PERMANENT HEARING LOSS

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 [198]. In response to concerns regarding the late age of diagnosis of congenital hearing losses (average age 35.1 months when screening was based on the presence of risk factors [199]), the Government in its 2006 Budget, announced a funding package ($16 million over four years) to establish a Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP). The Programme has been rolled out over a three year period (2007–2010) [200] and now that the Programme is fully implemented, the Ministry of Health has recently begun producing Programme monitoring reports. These reports describe the proportion of babies undergoing newborn hearing screening in each DHB, as well as the proportion referred for further audiological investigation [201].

The following section reviews the most recent data available on newborn hearing screening in New Zealand. Because at the time of writing, time series information on the number of babies diagnosed with permanent hearing loss is not available from UNHSEIP data, the section begins by briefly reviewing some historical and contemporary data from the Deafness Notification Database, the only other national source of information on the number of children and young people with permanent hearing loss in New Zealand.

Deafness Notification Database

Background

The main purpose of the Deafness Notification Database (DND), which was funded by the Ministry of Health between 1982 and 2005, was to collect and report on new cases of hearing loss diagnosed in New Zealand-born children and young people. The database was not operational during 2006–2009, but in 2010 it was re-launched by the NZ Audiological Society. Although a number of changes have been made to the way in which the data are collected and reported, the Society has tried to maintain as much continuity as possible between the two periods [202].

Old and New Notification Criteria

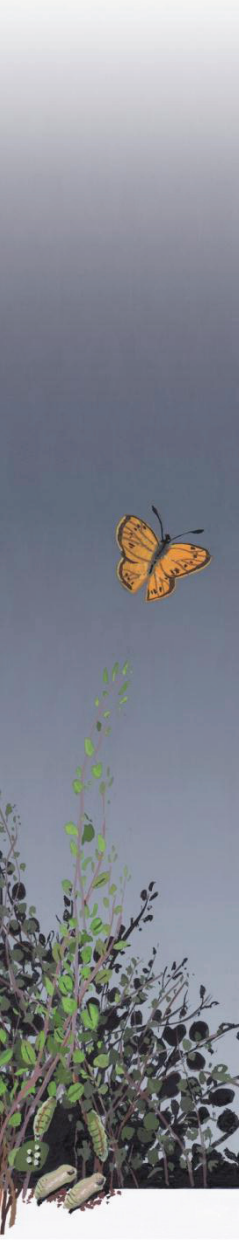
During 1982–2005, when the DND was managed by the National Audiology Centre, it collected information on children meeting the following criteria [199]:

1. Children needed to be <18 years of age, with congenital hearing losses or any hearing loss not remediable by medical or surgical means which required hearing aids and/or surgical intervention.
2. Children needed to have an average bilateral hearing loss (over 4 audiometric frequencies 500–4000 Hz) of >26 dBHL in the better ear.
3. 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 hearing losses via a new online form. Following consultation, the database was extended to also include [202]:

1. Children with an average hearing loss (over 4 audiometric frequencies 500–4000 Hz) of >26 dBHL in ONE ear (i.e. unilateral losses).
2. Children who were born outside of New Zealand [202].

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. This led to 180 notifications meeting the new criteria in 2010. Indications from previous DND data suggest that the



2010 dataset was likely to have captured 50–80% of new cases of hearing loss diagnosed in New Zealand children aged 0–17 years during 2010 [202].

Data Sources and Methods

Indicator

1. *Notifications to the New Zealand Deafness Notification Database*

Numerator: Children Aged 0–17 Years notified to the Deafness Notification Database who met the Database's notification criteria (see text above for criteria used).

All of the data in this section was derived from the National Audiology Centre's Annual Deafness Notification Database Reports 1998–2004 [199] or from the Deafness Notification Report 2010 produced by Digby et al for the NZ Audiological Society [202].

Notes on Interpretation

Note 1: The hearing loss severity scale used by the DND during 1996–2005, and the likely clinical implications of such hearing losses are briefly outlined below.

