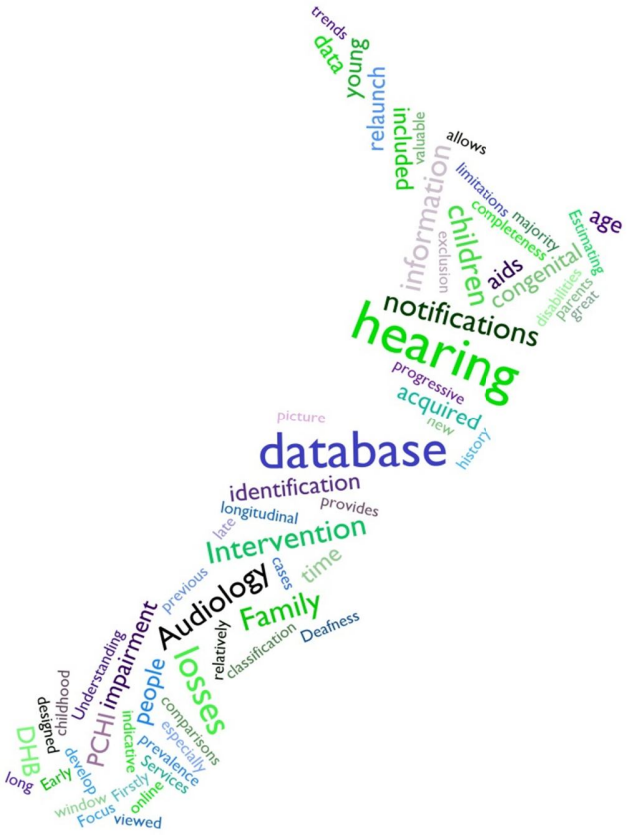


Deafness Notification Report (2010)

Notified cases of hearing loss (not remediable by grommets) among New Zealanders under the age of 19

Janet Digby, April 2011

NEW ZEALAND Audiological Society

Citation: Digby JE, Kelly AS, Purdy SC (2011) Hearing Loss in New Zealand Children: 2010, New Zealand Audiological Society, Auckland, New Zealand.

This and previous reports are available on the New Zealand Audiological Society website:
http://www.audiology.org.nz

Report summary

History and re-launch of the Database

The Deafness Notification Database (DND) was in operation from 1982-2005. The purpose of this database was 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. Operation of the Database was funded by the Ministry of Health during this time.

From 2006-2009 the Database was not in operation. In 2010 the DND was re-launched, with audiologists around the country encouraged to notify newly diagnosed hearing losses through a new online form. A number of changes have been made to the information collected, although every effort has been made to retain as many questions as possible to enable longitudinal comparison. This re-launched database has been funded by the New Zealand Audiological Society with help from Janet Digby.

Eligibility criteria have been revised, and now include those children born overseas and those with unilateral hearing losses.

Notifications received

In 2010, 180 notifications met the revised criteria for the DND, from 18 of the 20 district health boards. 91 notifications were also received retrospectively for the 2009 reporting period.

Without prevalence information for hearing loss among New Zealand children and young people there is no way of knowing what proportion of the total new diagnoses have been notified to the database during a given year.

Indications from previous data from the DND and from prevalence estimates suggest the 2010 dataset is likely to have captured between 50% and 80% of the new cases of hearing loss diagnosed in the 2010 calendar year among those under the age of 19.

Other key points about the children and young people notified include:

- The majority (81%) were known to be born in New Zealand
- Fewer children were reported as having other disabilities present when compared with previous years
- Fewer families identified as being European than in the general population of children and young people, while more identified as Māori

Other information

There are many systems for classifying the severity of hearing loss, including a number of current and historic systems used in New Zealand and overseas.

Anecdotal evidence from audiologists has for some time pointed to a higher proportion of less severe hearing losses among New Zealand children and young people. Although comparison with overseas data is problematic, there are some indications that the proportion of children in NZ with more severe losses may be lower than in other jurisdictions. Additional data will be required to form any conclusions on this issue.

Other key points:

- One third of notifications made to the database were for unilateral hearing losses

- Children who are identified as European are under-represented within the database, and Māori over-represented
- Fewer newly identified hearing losses (2010) were reported to have a known cause compared with previous data
- 28% of children and young persons identified with a hearing loss in 2010 had a known family history of hearing loss. The most common known cause was 'acquired'.
- 92% of children/young people diagnosed in 2010 were expected to receive one or two hearing aids

Identification of hearing losses

- Parents were, as in previous years, the most likely to first suspect a hearing loss
- The average age at identification (diagnosis) for all 2010 notifications is 68 months
- When those with unilateral hearing losses and those born overseas or with acquired hearing losses are excluded, as they have been for previous DND analyses, comparisons of the average age at identification can be made with previous data. These data show the age at confirmation for this group has risen to 51 months
- Previous DND reports showed children of Maori and Pacific ethnicity were identified later than European children – this effect is not visible in 2010 data
- Children born more recently are more likely to have a significantly lower age of identification but the number of referrals from newborn hearing screening is low
- Unilateral and mild hearing losses and those with an acquired cause, along with hearing loss in children born overseas are on average identified later than other hearing losses

Delays in diagnosis

There is a significant average delay between suspicion and confirmation of the hearing loss through diagnosis. Average delay (where suspicion information was recorded) in 2010 was 20 months.

Common causes of delay include those caused by difficulty in obtaining a confirmed diagnosis, parents not attending appointments, difficulty getting a referral to audiology and waiting times to see hearing professionals.

Contact details

Feedback on this report should be directed to the primary author of the report, Janet Digby. Janet can be contacted at: Levare Limited: PO Box 32 374, Devonport, or by email: janet@levare.co.nz or telephone (09) 445-6006.

2011 Notifications

Audiologists (including non MNZAS members) are encouraged to make future notifications to the database by following this link: http://www.audiology.org.nz/Prof/Deafness%20Notification%20Database.aspx

- Please read the online form carefully when making your notifications and provide as much specific information as possible in the spaces provided.
- Notifications to the database can only be made online – **please do not submit paper forms for inclusion**.
- Please remember to submit your 2011 notifications by the end of January 2012.

Contents

Introduction

Background

The Deafness Notification Database (DND) was New Zealand's annual reporting system for new cases of hearing loss among children and young people from 1982 to 2005. This system included data on the number and age of children diagnosed with permanent hearing loss and annual reports describing collected notifications were completed. The database was managed by the National Audiology Centre on behalf of the Ministry of Health and later by the Auckland District Health Board.

The database provided the only source of information from which the prevalence of permanent hearing loss could be estimated, and from which the characteristics new cases of hearing loss among children and young people could be understood.

In 2006, the Auckland District Health Board discontinued their contract to provide services associated with this database. No new provider was sought by the Ministry of Health. Prior to this, the National Audiology Centre held national contracts for a number of projects, including the collection and reporting of deafness data.

Between 2006 and 2009, NZAS and other groups such as Project HIEDI expressed concern that information on the number and nature of new hearing loss diagnoses among children in New Zealand was no longer being collected.

The role of the DND was seen as being of even greater importance since the start of implementation, in 2007, of the Universal Newborn Hearing Screening and Early Identification Programme (UNSHEIP). Information from the DND was seen as providing an important measure of changes in the age of identification and as only way to identify potential false negatives within the screening programme.

Direct approaches to the Ministry of Health to request the database be restarted were unsuccessful, in part we understand this was because the 'line-item' for funding of data collection and analysis of newly diagnosed cases of hearing loss had been split and reallocated to each individual district health board.

In 2009, Janet Digby approached the New Zealand Audiological Society with a view to re-establishing the database, and the partnership which formed has resulted in the re-launch of the database from the 1st January 2010.

It is hoped that the Ministry of Health will in future bring the database back under its auspices, building on the work done for the re-launch of the database in 2010.

Improvements to the database

There have been a number of improvements made to the database for its re-launch in 2010. These include:

- moving the database online to reduce keying errors associated with data entry of paper forms
- use of a revised consent process to ensure all records collected are done so with the consent of the family, through use of a consent form which has been the subject of legal review
- input from professionals regarding how the data will be analysed and reported to better meet the needs of the stakeholders
- use of a rigorous data backup system (both local and survey provider) and improved data security through encryption and data access policies
- development of standardised methods for data analysis
- wide distribution of the annual DND report

Steps have been taken to ensure that key data is comparable with previous Deafness Notification Database (DND) reports where-ever possible, although in some cases individual questions have been amended to make these more specific and/or to reflect improved understanding in a particular area (such as family history) and/or to allow more useful comparisons with both local and overseas data (allowed for example by the collection of complete audiometric data) and as a result direct comparisons with earlier reports are not possible.

