PERMANENT HEARING LOSS

Introduction

In New Zealand each year, it is estimated that 135–170 babies are born with mild to profound permanent congenital hearing loss, representing an incidence of 3 per 1,000 births [1]. The symptoms likely to be experienced by children with such hearing losses [2] are briefly outlined below.

Mild Losses (26–40 dBHL)
This may result in some difficulties in hearing soft speech and conversations (people sound as if they are mumbling) but children can often manage in quiet situations with clear voices. Speech and language usually develop normally if the child is fitted with hearing aids early.

Moderate Losses (41–65 dBHL)
This may result in difficulty understanding conversational speech, particularly in the presence of background noise. The volume of the TV and radio will need to be turned up to be heard. Speech and language will generally be affected if a hearing aid is not provided early. A hearing aid will assist most hearing difficulties if speech discrimination is good and the listening environment is not too noisy.

Severe Losses (66–95 dBHL)
This will result in normal conversational speech being inaudible and only raised voices at close distance being understood. Speech and language will not develop spontaneously in children with severe hearing loss. Hearing aids will amplify many speech sounds and will greatly assist children in developing speech, although speech quality is likely to be affected. Some children may benefit from a cochlear implant.

Profound Losses (96+ dBHL)
Learning to speak without significant support is very difficult, although there is individual variation. There is greater inconsistency in benefit derived from hearing aids: some children can understand clear speech in quiet conditions when wearing a hearing aid, while others derive little benefit. Children with losses in this range should be considered for cochlear implants, as there is evidence that these are of benefit, especially if implanted at a young age.

The Identification of Hearing Loss in New Zealand

In response to concerns regarding the late age of diagnosis of congenital hearing losses in New Zealand, which occurred at an average age 35.1 months when screening was based on the presence of risk factors [3], the Government, in its 2006 Budget, announced a funding package to establish a Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP). The UNHSEIP was rolled out during 2007–2010, and all DHBs now offer hearing screening to the families of newborn babies [4].

In tandem with this roll out, the Ministry of Health commissioned a series of UNHSEIP monitoring reports, which review the offer and uptake of newborn hearing screening at the DHB level, as well as the proportion of babies referred for audiology assessment, and the outcome of these investigations. Information on the number of babies diagnosed with permanent congenital hearing losses as a result of the UNHSEIP, as well as the average age at diagnosis, is also available from these reports [4].

Prior to the NZHSEIP, the only other source of information on children and young people with permanent hearing loss in New Zealand was the Deafness Notification Database, which collected information on children with hearing losses which met specific criteria [5]. Information from this Database is available as far back as 1998, although there was a gap in coverage during 2006–2009.

The following section begins by presenting a range of historical and contemporary data from the Deafness Notification Database on permanent hearing loss in children and young people, before reviewing information from the most recent NZHSEIP monitoring reports, on the number of babies diagnosed with permanent congenital hearing loss in recent years, and the ages at which these diagnoses occurred.

The Deafness Notification Database

Background

The aim of the Deafness Notification Database (DND) is to collect and report on new cases of permanent hearing loss diagnosed in New Zealand children and young people. The DND was funded by the Ministry of Health between 1982 and 2005, but was not operational during 2006–2009. In 2010 it was re-launched by the NZ Audiological Society, with Ministry of Health funding resuming from 2012. Although a number of changes have been made to the way in which the data are collected and reported (see notification criteria below), as much continuity in reporting has been maintained as possible between the two periods [5].

Data Sources and Methods

Indicator

Notifications to the New Zealand Deafness Notification Database

Data Source

NZ Deafness Notification Database

All of the data in this section were derived from the National Audiology Centre's Annual Deafness Notification Database Reports 1998–2004 [3], or from the 2010–2012 Deafness Notification Reports produced by Digby et al. [5]. These reports are downloadable at http://www.audiology.org.nz/deafness-notification-database.aspx

Changes to the DND Notification Criteria

During 1982–2005, when the DND was managed by the National Audiology Centre, children needed to meet the following criteria [3]:

- Be less than 18 years of age, and have a congenital hearing loss or any hearing loss not remediable by medical or surgical means which required hearing aids and/or surgical intervention.
- Have an average bilateral hearing loss (over 4 audiometric frequencies 500–4000 Hz) of >26 dBHL in the better ear.
- Children were excluded if their hearing loss was<26 dBHL, unilateral, acquired, or they were born overseas.

In 2010 the DND was re-launched by the NZ Audiological Society, with audiologists being encouraged to notify newly diagnosed cases via a new online form. Following consultation, the database was extended to include:

- Children with an average hearing loss (over 4 audiometric frequencies 500–4000 Hz) of >26 dBHL in ONE ear (i.e. unilateral losses).
- Children who were born outside of New Zealand.

Additional audiological guidance also suggested that while hearing losses arising from atresia, congenital ossicular fixation, meningitis and other acquired hearing losses should be included, hearing losses which could be fixed by the use of grommets (e.g. hearing losses associated with otitis media) should be excluded [5].

Additional Notes on Interpretation

DND data are reported by year of notification, rather than year of identification, with the degree of hearing loss assessed using the dBHL ranges outlined in the grey box above. As notification is not mandatory, these statistics may undercount the number of children with permanent hearing loss. In addition, the DND's notification criteria changed during the reporting period (as outlined above) and this must be taken into account when interpreting the data in this section.

New Zealand Distribution by Severity of Loss

In New Zealand during 2012, only 3% of notifications to the DND were for children with profound hearing losses. A further 1% of notifications were for children with severe hearing losses, while 42% were for children with moderate losses and 54% were for children with mild losses (**Table 1**). Note that the data in this table differs from that reported previously, due to a change in the way the authors of the 2012 DND report assessed the severity of hearing loss (in order to more closely align it with the way severity was calculated in 2005). This resulted in a lower proportion of children being reported as having severe or profound losses in the 2012 report, than in the 2010 report [5].