**Mild Loss (26–40 dBHL):* This may result in some difficulties in hearing soft speech and conversations (persons sound as if mumbling) but children can often manage in quiet situations with clear voices. Speech and language usually develop normally if the child is fitted early with hearing aids [203].

* *Moderate Loss (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 [203].

* *Severe Loss (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 [203].

* *Profound (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, with benefits being evident, especially if implanted at a young age [203].

Note 2: DND data are presented by year of notification, rather than year at first identification, with the degree of hearing loss assessed using the dBHL ranges outlined 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.

Note 3: Tests of statistical significance have not been applied to the data in this section, and thus the associations described do not imply statistical significance or non-significance (see **Appendix 2** for further discussion of this issue).

New Zealand Distribution of Hearing Loss by Type

In New Zealand during 2010, a total of 120 notifications were received by the DND for children with bilateral hearing losses of >26dB in the better ear. In addition, 60 notifications were received for children with unilateral losses, which previously would not have met DND criteria. While a number of notifications were also received for children with slight losses, these were not included in the analysis of DND data (**Table 134**).

Table 134. Deafness Notification Database Notifications by Type of Hearing Loss, New Zealand 2000−2005 and 2010

Type of Hearing Loss	Original Database						Re-Launched Database
	2000	2001	2002	2003	2004*	2005	2010
Bilateral Loss – Better Ear >26dB	**92**	**202**	**113**	**144**	**155**	**93**	120
Unilateral Loss	14	54	38	51	68	51	60

Source: Deafness Notification Database via Digby et al [202]; Note: 2001 figures include 44 retrospective notifications. In addition, as the result of an audit, 288 retrospective notifications were made in 2004, with 157 meeting the new criteria. They are not included here as they cannot be attributed to any one year; Numbers in bold indicate totals previously reported using old DND criteria.

New Zealand Distribution by Severity of Loss

In New Zealand during 2010, 15% of notifications to the DND were for children with profound losses. A further 6% were for children with severe losses, 37% were for children with moderate losses and 42% were for children with mild losses (**Table 135**).

Table 135. Notifications to Deafness Notification Database by Degree of Hearing Loss Using Old Criteria, New Zealand 2001−2004 and 2010

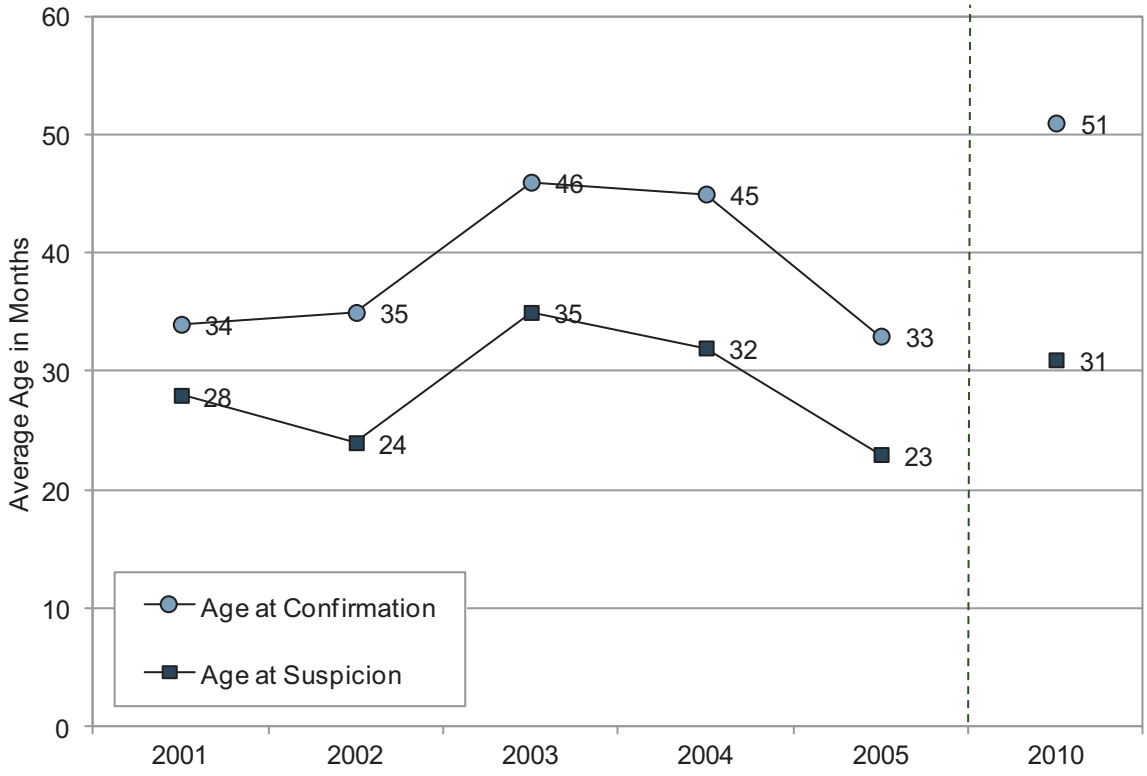
Degree of Hearing Loss	Proportion of Cases Notified (%)				
	2001	2002	2003	2004	2010
Mild	47	47	56	43	42
Moderate	35	39	33	34	37
Severe	10	9	6	15	6
Profound	8	5	5	7	15
Total	100	100	100	100	100