Although the Database has been restarted by the New Zealand Audiological Society, efforts have been made to publicise the database to non-members of the Society in an attempt to collect as many notifications as possible.

Changes to inclusion criteria

The original criteria for inclusion in the Deafness Notification Database was based on the Northern and Downs definition below and was applied to data until the end of 2005:

"Children under 18 years with congenital hearing losses or any hearing loss not remediable by medical or surgical means, and who require hearing aids and/or surgical intervention. They must have an average bilateral hearing loss (over four audiometric frequencies 500-4000Hz), greater than 26dBHL in the better ear (Northern and Downs classification 1984)[1]."

There was also a strong view among audiologists consulted that the previous definition (above) was 'medically focused' and didn't adequately acknowledge or include hearing losses, particularly mild and unilateral losses, where the family might not want hearing aids fitted or where hearing aids were may not be appropriate.

The criteria for inclusion have now been modified based on feedback from a small working group[2] providing advice on the database. The new definition still includes children and young people 18 years or younger but this database now includes children:

- with an average of hearing loss of 26dBHL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0kHz) in one or both ears[3]
- who are born outside of New Zealand

Specific guidance has been sent to audiologists to provide clarification of the type of cases which are included in the new database:

> Included within the database; atresia, congenital ossicular fixation, meningitis, acquired hearing losses.
>
> Excluded from the database; hearing losses which are can be fixed by the use of grommets (ventilation tubes), such as hearing loss associated with otitis media.

[1] Northern JL, Downs MP (1984) Hearing in Children, Williams & Wilkins.

[2] This group comprised: Professor Suzanne Purdy, Dr Andrea Kelly, Lesley Hindmarsh, Robyn McNeur and Mr Colin Brown.

[3] While cases of unilateral hearing loss have been technically excluded from the database previously, there were large numbers of notifications sent to the administrators of the database and these cases were routinely excluded from the database. Professionals consulted in the development of the re-launched database unanimously believed this group should be included within the database.

Weaknesses of the database

While every effort has been made to ensure the newly re-launched database improves our understanding of the characteristics of newly diagnosed permanent hearing losses among NZ children and young people, there are two main weaknesses associated with the database which reader should note:

- Incomplete dataset: There is no way of knowing how far away we are from having a complete 2010 dataset, but indications suggest there are likely to be a good number of notifications not included within the current dataset. There may be certain types of cases which are underrepresented and as a result inferences made from the data contained in this report should be taken as indications only.

  We hope that as time passes further efforts can be made to increase the proportion of notifications received, improving the capacity of the dataset to inform the Ministry of Health, Ministry of Education and other service providers about the number and nature of new diagnoses of hearing loss in New Zealand.

- Comparability: While efforts have been made to retain as many questions as possible from the previous notification form some questions have changed, and in some cases the context around questions has altered making comparability difficult or impossible for some items.

- Duplicate records: While every effort has been made to ensure no duplicate records were used within the analysis for this report there is a chance previous datasets contain children previously notified to the database who were notified again in 2010.

Possible future renaming of the database

The name of the database (Deafness Notification Database) is being reconsidered. A new name may better reflect the purpose and nature of the database, particularly as changes to the inclusion criteria mean cases of unilateral hearing loss re now included within the database.

If the reader of this report has any ideas on what the database might be called in future, these will be gratefully received by the primary author of this report, Janet Digby, email: janet@levare.co.nz.

Contributions

This report was made possible through funding from the New Zealand Audiological Society. Without this support, those working with children who are deaf and hearing impaired would have remained in the dark about the number and nature of new hearing loss diagnoses in New Zealand. The author of this report also donated significant time at no cost to this project.

In addition to thanking the New Zealand Audiological Society for their commitment to this project, the primary author of this report gratefully acknowledges the significant support and guidance of Dr Andrea Kelly and Prof. Suzanne Purdy. In addition, the support of the following individuals is also acknowledged: Lesley Hindmarsh, Robyn McNeur, Dr David Welch, Mr Colin Brown and Prof. Peter Thorne.

Thank you to all the families who provided consent for data to be included within the database. We hope providers will be able plan and provide better future services for families as a result of an improved understanding of the number and nature of new hearing losses diagnosed in New Zealand.

The time taken by individual audiologists around the country to make notifications and request consent from families is also very much appreciated, as are efforts of those who have completed the analysis for previous reports, which has its own unique challenges.

Notifications

General information

210 unique notifications were received for 2010 by the 31st of January 2011, of which 180 met the revised criteria[4]. These notifications were received from a total of 45 audiologists, representing 18 of the 20 district health board areas.

Slightly more notified cases were male (54%) than female (46%). This is a very similar ratio (1.18:1) of boys to girls as has been found elsewhere (≈1.2:1), with boys commonly found to have higher rates of hearing loss than girls within international research[5].

It is difficult to ascertain how long, on average, audiologists took to make each individual notification as some online forms were left open for a number of hours or even overnight. However, it is clear that many individual notifications took five minutes or less to make using the online form.

Retrospective notifications from 2009 were also collected during 2010. 99 notifications from 2009 were provided to the database, of which 91 unique notifications met the revised criteria. A short summary of data contained in these notifications can be found in *2009 Notifications to the database* on page 35 of this report.

Please note when reading this report:

- Unless otherwise specified, analyses within this report either duplicates the exclusions made in previous reports for the purposes of comparison or describes the full number of notifications 2010 shown in red in Table 1.
- No information on the presence of risk factors was collected for notifications in 2010 as the set of risk factors used locally and internationally is changing, and as we hoped the newborn hearing screening programme would be able to provide data on the proportion of cases identified with one or more risk factors.
- No information has been collected on the number of new diagnoses which could not be notified as the parents declined consent for the information to be provided.

Figure 1 on page 9 shows variability in the number of notifications provided to the original database, particularly in the last six years of its operation. From 2000, the number of notifications shown is split between those notifications included in the main analysis and those excluded from the main analysis as they did not meet inclusion criteria. Those included in the main analysis are shown in grey, with those excluded from the analysis in green.

Further detail can be seen in Table 1, on page 9, which shows the types of cases included in previous DND reports by period (calendar year) from 2000. Notifications included in main analysis are marked in red. For the 2010 and 2003 and 2004 figures, cases are sometimes listed in more than one group and hence totals are provided. (An additional 8 notifications were also removed from the 210

[4] Duplicate 2010 records were removed following consultation with advisors, where appropriate, as were cases where the hearing loss was classified as slight (less than 26dBHL averaged over four audiometric frequencies) and cases where the child or young person had mild losses with normal bone conduction thresholds.

[5] Mehra S, Eavey RD, Keamy DG, (2009) The epidemiology of hearing impairment in the United States: Newborns, children, and adolescents. Otolaryngology–Head and Neck Surgery, 140, 461-472.

notifications received, as these were mild losses with normal bone conduction; these are not included in Table 1.)

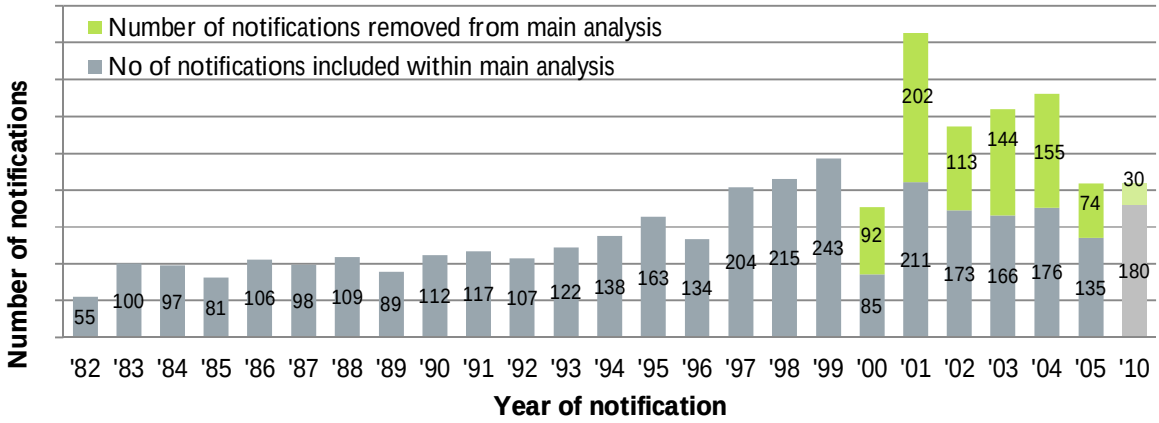


FIGURE 1: NOTIFICATIONS TO THE DEAFNESS NOTIFICATION DATABASE BY YEAR

While efforts have been made to remove duplicates from the 2010 dataset, without access to the original data it is not possible to know whether cases previously notified were mistakenly notified in 2010.

