation lasting 5 or more days

6.   Low Apgar Scores (0 to 4 at one minute or 0-6 at five minutes)

7.   Birth-weight less than 1500 grams

8.   Bacterial meningitis

9.   Infections such as rubella, herpes and toxoplasmosis and those associated with hearing loss

This method of identifying hearing loss in infants, based on referral for assessment following identification through risk factors, has not succeeded in reducing the average age of detection. (See section *8.1.1: Risk factors.*)

In addition to the use of risk factors, a parent questionnaire entitled 'Can Your Baby Hear?' has been available for use by WellChild providers for over 20 years, with very limited success in identifying children needing audiological assessment.

7.6.2   Existing regional UNHSEI programmes

Screening refers to the systematic application of a test to individuals where there is no reason to believe they have the condition. (This is in contrast to the application of a targeted approach, where individuals with a high index of suspicion are tested.) Universal screening refers to a test offered to all people in a defined population.

Over the last decade, a number of small universal newborn hearing screening programmes have been established in New Zealand hospitals. A number of these programmes have ceased operation as a result of a lack of sustainable funding or due to the reliance on a single individual's knowledge and drive (eg Whakatane, Southland). Gisborne's universal programme is the exception, as it has been in operation consistently for 7 years. In addition, a Neonatal Intensive Care Unit (NICU) based programme is operating in Christchurch Women's hospital and continues to operate despite difficulties with funding.

A small screening programme began operating at Wanganui Hospital in 2002 although this has not continued due of a lack of resources.

Many of these programmes have been driven by a single individual, often an audiologist, who has had to secure funding, review best practice to set protocols and implement the programme with limited assistance and often without long-term funding. Some programmes have utilised volunteers to reduce costs as funding for staff was unavailable.

Gisborne

A universal newborn hearing screening programme funded by the Tairawhiti District Health Board has been running consistently since 1997, the longest running programme in New Zealand. This programme, based at Gisborne Hospital, had screened 5,018 babies to May 2004, detecting 15 children with PCHI in that time (2.98/1000) only eight of whom (1.59/1000) would have been detected using the most aggressive risk factor approach[219].

This programme uses otoacoustic emissions as the screening method, with two hospital staff employed part-time as screeners, covering births seven days a week at the hospital. See section *8.2.2.1 Otoacoustic emissions (OAE)* for more information on this screening technology.

The programme receives assistance from a public health nurse and occasionally from plunket and midwives to help with follow-up for referrals and for the small proportion of infants who miss their first test. Coverage for this programme is 89% with 98-100% coverage for hospital births[219].

Christchurch

A research project to investigate screening was undertaken at Christchurch Women's Hospital, screening NICU babies from July 2001 to July 2002. During the 12 month period, 688 infants were admitted to the NICU. Hearing screening was offered to 438 mothers whose stay was more than 48 hours, and who lived in Canterbury. Of these, 435 infants (99.3%) were tested before discharge. 90.6% passed the Distortion Product Otoacoustic Emission test on the first occasion. 4.37% of infants were later referred to audiological assessment[220].

Two infants were identified as having bilateral moderately severe sensorineural hearing loss, one of whom had a risk factor. The complete results of this research project are to be published later this year by Flynn and colleagues[220].

Funding to continue the screening of NICU babies at Christchurch Women's Hospital has been declined, although limited testing will continue[220].

Waikato

Waikato District Health Board is funding a regional UNHSEI programme which began screening in February of 2004. This programme will screen approximately 3500 babies per year once fully rolled out, making it the largest programme established in New Zealand since the National Women's Hospital behavioural newborn screening programme in the 1970's. This programme began screening only NICU babies and now includes well babies and babies born at River Ridge and Waterford birthing units. This programme will eventually expand to cover all babies born in the Waikato region.

7.6.3   Average age of identification

The New Zealand Deafness Notification Database includes information on the age of suspicion and age of confirmation (comprehensive diagnosis) of hearing loss.

The average age of identification (confirmation) of children with moderate or greater hearing loss in New Zealand in 2002 was 35.1 months[188]. The median age of identification in 2002 was 30 months, with 25% of children being identified by the age of 15 months, and 75% by 58 months of age. New data from 2003 demonstrate further increases in this average age of identification, with the average moving up to 46.1 months for children with moderate or greater hearing losses.

Overall, the average age of identification has been increasing over the last 20 years regardless of category of loss. This average age is not comparable to the previous years' average age of 39 months as the definition for categories of loss used in the notification database has changed. See *16.4: Deafness notification data* for a more detailed description of the change.

Historically, the average age of identification of children with mild losses is later, at 66 months of age in 2002, reflecting the lower index of suspicion in cases of mild hearing loss[16-18, 20, 21, 209].

The average age of hearing loss identification in New Zealand is higher than in other countries without Universal Newborn Hearing Screening. For example the average age of identification in various US states before UNHS implementation was 19-36 months[221]. In the UK those screened only by the health visitor were identified at an average age of 26 months[222]. This average identification age does not compare favourably with the internationally recommended age for detection of hearing loss by 3 months[223].

The presence of risk factors in children with mild or moderate hearing impairment made little difference to the age these children were identified in 2002. In children with severe or profound hearing loss, those children with one or more risk factor(s) were identified significantly later than those without risk factor(s)[21].

Significant differences in the age of identification have been found between regions[16-21, 209]. Also data from regional audiology departments seem to suggest differences may exist. As an example, Waikato Hospital Audiology Department calculated their average age of detection for the 310 cases of bilateral sensorineural hearing impairment on their current list and found the average of all degrees of hearing impairment was 59.2 months, higher than the national average age of identification[224].

7.7   ETHNIC DIFFERENCES RELATING TO THE AGE OF IDENTIFICATION

7.7.1   Māori children with PCHI

Māori made up 23.5%*of the general population under 19 in 2001, yet they accounted for 49% of children notified to the Deafness Notification Database in the same year[20, 22]. The reason for the disproportionate notification rate is not clear and is made more difficult to assess as the cause of the hearing loss was unknown in the majority of cases[188]. There is speculation that the greater prevalence among Mäori may be the result of a greater prevalence of genetic hearing loss within this group[49].

The notification data also show Mäori are more likely to have mild (48%) or moderate losses (44%) and are less likely to

be diagnosed with severe (6%) and profound losses (2%) when compared to other groups[21].

Māori children have consistently been identified later than Non-Māori, with 80% of Māori children being identified by 70 months of age in 2002[21]. In comparison, 80% of European children were identified by 60 months of age[21]. The difference in average ages of Māori and Non-Māori was greatest in 1996, at 20.8 months, while similar ages of identification were reported in 1997[21]. In 2001, the average difference was 11.4 months later than European children, with it taking longer to confirm a suspected hearing loss in Māori for all years except 1997[225].

* This value differs from the proportion of Māori in the general population (19.5%) published in the Deafness Notification Data report [188].

7.7.2 Other ethnic differences

Figure 7 illustrates the differing ages at which three key groupings were identified with PCHI in 2002. The first bar shows the age by which 50% of children are identified, while the second shows the age by which 80% of children are identified. Pacific children are particularly disadvantaged, although they make up only a small overall proportion of notifications in 2002.
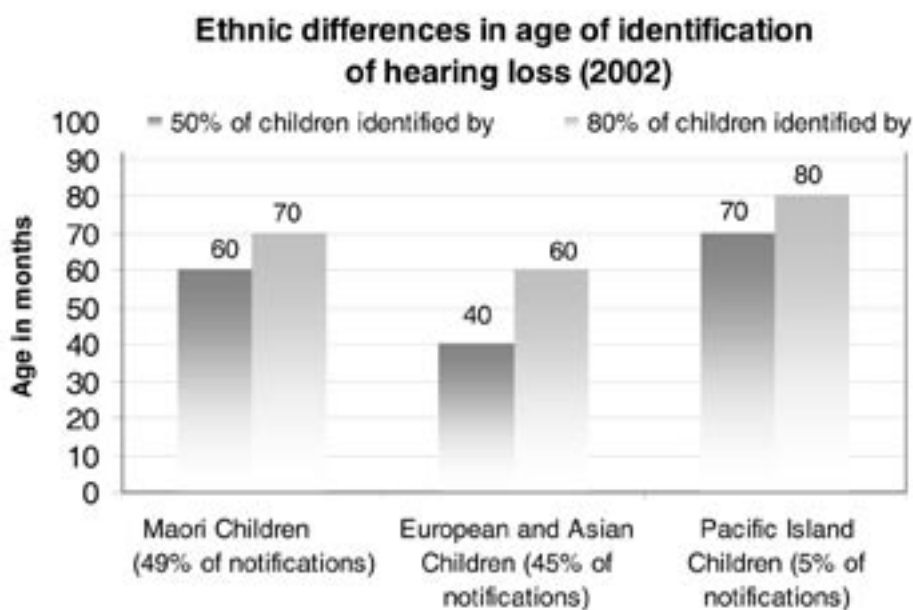
Figure 7: Identification of PCHI in Māori vs Non Māori 2002

7.8 DELAY IN CONFIRMATION OF PCHI

The Deafness Notification data show that in approximately half of all cases of PCHI it is the parent who initially suspects the hearing loss. Health care professionals such as Plunket nurses, GPs, midwives, paediatricians and public health nurses are not likely to first suspect hearing loss. Commonly, a parent will consult one of these practitioners with their concerns as deficits in the child's development build and become more visible. There is anecdotal evidence that parental concerns regarding their child's hearing are not always acted upon by some professionals, and so do not always result in prompt referral for audiological assessment.

Children with a suspected hearing loss are referred to an Audiologist for assessment. Diagnostic tests may include Auditory Brainstem Response measurements and/or behavioural testing depending upon the age of the child. A medical examination occurs after confirmation of the hearing loss.

In 2002, the average delay in confirming a hearing loss (i.e. from date of suspicion to date of confirmation by an audiologist) was 11 months. This is similar to previous years. The reason for these lengthy delays is not well understood, in part due to the nature of the question on the notification form and also because this section of the notification form has been 'poorly answered'[225]. These data highlight the problems with the use of a risk factor approach. This approach is not reliably applied.

7.9 AGE OF ENROLMENT IN INTERVENTION PROGRAMMES

No data is collected in this area, but it can be estimated that the average age of enrolment in an intervention programme will be similar to, or somewhat later than, the age of confirmation.

It would be useful to collect information on the age of enrolment in intervention programmes. This information is often collected overseas and it provides a key measure of the effectiveness of policy decisions as new approaches are implemented to attempt to reduce this age.

7.10 INTERVENTION AND MANAGEMENT SERVICES

Advisers on Deaf Children (AODC)

Once a child is diagnosed with a hearing loss the family is referred to an Adviser on Deaf Children, who usually contacts them that day or very soon after. Advisers work with children from the time of detection to the end of their formal school years. The AODC system is unique to New Zealand and parents often see Advisers as their key resource and link with the many professionals involved in their child's care, education and support.

In those first days the AODC talks through issues and concerns with the family and may offer to link the family to organisations and individuals such as:

- A member of the New Zealand Federation for Deaf Children for parent-to-parent contact
- A deaf adult
- Needs Assessors
- WINZ for the Child Disability Allowance

And provide the following key information:

- A folder of information 'Parent Information Folder'
- 'Parent pack' through a request to the Federation for Deaf Children

Broadly, the following functions are typical and are customised to meet the needs of each individual family:

- Liaising with audiology and specialist services
- Providing unbiased information to parents to assist informed decision making
- Providing support to parents and assisting parents to solve and manage issues that arise
- Liaising and working with educational intervention programmes (eg Deaf Education Centres, Hearing House)
- Providing information to and liaising with a wide range of professionals in the Health and Education sectors
- Monitoring educational progress and assessing audition, language and learning needs for the Individual Education Plan (IEP), in conjunction with other professionals
- Trial of and arranging supply, monitoring, and organising repair of hearing aid and FM equipment
- Assisting educational staff in their application for funding of listening equipment and/or educational support
- Assisting teachers, teachers aides and special needs coordinators in the selection of goals and strategies which will allow the child improved educational access

- Demonstrating and suggesting communication strategies that can be adapted into the family's day to day communication with the child, encouraging improvement in the child's knowledge of language

Children from diagnosis to school-age are included in the caseload of an AODC – falling under the Early Intervention Sector of Group Special Education (GSE). They will therefore have access to other disciplines (psychologist, early intervention teacher, speech-language therapist, Māori team) if required. Parents may be encouraged to enrol children at early intervention centres. At the kindergarten, pre-school or crèche, the child may require the additional support of an educational support worker (ESW). Itinerant Teachers of the Deaf may also become involved from three years of age, in an educational setting.

Medical

Otorhinolaryngologists (ORL, or Ear Nose and Throat Surgeons) coordinate medical management – diagnosis, prognosis, surgical intervention in some cases (ORL and plastic surgeons), ongoing monitoring and treatment of middle ear conditions and surgery for cochlear implants.

Geneticists, for the investigation of the genetic causes of deafness.

Paediatricians examine children for any associated conditions or abnormalities.

Audiological

Audiologists conduct the hearing assessment, fit amplification devices or are involved in cochlear implant assessment and initial and ongoing tuning of cochlear implants. They provide the ongoing monitoring of hearing and hearing aids, and ongoing audiological management.

Other educational support and options

Itinerant Teachers of the Deaf, Deaf Resource Centres and Schools for the Deaf.

7.10.1   Types of support provided

Support may include some or all of the following:

- Ongoing parent guidance and habilitation by AODCs
- Regular speech, language, and auditory training
- Regular sign language classes for the child and/ or family
- Cochlear implantation and habilitation
- Amplification systems
- Services of an interpreter/communicator for those students who use signed communication (eg NZSL or Sign Supported English)
- Favourable seating in the preschool or school classroom to facilitate listening and/or speech reading
- Captioned films/videos
- Assistance of a note taker, who takes notes for the student with a hearing loss, so that the student can fully attend to instruction
- Instruction for the teacher and peers in alternative communication methods, such as sign language

In terms of communication modes, only a small proportion of hearing-impaired individuals rely solely on sign language. In a study of 47,973 hearing-impaired children and youth (1990-1991 school year) conducted by the Center for Assessment and Demographic Studies at Gallaudet College (United States), 39% of those surveyed used auditory/oral as their primary mode of communication; 58% used total communication and only 1.3% used sign language alone[177].

7.11 CONCLUSION

Using data on the number and type of cases of hearing loss identified in children, it is estimated that approximately 17,000 New Zealanders have some degree of hearing loss that was present at birth. Three cases of significant bilateral PCHI occur per thousand births. In 60% of cases, the cause of PCHI is unknown, and thus cannot be detected through the use of a risk factor approach to detection. On average, PCHI is not detected until 46.1 months of age in New Zealand[15], later than other developed countries. In Māori and Pacific children PCHI is detected even later, and occurs in greater numbers than their population would suggest.

Advisers on Deaf Children work with children diagnosed with PCHI and their families until the end of their formal school years. Their role includes working with the medical, educational and other professionals involved with the family, in addition to providing information, support and direct assistance.

A number of hospital based UNHSEI programmes have been established in New Zealand, although many of these have ceased operation due to lack of sustainable funding or management issues. These programmes screen less than 5% of births in New Zealand. The Gisborne programme has been operating for seven years, while Christchurch Women's Hospital screen NICU babies only. Recently, a new and comprehensive programme has been established in the Waikato, and will screen 3,500 babies per annum.

8. APPROACHES FOR IDENTIFYING PERMANENT CONGENITAL HEARING IMPAIRMENT

This section discusses the main technologies and techniques used for identifying permanent congenital hearing impairment (PCHI). These are divided into 'targeted' and 'universal' approaches and include the use of risk factors, behavioural, physiological and genetic tests.

8.1 TARGETED APPROACHES

8.1.1 Risk factors

As described earlier there are a number of factors that are associated with a higher risk of PCHI. See section *7.6.1: Approach to identification – the risk factor approach.*

Categorising newborns by the presence or absence of these risk factors has been an approach used to identify infants that are more likely to have PCHI and so require audiological assessment. It is thought that between 10-30% of newborns are known to have at least one risk factor and theoretically should be referred for audiological assessment[197, 226] under such an approach.

Numerous studies have shown that a targeted approach using risk criteria is not the most effective method for identification of hearing impairment among all newborns[192, 214, 222, 227, 228]. The major problem identified within these studies is that only a proportion of children with a permanent congenital hearing impairment have one or more risk factor(s) (36 to 67%)[41, 197, 205, 211, 214, 215, 226, 229-231]. Thus, the use of a risk factor approach alone can identify only a proportion of children with permanent congenital hearing impairment[223, 232].

In addition, many studies demonstrate that only a proportion (20-50%), of children labeled 'high-risk' actually receive an audiological assessment as a result of presence of a risk factor. Due mainly to such low attendances for assessment, detection rates as low as 0.5 per 1000 births have been observed in areas using a risk factor approach alone[229, 233, 234].

Gene abnormalities are a significant cause of PCHI. But because the majority (90%) of hereditary deafness is autosomal recessive, and therefore will not be present in all generations, there may not be any suspicion of hearing loss at birth because parents may not be aware of their family's history. This makes the use of family history as a 'prospective' risk factor unreliable[212].

Specificity and sensitivity of risk factor approaches are low for a number of reasons. First, a risk factor is not present at birth in a significant proportion of infants born with a PCHI. Second, some babies are not correctly labeled as being 'high-risk'. Third, assessments are often not completed on a high proportion of 'high-risk' infants. This low specificity of high-risk approaches often results in costly programmes, as diagnostic follow-up focuses on ruling out the large number of false positives among referred individuals rather than identifying true cases of PCHI[124, 215, 228].

Key measures of the risk factor approach demonstrate that although the age at diagnosis of Newborn Intensive Care Unit (NICU) or risk babies can be lower than those with no risk factors, the average age of identification in overseas programmes using this approach is still considered late at 18-20 months[37, 214, 231]. Harrison and Roush report that only 50% of hearing-impaired NICU babies or infants with risk factors were identified by 12 months of age[230, 235].

Despite this poor performance, some overseas studies have suggested that selective testing of high-risk neonates can have the potential to permit the introduction of robust systems and achieve a higher benefit-to-cost ratio when compared with universal screening of newborns[148, 236].

There is some evidence that inclusion of other factors or a system weighted toward the more significant risk factors could be utilised to further target congenital hearing impairment and improve the positive predictive value[213, 237-240]. Improvement of a risk factor approach however, cannot by its very nature, contribute to identification of children without risk factors. The use of risk factors may eventually be enhanced by the use of genetic screening as more of the specific genes for hearing impairment are identified.

Use of risk factors for identifying permanent congenital hearing loss in New Zealand

The risk factor approach is the main method for identification of PCHI in New Zealand. It is estimated that 10-15% of New Zealand babies fall into the high-risk group[241, 242]; that is, where one or more risk factors would be present.

Examination of the Deafness Notification Database shows that use of a risk factor approach (used for thirty years in NZ) has not been successful in lowering the average age of hearing loss detection in New Zealand children[16-21, 209]. Surprisingly, some children with identifiable risk factors in New Zealand have no better chance of early identification than those without. In 2002, children on the Deafness Notification Database with severe hearing losses who had risk factors were identified later (24 month average age) than those without risk factors (17 month average age)[21].

Pellow and colleagues examined the implications of long term use of risk factors in New Zealand and found that the average age of identification had risen from 20.9 months in 1991 to 37.4 months in 1996. (These figures are not comparable to current ages as there have been changes in the scheme for categorising hearing loss.) They concluded that a 'dramatic change' in policy was necessary for real advances in early identification of hearing loss[243].

The key contributing factor to the late age of identification is that a high proportion (60%) of children identified with moderate or greater hearing losses from 1995 to 2002 had no known risk factors when examined retrospectively. This is at the high end of the range reported internationally and reduces the potential effectiveness of this targeted approach.

In addition, as noted in the 2003 Deafness Notification report, not all children with risk factors are identified as having a risk factor at birth and therefore are not referred for diagnostic assessment.[21] Only 58% of those labelled as 'high-risk' from 1995-1998 at National Women's Hospital actually attended an audiological appointment[244]. Thus a high proportion of those infants labelled 'high-risk' are also lost at follow-up.

Battin and Rush examined hearing status among low birth weight infants in New Zealand and found a lower proportion (1.9%) than is reported overseas (3.0-9.0%)[244]. Thus, it would seem that New Zealand does not have the same rates of hearing loss in low birth weight, high-risk infants as have been described in overseas studies.

It is not known how many infant audiological assessments are conducted in New Zealand because of the presence of one or more risk factors, although this number would likely be greater than the number that could be expected under universal newborn hearing screening. However, it is important to note that if the risk factor approach was rigorously applied, approximately 10-15% of all babies born in New Zealand would need to be referred for audiological assessment. This large number of referrals is significantly greater than the number of referrals likely through a universal newborn hearing screening approach (less than 4%)[226, 242].