Table 1. Notifications to the Deafness Notification Database by Degree of Hearing Loss, Using the Old Notification Criteria, New Zealand 2001–2004 and 2010–2012

Degree of Hearing Loss	Proportion of Cases Notified (%)							
	2001	2002	2003	2004		2010	2011	2012
Mild	47	47	56	43		59	60	54
Moderate	35	39	33	34		33	28	42
Severe	10	9	6	15		4	5	1
Profound	8	5	5	7		5	3	3

Source: Deafness Notification Database via Digby et al. [5]; Note: Those with unilateral losses, who were born overseas, or who had acquired losses were removed in order to maintain consistency with earlier criteria

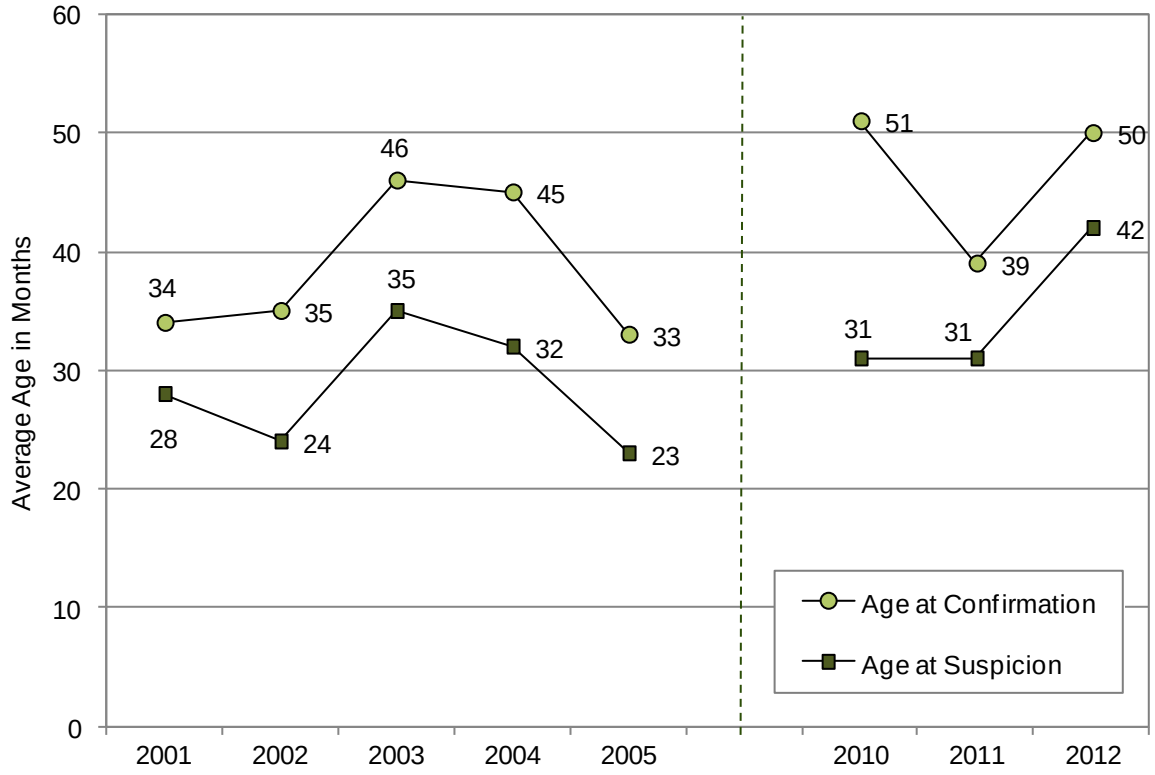
New Zealand Distribution by Ethnicity

In New Zealand during 2012, 103 children notified to the DND identified as European, 73 as Māori, 23 as Pacific, 12 as Asian/Indian and <3 as Middle Eastern/Latin American/African. As total response ethnicity was used, it was not possible to provide an overall breakdown of the proportions of children notified from each ethnic group.

Average Age at Suspicion and Confirmation of Hearing Loss

In New Zealand during 2012, when unilateral, acquired, mild, and overseas born cases were excluded (in order to ensure comparability with previous years) the average age at confirmation of a hearing loss was 50 months, although the average age of suspicion was much earlier (42 months) (**Figure 1**).

Figure 1. Average Age of Suspicion and Confirmation of Hearing Loss, New Zealand Deafness Notification Database 2001–2005 and 2010–2012

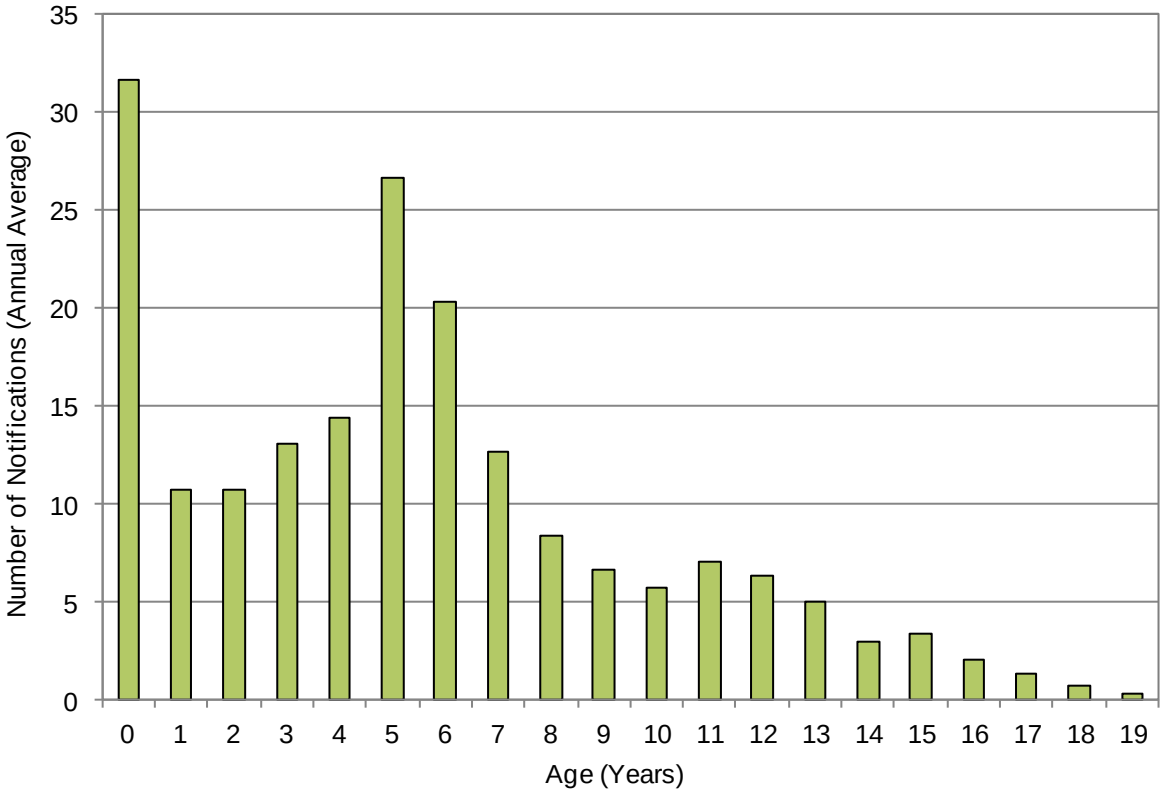


Source: Deafness Notification Database via Digby et al. [5]; Note: In order to ensure comparability with previous DND criteria, mild, acquired, unilateral, and overseas cases have been excluded

Number of Notifications by Age

During 2010–2012, the largest numbers of notifications to the DND were for babies under one year of age, likely as a result of newborn hearing screening. Numbers then dropped away during the preschool years. A second peak was evident at five years of age, likely as a result of the B4 School Check, with numbers then falling away again during mid-childhood. Note that this figure includes those with mild losses meeting DND criteria, those with acquired losses and those born overseas, all factors which may lead to a later age of diagnosis of permanent hearing loss (**Figure 2**). Further, in the 2012 DND Report [5] the peak in notifications in babies under one year increased during this period (2010 n=23; 2011 n=34; 2012 n=38) possibly as a result of the progressive roll out of newborn hearing screening [5].