The 2005 DND report notes that the dataset containing previous DND data contained duplicate entries, so the total number of notifications reported in each of the previous calendar years may be exaggerated somewhat. The lower number of notifications which met the criteria in 2005 was also attributed to removal of duplicate entries.

Work is continuing in 2011 to encourage audiologists to report new cases to the database and it is hoped that the number of notifications to the re-launched database will grow over time and will begin to approach 100% of all new diagnoses made in a given year.

	Re-launched database	Original database					
Case type	**2010**	**2005**	**2004**[6]	**2003**	**2002**	**2001**	**2000**
Children with bilateral hearing loss greater than 26dB in the better ear	120	93	155	144	113	202	92
Children with unilateral hearing losses	60	51	68	51	38	54	14
Children born overseas	13	24	14	36	25	34	5
Children with losses thought to be acquired (2000-2005) OR those confirmed as acquired (2010)	10	19	36	43	40	67	6
Children with slight hearing losses[7]	22	23	63	73	70	56	53
Total Notifications	**210**	**209**	**331**	**310**	**286**	**413**	**170**

TABLE 1: NUMBER OF NOTIFICATIONS RECEIVED IN EACH REPORTING PERIOD, BY CATEGORY

[6] 288 retrospective notifications were made in 2004 as a result of the Children's Hearing Aid Fund Audit for cases identified over the previous 11 years. 157 of these notifications were found to meet the criteria. These notifications are not included in the table below as they cannot be attributed to a particular reporting period.

[7] Slight losses are those which do not meet the criteria of 26dB average over four audiometric frequencies.

Estimating the total number of new diagnoses per year

As there is no prevalence information for permanent hearing loss among New Zealand children and/or young people, it is not possible to accurately understand how close the database is to collecting data on all new cases of permanent hearing loss which meet the inclusion criteria.

We can however use: 1) the number of notifications provided to the database until 2005; and, 2) estimates of hearing loss prevalence among children and young people to provide some indication of the number of new diagnoses of hearing loss annually among children and young people:

1. Reviewing the number of notifications to the DND in recent years, and without knowing the extent of over-reporting due to cases being notified in more than one reporting period, it would seem reasonable to assume that between 50% and 80% of all new diagnoses may have been notified in 2010.

2. Although there are a number of difficulties using prevalence from overseas jurisdictions, estimates of overall prevalence in children and young people from international data[8] would suggest that given there are approximately 1.106 million children and young people in New Zealand up to the age of 18 years of age[9] there may be approximately than 245 new hearing loss diagnoses made annually which fit the new criteria. Using this method the notifications collected may comprise approximately 73% of the number of new diagnoses each year.

District Health Board Representation

The following table shows the percentage of notifications (2010) from each district health board (DHB) and compares this with the percentage of the population under the age of 20[10]. All but two DHBs provided one or more notifications which met the criteria for inclusion.

In addition to the natural fluctuations in the number of hearing losses diagnosed in a given year, other factors influencing notification may include; the number of FTE audiologists employed by each district health board; workload of these audiologists; and, their commitment to making notifications to the database.

	Percentage of notifications received 2010	Percentage of population under the age of 20 (NZ 2006 Census)
Auckland	5.6%	8.9%
Bay of Plenty	7.2%	4.9%
Canterbury	24.4%	10.8%

[8] Fortnum et at 2001 estimated that for every 10 children detected through newborn hearing screening with a hearing loss averaging 40DBHL an additional 5-9 cases of permanent childhood hearing impairment might be detected in the post-natal years. Fortnum's prevalence at birth is 1.06 per thousand births and this rate is very close to more recent prevalence estimates from programmes such as those in New South Wales and Victoria which report an approximate additional figure of 0.5 per thousand for unilateral hearing losses. Using this information and including an estimate for children with unilateral and mild hearing losses we can make a rough estimate of the prevalence of permanent congenital hearing loss among 0-18 year olds of 4 per thousand.

[9] Statistics New Zealand 2006 Census Data: QuickStats About New Zealand's Population and Dwellings – Table 3 http://www.stats.govt.nz/Census/2006CensusHomePage/QuickStats/quickstats-about-a-subject/nzs-population-and-dwellings.aspx accessed on March 22nd, 2011

[10] This group is used as an approximation of the size of the population under the age of 19.

Capital and Coast	13.3%	6.1%
Counties Manukau	13.9%	12.7%
Hawke's Bay	5.0%	3.8%
Hutt	2.8%	3.5%
Lakes	1.1%	2.7%
Midcentral	2.2%	4.0%
Nelson Marlborough	0.6%	2.9%
Northland	6.7%	3.9%
Southern	0.6%	6.6%
South Canterbury	0.6%	1.2%
Tairawhiti	1.1%	1.3%
Taranaki	3.3%	2.6%
Waikato	8.3%	8.9%
Wairarapa	0.0%	0.9%
Waitemata	2.2%	12.0%
West Coast	1.1%	0.7%
Whanganui	0.0%	1.6%

TABLE 2: PERCENTAGE OF NOTIFICATIONS (2010) COMPARED WITH PERCENTAGE OF POPULATION UNDER 20 YEARS OF AGE BY DHB

Other disabilities

12% of 2010 cases notified were not thought to have any additional disabilities at the time the notification was made although in 10% of cases there was uncertainty regarding whether the child or young person had an additional disability.

For the 21 children and young people reported to have additional disabilities, 27 specific conditions were listed. The most common of these were those were related to a specific syndrome (8 children), developmental delays (3 children) and vision problems (3 children).

This proportion is compared to previous data below.

Notification Year	Proportion of cases with a reported additional disability
2010	12%
2005	18%
2004	23%
2003	21%
2002	29%

TABLE 3: PROPORTION OF CASES WITH A KNOWN ADDITIONAL DISABILITY

Fortnum et al (2002) reported that 27.4% of UK children studied with hearing loss have at least one other disability from a sample of 17,169 children with hearing loss. The most common additional disabilities reported in this study were learning difficulties (11%) and visual impairment (6%).

The low rate of additional disabilities among children and young people diagnosed in 2010 may be explained in some measure by the result of a lack of definition for the term 'disability' within this question and/or changing perceptions of what conditions are included within this very broad term.

It is worth noting that children with hearing loss in New Zealand may not be routinely assessed by a pediatrician and hence other disabilities may be under-diagnosed. Potential under-diagnosis may be indicated by the high proportion of cases (10%) where there was uncertainty around whether the child or young person had an additional disability.

Place of birth

This is the first year children and young people born outside of New Zealand have been formally included within the database. Of the 180 cases included in the main analysis, 13 were known to be born outside New Zealand.

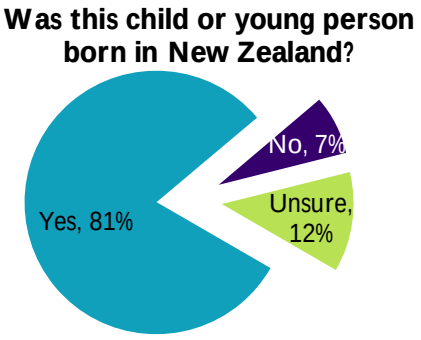


FIGURE 2: PROPORTION OF 2010 CASES BORN IN NEW ZEALAND

Notifications and ethnicity

The method used within this report to classify ethnicity is the total response method, where every person identifying with an ethnicity is included within that specific grouping. For example, if someone considers their child to be Samoan and Maori they are recorded under both these groups. This means the total number of ethnic groups selected by respondents is usually greater than the number of respondents.

This method provides a more accurate measure of the relative size of the groups identifying with a particular ethnicity when compared with older survey methods which required respondents to select only one ethnicity with which they mostly identified. Using this method also aligns the database with The New Zealand Census, which began explicitly instructing respondents that they could select more than one category for their ethnicity in 1996.

The New Zealand Census (2006) categorised respondents into five major groupings as per the Statistical Standard for Ethnicity (2005) and this standard will continue to be used for the next Census. These groups are; Māori, Pacific Peoples, Middle Eastern/Latin American/African (MELAA), European, and Asian.

2010 notifications (including those children and young people born overseas) are reported in the table below using this classification system. As a high proportion of those notified to the database are younger children data on the proportion of under 5 year olds recorded in the Census under each of ethnic group.