In conclusion, utilising the risk factors that exist currently will at best result in the identification of 40% of children born in New Zealand with PCHI, even with the most aggressive application. Universal newborn hearing screening requires fewer audiological assessments than a properly applied risk factor approach, and potentially fewer than the number currently completed.

8.2 UNIVERSAL APPROACHES

Universal approaches involve the testing or screening of all babies within a given population, often a region. This section outlines issues with behavioural testing, and examines commonly used objective tests as an alternative.

8.2.1 Behavioural testing

Behavioural hearing tests have been used for assessing newborn hearing. With babies less than 6 months of age, these tests rely upon an automatic response to sound. As an example, Behavioural Observation Audiometry (BOA) uses observations of the babies responses to sound, such as eye blinks, changes in heart rate, respiration and body movements to indicate hearing ability[245]. In the 1970's, two automatic screening techniques based on these behavioural responses (the Crib-O-Gram and the Auditory Response Cradle) were introduced as possible mass screening technologies. These tests were not able to accurately diagnose PCHI in babies.

There is overwhelming evidence that behavioural methods are not an accurate method of hearing screening in this young age group, even when carried out by a skilled paediatric audiologist[222, 246, 247]. This is because it is difficult, if not impossible, to control the many extraneous factors that affect the result, such as babies' state at the time of the test, the type of stimulus used, observer bias, and response habituation. Even children with normal hearing (eg teething children) demonstrate a large variation in responses to behavioural stimuli. Behavioural screening tests have high false positive and false negative rates and are therefore not able to accurately detect PCHI in children[43, 197, 222, 229, 248, 249]. It is also difficult to audit subjective behavioural methods compared to objective methods where the results are recorded and include set thresholds requiring little interpretation from the screener[250].

Once babies reach approximately 6 months of age, they can be tested through observation of voluntary head turns in response to selected sounds. This is also referred to as distraction testing. Observation of conditioned head turns can also be used, and is known as Visual Reinforcement Audiometry [VRA]. Distraction testing was once used for screening 9 month old 'high-risk' infants born at National Women's Hospital in Auckland, and has been used extensively in the UK as a hearing screening method for 6-9 month old infants[222]. Davis and colleagues reviewed the use of the Health Visitor Distraction Test in the United Kingdom. At the time of data collection (1993/4) this test was conducted in 90% of 7-8 month olds. These tests referred 9.3% of infants tested in 1994, identifying 26-28% of the children thought to have PCHI[222]. The review concluded that the HVDT was not playing as large a role as was previously thought, and that coverage fell below acceptable levels in about half the districts surveyed. In addition, the referral rate was not acceptable, being greater than 10% in half the districts, with the number of cases identified as a result being lower than the 35% (approximately) that could be expected using a targeted approach[222] (eg 'high-risk'). These behavioural tests are also not able to detect PCHI before the child reaches 6 months corrected age[250].

8.2.2   Objective tests

A number of physiological techniques used clinically to objectively measure auditory function have been adapted for rapid, low cost screening tests for newborns. This has made it feasible to implement universal screening programmes (population screening programmes) for congenital hearing loss, usually during the birth hospitalisation. Since these technologies were first applied in this way, they have become more accurate, less expensive and easier and faster to administer, with current tests enabling automated interpretation of results.

Two types of test are commonly used to screen babies for PCHI: otoacoustic emissions (OAEs) and automated auditory brainstem response (aABR). Both these tests are quick, non-invasive and safe for the baby[223] and require a sleeping or quiet child. They are easy to administer and require minimal training and test interpretation. Overseas, these tests have been successfully administered by both medical and paramedical personnel. Diagnostic versions of these tests have been used in New Zealand for some years to confirm diagnoses in infants with suspected hearing loss.

Other tests that are available or under development are the Steady State Evoked Potential (SSEP) and Post Auricular Muscle Response (PAMR). Reliable data on the effectiveness of these screening technologies is not available as the cohorts studied have been small[251]. However, these methods may be developed for use in newborns in the future.

8.2.2.1   Otoacoustic emissions (OAE)

Sensory cells in the cochlea of the healthy inner ear oscillate in response to an external sound. These oscillations, which are part of the normal hearing process, generate a small amount of energy (an 'echo') which passes from the inner ear to the ear canal where it can be detected as sound. The sounds or echoes recorded are known as otoacoustic emissions (OAEs) and their presence is a sign that the ear is functioning normally (Figure 8B). Measurement of OAEs is used clinically as an objective test of normal cochlear function at all ages and it has been adapted as a test of cochlear function in newborns.

For this test, a miniature earphone and microphone are placed in the ear, to both play sound and to record the response from the ear. If a baby has a normally functioning inner ear, an otoacoustic emission (echo) is produced and can be picked up by the microphone in the ear-canal. If a baby's ear is not functioning normally, no echo can be measured and follow-up testing is required to determine if a hearing loss exists.
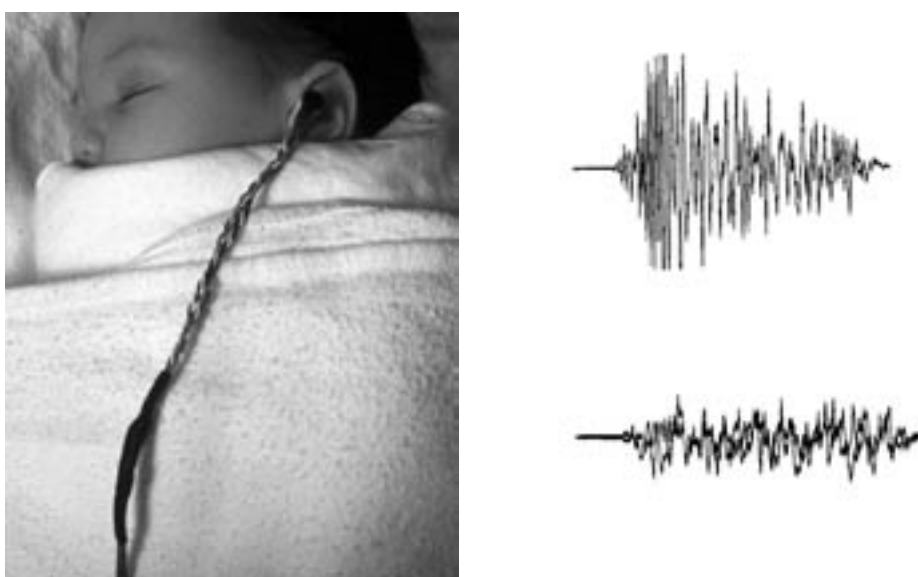
Figure 8 : A) An infant undergoing an OAE test. B) Otoacoustic emissions from a hearing (above) and hearing-impaired infant (below).

Figure 8A shows a baby undergoing an otoacoustic emission test in which the small probe is placed in the ear canal while the baby sleeps. The responses recorded in the normally-hearing baby show a relatively large burst of sound energy (Figure 7B, top trace) whereas a baby with limited or no inner ear function shows the lack of any obvious response (Figure 7B, bottom trace).

There are several different types of OAE, based predominantly on the type of sound stimulus used to evoke the emissions. Transient evoked otoacoustic emissions (TEOAEs) are generated in response to a click (a brief acoustic stimulus approximately 0.1msec in duration) at 70-80dBSPL (Decibel Sound Pressure Level). Another type of OAE is the Distortion Product Otoacoustic Emission (DPOAE) which occurs in response to paired tones[252].

TEOAEs in response to a 70-80dBSPL click are present in almost all normally hearing adults. With hearing losses in the audiometric frequencies of 30dBHL or greater there is a very high probability that TEOAE will be absent. Similarly the DPOAE otoacoustic emissions are present in the normal ear but are absent or of very low amplitude in people with hearing losses of greater than 30-40dBHL. Thus the absence of OAEs is a strong indicator of a mild or greater hearing loss[252].

The OAE is highly sensitive to a loss of function of the outer sensory hair cells (a population of the auditory sensory cells in the cochlea). These cells are responsible for the detection of low-level sounds and almost all congenital hearing losses include damage to these cells. However, because the test relies upon sound transmission through the middle ear it is also affected by middle ear pathologies or amniotic fluid present in the middle ear after birth. It can therefore give higher false positive rates especially in the first few days after birth[253] although the false positive rate can be improved by ensuring good probe fit and effective screener training[254]. By definition, OAEs will not provide information on the integrity of the auditory nerve, higher auditory brain centres or inner sensory hair cells. Testing for the presence of OAEs therefore will not reveal potential hearing losses due to abnormalities of the auditory nerve (such as auditory neuropathy) and auditory centres in the brain, unless these include peripheral damage.

8.2.2.2   Automated auditory brainstem response (aABR)

The Auditory Brainstem Response (ABR) is a series of electrical waves that can be recorded from electrodes on the scalp, in response to brief sounds played into the ear. These waves represent the electrical activity of different parts of the auditory nerve pathways from the cochlea to the mid-brain. The presence of these waves with changing sound intensity is highly correlated with behavioural hearing thresholds. The ABR is used clinically as an objective, diagnostic measure of the integrity of the ear and auditory nerve pathway up to the brainstem. The use of ABR for the assessment of newborn hearing has been recommended since 1979[255].

The ABR measure has been adapted as a neonatal screening test by automating the procedure, reducing the test time and establishing an estimate of auditory thresholds. This is the aABR test, where sounds in the form of clicks or tones are played to the baby through a probe inserted into the ear and 'band-aid' like sensors or electrodes are placed on the baby's head to detect this electrical activity. The sound is delivered at an intensity of 30-35dBHL and computer algorithms compare the waveform recorded with normal expected waves to determine the presence or absence of the waves. There are a number of aABR machines on the market. They all use different detection algorithms, some of which are reported to be better validated than others. Research indicates a high level of agreement between conventional ABR and aABR in the measurement of thresholds[256, 257].
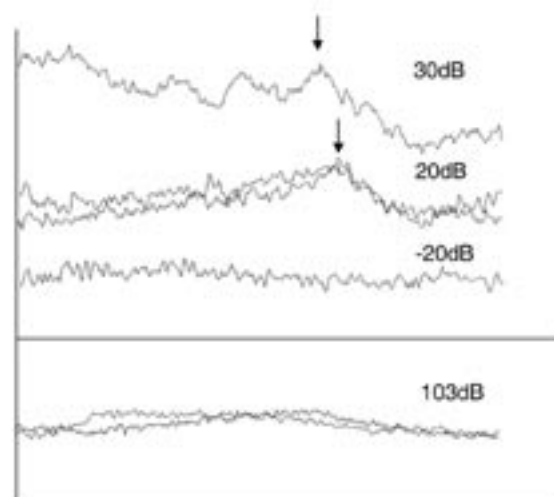
Figure 9: The normal ABR response shows a series of waves when recorded in response to a click or tone stimulus. The responses to a click sound (shown by the arrow) can be seen in the normally hearing baby at low sound levels of 20 or 30dBHL. However, no clear responses to a click are seen in the hearing impaired infant even at a level of 103dBHL.

8.2.3   Genetic testing

There have been substantial advances in knowledge of the genetic basis of deafness over the last decade[258]. Approximately half of all cases of PCHI are thought to be due to genetic disorders[259].

There are two main types of genetic deafness: Syndromic deafness in which there are other medical problems in addition to deafness, and non-syndromic deafness in which the hearing loss is the only condition. The majority of genetic hearing loss falls into the non-syndromic category and most are recessive, which means the mutations in the same allele must exist in both parents for the genetic disorder to appear in the infant. Thus a person can be a carrier without the hearing loss, and hearing loss from genetic causes will not appear in every generation. The identification of genes associated with deafness is increasing rapidly. By the middle of 2003 over 100 different loci (specific location on the human chromosome) and mutations of more than 30 genes had been identified as responsible for hearing loss. Among these genes a mutation of the CX26 gene is the most common and it is believed to be responsible for 10-20% of non-syndromic deafness[259]. It should be noted in the context of newborn hearing screening that not all of these are related to hearing loss at birth. For example the A1555G mutation may lead to hearing loss if someone is exposed to certain antibiotics[260, 261].

The identification of deafness genes clearly heralds the beginning of technology for genetic testing for undiagnosed deafness. At present, tests are available for at least five of the genes, but the tests are expensive[262].

There is the potential for genetic testing to be a major method of screening for the prevalence of PCHI in the future. As tests become available for detection of these genetic markers, the future of universal newborn hearing screening may include genetic tests at birth. These tests will only ever be helpful in the detection of a proportion of PCHI and this technology is too expensive for use as a universal screening test[263].

8.3   CONCLUSION

There is strong evidence that targeted and behavioural approaches to identification of PCHI in children have not been successful in lowering the age of identification to allow intervention to occur at the optimal time. Objective tests such as OAE and aABR are regularly used to indicate hearing function in newborns. In the future, there is potential for genetic testing to be used for this purpose, although these tests will never be able to detect all cases of PCHI.

Screening is defined by the New Zealand National Health Committee as 'a health service in which members in a defined population, who do not necessarily perceive they are at risk of, or are already affected by, a disease or its complications, are asked a question or offered a test to identify those individuals who are more likely to benefit than be harmed by further treatments to reduce the risk of disease or its complications'[44].

Many terms, such as EDHI (Early Hearing Detection and Intervention) and UNHS (Universal Newborn Hearing Screening) are used to describe programmes providing universal hearing screening for newborns and early intervention to diagnosed infants. Within this document we use the term UNHSEI (Universal Newborn Hearing Screening and Early Intervention) to refer to these programmes.

Universal newborn hearing screening and early intervention programmes have been established in many countries to enable early detection of permanent congenital hearing impairment (PCHI). Groundbreaking universal screening programmes were instituted in Hawaii and Rhode Island after legislative mandates were passed in 1990 and 1992 requiring newborn hearing screening for all babies born in those states. From this time a large number of programmes have been established, both in the USA and other countries. This section provides a review of the status and outcomes of such programmes.

9.1   TECHNOLOGIES AND PROGRAMME STRUCTURE

The early programmes utilised OAEs as a screening test but programmes now appear to be more commonly taking advantage of aABR technology, which is becoming cheaper and easier to use. In some cases, a two tier approach is employed, screening first with OAEs or aABR and then following that with a second OAE or aABR screening test. Among US programmes surveyed by White in 2003, 52% were using OAE and 67% were using aABR in some way[26]. The SWISH programme in New South Wales, Australia which was established in late 2002 uses aABR technology for screening as will the new Queensland programme, universal newborn hearing screening and early intervention (UNHSEI) programmes now commonly employ aABR, OAEs or a combination (two-tier) approach[264]. The UK programme adopts a two-tiered approach, screening first with OAEs, followed by aABR screening.

Figure 10 illustrates a typical process for a two-tier UNHSEI programme as an example of a screening programme structure. In this programme babies are screened, using OAE and those with a positive test are tested using aABR during a single session before discharge. Although various protocols have been used, on the whole children who screen 'positive' are then brought back for diagnostic assessment between two and eight weeks after discharge. Babies with a risk factor present are tested using aABR only,
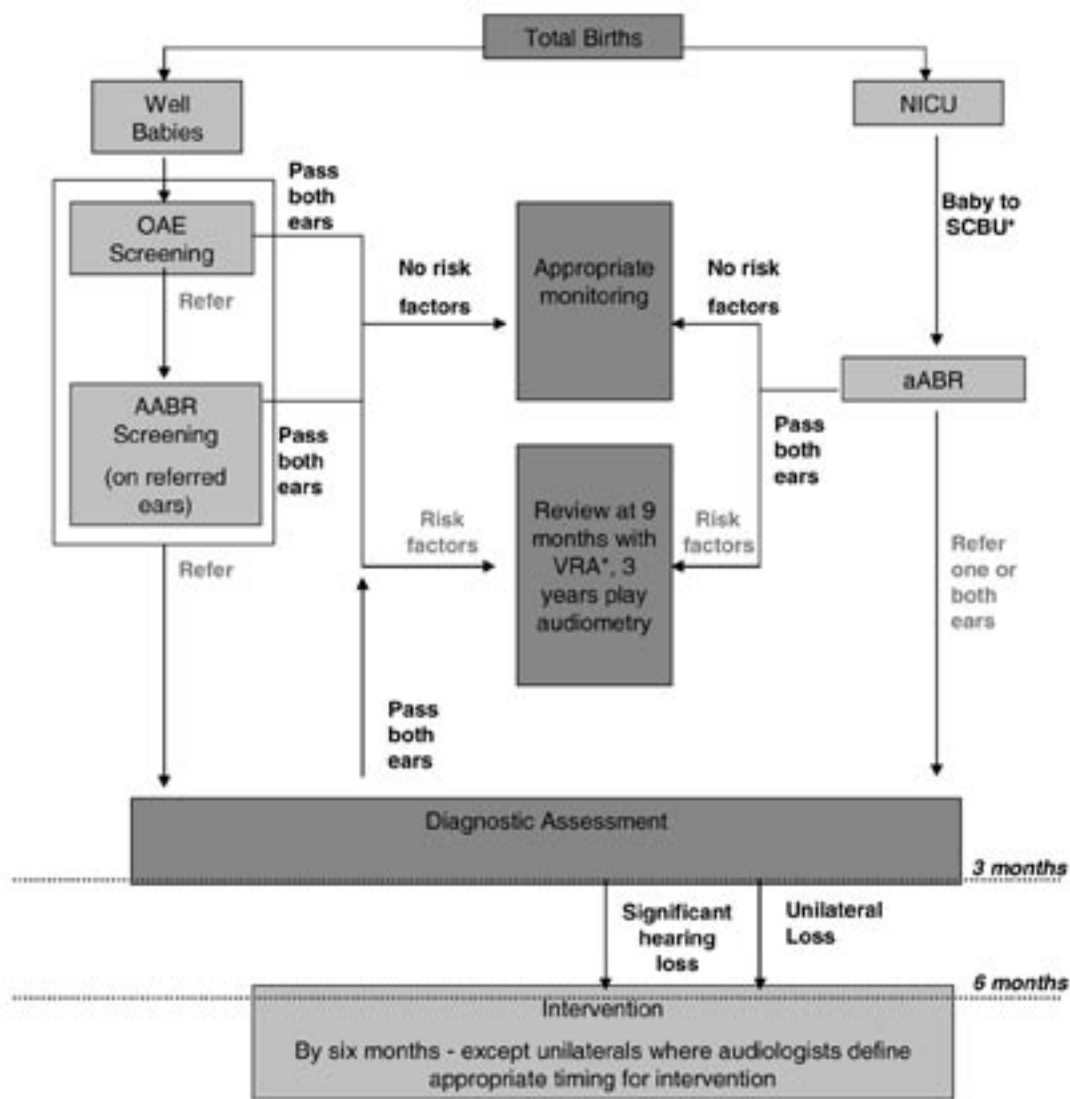
Figure 10: Example flow diagram of a screening programme (VRA – Visual Reinforcement Audiometry, SCBU – Special Care Baby Unit, NICU – Newborn Intensive Care Unit)

Children with positive screening results are referred for further testing using a combination of audiologic (diagnostic ABR) and otolaryngological consultation, to finally determine whether a hearing loss is present. Once the diagnosis is confirmed, the child can then be enrolled in an intervention programme. Depending on the programme, pediatricians, audiologists or other professionals (such as Advisers on Deaf Children in New Zealand) may be designated as the coordinator, having overall responsibility for involving all professionals and forming an individualised plan for the child and family.

There are significant differences in the way various studies and programmes report their findings which make comparison across programmes difficult. Firstly, within a screening programme, a 'Refer' result is followed by a diagnostic test to establish whether the condition is present. But many programmes provide referral data on more than one test event, as opposed to data from one or more tests at one testing event. (Referral data from more than one screening test is reported mostly from OAE screening, where repeated tests are often conducted to reduce referral rates.) Secondly, some combination programmes report referral data from the first test and then provide second test referral data, while others have only one referral rate. Thirdly, a number of programmes include data from risk babies when technically screening takes place only within the well baby population. Finally, some programmes include only cases that attend the second event (diagnostic test) and some count inconclusive results. As methods, definitions and objectives vary considerably it is difficult to directly compare data from various programmes and studies. Specificity data is especially difficult to compare for these reasons.