Figure 2. Number of Notifications to the Deafness Notification Database by Age, New Zealand 2010–2012



Source: Deafness Notification Database via Digby et al. [5]

Distribution by Region

Table 2 reviews the number of notifications received by the Deafness Notification Database by region using its old criteria during 1998–2004, while **Table 3** reviews the number of notifications received by DHB using the new criteria during 2010–2012.

Table 2. Number of Notifications Meeting the Old Criteria for Inclusion in the Deafness Notification Database by Region of Residence, New Zealand 1998–2004

Region of Residence	Notification Year						
	1998	1999	2000	2001*	2002	2003	2004+
Northland	10	8	11	10	5	7	10
Auckland Region	21	35	40	74	36	52	37
Waikato	7	13	9	19	10	9	15
Lakeland	3	<3	0	3	3	3	6
Bay of Plenty	10	6	4	21	6	12	9
Tairawhiti	3	0	<3	3	<3	<3	5
Taranaki	<3	<3	<3	<3	3	3	8
Hawke's Bay	<3	<3	<3	31	5	4	5
Manawatu	3	3	0	12	7	12	24
Wellington	3	10	5	8	12	17	5
Nelson Marlborough	<3	3	<3	<3	3	4	4
West Coast	0	0	0	0	<3	<3	<3
Canterbury	0	3	7	10	12	9	10
South Canterbury	0	<3	<3	4	<3	3	3
Otago	0	<3	8	5	5	3	7
Southland	<3	3	<3	0	3	4	6
New Zealand Total	65	90	92	202	113	144	155

Source: National Audiology Centre [3]; Note: *2001 figures include 44 retrospective notifications; +During 2004 an additional 157 retrospective cases were added to the database, but are not included in this total

South Island DHBs Distribution

In the South Island during 2012, 10 Nelson Marlborough, 7 South Canterbury, 23 Canterbury and 23 Southern children were notified to the Deafness Notification Database (**Table 3**).

Table 3. Number of Notifications Meeting New Criteria for Deafness Notification Database by District Health Board, New Zealand 2010–2012

DHB	2010	2011	2012
Notifications to Deafness Notification Database			
Northland	12	5	15
Waitemata	4	9	6
Auckland	10	15	7
Counties Manukau	25	20	26
Waikato	15	13	15
Bay of Plenty	13	12	3
Lakes	<3	9	4
Tairawhiti	<3	<3	<3
Taranaki	6	5	8
Hawke's Bay	9	11	13
MidCentral	4	<3	6
Whanganui	0	<3	3
Hutt Valley	5	4	10
Capital & Coast	24	24	8
Wairarapa	0	5	<3
Nelson Marlborough	<3	4	10
South Canterbury	<3	3	7
Canterbury	44	27	23
West Coast	<3	<3	0
Southern	<3	15	23
New Zealand	180	187	191

Source: Deafness Notification Database via Digby et al. [5].

Newborn Hearing Screening

Background

In response to concerns regarding the late age of diagnosis of congenital hearing losses, in 2006 the Government announced funding for the development of a Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP). The UNHSEIP was implemented over the three year period 2007–2010, with a view to ensuring that [4]:

- Babies are screened for hearing loss by one month of age
- Audiology assessments are completed by three months of age
- Initiation of appropriate medical, audiological and early intervention education services occurs by six months of age.

The UNHSEIP is jointly overseen by the Ministry of Health, which is responsible for screening, audiological diagnosis and medical interventions; and the Ministry of Education, which is responsible for early intervention services. DHBs in turn, are the main providers of newborn hearing screening, follow-up audiology services and medical interventions. DHBs must offer newborn hearing screening to the family/whānau of all eligible babies in their region, whether they are born at hospital or home, using a framework of nationally consistent policies, standards and guidelines [4].

Screening is usually undertaken while the baby is asleep or quietly resting. Two types of screening are available:

- *Automated Otoacoustic Emissions (AOAE):* Sensory cells in the cochlea of the inner ear oscillate in response to an external sound. These oscillations generate an 'echo', which passes from the inner ear to the ear canal, and which can be detected as sound. These sounds, known as otoacoustic emissions (OAEs), are a sign that the ear is functioning normally and the measurement of OAEs can thus be used to test normal cochlear function in the newborn. Testing involves placing a small earphone and microphone in the ear, playing a sound and recording the response from the ear. If a baby has a normal functioning inner ear, an OAE is produced and this can be picked up by the microphone in the ear-canal [2].
- *Automated Auditory Brainstem Response (AABR):* The AABR is a series of electrical waves that can be recorded from electrodes on the scalp, in response to brief sounds being played into the ear. The presence of these waves with changing sound intensity is highly correlated with different hearing thresholds, with the AABR being used to assess the integrity of the ear and auditory nerve pathways to the brainstem in newborn babies [2].

The following section reviews data from the UNHSEIP's monitoring reports [4], with the most recent including data for the 6-month period from 1st October 2011–31st March 2012.

Data Sources and Methods

Indicators

1. Proportion of eligible newborns whose parents/guardians consent to newborn hearing screening

Numerator: Number of eligible newborns whose parents/guardians consent to newborn hearing screening

Denominator: Number of eligible live births

2. *The proportion of eligible newborns that complete the UNHS screening protocol by one month of age*

Numerator: Number of eligible newborns who complete newborn hearing screening by one month of age

Denominator: Number of eligible newborns who complete newborn hearing screening

3. Proportion of newborns who do not pass hearing screening and are referred to audiology

Numerator: Number of eligible newborns who complete screening with a referral for audiology assessment

Denominator: Number of eligible newborns who complete screening

4. Proportion of newborns that pass screening but have risk factors for developing late onset or progressive hearing loss

Numerator: Number of newborns that pass screening but have risk factors for developing late onset or progressive hearing loss (e.g. family history, craniofacial anomalies, jaundice, NICU >5 days, intrauterine infections, meningitis)

Denominator: Number of eligible newborns who passed screening.

5. *Proportion of newborn babies who completed audiology by 3 months of age*

Numerator: Number of newborns who completed audiology by 3 months of age

Denominator: Number of newborns who completed audiology

6. Number of newborns with a permanent congenital hearing loss

Number of newborns who had a permanent hearing loss confirmed by audiology AND where the aetiology was an auditory neuropathy, mixed or sensorineural in at least one ear AND where the baby was referred through the UNHSEIP

7. Number of newborns with a conductive hearing loss

Number of newborns who had hearing loss confirmed by audiology, where the aetiology was NOT an auditory neuropathy, mixed or sensorineural AND where the baby was referred through the UNHSEIP. The majority of babies were identified as having a temporary conductive hearing loss.