Ethnic group (grouped total responses)	Percentage 2006 Census, under the age of 20[11]	Percentage 2006 Census under the age of 5	Number of cases 2010 DND[12]	Percentage of cases by ethnic group 2010 DND
European	62%	62%	101	51.5%
Māori	22%	24%	59	30.1%
Pacific Peoples	11%	13%	18	9.2%
Asian	9%	9%	13	2.6%
Middle Eastern / Latin American / African[(5)]	1%	1%	5	2.6%

TABLE 4: ETHNICITY GROUPINGS (2010) COMPARED WITH PROPORTION OF THE POPULATION UNDER THE AGE OF 20

All but 9 notifications for 2010 contained one or more ethnicity codes. Of those with one or more ethnicity code, 87% of respondents' selected one code for their child's ethnicity, while 11% selected 2, and 2% selected three codes.

The proportion of notifications within each ethnic group was calculated differently in previous reports with respondents being coded as belonging to one ethnic group. Categories used have also changed. As a result, direct comparison is not possible.

[11] Statistics New Zealand 2006 Census Data: QuickStats about Culture and Identity – Table 3 http://www.stats.govt.nz/Census/2006CensusHomePage/QuickStats/quickstats-about-a-subject/culture-and-identity.aspx, accessed on March 22nd, 2011

[12] Please note that as more than one ethnic group may be selected for a specific notification the number of cases in each ethnic group for 2010 total to more than the number of notifications

Severity of hearing losses

Audiometric data

Audiometric data was requested for both right and left ears. Examining the four data points for each ear shows that this data was provided for 150 and 178 of the 180 cases notified to the database, for right and left ears respectively.

Audiologists notifying cases to the database were asked to provide air conduction thresholds from the pure tone audiogram. In cases where the young age of the child meant the audiologist was unable to obtain audiogram data, audiologists were asked to estimate thresholds from the ABR using correction factors of 5, 5, 0, and -5dB for 0.5kHz, 1.0, 2.0 and 4.0kHz respectively.

Where a significant air bone gap was present, bone conduction thresholds at the appropriate frequencies were also collected and correction factors of -5 for 0.5kHz and 2.0kHz were provided to within the online notification form[13].

79% of cases notified contained data estimated from the pure tone audiogram, with the remaining 21% estimated from the ABR. This figure is an indication that children being assessed are old enough, in large part, to have their hearing assessed behaviourally. We would hope to see this figure drop in future years as newborn hearing screening programme coverage rates increase.

The overall four frequency average hearing threshold across the 2010 cases was 45.67dB HL for the right ears and 45.90dB HL for the left ears. Figure 3, below, shows the average audiometric data for each frequency for both left and right ears; there is very little difference in profiles between left and right ears. Figure 3 also indicates that severe losses are more prevalent at higher frequencies.

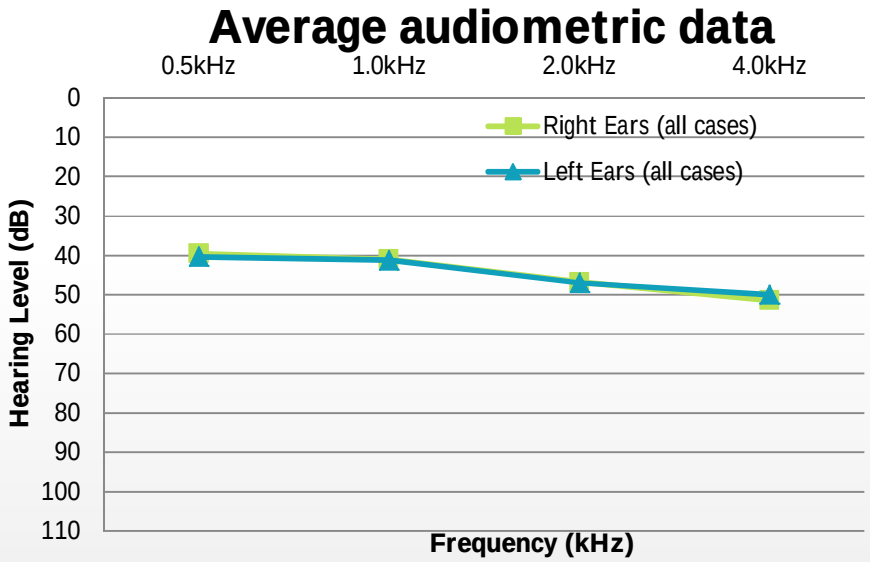


FIGURE 3: AVERAGE AUDIOMETRIC DATA FOR RIGHT AND LEFT EARS (2010)

[13] Correction factors for ABR and bone conduction were provided within the online notification form. These are from National Screening Unit (2009) Universal Newborn Hearing Screening and Early Intervention Programme National Policy and Quality Standards Appendix F Diagnostic and Amplification Protocols June 2010 and were accessed from http://www.nsu.govt.nz/health-professionals/2940.asp on the 22nd of March 2011.

Classifications

A large number of classification systems are used to categorise hearing loss severity, locally and in overseas jurisdictions. These differing systems make it difficult for meaningful direct longitudinal and geographical comparisons of the proportion of children in a particular severity category.

There does not seem to be a clear standard developing internationally for classifying hearing loss, or a consistent definition for where a hearing loss begins for the purposes of epidemiological comparison. In addition these systems, by and large, do not acknowledge any differences which may exist between the way hearing losses in children, young people and adults might best be categorised i.e. there is one system of classification for all groups.

While the New Zealand Deafness Notification Database (DND) collected some audiometric data for a number of years until the end of 2005, this data was problematic and did not allow comparisons to be made easily with data from overseas. The newly re-launched database contains full audiometric data and as a result more meaningful comparisons can now be made with overseas data.

The table below shows some of the differences between local and overseas severity classifications. (These systems use an average of the pure-tone thresholds at 0.5kHz, 1kHz, 2kHz and 4.0kHz.) Please note that audiologists in New Zealand are commonly using the 1981 Clark (ASHA) classifications within their clinical practice.

	1996-2005 NZ DND	1982-1996 NZ DND	Clark 1981 (ASHA website)[14]	Jerger and Jerger (ASHA website)[15]	World Health Organisation[16]	CDC[17]
Normal			-10-15dBHL		≤25dBHL	
Slight			16-25dHBL	0-20dBHL	26-40dBHL	
Mild	26-40dBHL	30-55dBHL	26-40dHBL	20-40dBHL		21-40dBHL
Moderate	41-65dBHL		41-55dHBL	40-60dBHL	41-60dBHL	41-70dBHL
Moderately Severe		56-85dBHL	56-70dHBL			
Severe	66-95dBHL		71-90dHBL	60-80dBHL	61-80dBHL	71-90dBHL
Profound	>95dBHL	≥86dBHL	≥91dBHL	≥81dBHL	≥81dBHL	≥91dBHL

TABLE 5: COMPARISON OF AUDIOMETRIC SEVERITY CLASSIFICATION SYSTEMS

[14] Clark JG, (1981) Uses and abuses of hearing loss classification. ASHA, 23, 493–500

[15] ASHA (2010) Information Series- Hearing Loss Types from http://www.asha.org/uploadedFiles/aud/InfoSeriesHearingLossTypes.pdf, accessed on 1st February 2011.

[16] WHO Prevention of blindness and deafness – grades of impairment from http://www.who.int/pbd/deafness/hearing_impairment_grades/en/index.html accessed on 1st February 2011.

[17] CDC-EHDI Hearing Screening and Follow-up Survey (HSFS): 2006 Explanations (Version B) accessed on 21st March 2011 from http://www.cdc.gov/ncbddd/hearingloss/2006-data/2006_HSFS_Explanations_B.pdf

Comparisons with previous data

By categorising the notifications using the severity scale used by the DND from 1996-2005, a comparison of the proportion of children in each group is possible. Table 6, below, shows the proportion of hearing loss notifications in each category in 2010 and compares this with data from 2001-2004[18]. 2010 figures shown here exclude those children born overseas and those with acquired hearing losses as previous data included also removed these cases.

We will be watching this data closely next year to see whether the current severity profile remains or whether this profile is an artifact of an unrepresentative 2010 sample.

Proportion of cases notified by degree of hearing loss	2001	2002	2003	2004	Average 2001-2004	2010
Mild	47%	47%	56%	43%	**48%**	42%
Moderate	35%	39%	33%	34%	**35%**	37%
Severe	10%	9%	6%	15%	**10%**	6%
Profound	8%	5%	5%	7%	**6%**	15%

TABLE 6: NOTIFICATIONS BY DEGREE OF HEARING LOSS USING 1996-2005 CLASSIFICATION SYSTEM

It is interesting to note that vaccination programmes have reduced rates of meningitis in New Zealand's and this reduction is expected to have led to a reduction in rates of (more severe) hearing loss[19]. A reduction in the number of cases may not be visible in the overall DND as the numbers are small and also as specific data on the number of cases of meningitis has not been collected in previous versions of the DND.