Source: Deafness Notification Database via Digby et al [202]

Average Age at Suspicion and Confirmation of Hearing Loss

In New Zealand during 2010, when unilateral, acquired and overseas born cases were excluded (in order to ensure comparability with previous years) the average age at confirmation of a hearing loss was 51 months, although the average age of suspicion was much earlier (31 months) (**Figure 130**). It is unclear however the extent to which changes in hearing screening (both newborn and at 4–5 years) and year to year differences in the completeness of the DND contributed to the variations in age of confirmation seen during this period.

Figure 130. Average Age of Suspicion and Confirmation of Hearing Losses, New Zealand Deafness Notification Database 2001−2005 and 2010



Source: Deafness Notification Database via Digby et al [202]. Note: In order to ensure comparability, 2010 cases were only included if they met earlier DND criteria (i.e. acquired, unilateral and overseas cases excluded)

New Zealand Distribution by Ethnicity

In New Zealand during 2010, 101 children notified to the DND identified as European, 59 as Māori, 18 as Pacific, 13 as Asian/Indian and 5 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 [202].

Distribution by Region

Table 136 reviews the number of notifications received by the Deafness Notification Database by region using its old criteria during 1998–2004, while **Table 137** reviews the number of notifications received by DHB using the new criteria during 2010.

In the South Island DHBs during 2010, a total of 49 children were notified to the Deafness Notification Database (**Table 137**).

Table 136. Number of Notifications Meeting the Old Criteria for Inclusion in 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 [199]; Note: 2001 figures include 44 retrospective notifications; During 2004 an additional 157 retrospective cases which had not been notified over an 11-year period were added to the database, but are not included in this total.

Deafness Notification Database Summary

In New Zealand during 2010, 120 notifications were received by the Deafness Notification Database for children with bilateral hearing losses of >26dB in the better ear and 60 notifications were received for children with unilateral losses. During 2010, 15% of children notified to the DND had profound losses, 6% had severe losses, 37% had moderate losses and 42% had mild losses. When unilateral, acquired and overseas born cases were excluded, the average age at confirmation of a hearing loss in 2010 was 51 months, although the average age of suspicion was much earlier (31 months).

In the South Island DHBs during 2010, a total of 49 children were notified to the Deafness Notification Database.