9.1.1 Screening parameters

The purpose of a UNHSEI programme is to identify children with PCHI, in order that they can meet their potential in terms of communication, social development, education and employment. Ideally, a screening programme would be assessed against these outcomes, but this is difficult due to the compounding factor of later onset hearing loss, the long time-frame over which outcomes will become apparent and the expense involved in conducting large longitudinal studies. Therefore, UNHSEI programmes generally have used measures of the screening process (eg sensitivity and specificity) to define the effectiveness of the programmes and other surrogate measures such as age of intervention as there is evidence to link age of intervention to later outcomes. Table 6 outlines the reported key outcome measures of UNHSEI programmes internationally. A full listing of key metrics can be found in section *16.2: Programme performance indicators.*

Sensitivity and specificity

Sensitivity is a measure of the proportion of people in the population being screened that have the condition in question, and who are correctly identified as having the condition. This percentage is related to the percentage of those who have the condition and are not identified, which is often referred to as the false negative rate. Specificity is a measure of the proportion of people in the population being screened that do not have the condition in question, and who are correctly identified as not having the condition. This percentage is related to the percentage of those without the condition who are incorrectly identified, which is called the false positive rate (i.e. the lower the specificity of a screen, the higher the false positive rate). The referral rate for a screening programme is the number or percentage of individuals who have a positive screen result and are referred for diagnostic assessment. A summary of the range of sensitivities, specificities and referral rates from various UNHSEI programmes is shown in *Table 6.* Direct comparison of these rates is subject to the limitations listed in the introduction to this section.

As with any screening test attaining high specificity within a UNHSEI programme is important to reduce anxiety and stress related to false positives. Specificity rates vary considerably in the literature, depending on the technology and protocols used and the length of time the programme has been operating. However, in focusing solely on specificity (reducing the rate of false positives) there is the danger that sensivity is compromised. In developing and changing protocols, programmes must consider the impact on both sensivity and specificity.

The American Academy of Pediatrics suggests that the false positive rate of any UNHSEI programme should be less than or equal to 3%, and many programmes seem to have adopted this recommendation[223]. Programmes that use a two-tier protocol often describe acceptably low false positive rates, from 3.5% false positives with the first screen to 0.2% after repeat screens[192]. aABR programmes also demonstrate low false positive rates, with recent results showing rates as low as 0.56%. Although higher rates are more common in OAE-based programmes some programmes that use OAEs alone describe similarly low false positive rates in situations where re-screening is used (for example a low of 0.98%, De Capua and colleagues[265]). False positive rates in OAE based programmes are shown to improve through the use of experienced staff, long average hospital stays, and extensive re-screening of babies who fail their first test[266].

The true sensitivity of newborn hearing screening is difficult to assess unless one or both diagnostic ABR and behavioural tests (the gold standards) are performed soon after the screen on all babies. Otherwise it is difficult to separate progressive hearing losses from those present at birth[267]. There is a significant cost and time involvement in performing diagnostic ABR. Accurate determination of behavioural thresholds using Visual Reinforcement Audiometry (VRA) cannot be undertaken until the child is 6-12 months of age. These issues place constraints on the assessment of screening sensitivity, particularly as large numbers of children must be assessed to determine the true sensitivity. Although a number of studies claim to present data on sensitivity and claim up to 100% sensitivity[265, 268], these studies have not compared the screening outcomes with ABR or behavioural testing, but instead have re-screened those that passed the initial test. Davis and colleagues[267] examined some of the factors that influence specificity and sensitivity of UNHSEI programmes. They noted that there was a difference between sensitivities and specificities achieved within an experimental context, compared to those within operating programmes or realistic field trials[267].

An important investigation by Norton and colleagues provides some indication of the outcomes of screening[268]. Results

from TEOAE, DPOAE and aABR screening were compared with those from Visual Reinforcement Audiometry (VRA) at 8-12 months corrected age in 4911 infants. This study was therefore able to compare behavioural thresholds with objective screening outcomes at relatively close intervals, providing a unique insight into the effectiveness of the three commonly used screening technologies. 64% of subjects returned for VRA testing, and 95.6% completed this assessment. The study showed that test performance was similar for all three methods when compared to the VRA results, although aABR was better at measuring performance at a frequency of 1 kHz. Although the number of cases of hearing impairment within the sample was still small, the study demonstrated that a number of cases of mild hearing loss were likely to be missed with all three key technologies. Only very small numbers of significant hearing loss were not detected. The authors evaluated the data using ROC (receiver operating characteristic) analysis across different technologies and frequencies and found that with very low false positive rates the true hit rate (sensitivity) was between 0.8 and 0.9. Regardless of the proportion of referrals 100% sensitivity was never achieved[269].

The results from Norton and colleagues are similar to those found by Mason and others[270] in which screening ABR results were compared to the eventual diagnostic results on 51 hearing-impaired children. Two cases of suspected progressive hearing loss were included within the false negative results. 46 of the 51 children failed the screening ABR on one or both ears, while 42 failed on failed on both ears. This relates to field sensitivities of 90% and 82% respectively. Specificity was 93%[270].

Thus, although a number of programmes report perfect or near perfect sensitivity, these reports should be viewed with caution. These views were echoed by Johnson and others at the recent International Conference on Hearing Screening and Early Intervention in Como, Italy[271]. These authors reported on a study, funded by the Centers for Disease Control and Prevention, on 1557 infants (of a total population of 88,252) who had detailed diagnostic evaluations conducted at 8-12 months and two-tier screening (OAE and/or aABR) soon after birth. The study also collected detailed enrollment and diagnostic data for babies born during the same period, at the same hospitals and who were referred for diagnostic assessment following OAE and aABR screening. The authors found that a two-tier approach could potentially miss 0.59 cases per thousand, although these are mostly mild hearing losses[271]. Providing audiological evaluations at 8-12 months is thought to optimize the likelihood of finding true cases, however it is impossible to determine whether these cases are true PCHI or whether the hearing loss developed in the period between the screening and 8-12 months. As a result, these types of studies may underestimate the sensitivity of UNHSEI programmes. Aspects of programme organisation also impact on sensitivity. For example, follow up rates and coverage.

Another example of the relatively low false negative rate comes from an examination of the rate of later detections in some of the international programmes. For example in the Flanders programme, where about 56,000 babies are screened each year, there have been no reports of children that have been identified later as having a hearing loss that was thought to be present at the time of screening[272].

On the basis of this background the number of babies that are potentially missed in a high quality screening programme appears to be small (and is likely to consist mostly of mild hearing losses) although claims of 100% sensitivity should be viewed with caution[268, 272]. A key challenge of the programmes over the past few years has been to increase specificity without sacrificing programme sensitivity[273].

Referral rates

Referral rates are the total proportion of screened infants referred (refer result) from a UNHSEI programme to diagnostic assessment. Again, reported rates differ depending on a number of factors. A number of important professional bodies (such as the American Academy of Pediatrics and the Joint Committee on Infant Hearing in the USA) have recommended that the referral rate must be lower than 4%[26, 223] primarily to reduce parent anxiety.

White recently conducted a thorough review of the state of Early Hearing Detection and Intervention (EHDI) programmes operating within the United States. As the number of hospitals conducting newborn hearing screening experienced a 20-fold increase in the 5 years to 1998, there had been many changes since his last review in 1995. White examined expected refer rates for three major protocols, and for these data calculated that screening 1000 babies would result in the following expected refer rates:

OAE followed by OAE – 920 pass, 80 referred then 72 pass on second screen and 8 would be referred for diagnosis of whom 5 would have normal hearing and 3 would have a hearing loss. (8.0% initial refer rate, 0.8% final refer rate)

aABR - 960 passed, 40 referred for diagnosis, of whom 37 would have normal hearing and 3 would have hearing losses. (4% refer rate)

OAE followed by aABR – 980 passed and 20 referred for diagnosis, of which 17 would have normal hearing and 3 would have hearing losses. (2% refer rate)[26]

Another review of US programmes conducted in 1998 reports refer rates for 64 OAE-based programmes at 8.4% compared to 56 aABR-based programmes at 4%[274].

	Sensitivity	Specificity	Referral Rate
Two -Tier	(No data available)	99%+ [275]	0.7% [276] -4.67% [277]
OAE	90.5% [278] - 100%[279]	65%[266]– 99.02% [265]	0.8% on final re-screen[280]-47.5% (after one initial screen and depending on the length of stay in hospital [281])
aABR	98% compared to diagnostic ABR[256]	99.8% after second screen [192] to a low of 96.5% after first screen[192]	0.56% [272] – 11.5% [282]

Table 6: Summary of key performance indicators from international UNSHEI programmes. N.B. Some of the specificities listed above were calculated from reported false positives.

Coverage

In order to calculate a true coverage rate, the denominator (total number of actual births) must be known. In many countries however, including New Zealand, this number is not accurately known. A reasonable surrogate denominator for a new or emerging New Zealand programme is the number of babies having metabolic screening.

Reported coverage rates among UNHSEI programmes vary considerably, although many programmes are now achieving high coverage rates (close to 100%)[192, 272, 283, 284]. These results must be viewed conservatively as not all programmes are likely to have a true understanding of the number of births in their target population.

Examples of programmes achieving high rates include the 97% coverage achieved in Hanover with OAE-based screening[283], 99.7% in Singapore using OAE/aABR[284] and 99.5% in Flanders in 2000 using aABR (mostly outside the hospital setting)[272]. Similar coverage rates were achieved in Honolulu, with 10,372 infants screened over a 5 year period and an average 98% coverage rate[192]. This rate had risen to 99.9% in year 5, with the remainder of babies screened as outpatients, bringing the final coverage to 100%[192].

A survey of 120 US screening programmes in 1998 reported that 94.9% coverage was average for OAE programmes (64 programmes) with 96.2% average coverage for aABR based programmes (56 programmes)[274]. More recently, a State-by-State Report Card[285], examining the coverage of various American universal newborn hearing screening programmes and their funding sources reported that 74% of US states had 94% or greater coverage. Nine of those states were achieving 95-100% coverage while only 3 screened 80% or less of the target population.[285]

Other examples of coverage include only 87% coverage attained in the Wessex Trial[35]; 90.6% in a Brazilian TEOAE based programme[286] and 91.5% of 11,606 screened using TEOAEs in East London[273].

Various approaches have been employed to improve coverage rates. Some programmes for example, screen infants as outpatients that were missed before discharge. Messner reports increasing US, volunteer-based aABR programme coverage rate from 91% to 95% by using an outpatient follow-up approach[287].

Coverage in the Tairawhiti hospital based programme is 89%. Expected data from the Waikato universal newborn hearing screening programme will provide a better indication of what can be expected from a regional UNHSEI programme in New Zealand.

Positive predictive value

The positive predictive value (PPV) is the probability that an individual with a positive screening result has the condition that the screen aims to detect. The PPV is sometimes given for the overall programme, or for high or low-risk groups. The authors of this report could not determine how this statistic was calculated in some studies. As a result, only PPVs from studies and programmes where this statistic seemed reliable are presented.

The 2001 Systematic Evidence Review prepared for the US Agency for Healthcare Research and Quality (AHRQ) compared the PPVs for high and low-risk groups from a number of programmes[288]. Predictably, a higher PPV was demonstrated for high-risk groups (low of 12.5% PPV for New York State and Honolulu, high of 75% Rhode Island) with low-risk PPVs ranging from 2.2% (Wessex, New York State and Honolulu) to 9.9% in Rhode Island and 22.2% following second stage OAE screening of these low-risk infants in Paris. The overall PPV from the Wessex trial was 6.7%[43].

Keren and colleagues (2002), calculated key metrics for a hypothetical cohort of 80,000 newborns containing 128 children with bilateral hearing impairment of 40dB or greater. They compared no screening, selective screening (eg risk factor approach) and universal screening, which involved a two-tier approach using TEOAE followed by aABR. Infants at increased risk were screened using aABR that was repeated on positive screens. The model used international data (on the number of cases where selective screening was not conducted, follow-up testing was not completed, true cases were missed and hearing loss was not likely to be confirmed before 6 months of age) to estimate yields and PPVs. Within the cohort, the authors estimated that the selective screening approach would identify 48.4% of the hearing-impaired infants in the population, referring 0.18% of all infants to diagnostic evaluation. This would give a PPV of 43%. Universal screening would identify 90.3% of the total hearing-impaired infants in the population, with 1.6% of all infants being referred for diagnostic evaluation, giving a PPV of 8.8%[38]. It is important to note that the targeted screening approach used in this study is different to the risk screening approach used in New Zealand so these results cannot be compared to the current situation in New Zealand.

In 1998, Mehl and Thomson studied 41,796 infants screened as a part of Colorado's UNHSEI programme between 1992 and 1996 using ABR, aABR or OAE, although the results were not separated according to technology. The PPV of the abnormal screen was at least 5% but technology improvements over time had increased this up to 19%[289].

Positive predictive values in the New York State Demonstration Project were calculated at 12.5% in the NICU and 2.2% in the 'Well Baby Nursery' (WBN) and were reported to be similar to the values achieved with metabolic testing[290].

In summary, currently reported PPV values appear to be between 2.2% (for well babies) and 75% for risk babies. As improvements in screening procedures are leading to lower refer rates, average PPVs are likely to increase.

9.1.2 Other programme measures

Yield

Yield is defined as the total number of cases of the condition identified through a specific programme. In the case of UNHSEI, this is often reported alongside the number of infants that must be screened to identify one case of PCHI, known as the NNS (number needed to screen).

Yield rates vary significantly depending on the underlying population prevalence and factors such as the pass/fail criteria used, coverage and follow-up rates. Programmes in Rhode Island, Colorado, New York, Utah, Hawaii, and New Jersey, using a variety of different screening techniques and protocols, have reported yields of between 1.65 and 4.15 infants with PCHI per thousand screened[291].

The Systematic Evidence Review prepared for the Agency for Healthcare Research and Quality (AHRQ) in 2001 compared a number of metrics from 10 programmes and calculated the NNS to find one case of bilateral permanent hearing loss for seven programmes where data on yield was available. The NNS for Wessex, New York, Rhode Island, Texas, Whipps Cross, Paris and Honolulu programmes were; 925, 1422, 666, 855, 755, 711, and 864, respectively. Two of these, from Rhode Island and Texas, included cases of mild bilateral hearing loss, where the others only included moderate and greater bilateral hearing losses[288]. These statistics cannot be examined in isolation however, as follow-up rates can significantly influence the NNS numbers. Unfortunately a number of the programmes that were considered in this review did not focus on ensuring that those with positive screens were brought back for follow-up testing. As an example, 43.4% of the infants who screened positive (6.5% of those screened) were lost to follow-up in the New York programme, significantly increasing the number of infants that needed to be screened (to 1422) to detect one case[288].

In some areas where universal hearing screening programmes have been in place for 5 or more years, such as Utah, Rhode Island and Hawaii, the prevalence of acquired losses in young children is less than expected, thus reducing the programme yield. This may be due to the fact that many children previously identified as acquired losses actually had congenital mild or moderate progressive permanent congenital hearing impairment[291]. More research is needed to confirm this.

On the basis of the likely prevalence of PCHI in New Zealand (2.38 to 3.00) between 333 and 420 infants would need to be screened to identify one infant with moderate-to-severe hearing loss. (See section *7.2: Prevalence.*) Issues impacting this yield would include screening technology and proportion of infants screening positive that are lost to follow-up.

Follow-up

The follow-up rate represents the level of success within a programme in recalling infants with positive screening results for diagnostic assessment. Follow-up testing may comprise a 'repeat screen' or a diagnostic assessment. Low follow-up rates alone can prevent the programme from operating effectively, by limiting the number of complete diagnoses being conducted on those that have positive screening results.

Follow-up rates reported in the literature vary considerably from a high of 100%[292] down to a low of 48.8%[289]. Reaching the 95% minimum successful follow-up required for a UNHSEI programme to be considered effective by the American Academy of Pediatrics has proven a challenge for many programmes[223].

A growing number of programmes now report successful follow-up with at least 80% of the children failing their screen, exceeding the 70% threshold recommended by the Joint Committee on Infant Hearing[264]. Flanders[292] Hawaii[192], Honolulu[192] and Rhode Island[36] programmes report the best compliance in this respect with 0-13% of infants with a positive screen failing to return for further testing. The Flanders programme mentioned above is the first to achieve a spotless record in the area of follow-up with programme managers ascribing this success to a 'trace and chase' approach.[292]

A Brazilian study[286] reported that 82% had returned for follow-up tests while 86% returned for further testing in the Whipps Cross Hospital programme[273] as did the same percentage in an inner city US programme over a five year period[293]. Mehl and Thomson examined results from the Colorado screening project 1992-1999, and found that 76% of referred infants had documented follow-up testing to confirm or exclude congenital hearing loss[215], an improvement on the 48.8% of examined follow-ups in the years 1992 to 1996[289].

However, especially in the United States, very poor follow-up rates are common. New York State reports 43.4% of positive screens were lost to follow-up[193]. White reports that approximately 40% of US babies failing their first screen are lost to follow-up, and that many programmes do not keep track of the proportion of babies that are lost to the system in this way[26]. Some US programmes have had greater success in following up infants who screen positive, with an average of 71% of infants successfully followed up in the multi-site aABR programme study by Stewart and others, screening a total of 11,711 infants[294].

Data from metabolic screening programmes in New Zealand indicate follow-up rates as high as 100% can be achieved.

9.1.3 Achieving improvements in key metrics

A number of approaches are being employed to improve the efficacy of universal newborn hearing screening programmes. These include the establishment of quality assurance programmes, improvements to follow-up rates, minimisation of false positives, and steps to address skill shortages (eg paediatric audiologists). In the USA, 30 states have recently funded cooperative agreements to develop tracking and data management systems although in many cases these are still 2-4 years away from implementation[295].

In addition to these approaches for improvement, another key factor in programme efficacy is the length of programme operation. A number of studies have shown that universal newborn hearing screening programmes demonstrate improvements over time in key metrics such as false positive rates, coverage and the number of children lost to follow- up[293, 296]. White reported on the key obstacles to successful implementation of UNHSEI programmes in both 1998 and 2001, as defined by state coordinators. The top three obstacles identified were the shortage of paediatric audiologists, insufficient physician knowledge, and inadequate third party reimbursement[26]. Hospital opposition became less important between the 1998 and 2001 surveys, indicating an increase in hospital ownership of newborn hearing screening programmes over that time. 'Opposition by parents' rates the least serious obstacle in both 1998 and 2001[26].

Isaacson examined the effect of a number of variables on follow-up rates and achieved an improvement in rates from 61 to 75%[297]. This improvement was achieved through the provision of free daycare for siblings and assisting with the financial burden incurred by transportation improve the percentage of children who can be retested[297].

Minimising false positives

A number of studies have highlighted the importance of reducing false positives[298] and identified methods that are effective in minimising these rates within a UNSHEI programme. As previously mentioned, improvements in specificity must be considered in relation to sensitivity rates.

Through the selection of thresholds, screening programme managers can balance sensitivity and specificity at a refer rate acceptable to the general population. The type of screening technology used is the first and perhaps most obvious programme factor that impacts on referral and false positive rates. Vohr and others examined the costs and referral rates in programmes using either TEOAE, aABR, or two-tier protocols (OAE followed by aABR) among 12,081 newborns at 5 sites. They found that aABR had the lowest referral rate at discharge (3.21%) followed by a two-tier protocol (4.67%), with TEOAE alone last at 6.49%. Other studies (eg Gravel and colleagues[299]) have also demonstrated that a two-tier protocol achieves lower (approximately half) referral rates than TEOAE alone.

However, it can be difficult to compare refer rates, especially from the OAE-based programmes, as different programmes have different methods for calculating 'first screen' refer rates. Some 'first screen' data can involve up to three screening events over weeks or days, after which the referral rates can be reduced to 1-6% in the well baby population[195, 300] and 14% in the NICU[301]. Other programmes report higher refer rates, because they consider the proportion of babies referred following a single screening event, although this may involve one or more screens. Some reports do not specify which of these methods were used, or the actual practice within the programme may not be reflected in the calculation methods specified. As a result and where possible, higher refer rates should be considered within the context of the negative effects of repeated screening, such as increased parent anxiety.

Clemens and colleagues studied the effect of rescreening infants failing their first screen before discharge in a cohort of 5010 infants screened using aABR. The initial false positive rate was 1.9% and Clemens and others estimated this could be reduced to approximately 0.5% if all infants failing their first screen were re-screened before discharge[302]. Using this system the false positive rate was reduced to 0.8%[303].

Korres and colleagues studied how OAE pass rates increase with time after birth and found that they reached optimal rates in the third and fourth day after birth[304]. Diez-Delgado also reported an increase in pass rates from OAE-based screening,

finding a 87.5% of those newborns who failed the screen within the first 24 hours passed a repeat screen before discharge at 48 hours[305].

The collection of detailed data is also important for improving the quality of a UNSHEI programme, as it provides information by which quality can be monitored, issues can be addressed and the efficacy of improvements measured[264].