Of the cases of hearing loss diagnosed in 2010 and notified to the database six were listed as being the result of meningitis.

Comparisons with data from overseas

It was more difficult than expected to source information on the severity profile of children and young persons with hearing impairment, in part as overseas data often exclude children with mild hearing losses.

While the collection of audiometric data makes comparison with overseas data easier as cases can be re-coded by any number of classification systems, there are other issues to consider when making such comparisons.

The first of these is that severity profiles are likely to differ between studies completed pre and post implementation of newborn hearing screening[20,21]. As newborn hearing screening has been recently

[18] 2004 data is used as it is unclear from the 2005 report which figures relate to which of the ASHA categories.

[19] Turner N, (2011) personal communication to J Digby March 21st 2011.

[20] There was previously a view that a significant number of new cases of hearing loss would develop after the child was born (acquired and progressive or late onset hearing losses) but there are indications from recent research that this view may be changing.

implemented in New Zealand and coverage is not yet universal, there will likely be a mix of late and early detected hearing losses being diagnosed for some time, depending on how quickly coverage rates increase, how close to coverage gets to 100% and how many children are lost to follow-up.

The second issue making comparison with overseas data difficult is that many studies do not include milder hearing losses and therefore miss part of the picture with regards to the severity profile of children and young people diagnosed with hearing loss.

Previous comparisons with UK data (contained within the 2001 DND report) refer to data from the Trent region, described by Fortnum and Davies (1997). The DND report compared the severity profile of cases within this study with all children notified with bilateral hearing losses to the DND. This comparison indicated that there may be a lower proportion of more severe hearing losses among NZ children.

A more recent and very large study from the UK (Fortnum et al 2001)[22] examined bilateral hearing losses among children born from 1980 to 1995, and resident in the UK in 1998 with permanent childhood hearing impairment in the better ear of >40dB[23]. Notifications ascertained 17,610 individual children with hearing losses meeting the criteria from health and educational professionals. A subgroup was then identified (n=6,698) which matched the cohort from the Trent study and relative prevalence of moderate and greater degrees of hearing loss calculated.

It is important to note that there are issues with direct comparison between the DND data and data from these studies as the groups are not identical. DND data contains children and young people diagnosed in a given year includes those born overseas while the Fortnum study includes only children born within a specific date range, those living in the UK in 1998 and born between 1980 and 1995. In addition, the Fortnum study contains only bilateral hearing losses, while the DND collects both information about children and young people with unilateral and bilateral hearing loss.

The proportion of cases in each severity grouping from the 2010 DND data (bilateral hearing losses only) is compared in Table 7, below. Data from these UK studies indicates there may be fewer more severe losses among children and young people in NZ than for those who are resident in the UK.

		Fortnum et al (Trent 1997)		Fortnum et al (2001), subset		DND (2010)	
Average thresholds	Degree of loss	Sample size	Proportion	Sample size	Proportion	Sample size	Proportion
40-70dB	moderate	270	55%	3739	56%	75	66%
71-95dB	severe	104	21%	1379	21%	24	21%
>95dB	profound	113	23%	1580	24%	14	12%
	Total	**487**		**6698**		**113**	

TABLE 7: COMPARISON DND (2010) AND UK DATA (BILATERAL LOSSES ONLY)

[21] Newborn hearing screening will also miss some milder hearing losses. These are difficult to detect and may not be identified until the B4 School Check (for those who receive this) or until the children begin to display issues learning. Some mild hearing losses will progress to more severe hearing losses and will be detected when the impact of the hearing loss becomes more obvious to parents, educators and or health professionals.

[22] Fortnum HM, Summerfield AQ, Marshall DH, Davis AC, Bamford JM (2001). Prevalence of permanent childhood hearing impairment in the United Kingdom and implications for universal neonatal hearing screening: questionnaire based ascertainment study. *British Medical Journal* 323, 536-539.

[23] Averaged over four audiometric frequencies (0.5,1.0, 2.0, 4.0kHz)

Comparing the proportion of cases in each severity grouping from the 2010 DND data with data from The Centres for Disease Control and Prevention in the US also indicates there may be fewer severe losses among children and young people in NZ than in the US.

		CDC (2006)		DND (2010)	
Average thresholds	Degree of loss	Sample size	Proportion	Sample size	Proportion
41-70dB	moderate	1280	47%	67	64%
71-90dB	severe	595	22%	21	20%
>91dB	profound	808	30%	17	16%
		2683		105	

TABLE 8: COMPARISON BETWEEN NZ DND AND US CDC 2006 DATA (2008)

Breakdowns by ethnicity and degree of loss

Figure 4 shows the proportion of cases in each of the various degrees of loss which were notified to the database, split by ethnicity grouping. Severity is categorised by the ASHA Clark classification system in this figure. *Please note that the Asian and MELAA categories contain particularly small samples.*

If these notifications are representative of children diagnosed with hearing loss in 2010 there is a worryingly high proportion of severe and profound losses among Pacific and MELAA children. It will be interesting to see whether next year's data shows a continuance of this pattern.

The 2005 DND report noted that Māori children notified in 2005 and between 1990 and 2005 were more likely to have a mild hearing loss than other ethnic groupings. 2010 data seem to show a similar pattern, although it will be important to see whether 2011 data repeats this pattern.

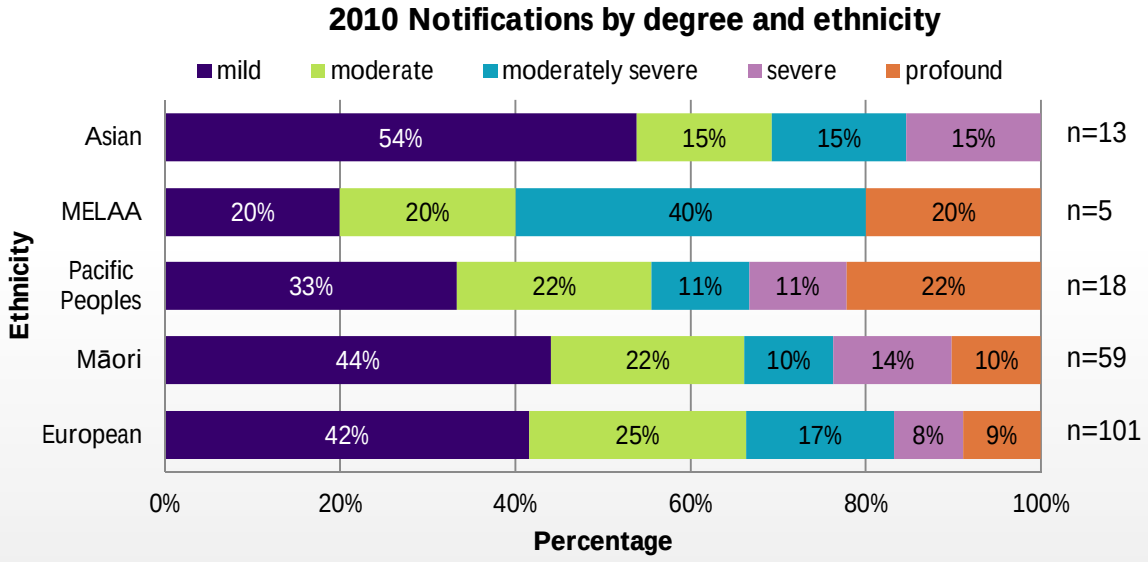


FIGURE 4: NOTIFICATIONS BY ETHNIC GROUP AND DEGREE

Unilateral hearing losses

Cases of unilateral loss within the database are defined as having hearing thresholds greater than 26dBHL in only one ear. It is particularly important to monitor children and young people with unilateral hearing loss as a significant proportion of these hearing losses progress over time[24].

Exactly one third of the notifications included within the main analysis were unilateral hearing losses. This is aligned with data from Colorado which reports that one third of hearing losses are unilateral[25].

Comparisons with previous DND data are problematic, as unilateral hearing losses were not within the original criteria for the database and therefore were likely to be under-reported. Some professionals are curious about whether New Zealand rates of unilateral loss could be reduced if immunization rates for conditions such as mumps could be increased.

Differences between the proportion of unilateral and bilateral notifications in each severity category are shown in Figure 5.