Table 137. Number of Notifications Meeting New Criteria for Deafness Notification Database by District Health Board, New Zealand 2010

DHB	Number of Notifications	Percent of Notifications
Northland	12	6.7
Waitemata	4	2.2
Auckland	10	5.6
Counties Manukau	25	13.9
Waikato	15	8.3
Bay of Plenty	13	7.2
Lakes	<3	1.1
Hawke's Bay	9	5
Tairawhiti	<3	1.1
Taranaki	6	3.3
MidCentral	4	2.2
Whanganui	0	0
Wairarapa	0	0
Hutt	5	2.8
Capital and Coast	24	13.3
Nelson Marlborough	<3	0.6
Canterbury	44	24.4
South Canterbury	<3	0.6
West Coast	<3	1.1
Southern	<3	0.6
New Zealand	180	100

Source: Deafness Notification Database via Digby et al [202]

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 goals of the Programme were to ensure that [201]:

- Babies were screened for hearing loss by 1 month of age
- Audiology assessments were completed by 3 months of age
- Initiation of appropriate medical, audiological and early intervention services occurred by 6 months of age.

As a result, for babies born in hospital, screening is now offered in most cases before the baby goes home, with those born elsewhere, or not managing to be screened prior to discharge, being able to access screening on an outpatient basis.

Screening is usually undertaken while the baby is asleep or quietly resting, with two types of screening being available:

- *Automated Otoacoustic Emissions (AOAE):* Sensory cells in the cochlea of the inner ear oscillate in response to an external sound, with these oscillations generating 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 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 [203].

- *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 [203].

During 2007–2010, the UNHSEIP was rolled out progressively across all DHBs, with the Ministry of Health being responsible for screening, the audiological diagnosis of hearing loss and medical interventions, and the Ministry of Education being responsible for Early Intervention Services [204]. The following section presents data from the UNHSEIP's most recent monitoring report, which reviews newborn hearing screening in participating DHBs for the period 1st April–30th September 2011.

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.

Notes on Interpretation

Note 1: All of the data in this section are derived from the UNHSEIP's second monitoring report [205], which covers the six month period from 1 April 2010 to 30 September 2010. All but one DHB (Southern) had implemented newborn hearing screening by the beginning of this period, and by the end of the period all DHBs were offering screening. Thus while reporting includes data from all 20 DHBs, data for Southern DHB is only from August 2010 onwards.

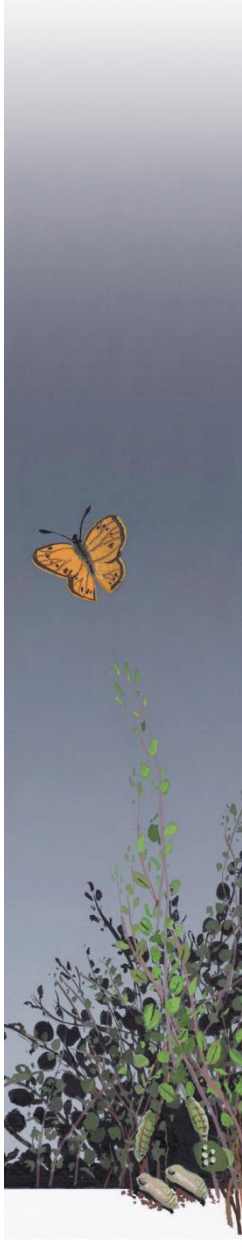
Note 2: All denominators in UNHSEIP reports are derived from the Birth Registration Dataset and include live births for the relevant period.

Note 3: Tests of statistical significance have not been applied to the data in this section, and thus the associations described do not imply statistical significance or non-significance (see **Appendix 2** for further discussion of this issue).

Distribution by DHB

In New Zealand during 1 April 2010–30 September 2010, the caregivers of 77.8% of eligible babies consented to newborn hearing screening, although this proportion varied considerably by DHB. Of those completing screening 94.0% did so within one month, with on average 2.4% of babies completing screening receiving an audiology referral. Of those babies who passed screening, a further 7.4% were deemed to have risk factors for delayed onset/progressive hearing loss (e.g. family history, craniofacial anomalies, intrauterine infections) which warranted follow up over time (**Table 138**).