9.1.4 Recommended programme guidelines

A number of major groups and organisations have formulated guidelines and principles for use by UNHSEI programmes. These include:

The American Academy of Paediatrics (1999) recommends that a hearing screening programme should-

- Detect at minimum all infants with significant bilateral hearing impairment, i.e. those with hearing loss >35 dB in the better ear

- Have a false positive rate of <3% and referral rate <4%

- Have a false negative rate of zero[223]

The Joint Committee on Infant Hearing (USA) recommend universal newborn hearing screening benchmarks include the following (abridged):

- Within 6 months of programme initiation, hospitals or birthing centres screen a minimum of 95% of infants during their birth admission or before 1 month of age. Programmes can achieve and maintain this outcome despite birth admissions of 24 or fewer hours.

- The referral rate for audiologic and medical evaluation following the screening process should be 4% or less within 1 year of programme initiation.

- Documented efforts to obtain follow-up on a minimum of 95% of infants who do not pass the hearing screening with an ideal return-for-follow-up rate of 70% of infants or more[264].

9.1.5 Conclusion

There is good evidence that a well organised and monitored universal hearing screening programme can achieve sensitivities approaching 100% with specificities of greater than 96% and coverage in excess of 95%[43, 192, 215, 293, 302, 303]. These metrics are dependent on the appropriate choice and use of screening technology, well-trained screening staff, adequate monitoring and management systems and crucial attention to processes to ensure follow-up of children who have a positive screen.

Currently, a number of programmes overseas are focused on finding ways to reduce false positives without damaging sensitivity and to improving follow-up rates. In addition, the screening technology is constantly being refined to reduce false positives and data management systems are being developed to ensure better follow-up and auditing of programmes. The trend toward establishing quality assurance programmes and the collection of programme data should assist in the task of continuous improvement. It is important to note that the implementation of a newborn hearing screening programme does not remove the need for later hearing surveillance.

9.2 POTENTIAL BENEFITS AND HARMS OF UNHSEI

9.2.1 Potential harms associated with UNHSEI

There are a number of potential harms associated with any screening programme in which a 'well' individual is screened for an abnormality. Clayton described the harms resulting from hearing screening as those associated with false positives, which include the potential effect on bonding, anger or resentment of the parents when a child is confirmed as having normal hearing, and lingering concern over child's hearing; and false negatives, which mainly relate to inappropriate

confidence in the hearing of a child leading to delay in diagnosis; and true positives, which may lead to emotional stress and inappropriate decisions regarding future reproduction[306].

Physical

Both commonly used methods for universal screening of newborns (OAE and aABR) pose almost no physical risk as long as equipment meets internationally recognised electrical safety standards for biomedical equipment, and attention is paid to standard methods for infection control. No reports of physical injury or infection resulting from the use of the common screening technologies were found in the literature.

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False negatives

False negatives can cause harm by leading parents to have inappropriate confidence in the hearing status of their child which can, in turn, lead to an increased delay in the diagnosis of the child's hearing impairment.

The number of false negatives can be minimised through the use of appropriate diagnostic protocols and by increasing adherence to these protocols through quality control systems. The impact of false negatives can also be reduced by educating parents about the signs of hearing loss. This can be done at the time of the newborn hearing screen.

For a number of reasons all screening programmes miss some cases of the disorder for which they screen and it is important that education of both parents and professionals should emphasise this and the importance of prompt follow-up if they have any concerns about a child's hearing.

Parental stress and anxiety

Universal newborn hearing screening has been shown in some studies to increase maternal anxiety[307, 308], while other studies have not found such effects. However, research into anxiety resulting from universal newborn hearing screening is limited in a number of ways. Firstly, anxiety is often not measured over time and so the long term effects of screening on parent anxiety levels are not well understood. Secondly, stress is reported without comparison to other kinds of stress and without comparison to stress levels present in the other groups such as new mothers whose babies are not screened. Thirdly, stress has not been measured in relation to accepted levels set by parent(s) in relation to their perception of the benefits resulting from having the screening.

Lasting emotional effects of false-positive tests were examined by Clemens and colleagues. The authors reported that 9% of mothers said they 'treated their child differently' (eg spoke louder, used hand clapping to get their attention) before outpatient re-screening, and 14% reported some kind of lasting anxiety after their child passed the outpatient repeat screen. These levels of stress were not related to those found in the general population, or to levels found in parents whose child failed the outpatient rescreen. The study also reported that 94% of parents whose child had a false positive screening result, approved of universal screening[302].

In the Wessex trial, involving 53,781 babies of whom 25,609 were screened for hearing loss, parental attitudes were measured 2-12 months after screening and both anxiety and attitudes toward the baby were found to be similar for both parents of screened and unscreened infants[43]. Stuart reported similar results following comparison of the relative stress of parents whose infants passed and parents whose infants failed their hearing screen. Equivalent stress levels were found in each of these groups of parents[309].

Generally, universal neonatal hearing screening appears to be well accepted by parents with reported refusal rates ranging from 0.4% to 3.32% and the observation that parents would want hearing screening for their future children[36, 272, 310-314]. It is of note that the 3.32% refusal rate reported by the Flanders programme[272] includes cases where the parents could not be contacted after repeated attempts and is therefore not a true refusal rate.

Luterman conducted a major study into parental attitudes and found that parents on the whole wanted to know whether their child has a hearing loss at birth. The general attitude among parents towards universal neonatal hearing screening was very positive in the sense that parents reported they felt reassured by having their infant screened[310].

Watkin and colleagues conducted two studies into the attitudes of parents towards newborn hearing screening. In their most recent study in 1997, they found that only 15% of the 288 mothers questioned reported having any anxiety at all, and less than 1% stated they were 'very worried'[315, 316]. Gregory, cited in Davis, reported 90% of parents of hearing-impaired children wanted identification to be as early as possible[222].

The Gisborne universal screening programme reports that refusals are rare, and the vast majority of parents are happy to have their child tested[219]. The Waikato District Health Board Newborn Hearing Screening Programme had screened 839 children to the end of May 2004, and only three parents had refused to have their child screened.[317]

Thus overall, the research in this area indicates that parents may experience some anxiety as a result of having their child screened for hearing impairment. This anxiety has not yet been measured relative to anxiety levels experienced under a targeted approach, or with no screening at all. Importantly, parents indicate that they would like early identification of hearing loss, as the refusal rate in most programmes is very low, a demonstration of this support. Any parental anxiety can be minimised through the use of informed consent processes, provision of quality information to parents at all stages of the process, and timely follow-up of infants with positive screening results[222].

Minimising negative psychological effects

The risk of disturbing the parent-child relationship or increasing stress on parents seems to be small, and could be further minimized by improved information, rapid and effective follow-up, sound diagnostic protocols and the consistent use of adequate parental education and information[318, 319].

Provision of clear, concise information for parents seems to be one key to reducing the negative psychological effects. A significant body of international literature[320, 321] and knowledge can be used as a starting point for the design of suitable parent information. A process is also required to ensure such communication with parents is culturally appropriate and accessible.

9.2.2   Potential benefits of UNHSEI and ethical considerations

The primary benefit of a UNSHEI programme is improved outcomes resulting from early intervention. This benefit is discussed in sections *9.4 Do UNHSEI programmes result in earlier identification and intervention?* and *9.5 Does early intervention lead to improved outcomes?*

In addition, there are other potential benefits such as equity of access to services, benefits relating to early intervention and indirect benefits of such programmes.

Equity of access to services and benefits resulting from early intervention

Age of intervention has been shown to be a critical determinant of developmental potential in children with hearing impairment. Differences in age of detection and intervention thus impose differential limits on the efficacy of intervention. As a consequence, there is inequality among children with PCHI. In New Zealand, these differences can be seen in differing ages of identification between ethnic groups and regions. This is in addition to the underlying inequality of access to personal development that exists already between hearing-impaired children and their hearing peers.

UNHSEI programmes remove these inequalities by providing access to early intervention, and minimise, as far as is currently possible, the inequality between children with PCHI and hearing children. Moreover, a well organised and sustainably funded national early detection and intervention programme with adequate monitoring and quality assurance systems is likely to result in higher (and hence more equitable) minimum service standards across the country than currently exist from region to region.

Parent views and perceptions on early versus late intervention have not been well studied (compared with their views on the acceptability of hearing screening). One study by Watkin and colleagues did, however, examine parent opinions of early versus late identification, by questioning 208 parents of hearing-impaired children who were not identified through a UNSHEI programme. The majority of respondents to the survey (89%) said they would have preferred a newborn screening programme to the method for identification they experienced. Only those parents whose child was identified before 18 months of age, or whose child had a mild hearing loss were satisfied with the age at which the hearing loss was confirmed[316].

The National Health Committee document on Screening to Improve Health in New Zealand, states that equity of access to quality services is important and that 'screening must reduce rather than exacerbate any inequalities that exist'[44]. As described in section *7: Permanent congenital hearing impairment in New Zealand*, there are ethnic differences in the age of identification and intervention.

Quality standards required by a national UNHSEI programme would ensure the same standard of care for all children screened. The current situation sees universal screening programmes operating in a small number of regions, often sporadically, and without commonality in protocols, standards and communications. A screening programme with high coverage rates would allow equal access to identification and treatment to newborns with PCHI, regardless of location, ethnicity or the presence of risk factors. Such a service would provide consistent screening service over time.

Other potential benefits

There are many additional benefits of UNHSEI programmes. These include potential benefits to families of children without PCHI, benefits to parents of children with PCHI, and benefits to society as a whole.

- Children identified with PCHI may benefit from the introduction of a universal newborn hearing screening programme as parents will be educated at the earliest possible time about hearing impairment, increasing their awareness of hearing loss and potentially improving vigilance in the identification of acquired or progressive hearing losses in childhood.

- As a group, parents of children with PCHI will also benefit through a reduced long term burden (intensity and length of habilitation) as a result of early intervention.

- Parents and/or hearing-impaired children may also benefit psychologically by avoiding regret in the future due to the delayed diagnosis and treatment of hearing impairment.

- A national UNHSEI programme is also likely to benefit society as a whole due to the reduced downstream costs (educational, benefit payments, mental health costs) associated with PCHI.

- Early identification can allow parents the opportunity to bond with their child as a child with a hearing loss, as opposed to having to change this perception later on[322].

- Early identification of PCHI can also reduce the need for sedation during diagnostic assessments, reducing risk to the infant and stress on the family.

- Early identification of unilateral losses resulting in regular monitoring of hearing may also ensure the earliest possible intervention for those losses that progress to become bilateral.

9.2.3   Notes on informed consent

In addition to the information provided to parents, international programmes have adopted a variety of protocols for gaining the consent of parents, including (1), the need for full written consent in all cases, (2), no requirement for written consent (as in 86% of US states in 2003[26]); or (3), written refusals from parents who do not give consent. White believes that the low number of US states requiring written consent indicates that universal newborn hearing screening is now seen as a routine part of newborn healthcare[26].

In New Zealand, informed consent would be required prior to screening as outlined in the Code of Health and Disability Services Consumers' Rights[323].

9.2.4 Conclusions

UNHSEI programmes result in many potential benefits for children with PCHI (such as equity of access to screening and early intervention services, and the resulting improved outcomes for the child) their families (education, reduced burden of intervention, reduced guilt) and the community at large. New Zealand parents would, like most overseas programmes, be asked to provide informed consent before their child is screened for PCHI.

9.3 QUALITY IN THE CONTEXT OF UNHSEI

The Institute of Medicine of the National Academies (United States) have described quality issues and defined six aims for healthcare delivery. These are that healthcare be safe, effective, patient-centred, timely, efficient and equitable[324]. These aims can equally be used to inform the design and to measure the quality of a UNHSEI programme as outlined in *Table 7*.

IOM quality aims	In the context of UHNSEI this refers to	Ideal features of a UHNSEI programme in order to meet these aims
Safe	Ensuring safe screening technologies and safe usage of these technologies for infants Minimising false positives and false negatives Complete and timely follow-up	A formal technology assessment framework Limited number of approved technologies selected for use Central procurement of screening technologies
Effective	Screening accuracy Minimising false positives and false negatives Complete and timely follow-up	A standardized training programme and ongoing evaluation of screeners A formal set of protocols for screening and diagnosis with ongoing monitoring and evaluation to ensure their consistent application
Patient-centred	Designed with parent input Utilising informed consent Providing consistent high quality service over time Effective and appropriate timely communication to parents	A formal policy for effective collaboration between screening, diagnostic and intervention services to minimise harmful delays and maximize outcomes Pilot programmes to inform best practice under various New Zealand circumstances eg maximising rural coverage
Timely	Screening at birth	A formal process for utilising local and overseas expertise in programme design
Efficient	Single protocols Reducing overall number of audiological assessments	A formal process for consumer involvement in programme design and evaluation Consistent service over time and across areas of the programme
Equitable	Universal application of protocols Single protocols Screening on-going	A standardised method for collection of and reporting on metrics

Table 7: Quality in the context of UNHSEI programmes

9.4   DO UNHSEI PROGRAMMES RESULT IN EARLIER IDENTIFICATION AND INTERVENTION?

Without universal newborn hearing screening, the average age of identification in western nations is generally reported to be between 19 and 36 months[47, 212, 221, 230, 231]. These ages are far later than the age recommendations from the Joint Committee on Infant Hearing and the National Institutes of Health[111, 325] which recommend children with PCHI should be diagnosed before 3 months of age, and that those with hearing loss should be enrolled in appropriate intervention programmes by 6 months of age[325]. These age thresholds have become commonly accepted standards, with many UNHSEI programmes adopting them as programme goals[215, 326-328].

It should be noted that in 2001, the United States Preventive Services Task Force (USPSTF) reported that without screening, the mean age of identification in the USA was 12-13 months of age. However others such as Yoshinago-Itano[329] have refuted this assertion stating that the Task Force had misinterpreted data and that the true average age of identification prior to the implementation of screening was actually 20 months[36].

The USPSTF claims that the age of intervention dropped from 13-16 months to 5-7 months after the introduction of UNHS[148] have also been disputed on the grounds that intervention could not take place until after the true reported age of identification of 20 months[329].

A number of overseas programmes have demonstrated that universal screening at birth does result in a significant reduction (often to six months or less) in the average age of identification and or intervention in children with PCHI[23, 37-39, 215, 330, 331]. Hayes reports that longstanding universal programmes in the United States typically see intervention initiated before the age of 6 months[23].

Although these data are encouraging, average ages of identification and intervention should be treated with caution, as reports of the average age of diagnosis and intervention within UNHSEI sometimes do not include the sometimes significant proportion of babies who screen positive, but are lost to follow-up. As an example, only 56% of babies referred from UNHSEI programmes in the US receive their diagnostic evaluation by three months of age[26]. White suggests this may be in large part due to the shortage of paediatric audiologists[26].

Timely follow-up for a high proportion of those infants returning after a positive screen is obviously crucial to take advantage of the earlier identification possible through UNHSEI programmes[325, 328, 332]. However, White reports difficulties in ensuring diagnosed children in the United States have access to early intervention services with Early Hearing Detection and Intervention (EHDI) coordinators reporting only 53% of babies identified with a hearing loss were enrolled in an appropriate early intervention programme by 6 months of age[26]. He attributes the low enrollment rate to a systemic failure in all but 5 states to provide a clear process by which children with developmental delays (such as those due to hearing loss) can receive assistance[26].

Yoshinaga-Itano reports a large improvement in the age of identification following implementation of screening in Colorado, with the average age of identification falling to 2.5 months of age, from the roughly two year average prior to implementation. These reductions in the age of identification have been demonstrated to flow through to earlier intervention in the Colorado programme, with enrolment occurring within the first four months after birth[47].

The New York State Demonstration Project was initiated to determine whether statewide universal newborn hearing screening should be established to screen the 250,000 births per annum in the state. As part of this project, eight hospitals were funded to screen newborns over a three year period and to follow-up identified infants. The authors reported that the median age of identification and enrolment in early intervention was 3 months of age within those hospitals (similar to those found by Mason and Herrmann[192] and Watkin[333]) with 80% of infants having their hearing loss diagnosed by 5 months of age. The average age at hearing aid fitting was 7.5 months, just over the Joint Committee on Infant Hearing (JCIH) age recommendation, but still considerably earlier than before newborn hearing screening was implemented[41]. The study concluded that early ages of identification and hearing aid fitting were achievable for all babies (risk and non-risk, well and newborn unit) through a large multi-centre universal newborn hearing screening programme.

The Hawaiian UNHSEI programme reported that as the proportion of infants screened increased, there was a corresponding decrease in the overall average age of identification of the hearing impairment and the fitting of hearing aids. At the point where 19% of infants were screened, the mean age of identification was 12 months, and amplification provided on average by 19 months. This average age of identification reduced to 3 months with intervention by 7 months once 95% of infants were being screened[40].

Some of the most reliable data on the improvements resulting from implementation of UNHSEI comes from the United Kingdom. The median age of confirmation has been provisionally reported at 7.3 weeks in those areas where screening has been implemented; with 80% of babies having their hearing loss confirmed by 3 months of age. This has reduced from an average age of identification of 22 months prior to implementation of universal screening (excluding cases of auditory neuropathy), with 25% of infants undiagnosed at 42 months of age[222, 334].

Also from the UK, the Wessex trial that occurred prior to the roll-out of the national programme, compared the ages of identification and intervention achieved with UNHSEI (followed by Hearing Visitor Distraction Testing [HVDT] at 8 months) to those achieved without newborn screening, or HVDT only at 8 months. Considering infants with moderate or severe hearing loss, screening resulted in highly signficant increases in the proportion of children whose hearing loss was confirmed and intervention had begun by 10 months of age. This trial found 'early confirmation and management of PCHI were significantly increased', with an average age at the time of treatment of 5.8 months[312] through screening.

In conclusion, results from a number of overseas programmes demonstrate that screening at birth leads to a significant reduction in the ages of identification and intervention in children with PCHI[23, 37-39, 215, 330, 331]. Although some statistics, especially those from the United States are likely to overstate the improvements resulting from UNHS[288], there is strong evidence that introduction of UNHSEI programmes leads to a significant improvement in age of detection and intervention. To maximise the benefit of these programmes, accurate data collection to enable identification and correction of problems, and a focus on follow-up is required.

9.5   DOES EARLY INTERVENTION LEAD TO IMPROVED OUTCOMES?

Calls for earlier diagnosis and intervention of hearing-impaired infants have been made for almost 60 years[335]. The primary intention of earlier intervention is to allow access to sound during the critical or sensitive period for language acquisition, as the ability to use this input meaningfully deteriorates with age due to physiological (deterioration of auditory pathways), and psychosocial (attention, practice, learning) factors[1, 8, 94-97]. In order to benefit from 'critical periods' of neurological and linguistic development, the identification of hearing loss, use of appropriate amplification and medical technology, and stimulation of hearing, needs to occur as early as possible, before language deficits develop[1, 7, 9, 10, 116, 336]. In addition to acoustic input, early intervention allows visual language to be taught, during the sensitive period for language acquisition. Where this learning is during the critical period for language acquisition, a spoken second language can then later be acquired.

Although access to sound during these critical developmental periods is clearly very important for children with severe or profound hearing losses (where auditory input is minimal), early appropriate intervention is also important for those children with mild or unilateral hearing losses.[153] These types of losses are now known to affect a child's speech and language development[337].

Interventions available for children with PCHI are designed to improve communication outcomes, hearing ability, and expressive and receptive speech and language. Audiological interventions incorporate amplification strategies, such as hearing aids, designed to take advantage of residual hearing, assistive devices such as FM systems to enhance speech detection over distance or in the presence of background noise (such as classrooms); and cochlear implants, which are prosthetic devices that directly stimulate the auditory nerve. In addition, sign language is a strategy for some children with limited residual hearing. Technological interventions are used in conjunction with various types of support and educational interventions (eg speech language therapy, signed communication lessons, auditory verbal therapy) to improve outcomes previously achieved for hearing-impaired children.

This section focuses on the effectiveness of *early* intervention on outcomes in children and only touches on evidence regarding the effectiveness of specific types of intervention. The type of intervention programme has significant effects on results, supporting the importance of recognizing individual differences in intervention choices[338].

Factors influencing the effectiveness of interventions include (1) developmental timing, (2) programme intensity, (3) direct learning, (4) programme breadth and flexibility, (5) recognition of individual differences, and (6) environmental support and family involvement[339-342].

Early intervention and its relationship to outcomes

Although to date there have been no prospective, controlled studies that have directly examined whether newborn hearing screening and the consequent earlier intervention result in improved speech, language, educational or career development, these longitudinal studies are in progress in the US and UK. The lack of this type of evidence is due to high cost of conducting such studies, the length of time such programmes have been operating, and the significant period of time it takes to reach measurable outcomes (eg approximately 5 years until school age, with other key measures such as whether a child undertakes tertiary training and whether their hearing loss affects vocational choice being still further distant in time).