Data Source

Universal Newborn Hearing Screening and Early Intervention Programme Monitoring Reports, available for download at http://www.nsu.govt.nz/health-professionals/4627.aspx

Notes on Interpretation

Note 1: The majority of data in this section were derived from the UNHSEIP's monitoring report covering the six month period 1 October 2011 to 31st March 2012. However, trend data on the number of babies with permanent congenital hearing losses is drawn from UNHSEIP monitoring reports going as far back as 1st April 2010–30st Sept 2010. While all but one DHB (Southern) had implemented newborn hearing screening by the beginning of this earlier period, the outcome of audiology referrals may have been less complete for some DHBs due to the time taken for babies to progress through the referral pathways.

Note 2: The denominators for earlier UNHSEIP reports were derived from the Birth Registration Dataset and included live births for the relevant period. For the past 18 months however, birth data has been sourced from the National Maternity Database (which combines live birth registrations from Births Deaths and Marriages with hospital discharge data and Lead Maternity Carer claims). It is thought that this provides a much more complete data set due to the often long lag time for reporting birth registration data.

Note 3: In the newborn hearing screening table DHB refers to DHB of birth, whereas for the audiology tables, DHB refers to the DHB where the testing took place. Further audiology information is incomplete, as some DHBs did not submit information, or the information submitted was incomplete. In the 1st October 2011 to 31st March 2012 report, audiology information was available for only 254 of the 408 babies referred to audiology.

Newborn Hearing Screening

New Zealand Distribution

In New Zealand during 1st October 2011–31st March 2012, the caregivers of 88.6% of eligible babies consented to newborn hearing screening. Of those completing screening, 92.8% did so within one month, with 1.5% of those completing screening receiving an audiology referral. Of those babies who passed screening, a further 5.1% were deemed to have risk factors for delayed onset/progressive hearing loss (e.g. family history, craniofacial anomalies, and intrauterine infections) which warranted follow up over time (**Table 4**).

South Island DHBs Distribution

In the South Island DHBs during 1st October 2011–31st March 2012, the majority (range 71.6%–99.6%) of the caregivers of eligible babies consented to newborn hearing screening. Of those completing screening, over 90% (range 95.0%–99.3%) did so within one month, with 1.6% of Nelson Marlborough, 0.7% of West Coast, 1.4% of Canterbury, 0.7% of South Canterbury and 1.2% of Southern babies who completed screening receiving an audiology referral. Of those babies who passed screening, a small proportion (range 2.6%–5.4%) were deemed to have risk factors for delayed onset/progressive hearing loss which warranted follow up (**Table 4**).

Table 4. Newborn Hearing Screening Indicators by District Health Board, New Zealand 1st October 2011–31st March 2012

District Health Board	Number of Births in Period	Consenting to Screening (%)	Completed Screening ≤1 Month*(%)	Referrals to Audiology* (%)	Targeted for Follow Up* (%)
Newborn Hearing Screening					
Northland	1,172	77.1	67.8	4.5	9.3
Waitemata	3,987	84.5	90.0	1.1	4.2
Auckland	3,331	90.1	94.8	1.7	4.2
Counties Manukau	4,327	71.5	88.5	1.8	6.1
Waikato	2,708	94.0	94.8	1.3	5.7
Lakes	779	96.8	97.9	0.9	3.4
Bay of Plenty	1,417	89.6	92.6	1.7	2.1
Tairawhiti	368	88.6	97.8	1.2	8.2
Taranaki	792	92.8	99.3	2.4	10.3
Hawke's Bay	1,110	95.2	98.8	0.9	5.7
Whanganui	429	87.4	97.8	0.3	7.3
MidCentral	1,112	73.8	70.9	1.1	7.6
Hutt Valley	1,054	99.5	99.5	0.6	3.4
Capital & Coast	1,943	99.3	97.9	1.6	4.6
Wairarapa	280	96.1	90.3	1.5	7.5
Nelson Marlborough	759	99.6	95.1	1.6	5.4
West Coast	211	71.6	96.6	0.7	3.4
Canterbury	2,980	97.6	95.8	1.4	3.8
South Canterbury	289	95.8	99.3	0.7	2.6
Southern	1,781	97.1	95.0	1.2	5.2
New Zealand	30,829	88.6	92.8	1.5	5.1

Source: National Screening Unit 2012 [4]; Note: *See Methods for Indicator Definitions

Audiology Referrals and Outcomes

New Zealand Distribution

In New Zealand during 1st October 2011–31st March 2012, 254 babies commenced an audiology assessment, with 85.9% of those who completed their audiology assessment doing so by 3 months of age. During this period, 30 babies were identified as having a permanent congenital hearing loss, while 73 were identified as having a conductive hearing loss (**Table 5**).

South Island DHBs Distribution

In the South Island during 1st October 2011–31st March 2012, 11 Nelson Marlborough, 21 Canterbury, 2 South Canterbury and 15 Southern babies commenced an audiology assessment, with the majority (range 76.2%–100%) of those who completed their audiology assessment doing so by 3 months of age. During this period, 2 Nelson Marlborough, 2 Canterbury, 1 South Canterbury and 1 Southern baby were identified as having a permanent congenital hearing loss, with a small number also being identified as having a conductive hearing loss (**Table 5**). No information was available from the West Coast.

Table 5. Newborn Audiology Indicators by District Health Board, New Zealand 1st October 2011–31st March 2012

DHB	Commenced Audiology (Number)	Completed Audiology ≤3 months (%)	Permanent Congenital Hearing Loss (Number)	Conductive Hearing Loss (Number)	Permanent Congenital Hearing Loss (% of Completed)	Conductive Hearing Loss (% of Completed)
Newborn Audiology						
Northland	38	71.1	0	7	0	18.4
Waitemata						
Auckland	40	100.0	1	12	2.6	30.8
Counties Manukau	17	83.3	0	0	0	0
Waikato	30	76.7	9	13	30.0	43.3
Lakes	5	100.0	2	2	40.0	40.0
Bay of Plenty	13	92.3	3	0	23.1	0
Tairawhiti						
Taranaki	17	88.2	2	8	11.8	47.1
Hawke's Bay	8	87.5	2	1	25.0	12.5
Whanganui	1	0	0	0	0	0
MidCentral	10	100.0	0	3	0	30.0
Hutt Valley	12	100.0	2	8	16.7	66.7
Capital & Coast	14	92.9	3	3	21.4	21.4
Wairarapa			0			
Nelson Marlborough	11	90.9	2	3	18.2	27.3
West Coast						
Canterbury	21	76.2	2	7	9.5	33.3
South Canterbury	2	100.0	1	0	50.0	0
Southern	15	80.0	1	6	6.7	40.0
New Zealand	254	85.9	30	73	12.1	29.4

Source: National Screening Unit 2012 [4]; Note: See Methods for Indicator Definitions

Trends in the Identification of Permanent Congenital Hearing Losses

New Zealand Distribution

In New Zealand, a total of 81 babies were identified as having permanent congenital hearing losses in the UNHSEIP's six-monthly monitoring reports spanning the period 1st April 2010–31st March 2012 (**Table 6**).