In the South Island DHBs (excluding Southern DHB where hearing screening commenced part way through the period), newborn hearing screening consent rates ranged from 60.8% to 98.7%, with the proportion of babies being referred for audiology assessment ranging



from 0% to 2.1% and the proportion being targeted for follow up ranging from 4.4% to 10.4% (**Table 138**).

Table 138. Newborn Hearing Screening Indicators by District Health Board, New Zealand 1 April 2010 to 30 September 2010

DHB	Number of Births in Period	Consenting to Screening (%)	Completed Screening ≤1 Month*(%)	Referrals to Audiology* (%)	Targeted for Follow Up* (%)
Newborn Hearing Screening					
Northland	1,157	46.4	66.0	4.6	14.6
Waitemata	3,862	48.8	93.5	1.2	6.3
Auckland	3,179	96.9	94.4	4.4	4.7
Counties Manukau	4,231	49.3	97.6	6.3	8.7
Waikato	2,675	101.0	95.7	1.7	7.1
Lakes	738	109.6	95.1	1.9	6.9
Bay of Plenty	1,394	99.0	95.9	0.9	5.4
Tairawhiti	341	105.3	97.7	0.9	6.3
Taranaki	764	100.3	88.4	2.3	11.1
Hawke's Bay	1,058	106.0	98.2	1.9	8.8
Whanganui	424	96.0	87.8	2.2	9.6
Mid Central	1,168	54.1	83.1	2.2	12.3
Hutt Valley	1,038	105.6	99.5	1.1	13.8
Capital & Coast	1,930	96.3	98.4	0.5	7.7
Wairarapa	247	78.5	97.4	0.5	7.3
Nelson Marlborough	844	94.8	90.9	1.0	8.6
West Coast	209	60.8	95.2	<0.1	10.4
Canterbury	3,231	94.9	93.5	2.1	4.4
South Canterbury	318	98.7	98.7	1.3	4.6
Southern*	1,753	38.0	85.9	2.8	8.3
New Zealand	30,694	77.8	94.0	2.4	7.4

Source: National Screening Unit 2011 [205]. Note: Data for Southern DHB is from August 2010 onwards; Consent rates in excess of 100% may arise as live birth denominators are based on DHB of domicile, while screening in a small number of cases may be carried out in a different DHB (see Monitoring Report [205] for further detail); See Methods for Indicator Definitions.

Distribution by Ethnicity, NZDep Index Decile and Birth Location

In New Zealand during 1 April 2010–30 September 2010, there were no marked differences by ethnicity or NZDep decile in the proportion of babies who completed screening within one month, although those who were born at home had lower completion rates than those born elsewhere. While there were some variations in audiology referrals and those targeted for follow up by ethnicity, NZDep decile and birth location, the significance of these differences remains unclear, as no assessments of statistical significance were available for this data (**Table 139**).

Outcome of Audiology Referrals

Of babies referred to audiology during April–September 2010, 40.5% started audiology assessment, although this varied by DHB (range 0% to 75%). This proportion should be interpreted with caution however, as some DHBs did not submit audiology forms to the NSU and there were delays in entering some data into the national database due to missing information. Of 563 babies who did not pass screening and were referred to audiology, audiology information was recorded in the national database for just 228 [205].

Of those babies with information in the national database, all that started audiology assessment completed the assessment, with 64% of those completing doing so by three months of age. Eleven babies (4.8% of those completing assessment) had a permanent/congenital hearing loss identified, with only one being a Neonatal Intensive Care (NICU)/Special Care baby Unit (SCBU) baby. A higher proportion were identified with a conductive or mixed hearing loss (24.1% of those who completed assessment). In terms of the age at which hearing loss was identified, in 9 cases this was by 4 weeks, in 13 cases by 8 weeks, in 16 cases by 12 weeks and the remaining 27 cases by over 12 weeks [205].