The number of retrospective studies examining the benefits of earlier intervention have increased dramatically over recent years although there is still a lack of high quality studies with large samples, good controls and well defined terminology[222]. There are a number of weaknesses in much of the currently available research. The first issue is that many of the studies in this area use convenience samples, with cases being included only where language assessments between 2 and 4 years of age are available[343]. This may introduce a bias as children who continue in intervention and therefore have language assessments available could have better results when compared to those who drop-out of the studies. Secondly, the dropout rates are often not reported, providing another possible bias within the sample as there may be differences in the outcomes between those that dropout and those that remain in the study[343]. Thirdly, due to the low prevalence of PCHI, many of the studies include relatively small numbers of children and who have been identified at different ages. For example, a number of older studies include children diagnosed as late as 3 years within the early identified group, broadening the early identified group[288]. As a result, it is difficult to separate the effects of early intervention from intervention at any stage in the child's development[196]. Finally, evaluation of the relationship between universal screening and outcomes can still be difficult as studies to date often focus on intermediate outcomes (such as enrolment in early intervention or expressive language scores) rather than 'outcomes of primary interest' such as communication, social functioning or occupational function in later life[288].

A significant amount of the research in this area has been generated from studies within the Colorado Home Intervention Programme (CHIP) programme. The Colorado Home Intervention Program (CHIP) began in 1969, and was later adopted by the Colorado Department of Public Health and Environment. It is unique in that it provides the only state-wide system of care in the US, serving children with hearing loss (from birth to three) and their families in their own homes. Colorado was the third state in the US to pass legislation requiring hearing screening of all children. The programme follows a 1, 3, 6 model (screening by one month, diagnosis by three, and intervention by 6 months of age).

CHIP research includes a study by Yoshinaga-Itano and others which examined 72 congenitally hearing-impaired children identified before six months of age and made comparisons with 78 children identified after that time. Both these groups received comparable ongoing intervention after diagnosis. Their cognitive function was measured and the children then had their language scores compared to a calculated cognitive quotient (indicating their age related cognitive potential). Results showed that children identified after 6 months performed on average 20 points lower than their cognitive quotient would predict, while those identified before 6 months performed to the level expected by their cognitive score. These results were independent of the mode of communication, gender, socio-economic background and degree of hearing loss[344]. Thompson and others highlight limitations of this study, in particular, that children in the later identified group had worse hearing loss, that they were more likely to use sign language, and to have mothers with lower educational achievement[343]. They claimed that all these factors were likely to impact negatively on the performance of the late identified group. However, Yoshinago-Itano and others later rebutted this argument, reporting that differences in communication mode and educational achievement were insignificant, and that differences in cognitive ability were addressed in their statistical methods[344].

Mayne and colleagues evaluated the factors related to expressive language development among 113 deaf and hard-of-hearing children in the CHIP programme. Identification of hearing loss by 6 months of age was one of the factors correlated with improved expressive language development, as was having a hearing loss as the only medical condition.

These results are also supported by studies from Apuzzo and Yoshinaga-Itano, who studied 69 high-risk hearing-impaired infants diagnosed between two and 25 months of age and enrolled in the CHIP programme[345]. These children were categorised into four groups with varying ages of identification 1) birth-2 months, 2) 3-12 months, 3) 13-24 months, and 4) 25 months or greater. These children were evaluated by their developmental quotient scores on the Minnesota Child Development Inventory (MCDI). The average expressive language scores were: identified at 0-2 months, 87.18; children identified at 3-12 months,

58.21; children identified at 13-24 months, 68.43; and children identified 25 months or greater, 58.77[345]. These results show that children identified earliest develop better expressive language than children in the other groups. However, this study has various limitations. Paradoxically, the later identified children were more likely to have more severe hearing impairments and follow-up rates were not reported. Other potential confounders such as socio-economic status and family involvement were not included as adjustments within the statistical analysis[343].

Another study of children in the CHIP programme by Yoshinaga-Itano and others is the only published study that directly compared the language performance of children born in hospitals with UNHSEI with those born in hospitals without UNHSEI. They found that hearing-impaired children identified through UNHSEI had expressive, receptive and total language scores that were within the normal ranges and that were 18 to 21 points higher than the mean (more than one standard deviation) for a group of unscreened infants. More than twice as many (56%) of the children in screened group attained language levels within the normal range compared with the unscreened group (24%). This study also found that regardless of whether the child was screened, those children identified before 6 months of age demonstrated a smaller gap between their language ability and cognitive ability than those identified after this time[346]. In a review of newborn hearing screening Thompson comments that although this study controlled for several important confounders and used validated, relevant measures of language outcomes, there were issues in the design of the study. As with many studies in this area the availability of language outcome tests determined whether each individual was selected. In addition, the samples were drawn from different areas and hospitals, which could have led to biases not controlled for in the study. Finally, the study did not use blind selection or assessments and reasons for exclusions are not reported[343].

Additional evidence for the positive effect of early identification and treatment comes from the findings of Moeller who conducted a retrospective study of 112 children enrolled for at least 6 months in a diagnostic and early intervention programme in Lincoln, Nebraska. Mean adjusted vocabulary scores were in the normal range for children identified before 11 months and were signficantly lower for children identified later. This study is unique as the regresssion analysis controlled for family involvement. In all, the age of identification explained 11.5% of the variance in vocabulary at age 5, compared to 57% explained by family involvement[347]. Thompson again comments on the similar potential for bias within this study, particularly as drop-out rates were not reported and early identified children may have more opportunity to drop-out[343].

Calderon and Naidu conducted a retrospective study of 80 children enrolled in a home intervention programme. The children were grouped by age of intervention (less than one year, one to two years, greater than two years) and assessed for receptive and expressive language. The age of entry to intervention programmes explained 43.5% of expressive language scores and 49% of the variance in receptive language[348]. Again, there were differences between the groups, which may have affected the results, as later identified children had less severe losses and early identified children had greater opportunity to drop-out of the intervention programme[343].

Downs examined the expressive language of earlier (enrolled in intervention before three months of age) and later identified (enrolled between 3 and 12 months of age) children detected through the Colorado screening programme. The results showed that at 40 months of age, earlier enrolled children performed to 87% of normal on the expressive language section of a standardized test, compared with 66% for later identified children[349].

Ramkalawan and Davis examined the literature relating to the effect of intervention age on language, and found that the lower the age of intervention, the better the outcome measures for language[350]. Current evidence suggests that language improvements resulting from early intervention continue for at least 9 years[347, 348, 351-354].

Robinshaw found that development of language in profoundly deaf children can proceed well or even normally with early appropriate intervention, although reported that limited assertions can be made with regard to early versus later intervention[355-358].

Yoshinaga-Itano and colleagues state that early identification creates the opportunity for improved speech ability, as distinct from being the primary predictor of speech ability[359].

In 1994 Bess and Paradise argued against the establishment of universal newborn hearing screening programmes in the United States on the basis that there was a lack of empirical evidence at that time to support the contention of better outcomes for children who are identified earlier. However, they later report that all children with significant losses should be identified as

early as possible[222, 360]. The Cochrane Collaboration published a systematic review of the evidence of the effectiveness of UNHSEI compared with targeted or opportunistic 'screening' and concluded that the 'long-term effectiveness of universal newborn hearing screening programs has not been established to date'. They report on the need for long term data, including randomised and controlled trails and on the lack of evidence that early diagnosis through UNHSEI leads to early treatment[361]. It is clear that there is a need for long-term prospective studies, to provide a clearer understanding of the efficacy of UNHSEI. However, Yoshinaga-Itano suggests that to conduct a randomised controlled trial in this area would require a very large number of infants (as many as 240,000) to be followed over at least 10 years[329]. Further discussion on the link between early diagnosis and treatment is contained in section *9.4: Do UNHSEI programmes result in earlier identification and intervention?*

In addition to the data presented above there is evidence that early intervention through the use of amplified residual hearing permits hearing-impaired children to become independent, speaking, reading, and contributing members of mainstream society. Many achieve age appropriate speech and language[11, 362-366]. The age of hearing aid fitting is also known to affect outcomes, with earlier fitting significantly improving speech intelligibility. Markides examined the effect of age at fitting of hearing aids on achievements in speech intelligibility among four groups of hearing-impaired children (fitted before 6 months of age, 6-12 months, 1-2 years and 2-3 years). Teachers rated speech intelligibility on a 7-point scale and results showed that the speech intelligibility of the children in the group aided earliest was significantly superior to the speech intelligibility of the children in the other three groups[367].

Finally, a number of studies have found that earlier cochlear implantation results in improved language and learning outcomes[368] although as Miyamoto points out, there is variability among subjects[369]. Cochlear implants provide auditory stimulation by electrically stimulating the auditory nerve in response to sound. These are used predominately in profoundly deaf children and adults whose deafness is due to loss of the sensory cells of the cochlea. Cochlear implants greatly improve auditory input which has been shown to enhance the rate of language development compared to children with amplification through hearing aids[370]. Govaerts examined the relative value of cochlear implantation at various ages, and found that all children implanted before the age of 6 years benefited from the implant. He reports that intervention before 4 years of age seems critical to avoid irreversible losses in auditory performance and that implantation before 2 years of age seems optimal[30]. Miyamoto and colleagues studied three groups of children who were implanted at differing ages. Those children who were implanted earlier demonstrated significant improvements on measures of speech perception and speech intelligibility, compared with the others. Miyamoto concludes that earlier implantation promotes the acquisition of speaking and listening skills[115].

Geers reports that a significant proportion of children with cochlear implants who are introduced to syntactic development between 2 and 4 years old develop age appropriate syntax and grammar[371]. There is also evidence that the rate of language development is set in the first year of development and is resistant to change throughout the first 60 months of life[47].

Notes on 'mainstreaming'

Placing severely hearing-impaired children within residential settings is said to provide benefit to the deaf community, but this is sometimes coupled with a poor record of achievement, compared with mainstream education, which produces better achievement levels but results in increased social isolation[372, 373]. One of the aims of interventions for hearing impairment is to assist the child to develop age appropriate skills in speech and language thus allowing the child to participate in the most appropriate and least restrictive learning environment. Successful mainstreaming is often seen as the final measure of success and studies that have examined the impact of the intervention on mainstreaming have demonstrated that the earlier the intervention, the greater its success[374 375].

However, a large number of confounding variables must be controlled when determining whether good communication skills are more likely to lead to mainstream (or the least restrictive and most appropriate) placement. Studies with children in whom hearing aids were fitted late are limited although research suggests that hearing-impaired and deaf children who are aided early can be successfully mainstreamed[376]. As an example, mainstreaming is common among hearing-impaired children in Hawaii as a result of early intervention through the Hawaiian UNHSEI programme[40].
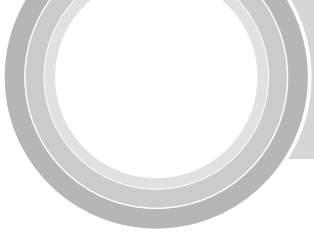
9.6   CONCLUSION

Overall, UNHSEI programmes are successful in reducing the age of detection for children with PCHI, enabling access to early intervention and improving outcomes. Despite difficulties with direct comparisons, many programmes are now attaining acceptable results in key programmes measures such as sensitivity, specificity, coverage, follow-up and false positive rates.

10. UNHSEI PROGRAMMES: COSTS, BENEFITS AND COST EFFECTIVENESS

This section will review information on the costs and cost-effectiveness of UNHSEI programmes. A number of cost perspectives must be considered; costs and benefits to the individual, the healthcare system and society as a whole. Forty three papers containing cost information were reviewed for this section. The authors of this report have made no attempt to assess the relative quality of these papers, but rather have provided an overview of the conclusions provided within.

In addition to the information presented below, there is a great deal of literature on the cost-benefit ratios for hearing-impaired children who are identified and are provided with interventions such as cochlear implantation[30, 115, 375, 377-381]. As these studies are only indirectly relevant to the discussion of costs and benefits coming from UNHSEI they are not examined in detail within this document. These studies often refer to the cost per quality adjusted life year (QALY), and there is general agreement that the costs associated with cochlear implantation are either acceptable or very acceptable under most circumstances[371, 382-385].

10.1 INTERNATIONAL PROGRAMMES

Most international cost benefit studies have focused on severe or profound hearing impairment as costs and benefits associated with less severe hearing impairment are more difficult to calculate. As pointed out by NCHAM (National Center for Hearing Assessment and Management, USA), the overall analysis of the cost benefit of hearing screening is difficult and the cost benefit studies suffer from major weaknesses such as they:

- are often based on assumptions or estimates of cost information as opposed to actual expenditure[386]

- are incomplete and do not consider some important costs and/or benefits[386] (eg depreciation, discount rates, overheads)

- consider only a part of the process (usually focusing on detection and ignoring intervention)[386]

- often do not include a comparison of the costs and benefits that would occur if a screening programme was not in place

An analysis of the existing literature in this area proved difficult because there were substantial differences in currency, the age of the studies, technologies used, and programme design, technology and programme screening protocols, the types of costs included (eg screeners' time, overheads, depreciation, capital expenditure etc.), processes involved in the study (eg screening process only vs. screening and diagnosis) and other programme specifics (eg use of volunteer personnel, existing equipment in use etc.). In addition, no studies reviewed included non-dollar costs within their calculations (eg cost of false positives).

The following is a review of studies in this area but the caveats outlined above need to be considered when comparing the results and assessing their validity.

10.1.1 Review of cost information

Forty-three papers containing cost information were surveyed and the more recent papers provide the main focus.

The reported cost per child screened ranges from a low of US$7.42 per infant (Utah, Weirather, 1997[387]) to US$33.05 (USA, Vohr, 2000[277]) or approximately £25 (UK, Bamford, 2003[39]). A number of programmes have reported very similar cost of US$25.00 (Nottingham, Mehl,1998[289], Netherlands, van Straaten, 1999[256], Washington, Kezirian, 2001[388], Colorado, Downs,1995[349], Washington, Kanne, 1999[194]).

Key cost studies:
- In 1995, The Rhode Island Hearing Assessment Programme was the first to calculate costs using actual expenditures and found a cost of US$26.05 per child screened, based on screening 4253 newborns[389].

- In 1997, a multi-center study estimated the cost of newborn hearing screening programmes in 6 hospitals in six states, using both aABR and OAE, using self report questionnaires and with some validation of this data by the authors. Costs were US$17.96 per baby screened using TEOAE and $26.03 per baby screened using aABR. This study did not include the staff time involved but did use recognised methods for the calculation of other costs[390].

- In Britain, Stevens and colleagues reported costs for ten different hearing screening programmes (a mix of targeted

and universal programmes). Costs per baby screened were lowest for risk factor programmes (US$8) and US$22 for the universal programmes[391]. These results are thought to be credible, despite a lack of information on how the costs were obtained[392].

The cost per child screened increases when costs of diagnostic evaluations are included. Diagnostic costs, although part of total programme costs, are incurred whether a UNHSEI programme is in place or not, as children are identified regardless and therefore require these services, but at a later time. This is demonstrated by one US study which included post discharge screening and diagnostic evaluation within its calculations. This study found that costs for a TEOAE based programme were US$58.07 compared with $45.85 for an aABR based programme (2002, Lemons, Indiana[393]).

Educational and lifetime costs of pre-lingual deafness

Few attempts appear to have been made to calculate the long-term cost savings of early identification and intervention. Keren and colleagues (USA) 2002, compared no screening, selective screening (eg risk factor approach) and universal screening for a hypothetical cohort of 80,000 newborns containing 128 children with bilateral hearing impairment of 40dB or greater. In their model the lifetime societal costs of congenital deafness included lost productivity, special education, vocational rehabilitation, medical costs, and the cost of assistive devices[382]. Mohr and colleagues lifetime cost estimate for pre-lingually deaf individuals of US$1.1 million dollars was used[394]. They assumed that improved language outcomes resulting from early intervention would result in a 75% decrease in lost productivity[382] and that improved language outcomes would result in a 10% decrease in special education needs and a 75% decrease in vocational rehabilitation needs. The total reduction in costs was therefore estimated at US$430,000 per deaf individual. The authors concluded that UNHS has the potential for long-term cost-savings when compared with selective hearing screening or no screening. It is important to note that the selective screening methods assumed in this study involved referring babies with a risk factor for aABR testing. This approach is likely to involve significantly less cost than the targeted approach used in New Zealand, where all babies with one or more risk factors should be referred for diagnostic assessment.

In addition, The U.S. Department of Education calculated that the annual cost of education in a regular mainstream classroom in 1990 was $3,383, while the annual costs for a hearing-impaired child in a self-contained classroom or residential placement was $9,689 and $35,780, respectively[395]. On this basis there would be substantial savings over the educational lifetime of a hearing-impaired child if the most appropriate educational setting for the child was more mainstream as a result of early detection and intervention. UNHS followed by diagnostic physiological and behavioural audiological follow-ups for infants with positive tests, and appropriate intervention, should significantly reduce this cost.

Despite difficulties in generalising the results of these studies to the New Zealand environment, cost reductions are believed to result from the early identification and intervention made possible through UNHSEI. 13 recent studies (post 1998) in which clear conclusions on cost benefit were reached were selected and conclusions are grouped below:

Cost effectiveness and feasibility

- 'UNS offers the most cost effective overall approach with alternative systems in place to identify late onset permanent hearing losses.' (UK, Stevens, 1998[391])

- 'Universal hearing screening with evoked OAE is logistically and economically feasible.' (Spain, Diez-Delgado, 2002[305])

- 'Universal newborn hearing screening using TEOAEs proved to be a cost effective and feasible method of identifying congenital hearing loss in Taiwan.' (Taiwan, Lin 2002[24])

- 'Hearing screening in a hospital-based newborn population is both feasible and cost effective.' (Alberta, Dort, 2000[396])

- 'These data lead us to conclude that all infants can be screened in a cost-effective manner.' (Nebraska, Gorga, 2001[397])

- 'Auditory brainstem response hearing screening of newborns at risk for significant hearing loss is a clinically efficient and cost effective approach to early detection of significant hearing loss.' (USA, Van Riper, 1999[398])

Justification and comparison to other forms of screening

• 'Universal newborn hearing screening is feasible, beneficial, and justified, as indicated by the frequency of the disease, the accuracy of screening tests, the ability to provide early intervention, the improved outcomes attributable to early amplification, and the recovery of all screening costs in the prevention of future intervention costs. …comparable cost per case diagnosed when compared with hypothyroidism or phenylketonuria, for example.' (Nottingham, Mehl, 2001[289])

• 'A Universal Newborn Hearing Screening is necessary and can be executed with calculable effort with concern to personnel, time and finance.' (Denmark, Bretschneider, 2001[399])

• 'Initial costs range from US$15 to US$25 per test, which is similar to neonatal screening for metabolic diseases. In addition to individual healthcare savings, early diagnosis may lead to savings on costs of intensive speech-language intervention and educational facilities.' (Denmark, van Straaten, 1999[256])

Potential for long term cost savings

• 'The short-term cost-effectiveness of UNHS is comparable to the cost per case diagnosed of other newborn screening programs and could be improved by increasing the rate of follow-up to diagnostic evaluation after positive screening test results. If early identification results in improved language abilities, lower educational and vocational costs, and increased lifetime productivity, then UNHS has the potential for long-term cost savings compared with selective hearing screening and no screening.' (Boston, Keren, 2002[38])

• 'With the present assumptions, it is shown that initially, the costs of UNHS exceed its benefits. However, after only four years of operation, UNHS programs will result in a net savings to society.' (Nebraska, Gorga, 2003[400])

• 'The particularly high costs associated with pre-lingual onset of severe to profound hearing impairment suggest interventions aimed at children, such as early identification and/or aggressive medical intervention, may have a substantial payback.' (USA, Mohr, 2000[394])

Relative cost

• 'Universal screening detects more cases of congenital hearing loss, at the expense of both greater cost and more false-positive screening results…[compared with targeted screening]' (North Carolina, Kemper, 2000[401])

Conclusions regarding cost effectiveness are highly situation dependent. In addition, it is also possible that there may be other ways to provide equally positive outcomes. Despite these considerations, international experience suggests in many contexts UNHSEI is considered cost effective and feasible. Examination of the potential cost effectiveness within the New Zealand situation is warranted.

10.2   INTERNATIONAL DATA ON TARGETED APPROACHES

Cost per child identified

Targeted 'screening' (risk factor approach) in one North Carolina study identified 51 of 110 cases, at $3,120 per case identified[401]. Keren and others found targeted screening identified 48.4% of the hearing-impaired infants within a hypothetical cohort of 80,000 births with a total cost per identified infant with normal language outcomes of $1,978,100 compared with $1,796,300 for universal screening.