South Island DHBs Distribution

In the South Island DHBs, 7 Nelson Marlborough, 8 Canterbury, 3 South Canterbury and 1 Southern baby were identified as having permanent congenital hearing losses in the UNHSEIP's six-monthly monitoring reports spanning the period 1st April 2010–31st March 2012 (**Table 6**). No information was available from the West Coast.

Table 6. Number of Babies Identified by Newborn Hearing Screening as Having Permanent Congenital Hearing Losses by District Health Board and Monitoring Period, New Zealand 1st April 2010–31st March 2012

DHB	6 Month Monitoring Period			
	1st April 2010–30th Sept 2010	1st October 2010–31st March 2011	1st April 2011–30th Sept 2011	1st October 2011–31st March 2012
Permanent Congenital Hearing Losses Identified by Newborn Screening				
Northland	0	0	2	0
Waitemata				
Auckland	1	2	3	1
Counties Manukau	1	0	0	0
Waikato	2	4	4	9
Lakes	0	1	0	2
Bay of Plenty	0	3	0	3
Tairawhiti	1			
Taranaki	0	1	1	2
Hawke's Bay			1	2
Whanganui				0
MidCentral	0	0	0	0
Hutt Valley	0	2	1	2
Capital & Coast	1	0	7	3
Wairarapa			0	0
Nelson Marlborough	1	2	2	2
West Coast				
Canterbury	4	1	1	2
South Canterbury	0	0	2	1
Southern*	0	0	0	1
New Zealand	11	16	24	30

Source: National Screening Unit [4]; Note: *Data for Southern DHB is only from August 2010 onwards

Local Policy Documents and Evidence-Based Reviews Relevant to the Identification and Management of Congenital Hearing Loss

In New Zealand there are a range of policy documents which consider newborn hearing screening. These are briefly summarised in **Table 7**, along with a number of other publications which consider congenital hearing losses in the overseas context.

Table 7. Local Policy Documents and Evidence-Based Reviews Relevant to the Early Detection and Management of Permanent Hearing Loss in Children

Ministry of Health Policy Documents
Ministry of Health. 2013. **Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP): National policy and quality standards.** Wellington: Ministry of Health. http://www.nsu.govt.nz/files/UNHSEIP-national_policy_and_quality_standards-jun13.pdf These National Policy and Quality Standards form part of the contract between the Ministry of Health and District Health Boards (DHBs) for the provision of services for the Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP). The standards are intended to increase knowledge about the programme, outline requirements of services, and assist DHBs to achieve high standards of practice.
Ali W, O'Connell R. 2007. **The effectiveness of early cochlear implantation for infants and young children with hearing loss**. NZHTA Technical Brief June 2007*,* 6(5). http://www.otago.ac.nz/christchurch/otago014007.pdf This Technical Brief produced by New Zealand Health Technology Assessment was commissioned by the New Zealand Ministry of Health. It compared the effectiveness of cochlear implantation at earlier and later ages. No eligible systematic reviews were found so 15 studies that were cross-sectional, case control or cohort studies were appraised. Implantation at less than 24 months of age was found to be more effective in terms of audiological performance, communication outcomes, educational achievement and quality of life than implantation at more than 24 months but it was unclear whether implantation at less than 12 months was more effective than implantation at more than 12 months.
International Guidelines
Muse C, et al. 2013. **Supplement to the JCIH 2007 position statement: principles and guidelines for early intervention after confirmation that a child is deaf or hard of hearing.** Pediatrics*,* 131(4), e1324-49. http://pediatrics.aappublications.org/content/131/4/e1324.full.pdf+html This best practice statement from the US, advocates for the implementation of coordinated state wide systems with the expertise to provide individualized, high-fidelity early intervention programmes for children who are deaf or hard of hearing and their families. It outlines comprehensive guidelines for early hearing detection and intervention programmes and establishing strong early intervention systems with appropriate expertise to meet the needs of children who are deaf or hard of hearing. Consistent monitoring of child and family outcomes is an essential step toward ensuring optimal outcomes. The establishment of practice standards, implementation of developmentally appropriate protocols for monitoring of outcomes, and commitment to research collaborations are critical steps toward this goal.
Ramsden JD, et al. 2012**. European Bilateral Pediatric Cochlear Implant Forum consensus statement.** Otol Neurotol*,* 33(4), 561-5. http://www.audiovestibologia.it/audiovestibologia/IMPIANTI_COCLEARI_files/2012%20EUROPEAN%20BILATERAL%20PEDIATRIC%20COCHLEAR%20IMPLANT%20FORUM%20CONSENSUS%20STATEMENT.pdf This is a consensus statement on paediatric cochlear implantation by the European Bilateral Pediatric Cochlear Implant Forum, as determined by a review of the current scientific literature. "Currently we feel that the infant or child with unambiguous cochlear implant candidacy should receive bilateral cochlear implants simultaneously as soon as possible after definitive diagnosis of deafness to permit optimal auditory development; an atraumatic surgical technique designed to preserve cochlear function, minimize cochlear damage, and allow easy, possibly repeated re-implantation is recommended."

National Institute for Health and Clinical Excellence (NICE). **Cochlear implants for children and adults with severe to profound deafness. London (UK): National Institute for Health and Clinical Excellence (NICE)**; 2009 Jan. 41p. (Technology appraisal guidance; no. 166). http://www.nice.org.uk/nicemedia/pdf/ta166guidancev2.pdf

These NICE guidelines evaluate the clinical effectiveness and cost-effectiveness of cochlear implants in England and Wales for children (12 months to 18 years) with severe to profound sensorineural hearing loss and for adults with severe to profound deafness. Thirty-three papers were included to evaluate clinical effectiveness, and 17 published studies were evaluated to assess cost-effectiveness. The major recommendations were:

- Unilateral cochlear implantation is recommended as an option for people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.
- If different cochlear implant systems are considered to be equally appropriate, the least costly should be used.
- Assessment of cost should take into account acquisition costs, long-term reliability and the support package offered.
- Simultaneous bilateral cochlear implantation is recommended as an option for the following groups of people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids:
  - Children
  - Adults who are blind or who have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness
- Acquisition of cochlear implant systems for bilateral implantation should be at the lowest cost and include currently available discounts on list prices equivalent to 40% or more for the second implant.

Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.

King AM. 2010. **The national protocol for paediatric amplification in Australia**. International Journal of Audiology, 49 Suppl.1, S64-9. http://informahealthcare.com/doi/abs/10.3109/14992020903329422?genre=article&id=doi%3A10.3109%2F14992020903329422

This is the Australian national protocol for amplification for hearing impaired children. It gives guidelines for selecting candidates for hearing aid fitting or referral to cochlear implant programmes and also covers management of children who have auditory neuropathy spectrum disorder and children who have mild and unilateral hearing loss. It describes the protocol for selection of hearing aids, hearing aid fitting and verification procedure and hearing-aid evaluation and also the criteria for supplying personal frequency modulated (FM) systems.