Table 139. Newborn Hearing Screening Indicators by Ethnicity, NZ Deprivation Index Decile and Birth Location, New Zealand 1 April 2010 to 30 September 2010

Variable	Completed Screening ≤1 Month* (%)	Referrals to Audiology* (%)	Targeted for Follow Up* (%)
Newborn Hearing Screening			
Ethnicity			
Māori	92.8	2.8	9.2
Pacific	95.5	4.4	6.6
Asian/Indian	96.2	2.7	4.4
European	93.8	1.7	7.3
Other	95.7	2.9	6.3
NZ Deprivation Index Decile			
Decile 1–2	95.2	1.7	7.1
Decile 3–4	93.7	1.9	6.3
Decile 5–6	94.0	1.9	6.6
Decile 7–8	93.4	2.4	7.9
Decile 9–10	94.2	3.3	8.2
Birth Location			
Public Hospital	94.4	2.4	7.4
Private Hospital	95.6	1.1	3.3
Home	74.0	2.7	8.1
Other Location	88.2	<0.1	17.6
New Zealand Total	94.0	2.4	7.4

Source: National Screening Unit 2011 [205]. Note: See Methods for indicator definitions

Newborn Hearing Screening Summary

In New Zealand during 1 April 2010–30 September 2010, the caregivers of 77.8% of eligible babies consented to newborn hearing screening, although this proportion varied considerably by DHB. Of those completing screening 94.0% did so within one month, with on average 2.4% of babies completing screening receiving an audiology referral. Of those babies who passed screening, a further 7.4% were deemed to have risk factors for delayed onset/progressive hearing loss which warranted follow up over time.

In the South Island DHBs (excluding Southern DHB where hearing screening commenced part way through the period), newborn hearing screening consent rates ranged from 60.8% to 98.7%, with the proportion of babies being referred for audiology assessment ranging from 0% to 2.1% and the proportion being targeted for follow up ranging from 4.4% to 10.4%.

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

In New Zealand a range of policy documents focus on the early identification of permanent hearing loss, and these are briefly summarised in **Table 140**, along with a number of evidence-based and other reviews which consider the identification or management of permanent hearing loss in the child and youth population.

Table 140. Policy Documents and Evidence-Based Reviews Relevant to the Early Detection and Management of Permanent Hearing Loss in Children and Young People

Ministry of Health Policy Documents
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 and 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.
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://nzhta.chmeds.ac.nz/index.htm This Technical Brief produced by New Zealand Health Technology Assessment (NZHTA) was commissioned by the Ministry of Health. It aimed to compare 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.
Project HIEDI. 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/?t=56 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.”
Systematic and Other Reviews From the International Literature
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 so that infants with hearing loss can receive the earliest possible intervention to maximise their opportunities to develop linguistic, literary, cognitive and social-emotional competence, 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 and reports on current challenges, opportunities, and future directions in the field.

McKay S, Gravel JS, Tharpe AM. 2008. **Amplification considerations for children with minimal or mild bilateral hearing loss and unilateral hearing loss**. Trends in Amplification, 12(1), 43-54.

This review suggests there is limited evidence on which to base the decision as to whether or not to provide hearing technology for children who have minimal or mild bilateral hearing loss or unilateral hearing loss. These children have a greater risk for academic, speech-language, and social-emotional difficulties than their normal hearing peers but it is unknown how to identify those at greatest risk for these difficulties and whether the provision of early amplification assistance will help prevent them. The current hearing technology options for these children are also reviewed.