The reported costs to diagnose one hearing-impaired infant also vary considerably due to the method of calculation, protocols used and prevalence rates. Estimates of the cost to detect one hearing-impaired infant range from US$3000 (US 1999[398]), £3000-6000 (London 1988[402]), US$5100-$9500 (Washington 2001[388]), US$9600 (UK 2001[289]), US$11,650 (North Carolina 2000[401]), $14,310 (New York City 1987[403]) to $22,114 (South Dakota 2000[404]).

In studies examining the cost effectiveness of targeted approaches, the following clear conclusions were reached:

• 'It appears that the poor performance of the … protocol is due to low specificity and sensitivity of the high risk

register. This generates a costly and ineffective program as follow-up exams focus on ruling-out false-positives rather than correctly identifying true hearing losses.' (NY City 1996[228])

- 'Reasonable outlay' (UK, 1996[405])

- 'Screening for hearing loss in high-risk neonates is highly reliable and cost effective.' (1992, Belfast[406])

10.3   COST BENEFIT FOR THE NEW ZEALAND SITUATION

Although different cost structures in New Zealand mean that the findings from overseas studies cannot be directly applied to this country[407] the studies do indicate a potential for cost savings in New Zealand were a UNHSEI programme to be implemented. A cost benefit study would need to be broad in order to consider the impact of early detection and intervention on the requirements for educational funding, support packages, disability allowances, productivity and quality of life for those children diagnosed. Ideally these implications would be considered for appropriate time horizons of 5 years and total lifetime to ensure downstream cost savings are included[408].

Table 8 shows some of the costs and benefits that would need to be considered when performing a cost benefit analysis of a UNHSEI programme here in New Zealand. It is a synthesis of those considered in a Request for Proposal from Project HIEDI, and those included in a scoping document written by R Milne of Health Outcomes Associates Limited.

Without screening	With screening	
Direct Economic Costs • Specialist care • GP care • Productivity losses • Special education • Income Support payments **Societal Costs** • Stress to individual • Loss of potential • Stress to family • Out of pocket costs to family • Time costs to family • Increased costs of stress on other government systems and NGO's	**Establishment costs** • Staff to coordinate and manage • Temporary increase in assistive devices and habilitation • Staff to train screeners • Equipment costs for screen • Design and production of parent communications • Database design and setup • Automated follow-up system eg letter generation etc • Temporary increase in cochlear implantation • Temporary bulge in number of hearing aids **Ongoing costs** • Staff to screen • Staff to coordinate and manage • Equipment renewal and replacement • Follow-up costs • Measurement of success • Improvements to system • Support staff • Training • Other employment costs • Occupancy costs • Overheads • Consumables • Administration • Additional audiology visits	**Cost Offsets** • Educational access funding (ORR's) • Child disability allowance • Adult disability allowance • Vocational support • Residential places in Deaf Education Centres (Kelston, Van Asch) • Reduced costs for accidental injury • Reduced speech/language training at school age • More rapid entry into the workforce with productivity gains (age 16+) • Reduced travel to specialised learning facilities

Table 8: Some costs and benefits associated with universal newborn hearing screening and early intervention[408, 409]

10.4   NEW ZEALAND PROGRAMMES: CURRENT AND PLANNED

The following information is intended to give the reader an idea of the costs associated with New Zealand universal newborn hearing screening programmes. It may be subject to the same limitations described for overseas programmes in section *10.1: International programmes.* Please note that these are direct costs only.

10.4.1   Gisborne

The overall cost of the Gisborne programme is approximately $11,000 per year for an OAE-based programme screening approximately 800 infants per year. This means an average cost per child screened of approximately $14.50[410]. This includes costs up to the time of diagnosis but is exclusive of equipment costs. These figures are approximations as the costs included are limited. (The original OAE screening equipment was donated to the hospital. A replacement computer has recently been purchased for approximately $3000.) The cost of detection for each of the children identified with PCHI is approximately $5075[410].

10.4.2   Waikato

The total budget to screen 2700 infants in the first year is approximately $144,000, including capital costs and first year implementation costs. This includes equipment costs of $42,000 and costs for audiology, technician, coordination and administration. [411] These costs relate to a two-tier programme (TEOAE and aABR) with a maximum of 5% of infants referred to audiology and approximately 7 children requiring hearing aids[412]. No cost has been included in programme budgets for audiology related cochlear implant intervention. These diagnostic and intervention related costs would be incurred regardless of whether a programme is in place.

10.4.3   Christchurch Women's Hospital NICU screening

The average cost of the DPOAE test in Christchurch Women's hospital was estimated at $25, including time required for testing and costs of the test[220]. This compares to an estimated cost of an diagnostic ABR of $250, including the time taken to test and the cost of consumables[220].

10.5   CONCLUSION

It is clear from overseas research that in many jurisdictions, UNHSEI programmes have been proven to be cost effective and have contributed long term cost savings. However, due to the large number of variables a comparison of costs across programmes and countries was not possible within the scope of this review. As an assessment of cost and cost benefit is only directly relevant to the specific circumstances and population studied, portability to the New Zealand situation is only indicative. A local study needs to be undertaken to fully explore the costs and benefits in the New Zealand environment.

11. LOCAL SUPPORT AND ENDORSEMENTS

11.1 SUPPORT FOR UNHSEI IN NEW ZEALAND

The early identification of hearing loss in infants has been a priority for hearing related health care professionals, support groups and government departments since the early 1980's. In the mid-1980's, the risk factor approach was implemented nationally. At the time this was felt to be the best way to detect hearing loss in infants and early studies showed high rates of hearing loss in 'high-risk' infants who had risk factors for hearing loss. In addition to the use of risk factors, WellChild Providers were also charged with identifying and referring children suspected of having a hearing loss.

Concern at New Zealand's poor performance in the early detection of hearing loss has been growing, with parent groups such as the New Zealand Federation for Deaf Children voicing protest at the consistently late average age of identification. Both the Ministries of Health and Education have also acknowledged the importance of early detection and intervention for children born with PCHI[139, 413, 414].

In 1998, a submission was made to government on the need for a programme to screen newborns for hearing loss by the National Foundation for the Deaf, Hearing House and the New Zealand Federation for Deaf Children. A delegation, led by Sir Peter Tapsell, met with representatives from Ministries of Health, Education and Social Welfare. This proposal was well received and there was an acknowledgement of the need for screening and intervention by government at that time. Whilst this may have facilitated the inclusion of early detection in a number of child health and education documents, no obvious movement toward implementation has taken place.

Throughout this time, a growing number of professional and community groups have been interested in seeing a national programme of UNHSEI established in NZ. In 2001, a group comprising representatives of consumer, voluntary and professional organisations and government ministries was formed to further this aim. (See section *2.3: Consultative Group* for a list of Consultative Group members.) This Newborn Hearing Screening Consultative Group comprises representatives from a broad range of stakeholder groups including parents of hearing-impaired children, societies of hearing-impaired and deaf people, educators of hearing-impaired children, and health professionals in the areas of child health and deafness.

In addition to broad sector support, there have been a number of local universal hearing screening programmes established, with one in Tairawhiti operating continuously since 1997. A number of other programmes have ceased operations due to a lack of sustainable funding. More recently, the Waikato District Health Board has established a hospital-based universal hearing screening and early intervention programme in the region. This programme will eventually screen 3500 babies annually, making it the largest programme of its type in New Zealand. It began in February 2004 and has the potential to be a pilot for a national programme.

Project HIEDI (Hearing Impairment: Early Detection and Intervention) was formed in November 2002 to extend the work of the Consultative Group, to raise awareness about the need for early detection and intervention in New Zealand and to review the evidence for a national newborn hearing screening and early intervention programme. A Project Manager was appointed at this time and a Steering Team formed which has been responsible for the preparation of this document.

11.2 ENDORSEMENTS

The following organisations and individuals had endorsed the Newborn Hearing Screening Consensus Statement (This statement can be seen in section *11.2.1.*) at the time of publication. Further endorsements are pending.

- Association of Advisers on Deaf Children – still to come
- Association of Teachers of the Deaf
- North Island Cochlear Implant Programme
- Deaf Education Aotearoa New Zealand (DEANZ)
- Hearing House
- Immunisation Advisory Centre, University of Auckland
- Itinerant Teachers of the Deaf
- Kelston Deaf Education Centre

- National Ear Nurse Specialist Group

- National Foundation for the Deaf

- New Zealand Audiological Society

- New Zealand Federation for Deaf Children

- New Zealand Society of Otolaryngology Head and Neck Surgery

- The Paediatric Society of New Zealand

- New Zealand Vision Hearing Technicians Society

- Ngati Awa Society and Health Services

- NZ Speech Language Therapists Association

- Royal NZ Plunket Society

- The Southern Hearing Charitable Trust

- Van Asch Deaf Education Centre

Endorsements from non Consultative Group organisations:

- Brainwave Trust

- The Royal New Zealand College of General Practitioners

11.2.1   New Zealand Consensus Statement

New Zealand Consensus Statement on Early Identification and Intervention of Hearing Loss in Newborns

This Consensus Statement was endorsed by the Newborn Screening Consultative Group meeting on 6th July 2002. Formal endorsements were collected from February 2003. This Consensus Statement is based on a similar statement from the Australian National Hearing Screening Committee and their approval to use this as a foundation for the New Zealand consensus statement is acknowledged.

The Consultative Group notes:

Hearing impairment is a major congenital condition in newborns. Permanent hearing impairment affects 2-3 per 1000 live births (approximately 120-180 infants) in New Zealand each year. This is more frequent than other conditions for which infant screening currently occurs. (eg cystic fibrosis 0.5 per 1000 live births)

A significant bilateral hearing impairment, if undetected and not managed, will impede speech, language, cognitive development, and emotional and social well-being. Unilateral and mild hearing impairments can also have significant educational impacts.

International research shows that babies whose hearing impairment is detected early (before six months of age) and those who receive appropriate early intervention have significantly better language levels than those children identified after the age of six months.

The average age of detection of moderate-severe hearing impairment in children is currently 33 months in New Zealand. The current method of identifying hearing loss in newborns - referral for assessment after identification using the risk factor approach, is not working. Approximately 60% of babies identified with hearing loss have no known risk factor. For hearing losses in the mild-moderate range 30-50 dBHL hearing loss is not detected until 4-6 years.

Māori are significantly over-represented in hearing loss statistics. The 2000 Census data for children under 19 show Māori

to be 19 % of the population and in 2001 they comprised 48% of deafness notifications. Currently Māori and Pacific children are identified significantly later than other ethnic groups.

Early identification (before the age of 3 months) is now feasible using objective, physiological screening techniques (otoacoustic emissions and automated auditory brainstem response) at birth, followed by diagnostic physiological and behavioural audiological tests in babies who do not pass the initial screening. Overseas programmes have shown it is possible to achieve coverage rates approaching 100% and false positive rates as low as 2%.

Centres in the USA where newborn screening has been implemented have reduced the detection and diagnosis time to below 3 months.

The American National Institutes of Health Consensus Statement, 1993, the European Consensus Statement, 1998, the American Academy of Pediatrics, 1999, the US Joint Committee on Infant Hearing, the Australian Newborn Hearing Screening Committee 2002 have all supported the introduction of screening. It is mandatory to offer newborn screening in most states of the USA. Universal newborn hearing screening is being implemented nationally throughout England and Wales. A large scale trial of newborn hearing screening is currently under way in Western Australia and one is about to begin in New South Wales.

The Newborn Hearing Screening Consultative Group proposes that:

1. Universal newborn hearing screening of infants is feasible, beneficial, and justified.

2. Principles of equity and efficiency demand the establishment of a high quality programme of universal newborn hearing screening in New Zealand as soon as possible. This will reduce current inequities in age of detection for different ethnic groups.

3. Prompt audiological assessment must be achieved for all neonates identified by hearing screening and effective intervention must follow for those in whom the impairment is confirmed.

4. To be effective, a newborn hearing screening programme should be culturally appropriate for all groups and in keeping with the principles of the Treaty of Waitangi

5. To be effective a newborn hearing screening programme should be universal (i.e., includes all newborns), since selective screening based on high-risk criteria in practice detects at most, half of all infants with congenital hearing loss.

6. To be effective a newborn hearing screening programme should achieve high coverage and follow-up rates, relative to the total number of births in the population.

7. To be effective a newborn hearing screening programme should be co-ordinated and comprehensive in its approach. That is, it should include training and supervision of personnel, full and accessible information for parents at all stages of the programme, quality assurance, the follow-up of identified children, systems for reporting and monitoring outcomes, and counselling for parents of children with hearing impairment.

8. Models for the delivery of a newborn hearing screening programme need to be designed to take account of New Zealand patterns of population distribution and service delivery.

9. Effective universal newborn hearing screening will not replace the need for vigilance and for continued surveillance of hearing behaviour and language development to detect hearing impairment in children who have not received newborn screening or who develop permanent hearing loss at a later age.

The Newborn Consultative Group resolves that:

1. A programme of universal newborn hearing screening should be introduced throughout New Zealand in order to detect children with hearing loss at the earliest possible age.

2. A Hearing Screening Committee should be established and with the Ministry of Health should work with the stakeholder groups and community to establish a coordinated screening programme. This process should be initiated by establishing pilot programmes around the country.

3. A universal hearing screening programme must be sufficiently resourced to enable high quality monitoring and evaluation.

4. A nationally co-ordinated approach will be necessary to achieve effective and efficient universal newborn hearing screening programmes for all children in New Zealand.

5. Clear timelines should be specified for the planning and implementation of universal newborn hearing screening in New Zealand.

6. The Ministries of Health and Education and the stakeholder groups should work together to ensure audiological assessment, diagnosis and habilitation occurs at the earliest possible age, and that excellent support should be achieved for all New Zealand children identified with hearing impairment.

11.3 CONCLUSION

Strong support exists among parents, educators and health professionals for a national UNHSEI programme to address the worsening problem of late detection of PCHI in New Zealand. District Health Boards have made numerous attempts to address this issue through the establishment of hospital UNHSEI programmes, although many of these have ceased operation due to lack of sustainable funding. Project HIEDI has collected formal endorsements from a large number of key sector groups, who would like to see action taken urgently to ensure improved outcomes for PCHI through the establishment of a national UNHSEI programme.

12. INTERNATIONAL SUPPORT AND EXPERIENCE

12.1   KEY EVENTS AND SUPPORTING BODIES

The following three key events that occurred between 1990 and 1994 in the USA impacted greatly on the development of policy on early detection of infants with hearing loss and contributed to a large increase in the number of universal screening programmes in operation:

- 1990: US Department of Health and Human Services specified a goal to lower the identification age of hearing loss to less than 12 months.

- 1993: the National Institutes of Health (NIH) issued a consensus statement recommending that universal hearing screening using OAEs and ABR be implemented for all newborns.

- 1994: The Joint Committee on Infant Hearing released a position statement advocating identification of hearing impairment by 3 months and enrolment in intervention programmes by 6 months of age. This statement was developed and approved by the American Speech and Hearing Association (ASHA), the American Academy of Otolaryngology - Head and Neck Surgery, the American Academy of Audiology, the American Academy of Pediatrics (AAP) and the Directors of Speech and Hearing Programs in state health and welfare agencies.

These events are thought to have contributed to a 20 fold increase in the number of UNHSEI programmes (termed Early Hearing Detection and Intervention or EDHI) in the United States from 1993 to 1998[26], and to an increase in research on their implications[415].

There is a continuing trend for many medical and educational bodies throughout the world to endorse UNHSEI. Organisations and bodies supporting the establishment of these programmes now include:

- The National Institutes of Health (USA) (NIH Consensus Statement), 1993

- European Consensus Statement on Neonatal Hearing Screening, 1998

- American Academy of Pediatrics, 1999

- US Joint Committee on Infant Hearing

- Australian Newborn Hearing Screening Committee, 2002

- American Speech and Hearing Association

- American Academy of Audiology

- Health Technology Advisory Committee (1997, Minnesota)

- Healthy People 2010: National Disease Prevention and Health Promotion Objectives (U.S. Department of Health and Human Services/Public Health Service, 2000)

These bodies all make similar recommendations, stating that the latest age for diagnosis should be three months of age, and that diagnosed infants should be enrolled in appropriate intervention programmes before six months of age. The Joint Committee on Infant Hearing and the National Institutes of Health Consensus Statement recommend screening all infants for hearing loss, preferably during the newborn period[325]. The American Academy of Paediatrics (1999) is typical in its recommendations which state that a hearing screening tool should-

- detect at minimum all infants with significant bilateral hearing impairment ie: those with hearing loss >35 dB in the better ear.

- have a false positive rate of <3% and referral rate <4%.

- have a false negative rate of zero[223].

Healthy People 2010 (U.S. Department of Health and Human Services/Public Health Service 1990) also concur that without early identification of hearing impairment 'it is difficult, if not impossible, for many of them (hearing-impaired) to acquire the fundamental language, social, and cognitive skills that provide the foundation for later schooling and success in society.' This body also states that early intervention improves progress in hearing-impaired children making them more successful in school and more productive members of society[235].

In contrast a few organizations or groups have expressed concern or reservations over newborn screening. The American College of Obstetricians and Gynaecologists recommends screening for hearing loss only in neonates with particular risk factors.

The United States Preventative Services Task Force examined newborn hearing screening and commented that it was not able to identify any evidence that would allow it to assess the magnitude of potential benefits or determine whether they alone were sufficient to offset the potential harms of screening[148]. This group concluded that 'not enough data exists to support the assertion that newborn screening leads to improved language outcomes'. This position has been criticised[329] as it does not acknowledge the crucial role of intervention in leading to these improved outcomes, which provides an opportunity through which improved outcomes can be achieved. In addition, the group has been criticised for requiring evidence from controlled prospective studies, which could be considered unethical to conduct and which would necessitate prohibitively large numbers of participants, especially if it attempted to control the numerous intervention variables that would be needed to truly determine the result of screening on outcomes.[329] (Further criticism of the USPSTF is discussed in section *9.4: Do UNHSEI programmes result in earlier identification and intervention?).* The Task Force did state that implementation of universal newborn screening is feasible and does reduce the average age of identification[295], although the Task Force's conclusions are often reported as being wholly against universal newborn hearing screening, or are given out of context.

12.2   TREND TOWARDS UNHSEI

Technological developments have made it possible to screen all newborns for hearing impairment in an accurate and relatively low cost manner. This has led to growing support for UNHSEI across many developed western nations and increasingly in countries considered 'second world', leading to its description as an international standard of care[23-25].

Currently, universal newborn hearing screening is mandated by law in 37 States[26] plus the District of Columbia in the USA 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). Programmes exist or are being implemented in Sweden, Scotland, Denmark and Croatia[28]. Regional or hospital-based screening programmes also exist in a number of other countries including Russia, Lithuania, Romania, Hungary, Brazil, Poland[416], Italy[417], Belgium[417] and Austria [418].

A number of reviews are being conducted at present into the potential value of universal newborn hearing screening. These include a Medical Services Advisory Committee (MSAC) full health technology assessment of the safety, effectiveness, and cost effectiveness of a universal newborn screening programme in Australia, following a call for the establishment of an Australian programme by the Queensland Health Minister in 2002. A review is also expected this year from the Canadian Working Group on Childhood Hearing. In addition, the American Academy of Family Physicians (AAFP) and the Canadian Task Force on Preventive Health Care are currently reviewing their positions on UNHS.

12.3   CONCLUSION

Internationally, there has been a rapid increase in the number of babies identified with PCHI through UNHSEI programmes, leading to its description as an international standard of care. This increase has been driven by the vocal support of significant medical and educational bodies and the growing evidence that UNHSEI programmes are an important tool in ensuring infants with PCHI are enrolled in early intervention programmes by the 6 month recommended age.

13. CONCLUSION

Hearing impairment contributes significantly to the national burden of disease. Permanent congenital hearing impairment (PCHI) is an important public health issue. There are varying degrees of PCHI and all affected individuals benefit from intervention.

Language development is delayed or prevented in children with PCHI. As a result, cognitive development, communication skills, educational achievement, employment, social functioning and mental health are negatively affected. Numerous studies have shown the benefits of early intervention on language development.

Children currently suspected of having PCHI are referred for diagnostic tests to confirm the deficit. At present these tests are offered to children that have one or more risk factor(s) for PCHI and to those children whose behaviour indicates to parents or healthcare professionals there may be a problem. A small number of infants are identified by the hospital based newborn hearing screening programmes currently operating in Christchurch, Gisborne and the Waikato.

About 250 children with varying degrees of PCHI are notified to the National Audiology Center each year. The median age of identification of moderate or greater bilateral PCHI was 46.1 months in 2003, much later than the average age in similar countries and the recommended international median age of 3 months. Paradoxically, some groups of children with identified risk factors are notified later than those without. Māori children are identified significantly later than other groups. The age of identification is unacceptably late in New Zealand compared to other countries and seriously affects the outcomes in these infants. However this creates great potential for improvements as a result of a universal newborn hearing screening and early intervention (UNHSEI) programme.