American Academy of Pediatrics, Joint Committee on Infant Hearing. 2007. **Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs**. Pediatrics, 120(4), 898-921. http://www.pediatrics.org/cgi/content/full/120/4/898

The position statement of the Joint Committee on Infant Hearing endorses screening of all newborns in order to ensure that infants with hearing loss can receive the earliest possible intervention with a view to maximising their opportunities to develop linguistic, literary, cognitive and social-emotional competence, and so that their educational and vocational attainment in adulthood can be as good as that of their hearing peers. It provides guidelines on screening protocols, evaluation of hearing impaired children detected by screening programmes, early intervention programmes, continued surveillance of infants and toddlers, protection of infant and family rights, information infrastructure, benchmarks and quality indicators. It also reports on current challenges, opportunities, and future directions in the field

Systematic and Other Reviews From the International Literature

Colgan S, et al. 2012. **The cost-effectiveness of universal newborn screening for bilateral permanent congenital hearing impairment: systematic review.** Acad Pediatr, 12(3), 171-80.

This systematic review considered the cost-effectiveness of universal newborn screening for bilateral permanent congenital hearing impairment. Twenty-two observational or modelled evaluations were identified of which only 2 clearly compared universal newborn hearing screening to risk factor screening for bilateral permanent congenital hearing impairment. Of these, the single evaluation that examined long-term costs and outcomes found that universal newborn hearing screening could be cost-saving if early intervention led to a substantial reduction in future treatment costs and productivity losses. There is a clear need for further research on long-term costs and outcomes to establish the cost-effectiveness of universal newborn hearing screening in relation to other approaches to screening, and to establish whether it is a good long term investment.

Janssen RM, et al. 2012. **Bilateral bone-anchored hearing aids for bilateral permanent conductive hearing loss: a systematic review.** Otolaryngol Head Neck Surg, 147(3), 412-22.

This systematic review evaluated the outcome of bilaternal versus unilaternal bone-anchored hearing aids (BAHA) for individuals with bilateral permanent conductive hearing loss in children and adults. Eleven observational studies were included. Bilaternal BAHA provided audiologic benefit and patient's subjective benefit compared to unilateral BAHA. The studies had small sample sizes and were limited in number.

Korver AM, et al. 2011. **Causes of permanent childhood hearing impairment**. Laryngoscope, 121(2), 409-16.

The causes of Permanent Childhood Hearing Impairment (PCHI) are often quoted as being hereditary in 50%, acquired in 25%, and unknown in 25% of cases. This population based study and systematic review investigated the evidence for the reported distribution of causes. In the study-population (n = 185) a hereditary cause was found in 38.9%, acquired cause in 29.7%, miscellaneous cause in 7.1%, and the cause was unknown in 24.3%. The systematic review (n = 9 articles) resulted in a weighted mean of 30.4% hereditary, 19.2% acquired, and 48.3% unknown causes of PCHI.

Cincinnati Children's Hospital Medical Center. Best evidence statement (BESt). **Effects of amplification on quality of life among school age children with single sided deafness.** Cincinnati (OH): Cincinnati Children's Hospital Medical Center; 2011. www.cincinnatichildrens.org/workarea/linkit.aspx?linkidentifier=id&itemid=88051&libid=87739

The objective of this evidence-based statement was to evaluate, among school age children with single sided deafness, if amplification bone conduction hearing aids versus no amplification improve quality of life. The authors concluded that amplification should be offered, noting that selected educational and family outcomes are important to monitor when amplification is used or if a decision is made not to provide amplification and that adequate information to families and the child are necessary for informed decision making regarding interventions.

Cincinnati Children's Hospital Medical Center. Best evidence statement (BESt). **Audiologic management for children with permanent unilateral sensorineural hearing loss**. Cincinnati (OH): Cincinnati Children's Hospital Medical Center; 2009 Aug 20 http://www.cincinnatichildrens.org/assets/0/78/1067/2709/2777/2793/9198/d385a2a5-e6d6-4181-a9df-f84ebd338c31.pdf

The objective of this statement was to evaluate whether amplification (i.e. digital hearing aid (HA), frequency modulation (FM) system, contralateral routing of signal CCROS) link aid etc.) compared to no amplification improves educational or functional performance in school-age children with severe to profound unilateral sensorineural hearing loss (USNHL) or mild to moderately severe USNHL.

In children with severe to profound USNHL it is recommended that school-aged children be fitted with an FM system as the first line of amplification technology, selecting an FM system with the most open fit to decrease occlusion in the good ear. It is recommended that provision of a HA in children with severe-profound UHL be on a case-by-case basis.

For children with mild to moderate sensorineural UHL it is recommended that children be fitted with a hearing aid (FM ready) as the first line intervention, and that provision of an FM system with or without a HA be discussed with the family. Note that the quality of evidence of UHL is moderate due to small numbers of studies and small sample sizes

Nelson HD, et al. 2008. **U.S. Preventive Services Task Force Evidence Syntheses, formerly Systematic Evidence Reviews**. Universal Newborn Hearing Screening: Systematic Review to Update the 2001 U.S. Preventive Services Task Force Recommendation. Rockville (MD): Agency for Healthcare Research and Quality (US).

This review was an update for the U.S. Preventive Services Task Force on universal newborn hearing screening (UNHS) to detect moderate to severe permanent, bilateral congenital hearing loss. Children with hearing loss who had UNHS had better language outcomes at school age than those not screened. Infants identified with hearing loss through universal screening had significantly earlier referral, diagnosis and treatment than those identified other ways.

Systematic and Other Reviews on Cochlear Implants

Eze N, et al. 2013. **Systematic review of cochlear implantation in children with developmental disability**. Otol Neurotol, 34(8), 1385-93.

This systematic review compared the outcome of cochlear implantation in children with developmental disability with children without developmental disability. Thirteen studies were included where expressive and/or receptive language outcomes were compared with children with cochlear implants and normal development. Seven studies demonstrated a worse outcome for children with developmental disability. Six articles showed no difference in the outcome between the 2 groups. Children with developmental disability may not benefit from cochlear implantation based on traditional assessment tools but appear to improve their environmental awareness and quality of life. More work is needed to define the term benefit when used in this context. Autistic children consistently had a negative outcome.

Incerti PV, et al. 2013. **A systematic review of electric-acoustic stimulation: device fitting ranges, outcomes, and clinical fitting practices**. Trends Amplif, 17(1), 3-26.