Centre for Allied Health Evidence review team. 2007. **A Systematic Review of the Literature on Early Intervention for Children with a Permanent Hearing Loss Volumes I and 2**. Brisbane: Queensland Health. http://www.health.qld.gov.au/healthyhearing/pages/publications.asp

The Centre for Allied Health Evidence was commissioned by Queensland Health to identify variables associated with successful early intervention for children aged 0–3 years with permanent hearing loss. The review found good evidence for the influence of age at detection of hearing loss, age at implantation (whether of hearing aids or cochlear implants), age at onset of hearing loss, and duration of hearing loss on outcomes. It states that whether children use hearing aids or cochlear implants they should be adequately supported through high quality, intensive programs to develop communication skills. It found a lack of high level, high quality research investigating the effectiveness of one particular communication approach over another. It identifies significant areas of ongoing research.

Corabian P, Eng K, Lier D, et al. 2007. **Screening Newborns for Hearing**. Edmonton: Institute of Health Economics. http://www.ihe.ca/publications/library/2007/screening-newborns-for-hearing/

This Canadian review considered three aspects of Universal Newborn Hearing Screening (UNHS) using Automatic Otoacoustic Emissions and/or Automated Auditory Brainstem Response: the social considerations, the published evidence of efficacy/effectiveness and safety, and cost effectiveness (via a review of the economic literature).The authors concluded that such screening is effective in detecting moderate to profound permanent congenital hearing loss but that there is as yet limited evidence for the safety and clinical efficacy of UNHS from well designed clinical trials or that early detection leads to more effective habilitation.

Puig T, Municio A, Meda C. 2005. **Universal neonatal hearing screening versus selective screening as part of the management of childhood deafness**. Cochrane Database of Systematic Reviews, 2005(2), Art. No.: CD003731. DOI:10.1002/14651858.CD003731.pub3.

This review compares the effectiveness of a universal neonatal screening and early intervention program with two alternatives: screening and treatment of high risk neonates only and opportunistic screening and treatment. The authors found no randomised controlled trials comparing universal newborn hearing screening with either of the other two options and concluded that the effectiveness of universal newborn hearing screening programs had not yet been established. This Review was withdrawn from Issue 1, 2010 of The Cochrane Library onwards because the review authors were unable to continue updating.

Other Relevant Publications

Grewal S, Merchant T, Reymond R, et al. 2010. **Auditory late effects of childhood cancer therapy: A report from the Children's Oncology Group**. Pediatrics, 125(4), e938-50.

Some forms of therapy for childhood cancer can cause hearing loss, particularly platinum compounds (used to treat neuroblastoma, hepatoblastoma, osteosarcoma, and germ-cell tumours) and radiation (for head and neck tumours).This report from the Auditory/Hearing Late Effects Task Force of the Children's Oncology Group had four aims: to review ototoxicity from childhood cancer therapy, to describe the cochlear pathophysiology and genetics of cisplatin-related hearing loss, to explain the impact of chemotherapy and radiation induced hearing loss and to provide recommendations for the evaluation and management of children at risk of hearing loss due to cancer treatment.

Lieu JE. 2004. **Speech-language and educational consequences of unilateral hearing loss in children**. Archives of Otolaryngology -- Head & Neck Surgery, 130(5), 524-30.

This review considered the effects of unilateral hearing loss on the development of speech and language and educational achievement. It found that for school age children with unilateral hearing loss there appeared to be increased rates of grade failures, need for extra educational help and perceived behavioural issues in the classroom. Some of these children had delays in speech and language and it was unclear whether these lessened with increasing age. It recommended further research in these areas.

Systematic and Other Reviews on Cochlear Implants

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, including 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.

Bond M, Elston J, Mealing S, et al. 2009. **Effectiveness of Multi-Channel Unilateral Cochlear Implants for Profoundly Deaf children: A systematic review**. Clinical Otolaryngology, 34(3), 199-211.