Reliable technologies (otoacoustic emissions and automated auditory brainstem response) suitable for mass testing of newborn babies have been developed in the last two decades. These have been used with great success by screening programmes, both in New Zealand and overseas. Although comparison between programmes is difficult for reasons of semantics and definition, coverage greater than 95% and sensitivity approaching 100% at a recall rate about 4% are now regularly being achieved. Screening dramatically improves age at diagnosis, hence age at intervention and outcome.

The cost and cost-utility studies done in New Zealand and elsewhere are of limited comparability and value in predicting the costs of a full UNHSEI programme, however many other jurisdictions are convinced of the value of screening and early intervention and have implemented programmes to achieve positive economic as well as health and educational benefits.

There is unanimous support for a New Zealand universal newborn hearing screening and early intervention programme; from consumer groups and health and education professionals working with hearing-impaired children and their families.

Implementation of a national UNHSEI programme would ensure that children are diagnosed at the earliest possible time, improving their educational, cognitive and social outcomes while reducing downstream educational costs. There would be particular benefit to Māori and Pacific children. Such a programme would not increase the number of children diagnosed overall with PCHI. Current health and education systems already deliver diagnostic and intervention services for hearing-impaired children. These systems may need to be refined to better deal with younger (earlier identified) infants.

UNHSEI meets the Criteria for Assessing Screening Programmes published by the New Zealand National Health Committee. Permanent congenital hearing impairment is relatively frequent and late diagnosis has severe impacts; there are suitable tests; there is proven benefit from early intervention; there is good evidence that screening is effective; benefits are considered to outweigh harms in jurisdictions currently screening, there is infrastructure in place for diagnosis and treatment (which may need improvement to support screening); socially and ethically hearing newborn screening is well accepted; and positive cost benefit is likely.

Screening is very well aligned with 'The New Zealand Disability Strategy' and meets many of its goals, while also making significant contributions to key Māori, Education and Health strategies. Implementation of such a programme is well supported within the sector, with both professional and consumer groups unified around this need.

Outcomes for New Zealand children with permanent congenital hearing impairment are worse than that in many countries including others not screening. It is time to redress this by providing access for all New Zealand infants to the best possible outcome through high quality newborn hearing screening and early intervention.

14. RECOMMENDATIONS

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.

- The following should be considered subsequent to any decision to implement a national programme, :

  - Addition of a small number of pilot programmes, in addition to existing regional programmes, which would be evaluated to inform 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.

15. APPENDICES: COMPLIANCE WITH DISABILITY, HEALTH, EDUCATION, AND MĀORI HEALTH STRATEGIES

15.1   THE NEW ZEALAND DISABILITY STRATEGY

Disability Strategy Objectives[419]	UNHSEI complies with the strategy by...
1. Encourage and educate for a non-disabling society.	Publicity and education resulting from a UNHSEI programme shares the message of minimal disablement for hearing-impaired individuals.
2. Ensure rights for people with disabilities.	Implementation of a UNHSEI programme sends a clear signal to deaf and hearing-impaired people and their families that their condition is an important public health problem and that early identification is a priority as it minimises the effects of their disability, enabling these groups to reach their potential. It also ensures the right of these groups to equal access to identification and intervention services, therefore ensuring full access to educational opportunities.
3. Provide the best education for people with disabilities.	A UNHSEI programme ensures fuller participation in education for all children born with PCHI through early appropriate intervention, enabling these children to reach their educational potential through access to language.
4. Provide opportunities in employment and economic development for people with disabilities.	Greater educational achievement resulting from earlier interventions will allow children born with PCHI to have broader vocational choice and employment opportunities, enhancing their independence and earning potential.
5. Foster leadership by people with disabilities.	Maximum benefit from education, improved cognitive development, mental health and self-esteem increases the possibility that those born with PCHI will take leadership in their chosen field.
6. Foster an aware and responsive public service.	Implementation of a UNHSEI programme will lead to improvements in the services available to hearing-impaired children and their families, and an awareness of hearing impairment both in the public service and general public.
7. Create long-term support systems centred on the individual.	Implementation of a UNHSEI programme will lead to improvements in the long-term support systems centred on the individual.
8. Support quality living in the community for people with disabilities.	Maximum benefit from education, improved cognitive development, mental health and self-esteem will lead to improvements in the quality of life of those with PCHI living in the community.
9. Support lifestyle choices, recreation and culture for people with disabilities.	Higher achievement resulting from early intervention will increase lifestyle, recreational and cultural opportunities for people with PCHI.
10. Collect and use relevant information about people with disabilities and disability issues.	A UNHSEI programme will include collection of information on the prevalence of PCHI here in New Zealand. It will also provide data that may be useful in measuring the effectiveness of early intervention services.

Disability Strategy Objectives[419]	UNHSEI complies with the strategy by...
11. Promote participation of disabled Māori.	Implementation of a UNHSEI programme will reduce current inequities in age of identification and delays in confirmation of Māori children with PCHI. Māori children form a higher proportion of hearing-impaired children than their population frequency, so the programme will be of particular beneifit to Māori. As it would be a universal programme, all efforts will be made to ensure participation by Māori, through their involvement at each stage of the design, implementation and evaluation of the programme.
12. Promote participation of disabled Pacific peoples.	Implementation of a UNHSEI programme will reduce current inequities in age of identification and delays in confirmation of Pacific children with PCHI. As it is a universal programme, all efforts will be made to ensure participation by Pacific groups, at each stage of the programme.
13. Enable disabled children and young people to lead full and active lives.	Maximum benefit from education, improved cognitive development, mental health and self-esteem will lead to improvements in quality of life for children and young people with PCHI.
14. Promote participation of disabled women in order to improve their quality of life.	Implementation of a UNHSEI programme will ensure equal participation of girls with PCHI, enabling improvements in their quality of life through early intervention.
15. Value families, whanau and people providing ongoing support.	Implementation of a UNHSEI programme will reduce the significant burden placed on families, whanau and people providing ongoing support that results from late identification of PCHI. Early intervention programmes will involve families and whanau.

Table 9: Compliance with The New Zealand Disability Strategy[419]

15.2   HEALTH STRATEGY

Health Strategy[420]	UNHSEI complies with the strategy by...
Seven fundamental principals	
1. Acknowledging the special relationship between Māori and the Crown under the Treaty of Waitangi	Māori children form a higher proportion of hearing-impaired children relative to their population, so the programme will benefit them proportionately more. Involvement of Māori in the implementation of a UNHSEI programme will be critical to ensure full participation and access.
2. Good health and well-being for all New Zealanders throughout their lives	Children detected with PCHI will receive special paediatric, otologic, audiologic and educational care. Fuller access to education, improved cognitive development, mental health and self-esteem will lead to improvements in the quality of life of those with PCHI.
3. An improvement in health status of those currently disadvantaged	Implementation of UNSHEI will increase awareness of the condition in the community and lead to improved services for management of the condition and hence to benefits for those already affected.
4. Collaborative heath promotion and disease and injury prevention by all sectors	A UNHSEI programme will require and reinforce collaboration between health and education professionals and agencies.
5. Timely and equitable access for all New Zealanders to a comprehensive range of health and disability services, regardless of ability to pay	Implementation of a UNHSEI programme would improve both the time of identification and early access to intervention for all New Zealand babies born with PCHI.
6. A high performing system in which people have confidence	A UNHSEI programme would incorporate quality parameters, regular audit and publication of results. This transparency would encourage public confidence in the programme.
7. Active involvement of consumers and communities at all levels	The success of a UNHSEI programme would require active involvement of consumers and communities at all levels, especially in the design of appropriate, accurate communications to be given to parents who are offered the screen for their babies. Consumer consultation is especially important during the implementation phase of a programme and could be facilitated through active links to the National Screening Unit which incorporates active consumer consultation.
To reduce inequalities in health status the Strategy will work to:	
• Ensure accessible and appropriate services for people from lower socio-economic groups	Being a universal programme, newborn hearing screening and early intervention would ensure accessible and appropriate services for newborns from all groups, including lower socio-economic groups, Māori and Pacific peoples. Programme audit would include assessment of relative participation by Māori and Pacific peoples.

Health Strategy[420]	UNHSEI complies with the strategy by...
• Ensure accessible and appropriate services for Māori • Ensure accessible and appropriate services for Pacific peoples	Being a universal programme, newborn hearing screening and early intervention would ensure accessible and appropriate services for newborns from all groups, including lower socio-economic groups, Māori and Pacific peoples. Programme audit would include assessment of relative participation by Māori and Pacific peoples.
In addition to those priority objectives there are 5 service delivery areas for short to medium term focus	
• Public health	The proposed programme involves universal screening of the well child population. Newborn hearing screening would be a valuable addition to the WellChild Tamariki Ora Schedule.
• Primary health care	Implementation of a UNHSEI programme will reduce the number of hearing-impaired children presenting for diagnosis to primary healthcare services. Early diagnosis will facilitate more appropriate primary health care for children with a PCHI.
• Reduce waiting times for public hospital elective services	Increased focus on early intervention is likely to encourage a reduction in cochlear implant waiting times for children.
• Improving the responsiveness of mental health services	
• Accessible and appropriate services for people living in rural areas	Being a universal programme, newborn hearing screening and early intervention would ensure accessible and appropriate services for newborns in rural areas. Metabolic screening which attains more than 99% coverage in the newborn population provides a useful model.
Relevant population health objectives	
1. WellChild	Implementation of a UNHSEI programme would contribute to the WellChild goals through improving access to education, cognitive development, mental health and self-esteem.

Table 10: Compliance with New Zealand Health Strategy[420]

15.3 EDUCATION STRATEGY

Education Priorities for NZ - May 2003[421]	UNHSEI complies with the strategy by...
1. Provide all New Zealanders with strong foundations for future learning.	Early detection and intervention for PCHI enables maximal language development and therefore maximal benefit from participation in education activities for deaf and hearing-impaired children through strong learning foundations. There is strong evidence that early intervention provides a positive foundation for improved educational outcomes.
2. Ensure high levels of achievement by all school leavers	There is strong evidence that without early detection and intervention hearing-impaired children are more likely to under-achieve throughout their educational career.
3. Ensure that New Zealanders engage in learning throughout their lives and develop a highly skilled workforce	Early intervention improves the opportunity for hearing-impaired people to become skilled participants in the workforce and lifelong learners.
4. Make a strong contribution to our knowledge base, especially in key areas of national development	Improved language and education in hearing-impaired individuals maximises their contribution to the national knowledge base.
Two overarching goals for the next three years:	
Goal One: Build an education system that equips New Zealanders with 21st century skills	
Goal Two: Reduce Systematic Underachievement in Education	
Through	
• More children participating in quality early childhood education, especially those from disadvantaged families, and making sound transitions to schooling	Hearing-impaired children with early detection and intervention will have specialist guidance concerning appropriate participation in early childhood education.
• Less underachievement at all stages of schooling, especially in literacy and numeracy, by lifting the performance of the bottom 25%	Where PCHI is detected late intervention is focused on remediation as language and learning deficits already exist. Early detection and intervention have been shown to significantly improve participation and performance, including literacy by this under-achieving group.
• Increased retention to senior secondary school, and all students leaving school with upper secondary qualifications and making successful transitions to further education, training or employment	Early diagnosis and intervention increases the educational achievement of hearing-impaired individuals and thus increases the likelihood that they will continue to higher education, both secondary and tertiary.

Education Priorities for NZ - May 2003[421]	UNHSEI complies with the strategy by...
• More adults with good levels of literacy, numeracy and other foundation skills, and increased achievement at higher tertiary levels by Māori and Pasifika peoples.	Early detection and intervention would optimise the opportunity for children born with PCHI to fully participate in the education system, improving performance by this group as children and therefore as adults. The introduction of a UNHSEI programme would lead to even greater improvements with Mâori and Pacific peoples, who are at the present time detected later (meaning they are in greater deficit by the time intervention is begun) and have longer delays in confirming diagnosis.
• **Specific GSE Vision:** Our vision is to have an educational context available to all children that maximises their well-being, their inclusion, their learning and achievement, while embracing their uniqueness, their creativity and their participation.	Early intervention is an essential prerequisite if achievement, participation and well-being are to be maximised.

Table 11: Compliance with New Zealand Education Strategy[421]

15.4 MĀORI HEALTH STRATEGY

Māori Health Strategy 2002[422]	UNHSEI complies with the strategy by...
Pathway One: Development of Whanau, hapu, iwi and Māori communities	
1.1 Fostering Māori Community development	
1.2 Building on Māori models of health	
1.3 Removing Barriers (to remove barriers to Māori with disabilities and their Whanau participating in NZ society, including Te Ao Māori)	The introduction of a UNHSEI programme would allow fuller participation by Māori with this disability in education (eg mainstream schooling, Kura Kaupapa, Kohanga Reo) and society.
Pathway Two: Māori participation in the health and disability sector	
2.1 Increasing Māori participation in decision making	
2.2 Increasing Māori provider capacity and capability	
2.3 Developing the Māori health and disability workforce	
Pathway Three: Effective health and disability services	

Māori Health Strategy 2002[422]	UNHSEI complies with the strategy by...
3.1 Addressing health inequalities for Māori	There appears to be is a disproportionately large number of Māori with PCHI. Māori children with PCHI are also currently identified later than other groups, with a longer delay in the confirmation of diagnosis. This disparity would be eliminated with the introduction of a UNHSEI programme which would ensure equity of access for all groups, regardless of geographic location, socio-economic status or ethnicity. Of all groups, Māori have much to gain from the introduction of such a system, as they are underserved by the present system.
3.2 Improving mainstream effectiveness	
3.3 Providing highest quality service	
3.4 Improving Māori health information	Information collected as a result of the establishment of a UNHSEI programme would provide important data on the numbers of Māori born with PCHI, as distinct from those who acquire hearing losses after this time. This information has never been available previously and could be used to research the characteristics (causes, changes over time) of PCHI in this population.
Pathway Four: Working across sectors	
4.1 Encouraging initiatives with other sectors that positively affect whanau ora	The establishment of a UNHSEI programme would require collaboration from Māori in addition to both health and education sectors. Early identification and intervention would not only benefit hearing-impaired Māori children but also their whanau reducing the economic, emotional, and time burden required in remedial work.
Hearing is one of eight Māori health priorities identified for progress by the Ministry of Health and DHB's.	

Table 12: Compliance with New Zealand Māori Health Strategy[422]

16.1   PREVALENCE

This section lists internationally reported prevalence rates for PCHI. These are provided 'per thousand births'. The rates are for prevalence at birth as reported or estimated by studies of universal newborn hearing screening and programmes.

Country/Region	Prevalence per 1000	Conditions/Sample
United States	**0.90-5.95**	
USA: Reports a range of current studies examining prevalence of congenital hearing loss[190].	0.90	Permanent bilateral hearing loss of more than 35 dBHL
	3.24	Bilateral hearing loss
	5.95	Unilateral and moderate hearing loss infants
Atlanta[191]	1.10	Moderate and greater losses
Rhode Island[36]	2.00	
New York State[41]	2.00	Prevalence of infants diagnosed with permanent hearing loss
Hawaii[192]	1.40	Bilateral loss requiring amplification
New York[193]	0.90	Well baby nursery
	8.00	Newborn intensive care unit
	1.96	Overall
Washington[194]	2.18	Combined incidence of conductive and sensorineural hearing loss
Utah State[195]	5.95	Unilateral or bilateral sensorineural hearing loss > 25 dBHL
Texas[423]	2.15	
Colorado[289]	2.56	
New Jersey[424]	3.30	
UK	**1.20-3.50**	
England[196]	1.32	
Trent region[197]	1.33	Moderate and greater losses
Southampton	3.50	
United Kingdom[198]	1.07 and 2.05	Adjusted for under ascertainment. Permanent childhood hearing impairment with hearing level in the better ear >40 dBHL averaged over 0.5, 1, 2, and 4 kHz.
Nottingham over the period 1983-1988[199]	1.20	Bilateral sensorineural hearing impairments of at least 40 dBHL

Country/Region	Prevalence per 1000	Conditions/Sample
Europe	**1.00-3.25**	
France[200]	1.40	
Denmark[201]	3.25	Permanent hearing impairment
Denmark[202]	1-1.50	Congenital permanent hearing impairment
Copenhagen[203]	1.97	Estimated prevalence of children provided with hearing aids
	1.50	Estimated prevalence of congenital hearing impairment
Northern Finland[204]	1.20	Overall prevalence of hearing impairments with PTA 0.5-4 kHz > or = 40 dBHL
Austria[205]	1.27	Prevalence rate of newborn hearing impairment
Norway[206]	1.00	
Other		
Brazil[286]	2.30	Confirmed hearing loss in babies

Table 13: Prevalence of PCHI

16.2 PROGRAMME PERFORMANCE INDICATORS

This programme shows key measures of various programmes and studies, ordered by measure, and listed by technology and then data within each.

Specificity	**Two Tier**
	Hunter (UK) 1994, Large maternity hospital, specificity of 99%[425].
	Kennedy (UK) 1991, a mix of low and high-risk infants. Achieved greater than 99% specificity[275].
	OAE
	DeCapua (Italy) 2003, 532 newborns tested using TEOAE, 99.02% specificity[265].
	Schonweiler et al (Germany) 2002, Comparison of novel ABR with DPOAE and TEOAE, using click evoked ABR as standard. Sensitivity for all three methods 100%. Step 2 specificity for left and right ears were 87.7/92.3% for Echoscreen, 82.4/84.4% for DPOAE and 82.4/89.1 for Evoflash[426].
	Bauman (Germany) 2001, 102 children, Echoscreen device (95.9%), Otoclass analysis software (94.2%), Echosensor device (77.3%)[279].
	Psarommatis et al (Greece) 2001, 60 children tested with OAE and compared to results of ABR, 91% specificity using 'Echocheck' when compared to ABR[427].
	Luppari (Italy) 1999, 500 children DPOAE, specificity was 84% when compared to ABR, the percentage of false positives was rather high (16.2%) and specificity was 84%[428].
	Guo & Yao (China) 1996, 132 high-risk infants tested with both OAE and ABR. The specificity of OAE compared to ABR was 95%[278].
	Watkin (UK) 1996, East London hospital screened 11,606 infants with an initial TEOAE test. 13% failed in both ears on first test with 1.75% failing both stages bilaterally[273].
	Brass et al (UK) 1994, tested 162 ears (3-6 weeks old) using narrow band stmuli against commercially available broad band TEOAE equipment. Specificity was 92%[268.]
Referrals and failure rates	**Two Tier**
	Almenar Latorre et al (Spain) 2002, 1532 newborns screened with OAE and followed by aABR. 97% passed OAE, 12% referred for ABR exploration and 0.7 % referred for full diagnostic assessment[276].
	Vohr et al (USA) 2001, clinical outcomes obtained retrospectively for 12,081 births, referral rate for two-step (TEOAE and aABR) 4.67%[277].
	Gravel et al (USA) 2000, Refer rates at discharge were twice as high using the one-technology protocol versus two-technology protocol, even when the best outcomes from programme year 3 were considered exclusively[299].
	OAE
	Jakubikova et al (Slovakia) 2003, TEOAE of 3,048 infants with no known risk factors. 4.5% failed initial screen, with 0.98% failing the second screen[429].

Referrals and failure rates continued	Diez-Delgado Rubio et al (Spain) 2002, bilateral EOAE taken in 458 infants. Until 24 hours after birth pass rate of 72.5 %, rising to 93.6% (second), 97.9% (fifth) and 94.7% (sixth) days [305].

Chapchap (Brazil) 2001, 4231 babies were tested with TEOAE before discharge. Stage one refer rate was 1.8%[286].

Liu (China) 2001, DPOAE screening of 2,998 newborns pre discharge. 90.4% newborns passed the initial OAE screen, with 91.3% of those passing the second screen after one month, making a total of 99.2% pass rate in total. (Initial fail rate of 9.6%, falling to .8% after second screen) [280].

Vohr et al (USA) 2001, clinical outcomes obtained retrospectively for 12,081 births, TEOAE referral rate 6.49% [277].

Stone et al (USA) 2000,1002 infants screened with DPOAE. 11.1% failing the initial screen with 0.1% failing the re-screen[404].

Kanne et al (USA) 1999, 2289 infants tested using TEOAE screening, 91.1% passed the first screen with a total of 2.34% referrals to audiological assessment after rescreening of those failing the first screen[194].

Paludetti et al (Italy) 1999, Screening 320 newborns without risk factors with TEOAE. 22.8% failed initial screen with 4.7% failing second screen[430].

A survey of 120 US screening programmes in 1998 (reported on the Infant Hearing website)[274, 431] 64 OAE programmes having an average pass rate of 91.6% at discharge[274].