This systematic review evaluated the range of acoustic hearing in the implanted ear that can be effectively preserved for an electric-acoustic fitting; what benefits are provided by combining acoustic stimulation with electric stimulation; and what clinical fitting practices have been developed for devices that combine electric and acoustic stimulation. Twenty-seven articles were identified. The effectiveness of combined electric and acoustic stimulation as compared with electric stimulation alone was consistently demonstrated, highlighting the potential value of preservation and utilization of low frequency hearing in the implanted ear. However, clinical procedures for best fitting of electric-acoustic devices were varied. There is a need for further investigation of fitting procedures aimed at maximizing outcomes for recipients of electric-acoustic devices.

Turchetti G, et al. 2011. **Systematic Review of the Scientific Literature on the Economic Evaluation of Cochlear Implants in Paediatric Patients**. Acta Otorhinolaryngol Ital, 31(5), 311-8.

In this economic evaluation of cochlear implant (CI) in children, nine studies were included. Cost analysis, cost-effectiveness analysis and an analysis of educational costs associated with CI were performed. The direct cost ranged between € 39,507 and € 68,235 (2011 values). The studies related to cost-effectiveness analysis were not easily comparable: one study reported a cost per QALY ranging between $ 5197 and $ 9209; another referred a cost of $2154 for QALY if benefits were not discounted, and $16,546 if discounted. Educational costs are significant, and increase with the level of hearing loss and type of school attended. This review shows that the healthcare costs are high, but savings in terms of indirect and quality of life costs are also significant. Cochlear implantation in children is cost-effective.

Forli F, et al. 2011. **Systematic Review of the Literature on the Clinical Effectiveness of the Cochlear Implant Procedure in Paediatric Patients**. Acta Otorhinolaryngol Ital, 31(5), 281-98.

This systematic review summarized the results of scientific publications on the clinical effectiveness of the cochlear implant (CI) in children. Studies suggest that children implanted within the first year of life present hearing and communicative outcomes that are better than those of children implanted after 12 months of age. For children implanted after the first year of life, all studies confirm an advantage and many document an advantage in children who received cochlear implants under 18 months of age compared to those implanted at a later stage. Studies demonstrate that bilateral CI offers advantages in terms of hearing in noise, sound localization and during hearing in a silent environment compared to unilateral CI. There is, however, a wide range of variability. The studies also document the advantages after sequential bilateral CI. The studies also indicate that CI is also suitable for children with disabilities associated with deafness.

Vlastarakos PV, Candiloros D, et al. 2010. **Diagnostic challenges and safety considerations in cochlear implantation under the age of 12 months**. International Journal of Pediatric Otorhinolaryngology, 74(2), 127-32.

This review considers the current knowledge on cochlear implantation in children aged less than 12 months, regarding diagnostic, surgical and anaesthetic challenges. The studies reviewed included 3 meta-analyses, 4 prospective controlled studies, 25 prospective studies, 21 retrospective studies, 1 guideline, 8 review articles and 4 books. Based on a meta analysis of 125 infants the authors conclude that there is not an increased anaesthetic or surgical risk associated with infancy. Detection of other developmental issues which may affect the likelihood of developing normal speech and language is challenging but there are appropriate evaluation techniques for reliable assessment of the prelexical domains of infant development.

Johnston JC, Durieux-Smith A, Angus D, et al. 2009. **Bilateral paediatric cochlear implants: A critical review**. International Journal of Audiology, 48(9), 601-17.

This review evaluated the published research on bilateral cochlear implants. Because of the relative newness of this topic the authors searched for Randomized controlled trials (RCTs), prospective cohort studies, retrospective cohort studies, case studies and series, reviews, and qualitative studies. Of the 29 studies that met the inclusion criteria there were no RCTs, 4 reviews, 1 national survey of Cochlear Implant centres in the US, 15 cohort studies and 2 case control studies and 7 case series or case studies. All of the studies had relatively small numbers of participants (less than 50). It found that sound localisation and speech recognition in noise seem to be improved with bilateral cochlear implants compared to a unilateral implant and that the greatest benefits occur when the second implant is done early. It recommended further research into cost-effectiveness, quality of life, speech, language and psycho-educational measures.

Bond M, Mealing S, Anderson R, et al. 2009. **The effectiveness and cost-effectiveness of cochlear implants for severe to profound deafness in children and adults: A systematic review and economic model**. Health Technology Assessment, 13(44), 1-96.

This review investigated whether it is clinically effective and cost-effective to provide a unilateral cochlear implant for severely to profoundly deaf people (who do or do not use hearing aids), and whether it is clinically effective and cost-effective to provide bilateral cochlear implants for severely to profoundly deaf people who have a single cochlear implant (who do or do not use a hearing aid as well). This study reported on a systematic review of the literature which found 33 suitable papers of which only 2 were randomised controlled trials.

All of the studies reviewed found that for children there were gains on all outcome measures when comparing one cochlear implant with non-technological support or an acoustic hearing aid. Earlier implantation in children produced the greatest benefits. From the Markov model base-case analysis the authors estimated that, for prelingually profoundly deaf children, the incremental cost-effectiveness ratio (ICER) for unilateral implantation compared with no implantation was £13,413 per quality-adjusted life-year (QALY).The best evidence for the benefits of bilateral cochlear implants was in understanding speech in noisy conditions. The authors conclude that unilateral cochlear implantation for children and adults is cost effective but state “decisions on the cost-effectiveness of bilateral cochlear implants should take into account the high degree of uncertainty within the model regarding the probable utility gain.”

Bond M, et al. 2009. **Effectiveness of Multi-channel Unilateral Cochlear Implants for Profoundly Deaf Children: a Systematic Review**. Clin Otolaryngol, 34(3), 199-211.

This systematic review evaluated the strength of evidence when comparing the effectiveness of unilateral cochlear implants with non-technological support or acoustic hearing aids in children with permanent bilateral hearing loss (PBHL) in the UK. Fifteen studies were identified. They were of moderate to poor quality and heterogeneity in design and outcomes precluded meta-analysis. However, all studies reported that unilateral cochlear implants improved scores on all outcome measures. Additionally five economic evaluations found unilateral cochlear implants to be cost-effective for profoundly deaf children at UK implant centres.

Papsin BC, Gordon KA. 2008**. Bilateral cochlear implants should be the standard for children with bilateral sensorineural deafness**. Current Opinion in Otolaryngology & Head & Neck Surgery, 16(1), 69-74.

This review looks at the literature on bilateral cochlear implantation in children and recommends simultaneous bilateral implantation when possible and if not then the shortest possible interval between implantation of the first and second ears. It recommends further research to determine the interval after which bilateral cochlear implantation provides so little benefit that it is not cost-effective.