This review considered the evidence comparing the effectiveness of unilateral cochlear implants with non-technological support or acoustic hearing aids in children with profound bilateral hearing loss. 15 suitable studies were identified but the degree of heterogeneity in design and outcomes precluded meta-analysis. However, all studies reported that for all outcomes measured, unilateral cochlear implants produced improvement. Five economic evaluations found that unilateral cochlear implants were cost-effective for profoundly deaf children at UK implant centres. The authors considered that the systematic review process gave greater weight to the positive findings of the15 papers reporting on this subject and noted that an RCT to prove the effectiveness of cochlear implants would be unethical.

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 research on bilateral cochlear implants. 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, 2 case control studies and 7 case series or case studies. All of the studies had small sample sizes (<50). The authors found that sound localisation and speech recognition in noise seemed to be improved with bilateral cochlear implants compared to a unilateral implant and that the greatest benefits occurred when the second implant was done early. They recommended further research on cost-effectiveness, quality of life, speech, language & psycho-educational measures.

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

Papsin BC, Gordon KA. 2007. **Cochlear implants for children with severe-to-profound hearing loss**. New England Journal of Medicine, 357(23), 2380-7. http://content.nejm.org/cgi/content/full/357/23/2380

In this review on cochlear implants, the authors state: “To our knowledge, large, randomized trials comparing cochlear implants with other forms of hearing assistance have not been performed.” However young children receiving a cochlear implant have normally been previously fitted with hearing aids and found to have received no benefit from them. There is a strong correlation between the successful development of language in children with early-onset deafness and cochlear implantation between 12 and 24 months of age. In the absence of auditory stimulation, changes occur in the central auditory system, which mean that the longer the child waits for a cochlear implant the less satisfactory the outcomes are. There is some uncertainty about the youngest safe age at which to perform implantation but interest in early implantation (<1 year) is increasing. There is evidence that bilateral implantation provides extra benefits with better hearing in noisy situations, an ability to discriminate between sounds at different locations, and evidence of binaural processing in the brainstem. Parental commitment to post operative therapy programs focusing on the development of auditory skills is important. Minor perioperative complications are relatively common (15-20%) and include peri-lymphatic fistula or cerebrospinal fluid leak, tinnitus, vertigo, facial-nerve weakness or paralysis, epidural hematoma, and cellulitis of the surgical flap. Major complications requiring further surgery, which tend to occur later, are rarer (2-5%) and include flap necrosis, otitis media, cholesteatoma formation, non-auditory stimulation of the facial nerve, and electrode extrusion. Long term device failure can occur for various reasons requiring re-implantation which although challenging to perform produces results as good as or better than the original implantation.

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 considered two issues: 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 systematic review found 33 suitable papers of which only 2 were randomised controlled trials. The authors also developed a state-transition (Markov) model of the main care pathways deaf people might follow and the main complications and device failures. 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 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 pounds 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.”

Barton GR, Stacey PC, Fortnum HM, et al. 2006. **Hearing-impaired children in the United Kingdom. IV: Cost-effectiveness of pediatric cochlear implantation**. Ear and Hearing, 27(5), 575-88

This study, based on a health utility questionnaire completed by the parents of 403 implanted children, and 1863 non-implanted children, looked at cost-effectiveness of cochlear implantation in hearing-impaired children considering 8 clinical and demographic variables. The authors concluded that in the U.K. implantation was a cost effective strategy with the greatest benefits associated with younger children and greater degrees of preoperative hearing loss.

This paper was part of a series of four by the authors in the same journal issue. The other papers considered the costs incurred by families, the costs to the education system, and auditory performance, communication skills, educational achievements, and quality of life.

Centre for Reviews and Dissemination. 2011. **Hearing-impaired children in the United Kingdom. IV: Cost-effectiveness of pediatric cochlear implantation** (Structured abstract). NHS Economic Evaluation Database (NHSEED), 2011(4).

The authors at the CRD reported that the methodology appears to have been generally appropriate although few details were given for the cost analysis and that the authors' conclusions appear to be valid within the limits of the study design. The study was based on children implanted before 2000 and therefore more recent improvements in cochlear implantation are not considered.

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 noted 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.