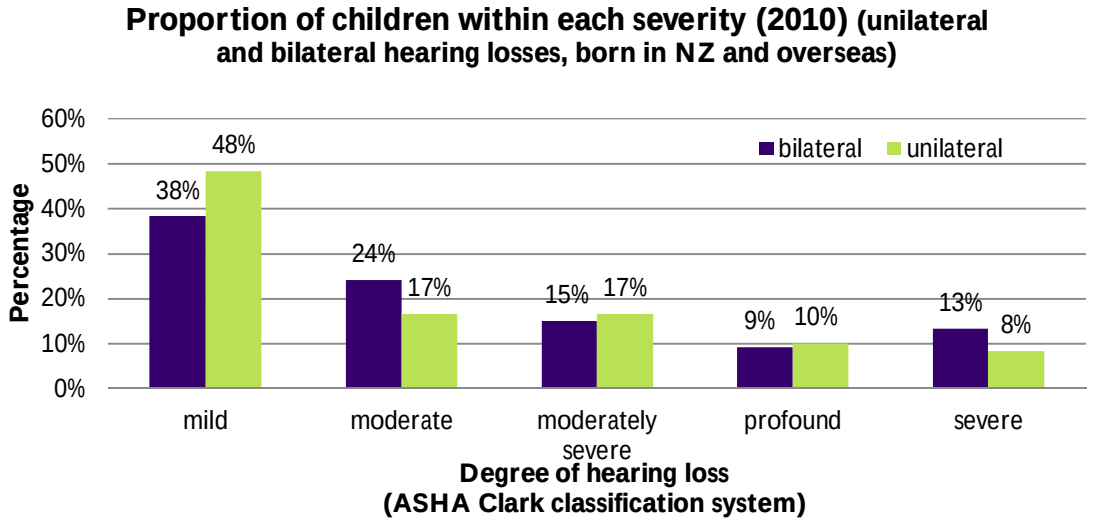


FIGURE 5: PROPORTION OF HEARING LOSSES FOR UNILATERAL AND BILATERAL HEARING LOSSES

Within Figure 6 on page 20, 2010 notifications are broken down by ethnicity and whether the hearing loss was unilateral or bilateral. Rates of bilateral hearing losses are significantly higher for Maori (and MELAA also, although this group is very small) than for European children. This is the first time this analysis has been performed on DND data and given the number of notifications provided in 2010 this effect would need to be demonstrated for a period of time before any conclusions could be drawn.

[24] Yoshinago-Itano C (2010) Personal Communication, J Digby, 7/4/2010.

[25] Yoshinaga-Itano C, (2009) Oticon Foundation Hearing Education Presentation. Raising the Bar for children with hearing loss. Powerpoint Presentation at the University of Auckland.

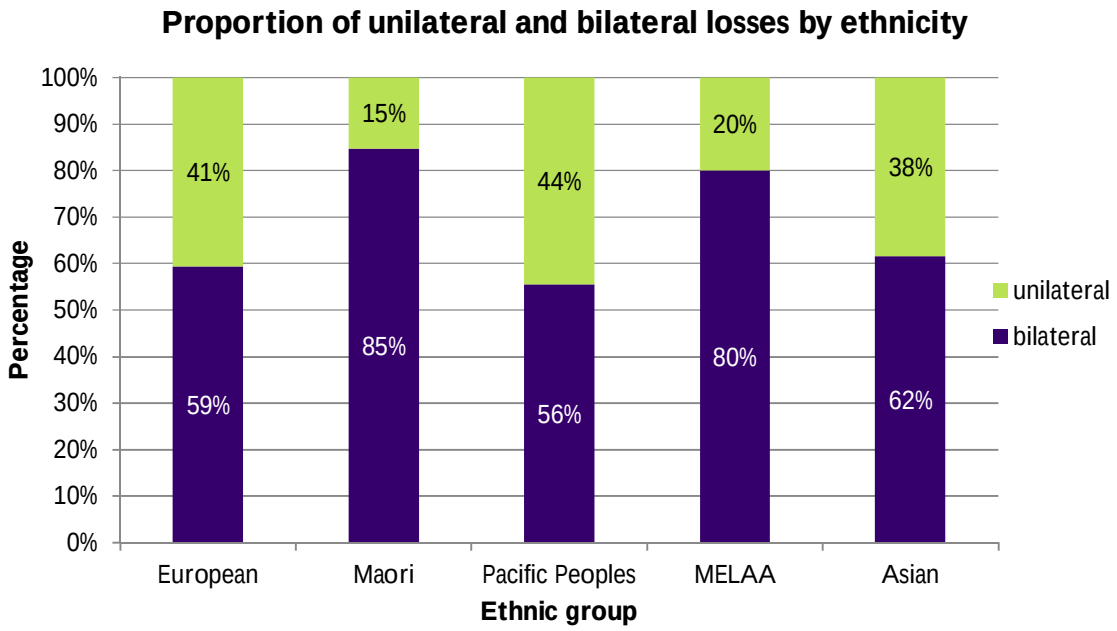
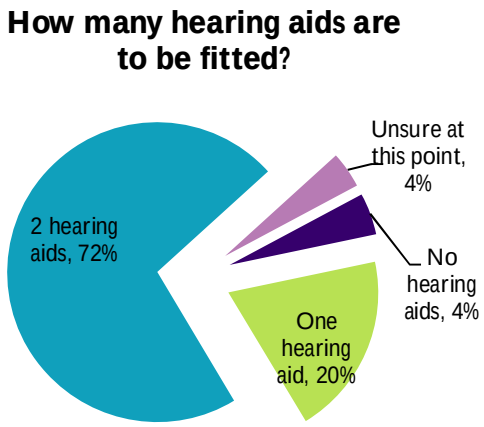


FIGURE 6: PROPORTION OF UNILATERAL AND BILATERAL NOTIFICATIONS (2010 BY ETHNICITY)

Hearing aids and cochlear implants

2010 data relating to the fitting of hearing aids is included in Figure 8 on page 22. The majority of children and young people with a hearing loss which was first diagnosed in 2010 are to be fitted with two hearing aids.



Further data regarding the severity classifications of those to receive hearing aids are found in Figure 8 on page 22.

It is worth noting that some children with unilateral hearing losses were reported to be receiving more than one hearing aid as although the average threshold for the better ear does not meet the 26dB average, one or more hearing thresholds are seen sufficiently poor to warrant amplification in the better ear. This is indicative of one of the limitations related to classification systems which average hearing thresholds and categorise children into severity groups – the result is a loss of richness in the data.

FIGURE 7: NUMBER OF HEARING AIDS TO BE FITTED (2010)

In this case unilateral hearing loss is used to indicate asymmetry, but it does not indicate that the child doesn't require help to ensure they can hear in the better ear.

For example, in one case reported to the database, the child's hearing loss was profound in the left ear (110+dB at each of the four frequencies 0.5, 1.0, 2.0 and 4.0kHz). While the average over the same four frequencies in the right ear was not enough (26dBHL or greater) to put this child's hearing loss into the bilateral category there was a 40dBHL loss at 4.0kHz and so the child was being fitted with two hearing aids.

Comparison of data from Australian Hearing

Some previous DND reports have compared the proportion of children in each category of hearing loss with data from Australian Hearing. However, the Australian Hearing data only includes those who have been fitted with hearing aids, and includes a larger age group (under the age of 21) and so previous comparisons have been problematic. Figure 8 below shows the proportion of children in each of the Australian severity categories who, in the case of Australia, have been fitted with hearing aids, and in the case of NZ, who expected to be fitted with hearing aids.

The graph below seems to indicate that a smaller proportion of New Zealand children with mild hearing losses are receiving hearing aids when compared with their Australian counterparts, although this may be the result of differences between the severity profile of NZ and Australian hearing losses. Incomplete notifications for mild hearing losses may impact on this data, but this is an interesting result, and it will be very interesting to see whether this trend is visible in 2011 data.

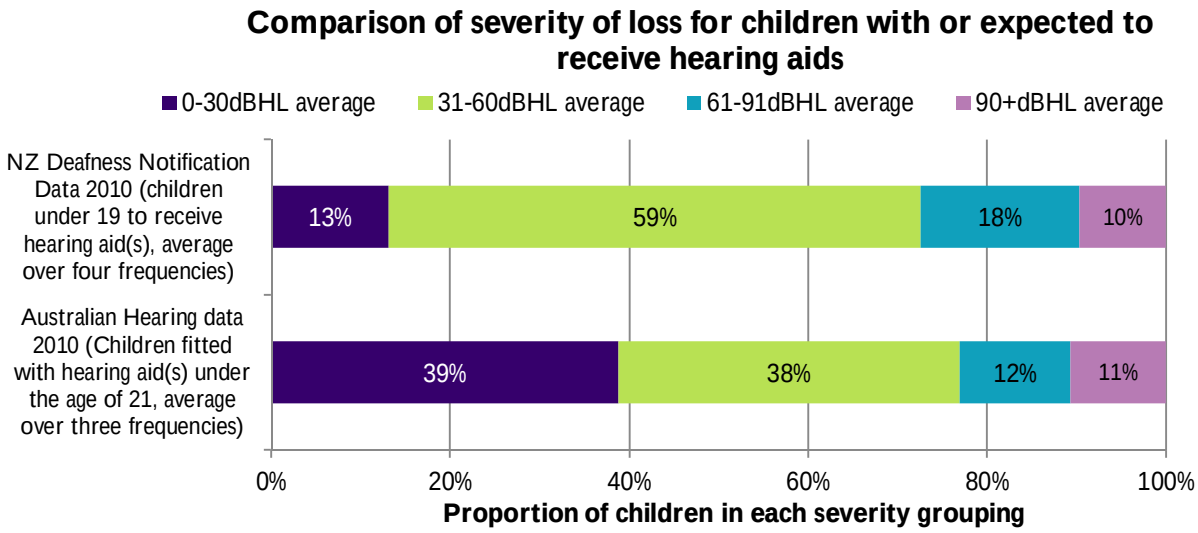


FIGURE 8: PROPORTION OF AIDED OR SOON TO BE AIDED CHILDREN BY SEVERITY - COMPARISON OF NZ AND AUSTRALIAN DATA[26]

Cochlear Implants

38 cases of severe or profound hearing loss (ASHA, Clark classification) were reported as newly diagnosed in 2010. These children are likely to meet audiometric criteria for implantation.