Other Relevant Publications on Cochlear Implants
Bird P, Botting A, Milburn J, et al. 2010. **An audit of referrals to the Southern Cochlear Implant Paediatric Programme**. New Zealand Medical Journal, 123(1313), 10-4. http://journal.nzma.org.nz/journal/123-1313/4077/content.pdf This paper reports on a review of 75 paediatric referrals the Southern Cochlear Implant Programme from March 2003-March 2008 (before the introduction of the newborn hearing screening programme). The mean age at referral was 17 months with a range of 1 to 203 months. The authors state that the age of referral has been unacceptably high and that children with known risk factors for significant sensorineural hearing loss have not been receiving early diagnosis.
Bird PA, Murray D. 2008. **Cochlear Implantation: a panacea for severe hearing loss?** Journal of the New Zealand Medical Association, 121(1280). http://journal.nzma.org.nz/journal/121-1280/3220/ This article provides a local perspective on the issue and points out that there is an increasing body of evidence on the benefits of bilateral implants in children. (Currently only one implant per child is normally publicly funded.)
Battmer RD, O'Donoghue GM, Lenarz T. 2007. **A multicenter study of device failure in European cochlear implant centers**. Ear & Hearing, 28(2 Suppl), 95S-99S. This study reports on cochlear implant failure in 27 European centres and notes that while overall cochlear implant systems are satisfactory there is considerable variation in the reliability of different systems. A common industry independent failure database using uniform reporting protocols would be beneficial to users and clinicians.
Documents Relating to the Universal Newborn Hearing Screening Programme in New Zealand
Ministry of Health. 2012. **Quality improvement review of a screening event in the Universal Newborn Hearing Screening and Early Intervention Programme.** Wellington: Ministry of Health. http://www.health.govt.nz/publication/quality-improvement-review-screening-event-universal-newborn-hearing-screening-and-early This review was undertaken by the National Screening Unit (NSU) with the input of an Incident Review Group of the Universal Newborn Hearing Screening and Early Intervention Programme following issues identified between July and November 2012 where babies were not screened correctly for permanent congenital hearing loss. The review outlines the findings and recommendations to improve DHB service provision and strengthen the leadership and surveillance of the programme by the NSU.
Universal Newborn Hearing Screening and Early Intervention Programme **Newborn Hearing Screening and Audiology Workforce Strategy and Action Plan June 2008** http://www.nsu.govt.nz/files/Universal_Newborn_Hearing_Screening_and_Early_Intervention_Programme.pdf This workforce development strategy and action plan was to address the development of the newborn hearing screening and audiology workforce required for implementing the UNHSEIP. The strategy outlines initiatives that would need to be implemented by the National Screening Unit, DHBs, professional bodies and educational institutions in a collaborative manner.
Universal Newborn Hearing Screening Advisory Group. 2005. **Universal Newborn Hearing Screening for New Zealand 2005: A Report of the Universal Newborn Hearing Screening Advisory Group to the National Screening Unit**. Wellington: Ministry of Health. http://www.moh.govt.nz/moh.nsf/0/D71ADADE4D79E24ECC2571210075DD7B/$File/universalnewbornfeb06.pdf This report contains the findings and recommendations of the Universal Newborn Hearing Screening Advisory Group to the National Screening Unit regarding high-level policy and implementation issues for a (then) future universal newborn hearing screening programme for New Zealand. It contains background information on congenital hearing loss, New Zealand statistics and summarises the benefits of lowering the average age of detection of hearing loss. It also addresses issues relevant to intervention services and the design and operation of screening services.
Project HIEDI (Hearing Impairment – Early Detection and Intervention). 2004. **Improving outcomes for children with permanent congenital hearing impairment. The case for a national newborn hearing screening and early intervention programme for New Zealand**. Auckland: Project HIEDI. http://www.nfd.org.nz/site_resources/library/OrganisationFiles/HIEDI_Evidence_Based_Case.pdf This very comprehensive report (with 435 references) includes information on hearing loss in general, the effects of permanent congenital hearing loss, New Zealand data, and issues relating to universal newborn hearing screening and early intervention programmes and international experience with them. The authors state “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.”

Other Relevant Websites
https://www.health.govt.nz/your-health/conditions-and-treatments/disabilities/hearing-loss Ministry of Health webpage related to hearing loss with information on symptoms, hearing checks for children and hearing services available in New Zealand. The webpage includes a section on cochlear implants
The National Foundation for the Deaf Incorporated http://www.nfd.org.nz/ The National Foundation for the Deaf Incorporated is a foundation with nine member organisations set up to promote the interests, advancement, independence and wellbeing of deaf and hearing impaired New Zealanders. The foundation also raises awareness of the health, social, educational, economic, environmental and cultural barriers encountered by deaf and hearing impaired people and their families. They facilitate communication and coordination among professional and community organisations working with deaf and hearing impaired people.
New Zealand Deafness Notification Database http://www.audiology.org.nz/deafness-notification-database.aspx The Deafness Notification Database (DND) was in operation from 1982-2005 to collect and report on the number and nature of new cases of hearing loss diagnosed among children and young people born in New Zealand. It was funded by the Ministry of Health. From 2006-2009 the database was not in operation but it was re-launched in 2010 funded by the New Zealand Audiological Society. Eligibility now includes those children born overseas and those with unilateral hearing losses. Reports are now available for 2010, 2011 and
The New Zealand Audiological Society http://www.audiology.org.nz The New Zealand Audiological Society is a self-governing body representing more than 300 audiologists that was incorporated in 1976. Their aim is to assist the hearing impaired community of all ages to participate as fully as possible is all aspects of life. The website has information for the public on hearing loss, screening, and hearing aids. There is also information for audiologists including standards of practice, events and other links.
Hearing Association New Zealand Te Kāhui Rongo o Aotearoa http://www.hearing.org.nz The Hearing Association New Zealand sets out to advance the interests and general welfare of all persons with a hearing loss, their family, whānau and all who support them, to improve their quality of life. This national body supports 32 separate associations which are dedicated to raising the profile of hearing issues, services and information. There are branches throughout New Zealand. Information on hearing screening, hearing aids and other products, various classes and other information sheets are available on the website.

Note: The publications listed were identified using the search methodology outlined in Appendix 1 (Search Methods for Policy Documents and Evidence-Based Reviews)

References

1. Universal Newborn Hearing Screening Advisory Group. 2005. Universal Newborn Hearing Screening for New Zealand 2005: A Report of the Universal Newborn Hearing Screening Advisory Group to the National Screening Unit. Wellington: Ministry of Health http://www.moh.govt.nz/moh.nsf/0/D71ADADE4D79E24ECC2571210075DD7B/$File/universalnewbornfeb06.pdf
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3. National Audiology Centre. 2004. New Zealand Deafness Notification Data 2004. Auckland: National Audiology Centre
4. National Screening Unit. 2012. Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP): Monitoring Report on Newborn Hearing Screening Service Provision October 2011-March 2012. Auckland: Ministry of Health http://www.nsu.govt.nz/health-professionals/4627.aspx
5. Digby J, Purdy S, Kelly A. 2013. Deafness Notification Report (2012) Hearing Loss (not remediable by grommets) in New Zealanders under the age of 19. Auckland: Accessable http://www.audiology.org.nz/deafness-notification-database.aspx