Doyle et al (USA) 1998, Comparison of pass rates for TEOAE and aABR in 116 newborns 5-32 hours old. 57% passed the (initial) TEOAE screen[432].

Doyle et al (USA) 1997, Comparison of pass rates for TEOAE and aABR in 200 newborns 5-120 hours old. Overall, 79% passed the TEOAE screen[433].

Doyle et al (USA) 1997, Investigating the relationship between middle and external ear factors and pass rates. 79% pass rate for TEOAE screening[282].

McNellis et al (US) 1997, Fifty healthy, low-risk newborns were tested with ABR, repeated EOAE and otoscopy. EOAE initial passing rate was 61%. The EOAE pass rate improved with each retest and approximated the ABR rate by the fourth test[300].

Maxon et al (USA) 1997, screened 1328 non NICU newborns, from 6 to 60 hours old using TEOAE before discharge. Those infants 6-9 hours old had a 90% pass rate while those 24-27 hours old had a 94% pass rate[254]. The mean refer rate was 6.9%[254].

El-Refaie et al (UK) 1996, compared OAE vs aABR in SCBU infants with external and middle ear abnormalities. TEOAE initial screen pass rate 52.5% with many more infants with no abnormalities passing this screen[281].

Watkin (UK) 1996, Hospital-based screening of 11,606 infants with an initial TEOAE test. Of those receiving an initial test, 13% failed in both ears. Only 1.75% of the cohort failed both stages of the TEOAE screen bilaterally[273].

Kennedy (UK) 1991, 370 low and high-risk infants – bilateral fail rates for automated OAE 3.0% [275].

aABR

Kind and Gezin (Child and Family) report 2001. Reports on key metrics from 55,999 screened babies in 2000. 0.56% refer after first test. (Of those 90.27% had confirmed hearing losses) Total of 3.94% retests[272]. |

Referrals and failure rates continued	Vohr et al (USA) 2001, clinical outcomes obtained retrospectively for 12,081 births, aABR referral rate 3.21%[277].
	Van Stratten (Netherlands) 1999, average referral rates of 4% for hospital based ABR programmes[256].
	A survey of 120 US screening programmes in 1998 reported on the Infant Hearing website [274, 431] found that out of 56 ABR-based programmes, on average 96.0% of cases passed by the time of discharge[274].
	Doyle et al (USA) 1998, comparison of pass rates for TEOAE and aABR in 116 newborns 5-32 hours old. Overall, 92% of ears passed the aABR[432].
	Doyle et al (USA) 1997, comparison of pass rates for TEOAE and aABR in 200 newborns 5-120 hours old. Overall, 88.5% of ears passed the ABR screen, and 79% passed the TEOAE screen[433].
	Doyle et al (USA) 1997, investigating the relationship between middle and external ear factors and pass rates . 88.5% pass rates for ABR screening[282].
	McNellis et al (US) 1997, fifty healthy, low-risk newborns were tested with ABR, repeated EOAE and otoscopy. The initial ABR passing rate was 98%[300].
	El-Refaie et al (UK) 1996, compared OAE vs aABR in SCBU infants with external and middle ear abnormalities. 100% pass rate for initial ABR screen[281].
	Kennedy (UK) 1991, 370 low and high-risk infants – bilateral fail rates for ABR, 3.2% with ABR, and 2.7% with aABR[275].
	The following specificities have been calculated by using the authors' false positive rates.
	aABR
	Stewart et al (USA) 2000, 11,711 infants screened using aABR. <2% refer rate at the time of discharge with specificity of 99.1% or 98.5% if those infants lost to follow-up are included within the false positives[294].
	Clemens (USA) 2000, retrospective analysis of 5010 infants screened using aABR. 98.1% specificity rate achieved using rescreening. If all infants failing the initial screen had been re-screened this specificity would have improved to 99.5%[302].
	Mason and Herrmann (USA) 1998, 96% of 10,372 infants born in 5 year period, screened with aABR in nursery. 96.5% specificity after the initial screen, rising to 99.8% after the second screen[192].
	OAE
	Barker et al (USA) 2000, 569 newborns tested with DPOAE with results compared to ABR. Specificity ranged from 65% to 89% depending on criteria used[266].
	Luppari (Italy) 1999, 500 children tested using DPOAE. The specificity was 83.8% compared to ABR[428].
	ABR/OAE (Not separated)
	Mehl and Thomson (USA) 1998, 41 796 infants screened as a part of Colorado's UNHS programme between 1992 and 1996 using ABR, aABR or OAE (not separated). Specificity on the initial screen was 94%, recently improving to as high as 98%[289].

Sensitivity	**OAE's**
	DeCapua (Italy) 2003, 532 newborns tested using TEOAE, 100% sensitivity[265].
	Bauman (Germany) 2001, 102 children, 100% sensitivity using OAE when compared to a control BERA[279].
	Psarommatis et al (Greece) 2001, 60 children tested with OAE and compared to results of ABR, 93% specificity using 'Echocheck' when compared to ABR[427].
	Guo and Yao (China) 1996, 132 high-risk infants tested with both OAE and ABR. The sensitivity of OAE compared to ABR was 90.5%[278].
	Brass et al (UK) 1994, tested 162 ears (3-6 weeks old) using narrow band TEOAEs against a commercially available broad band TEOAE machine. Sensitivity of the method was 100% [268].
	aABR
	van Straaten (1999) examined the use of aABR for use in newborns, finding automation has a 98% agreement with conventional ABR, which is considered the gold standard in diagnostic audiology[256].
	Other
	Mehl and Thomson (USA) 1998, 41,796 infants screened as a part of Colorado's UNHS programme between 1992 and 1996 using ABR, aABR or OAE (not separated). Sensitivity is shown to be at or near 100% [289].

Table 14: Key measures of UNHSEI programmes

16.3   MILESTONES IN DEVELOPMENT OF SPEECH AND LANGUAGE

The following table shows the early milestones for age appropriate development of speech and language. It is based on a version of the models developed by Epstein and Reilly in 1989[434] and incorporates the one used by the Audiology Department at Capital and Coast District Health Board 2003.

Age	Normal development of speech and language
Birth	• reduces activity when listening to sound • startled by loud sounds • awakened by loud sounds
Birth to 3 Months	• stops activity for unfamiliar voice • coos and gurgles (repeated over and over) • laughs and uses voice when played with • watches your face when spoken to
3 Months to 6 Months	• babbles (uses a series of sounds) • responds to changes in tone of voice • looks around for the source of new sounds • makes at least 4 different sounds when using voice • babbles to people when they speak
6 Months to 9 Months	• babbles using 'song-like tunes' • uses voice (not crying) to get your attention • uses different sounds and appear to be naming things
9 Months to 12 Months	• uses jargon (appear to be talking) • uses consonant sounds (b, d, g, m, n) when 'talking' • jabbers in response to a human voice, using changes in loudness, rhythm, and tone • recognises sounds for common items (eg cup, juice)
First true words appear between 12 to 15 months of age	
12 Months to 18 Months to 20 words	• gives one-word answers to questions • imitates many new words • uses words more than one syllable with meaning (eg 'bottle') • speaks 10 to 20 words
18 Months to 24 Months	• uses own first name • uses 'my' to get toys and other objects

Age	Normal development of speech and language
	• tells experiences using jargon and words • uses 2-word sentences (eg: 'my shoe', 'go bye-bye', 'more juice')
24 Months to 30 Months	• answers questions (What do you do when you are sleepy?) • uses plurals (example '2 books', 'dogs') • speaks 100 to 200 words
30 Months to 36 Months	• uses question forms correctly (who? what? where? when?) • uses negative forms (eg: 'it is not', 'I can't') • relates experiences using 4- to 5-word sentences
3-4 years	• hears calls from another room • understands simple 'Who?' 'What?' and 'Where?' questions • responds to whispered speech • uses approximately 1000 word vocabulary - 80% of which should be intelligible to strangers • uses sentences with more than 4 words
4-5 years	• hears and understands most of what is said • voice sounds like other children • tells stories • communicates easily with other children and adults • uses adult-like grammar

Table 15: Milestones for age appropriate development of speech and language

16.4   DEAFNESS NOTIFICATION DATA

The Deafness Notification Database reports are based on notifications that meet the following criteria: 'Children under 18 years with congenital hearing losses or any hearing loss not remediable by medical or surgical means, and which require hearing aids and/or surgical intervention. They must have an average bilateral hearing loss (over four audiometric frequencies 500-4000Hz), greater than 26 dBHL in the better ear (Northern and Downs classification 1984).' The criteria for classifying different degrees of hearing loss changed after 2001. 'Degree 1' is the system of classification for degree of hearing loss used by the database prior to 2001.

The new system, 'Degree 2', brings the database in line with the system of classification used by Audiologists in New Zealand clinical practice.

Degree 1	Degree 2
Mild: 30-55dBHL	Mild: 26-40 dBHL
Moderate to Severe: 56-85dBHL	Moderate: 41-65 dBHL
	Severe: 66-95 dBHL
Profound: Greater than 86 dBHL	Profound: Greater than 95 dBHL

Table 16: Classification of hearing loss in the New Zealand Deafness Notification Database

16.5   EFFECTS OF HEARING LOSS ON SPEECH DEVELOPMENT

The following negative effects of hearing loss on vocabulary, sentence structure and speaking are adapted from those published by the American Speech Language Association[435].

Vocabulary

- Vocabulary develops more slowly in children who have hearing loss.

- Concrete words are learned more easily than abstract words. Children with hearing loss also have difficulty with function words like 'the' 'an' 'are' and 'a'.

- The gap between the vocabulary of children with normal hearing and those with hearing loss widens with age. Children with hearing loss do not catch up without intervention.

- Children with hearing loss have difficulty understanding words with multiple meanings.

Sentence Structure

- Children with hearing loss comprehend and produce shorter and simpler sentences than children with normal hearing.

- Children with hearing loss often have difficulty understanding and writing complex sentences.

- Children with hearing loss often cannot hear word endings such as '-s' or '-ed'. This leads to misunderstandings and misuse of verb tense, pluralisation, non-agreement of subject and verb, and incorrect use of possessives.

Speaking

- Sounds that are more quiet, such as 's' 'sh' 'f' 't' and 'k' often can not be heard by the hearing-impaired. Without hearing these sounds they are often excluded from speech and so speech may be difficult to understand.

- Children with hearing loss often do not hear or clearly hear themselves when speaking. This can mean they speak too loudly or softly, have unusual pitch or rate of speech again making them difficult to understand. This is often interpreted as mumbling or lazy speech.

17. TERMS OF REFERENCE

The terms of reference for this document are as follows:

- To assess the international and local evidence for the efficacy of universal newborn hearing screening, the effect of early intervention and the impact of universal hearing screening on child development.

- To assess newborn hearing screening in the context of the government criteria for screening programmes and the Health, Education, Māori and Disability strategies.

- In particular to review:

  - The importance of hearing and impact of PCHI hearing loss on children

  - Hearing loss in New Zealand children

    - Incidence and prevalence

    - Severity

    - Hearing loss in Māori children

  - Screening approaches

    - Diagnostic technologies

    - The evidence of improved outcomes

    - Cost effectiveness

    - Universal screening and early intervention

      - History of newborn hearing screening in New Zealand

      - Current approach and compliance with screening criteria in New Zealand

      - Overseas trends

      - Benefits

      - Costs

      - Compliance with key New Zealand government strategies

      - Local support

ABR/AUDITORY BRAINSTEM RESPONSE: A non-invasive test that measures the brain responses to auditory stimuli. This test can indicate whether or not sound is being detected, even in an infant.

ACQUIRED HEARING IMPAIRMENT: Hearing impairment which develops after birth.

AMPLIFICATION: The use of hearing aids and other electronic devices to increase the loudness of sounds so that they may be more easily received and understood.

ATTACHMENT: A reciprocal bond between an infant and parent.

AUDIOGRAM: A graph on which a person's ability to hear different pitches (frequencies) at different volumes (intensities) of sound is recorded.

AUDIOLOGICAL ASSESSMENT: Assessment by an audiologist with diagnostic hearing tests to assess the type and degree of hearing impairment. May also include how well a child is hearing with amplification.

AUDIOLOGIST: An audiologist is a university-trained professional who is specially qualified to measure hearing, diagnose and advise on the management of hearing disorders, and supply and fit hearing aids and other hearing devices to suit individual needs. The New Zealand Audiological Society is the professional body in New Zealand responsible for the clinical certification of audiologists.

AUDITORY PROCESSING DISORDERS: Within the lower brain there are a number of parts that respond to sound and send response to the hearing centre in the auditory cortex. These brain pathways are all involved in the complex interpretation of the sound messages from the ear to provide our sense of hearing. Abnormalities of the auditory brain pathways lead to complex problems of processing, interpreting and understanding sound and speech.

BILATERAL HEARING IMPAIRMENT: A hearing impairment in both ears.

BINAURAL HEARING AIDS: Hearing aids worn on both ears.

BONE CONDUCTION: Sound conducted through the skull.

CHRONOLOGICAL AGE/ADJUSTED AGE: Chronological age is how old the infant or child is, based on his/her date of birth. It is referred to when comparing him or her to other children born at that same time. If a baby was born prematurely, however, his/her development may be measured at his/her adjusted age. Adjusted age takes into account the time between premature birth and the actual due date of a full term pregnancy. Calculating adjusted age provides a truer reflection of what the baby's developmental progress should be.

COCHLEAR IMPLANT: An electronic device surgically implanted to stimulate nerve endings in the inner ear (cochlea) in order for the person to receive and process sound and speech.

COGNITIVE: Refers to the ability to think, learn and remember.

CONDUCTIVE HEARING IMPAIRMENT: Impairment of hearing due to failure of sound waves to reach the inner ear through the normal air conduction channels of the outer and middle ear. In children, conductive impairment is typically medically correctable, and is most often associated with otitis media. This type of impairment usually affects all frequencies of hearing and does not usually result in severe losses. A person with a conductive hearing loss is usually able to gain significant benefit from a hearing aid or may be helped medically or surgically.

CONGENITAL HEARING IMPAIRMENT: Hearing impairment present at birth or associated with the birth process.

CRITICAL PERIOD: A period during which a specific stimulus is required for normal development of the system, and during which the organism is maximally vulnerable to environmental manipulation. (Adapted from Eggermont, 1986)

DECIBEL (dB): The unit of measurement for the loudness of a sound. The higher the dB level, the louder the sound.

DECIBEL HEARING LEVEL (dBHL): This represents the decibel hearing level relative to normative data from adults.

DECIBEL SOUND PRESSURE LEVEL (dBSPL): The sound pressure level referenced to a sound pressure of 20 micropascals.

DECIBEL PEAK SOUND PRESSURE LEVEL (dBpSPL): The peak sound pressure level referenced to a pressure of 20 micropascals.

DISABILITY ADJUSTED LIFE YEAR (DALY): This is a quantitative indicator of burden of disease that reflects the total amount of healthy life lost, to all causes, whether from premature mortality or from some degree of disability during a period of time. It is calculated for a disease or health condition as the sum of the years of life lost due to premature mortality (YLL) in the population and the 'years lived with disability' (YLD) for incident cases of the health condition.

EHDI: An abbreviation for Early Hearing Detection and Intervention. This term is used mainly in reference to universal newborn hearing screening and early intervention programmes, known as EHDI programmes.

ENT SURGEON: A medical specialist doctor, who specializes in the ears, nose and throat. Sometimes an ENT surgeon is referred to as an otorhinolaryngologist or ORL specialist.

EVOKED OTOACOUSTIC EMISSIONS (EOAE): A class of otoacoustic emission (OAE) that is produced by the healthy inner ear after stimulation with sound. The measurement of these emissions can be used as an objective, passive audiological test that verifies cochlear activity, generally through the use of 'click' stimuli. This test is often used in testing infants suspected of hearing impairment. A probe is placed in the ear canal for this measurement.

FREQUENCY: The number of vibrations per second of a sound. Frequency, expressed in Hertz (Hz), determines the pitch of the sound.

GAIN: Increase in sound pressure level due to amplification. For example, a child with unaided hearing who can not hear sounds less than 70 dBHL who, with amplification can hear at 30dBHL is experiencing a gain of 40 dB.

GENETIC COUNSELING: The provision of genetic information to individuals and families with birth defects/genetic disorders (eg hearing impairment) including recurrent risk information.

HEARING SCREENING (NEWBORN): Audiometric testing of the ability to hear in order to identify individuals who may benefit from intervention with the aim of minimising morbidity associated with hearing impairment.

HEARING AID: An electronic device that amplifies sound and conducts it to the ear.

HEARING-IMPAIRED: Applies to those whose hearing is not within the normal range. It may be used to refer to those for whom the primary receptive channel of communication is, even with deficits, hearing.

HEARING IMPAIRMENT: The following classifications are used in New Zealand audiology clinical practice to describe hearing loss.

These hearing levels are measured over a 4 frequency average:

> Mild: 26-40 dB

> Moderate: 41-65dB

> Severe: 66-95dB

> Profound: greater than 95dB

INCIDENCE: The rate of occurrence of new cases of a particular disease in a population. Usually expressed as cases per thousand of population per year.

INTENSITY: The loudness of a sound, measured in decibels (dB).

NICU: Neonatal Intensive Care Unit.

MAINSTREAMING: The concept that students with disabilities should be integrated with their non-disabled peers to the maximum extent possible. Mainstreaming is one point on a continuum of educational options. The term is sometimes used synonymously with 'inclusion'.

MIXED HEARING LOSS/ IMPAIRMENT: A combination of conductive and sensorineural hearing loss and indicates disorders in both the outer or middle, and inner ear.

OTITIS MEDIA: A condition where fluid is present with or without infection, in the middle ear and may cause temporary hearing impairment, which can evolve into permanent impairment. Children with recurring episodes of otitis media may experience fluctuating hearing impairment and may be at risk for speech and language delays.

OTOACOUSTIC EMISSIONS (OAE): Sounds that are produced by the healthy inner ear spontaneously or after sound stimulation. Various types of OAE are refered to as TEOAE, DPOAE and EOAE. The measurement of these sounds can be used as an objective, passive audiological test that verifies cochlear activity. This test is often used in testing infants suspected of hearing impairment. A probe is placed in the ear canal for this measurement.

OTOLOGIST: A physician who specializes in medical problems of the ear. (See ENT Surgeon)

OTORHINOLAYRNGOLOGIST: A physician who specializes in medical problems of the ear, nose and throat. (See ENT Surgeon)

PLASTICITY: Refers to brain mutability and flexibility, which underlies alteration of structure and function over time in response to change.

POSITIVE PREDICTIVE VALUE: The positive predictive value (PPV) is the probability that an individual with a positive screening result has the condition screening was aiming to detect.

PREVALENCE: The total number of instances of a specified condition in a given population at a particular time or during a specified period. Usually expressed as rate per thousand.

RESIDUAL HEARING: The amount of usable hearing that a person with hearing impairment has.

SEMANTICS/VOCABULARY: The meaning or content of words or combinations of words.

SENSORINEURAL: A type of hearing impairment caused by damage that occurs to the inner ear (cochlea) and/or hearing nerve. Sensorineural damage is usually irreversible. This type of hearing loss can range from mild to profound and usually affects particular frequencies more than others.

SENSITIVE PERIOD: A period during which the action of a specific stimulus is required for normal development of the system, and during which the organism is *maximally* vulnerable to environmental manipulation[91].

SENSITIVITY (DETECTION RATE): The proportion of people in the screened population who have the condition in question and who are correctly identified (by the screening test or programme) as having the condition.

SPECIFICITY: The proportion of people in the screened population who do not have the condition in question and who are correctly identified by the screening test or programme as not having the condition. Specificity is also another way of expressing the false positive rate, with 100 minus the false positive rate being equal to the specificity.

SPEECH ZONE (SPEECH BANANA): On an audiological graph measured in decibels and frequencies, the area wherein most conversational sounds of spoken language occur. This is sometimes called the 'speech banana' because of the shape this area depicts on the graph.

SYMMETRY: Symmetry relating to hearing loss refers to the degree to which the level and configuration of hearing loss are the same in each ear.

TOTAL COMMUNICATION: A philosophy focused on teaching hearing-impaired children to communicate using all techniques available to utilise hearing and vision. This may include the use of residual hearing, sign language, spoken language, gestures, lip reading, cued speech or a combination of these, depending on the abilities, nature and hearing status of the child.

TYMPANOGRAM: A graph of middle ear function carried out using tympanometry. Provides indirect information on how the ear canal, eardrum, Eustachian tube, and middle ear bones are working. A tympanogram is not a test of hearing.

UNILATERAL HEARING LOSS: A hearing impairment in one ear.

19. LIST OF FIGURES AND TABLES

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