Cochlear implants are provided to children and young persons by funds from the Ministry of Health. This funding is administered by two cochlear implant trusts. The Northern Cochlear Implant Trust covers the area northwards from a horizontal line extending roughly through Taupo and the Southern Hearing Charitable Trust covers south of this line.

During the 2010 calendar year there were 17 cochlear implants provided in the Northern Region and 18 in the Southern Region to children and young people under the age of 18.

[26] Australian Hearing (2011) Report on Demographics of Persons under the age of 21 years with hearing aids - 2010

Cause of hearing loss

As seen in Figure 9 below, the proportion of hearing losses where the cause was thought to be known has decreased significantly in 2010. At least some of this difference is thought to be the result of changes in the cause information requested as the form has been made more specific.

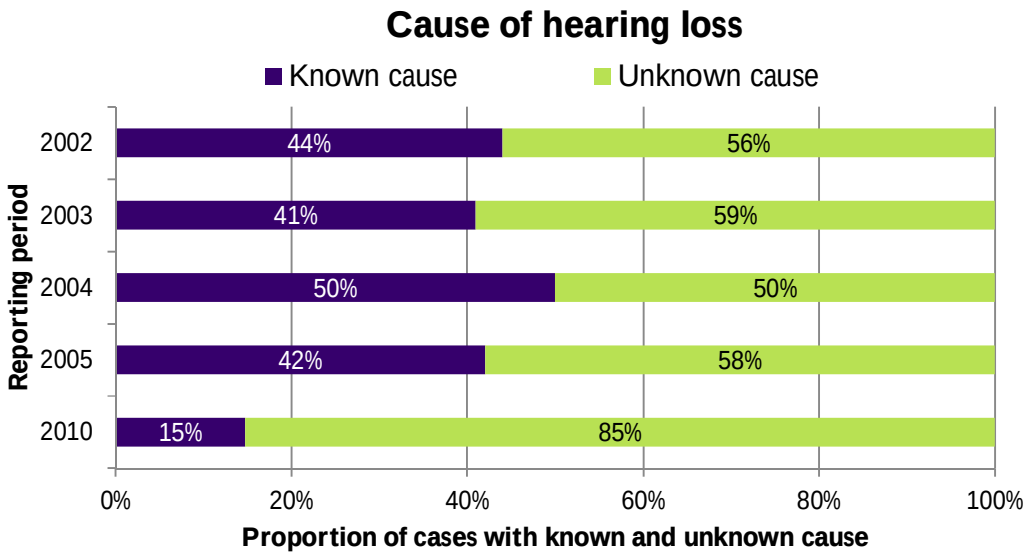


FIGURE 9: PROPORTION OF HEARING LOSSES OF KNOWN AND UNKNOWN CAUSE NOTIFIED TO THE DND BY YEAR

Perhaps unsurprisingly, the more severe the hearing loss, the more likely the aetiology is known. For example, only 13% of mild hearing losses (ASHA, Clark classification) were recorded as having a known cause compared with 32% of profound losses.

There is a drive among the ENT community in New Zealand to increase the proportion of hearing losses which have aetiological investigations such as genetic testing performed and it will be interesting to see whether the proportion of hearing losses with a known cause increases in 2011 notifications.

Known aetiology breakdown	
Acquired hearing loss	11
Other	6
Genetic cause (non syndromic)	3
Syndrome	3
Not listed	2

Table 9 below shows the breakdown of aetiology where this was known.

Known aetiology breakdown	
Acquired hearing loss	11
Other	6
Genetic cause (non syndromic)	3
Syndrome	3
Not listed	2

TABLE 9: BREAKDOWN OF KNOWN AETIOLOGY (2010)

Of the cases of hearing loss diagnosed in 2010 and notified to the database five were listed as being the result of meningitis.

Cases recorded as 'other cause' include those with cholesteatoma and permanent conductive hearing loss. Two of the cases where genetic cause had been confirmed were Connexin 26 related.

Family History

Previous reports note a relatively high proportion of cases were recorded as having 'family history' as the cause of the hearing loss. (Family history was reported as the cause of the hearing loss in 24-32% of cases between 2001 and 2005.)

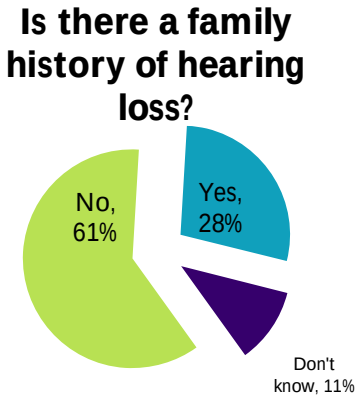


FIGURE 10: TABLE 10: INITIAL FAMILY HISTORY PRESENT (2010, ALL CASES)

Changes have been made to this question to try and gain more specific responses about the specific nature of the family history.

Of the 28% of cases where a family history was specified, 2% of these cases related to extended relatives (non-direct relatives).

Of the cases where the family member or members with hearing loss were or currently included the child young person's sibling(s) and/or parent(s):

- The direct relatives were reported as either still having the hearing losses in 89% of cases, while the family were unsure whether this loss was still present in 11% of cases
- The majority of these close relatives use (or used) a hearing aid or cochlear implant (71%), while the 21% did not and in the remainder of cases there was uncertainty regarding whether the relative used such a device
- The majority of these close relatives (67%) had their hearing loss from childhood while the remaining 33% were unsure when the hearing loss was first detected

Identification of hearing losses

Who first suspected the hearing loss?

Information on who first suspected the child or young person's hearing loss was recorded for 115 of the 145 children and young people known to have been born in NZ. (Information on place of birth was not recorded for 22 children and 13 children were born overseas)

Parents or caregivers were the most likely to first suspect the hearing loss, followed by Vision Hearing Technicians (VHTs), and medical professionals.

Previous DND reports also observed that parents were most likely to suspect a child's hearing loss. Parents first suspected hearing losses in between 34% and 52% of cases notified to the database between 2000 and 2005.

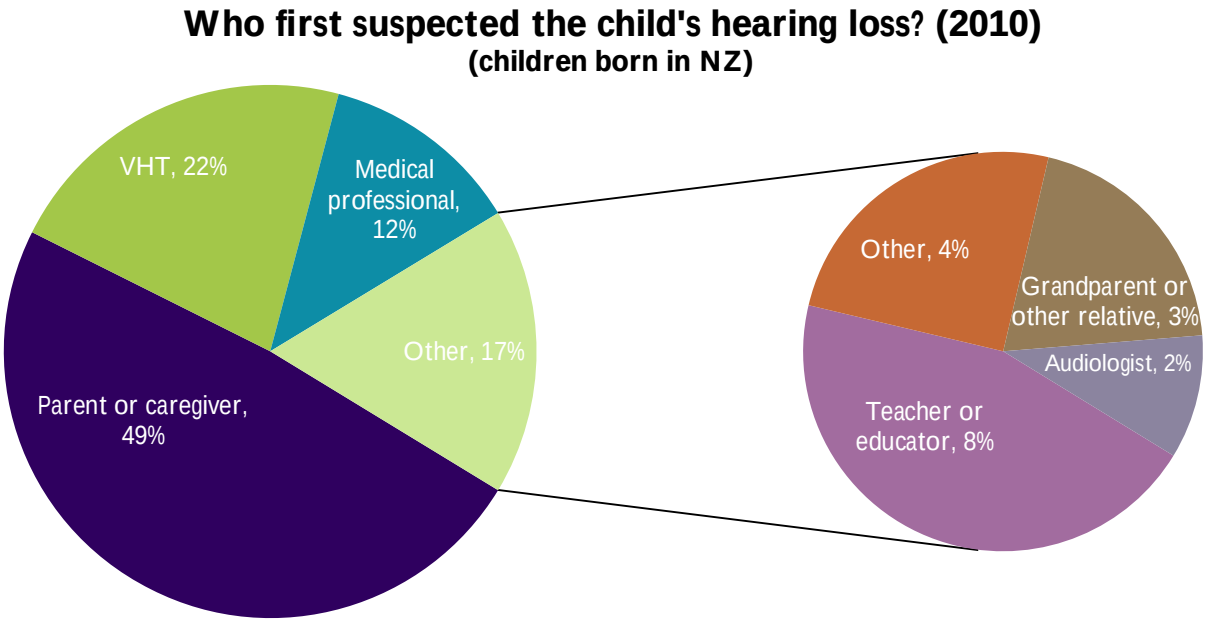


FIGURE 11: FIRST SUSPICION OF HEARING LOSS (CHILDREN BORN IN NZ) 2010

Age at identification

Figure 12 shows the number of cases by year of birth. Diagnoses in children born around 2005 may be higher due to diagnosis made due to hearing screening included within the B4 School Checks. These checks aim to screen children for hearing loss at the age of four, although some children are screened when they reach school.

The lower number of notifications for those born in 2008 may be due to a number of factors including: incompleteness in the number of notifications received for 2010; difficulties testing children around the age of two; or, a natural gap between identification of severe and profound hearing losses and the more difficult identification of mild and moderate hearing losses. The 2005 Deafness Notification Database Report contained a similar pattern although within that year's report there were fewer notifications three and four years before the year in which notifications were being collected.

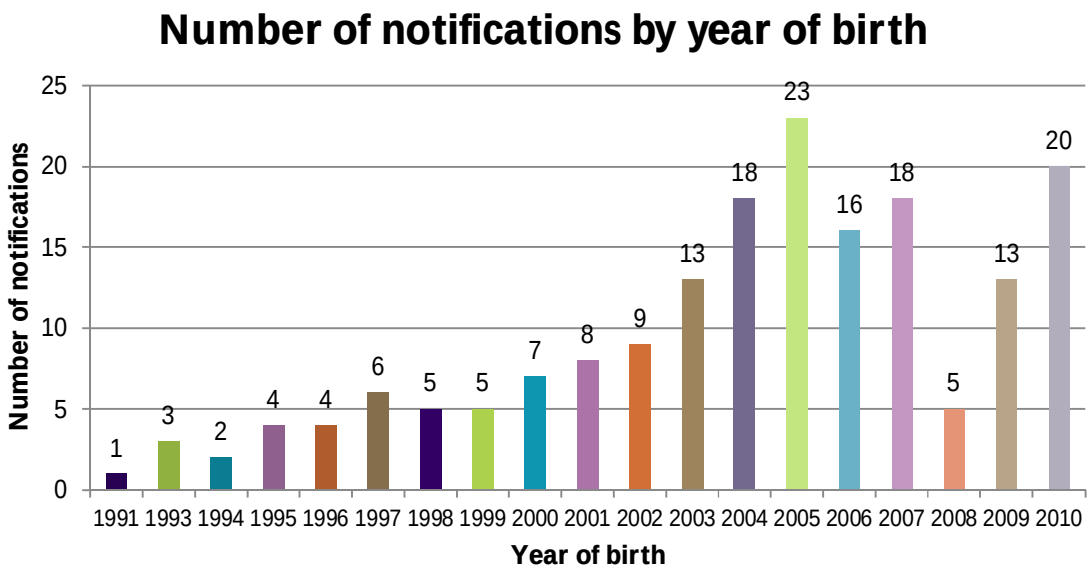


FIGURE 12: NOTIFICATIONS BY AGE AT DIAGNOSIS (2010)

Issues with using a single measure for 'age at identification'

There are a number of issues with reporting the average age at identification (diagnosis) for all groups of children. While this may be useful as it describes the age at which providers will begin working with children to provide interventions of some type, the average relates to all newly diagnosed children as there are difficulties attempting to separate out children with hearing losses which are late onset (such as progressives and acquired hearing losses).

Average age at identification for various groups

Keeping this in mind, the average age at diagnosis of children with all degrees of hearing loss, including those with acquired hearing losses and those born overseas is 68 months. While this does not show a reduction in the average age hoped for as a result of newborn hearing screening implementation, it is important to remember this average age includes all children diagnosed in 2010, including those born before screening was implemented and those with acquired or progressive hearing losses.

For the purpose of comparison with previous data, the average age at diagnosis is presented, but this average age has also been split by further subgroups to add further value to this measure. Figure 13 on page 28 shows the suspicion and confirmation ages of children notified in each calendar year. For consistency with previous data, this figure excludes 2010 cases where the hearing loss is unilateral, acquired or mild and those where the children are born overseas.

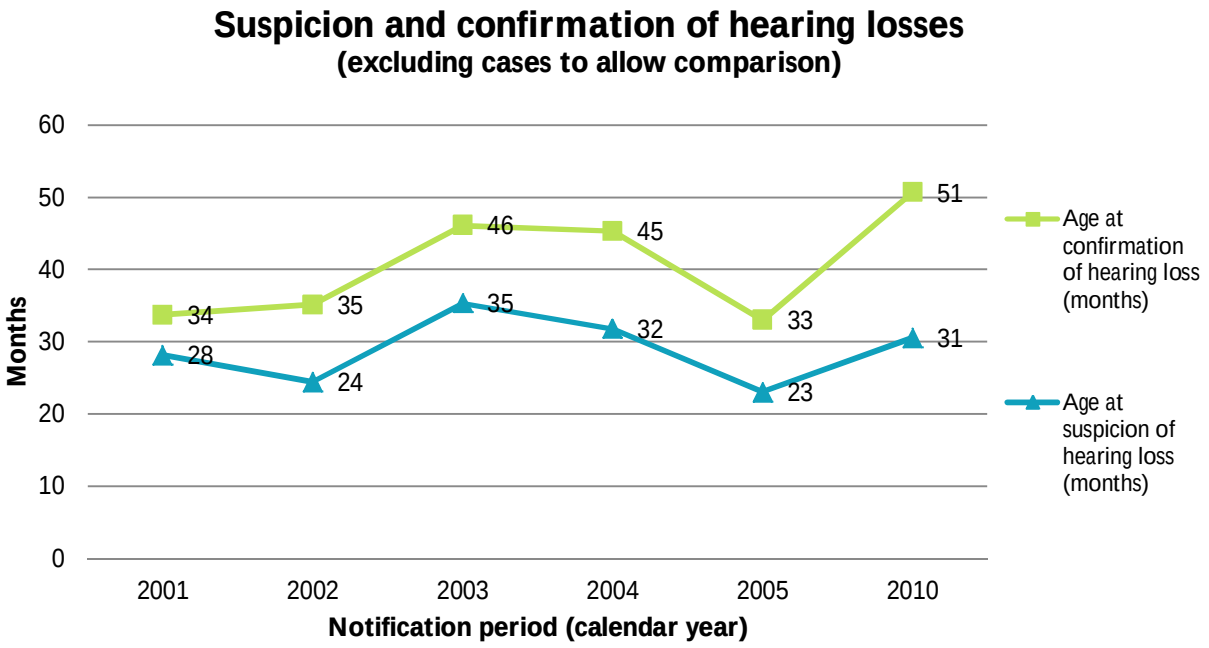


FIGURE 13: SUSPICION AND CONFIRMATION OF HEARING LOSSES - COMPARISON WITH PREVIOUS DATA (EXCLUDING APPROPRIATE CASES)

A number of groups are having their hearing losses identified later, including those with:

- unilateral hearing losses are, on average confirmed at 82 months of age compared to an average of 62 for bilateral hearing losses
- acquired hearing losses e.g. late onset, progressive and trauma related (76 month average)
- born overseas (95 months on average)
- mild and moderate hearing losses (89 and 71 months respectively)

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2010)
Mild	89
Moderate	71
Moderately severe	47
Severe	29
Profound	46

TABLE 10: AGE AT DIAGNOSIS BY DEGREE OF HEARING LOSS (2010)

Figure 14, on page 29, shows the spread of ages at diagnosis for each of these groups. The greatest variability in the age at diagnosis is for mild and moderate hearing losses, although there are a number of very late diagnoses for children and young people with profound hearing loss. Figure 15 also shows the variability within the age at diagnosis, this time by ethnic group. No MELAA group data is contained in this graph as there are only two cases within this sample.

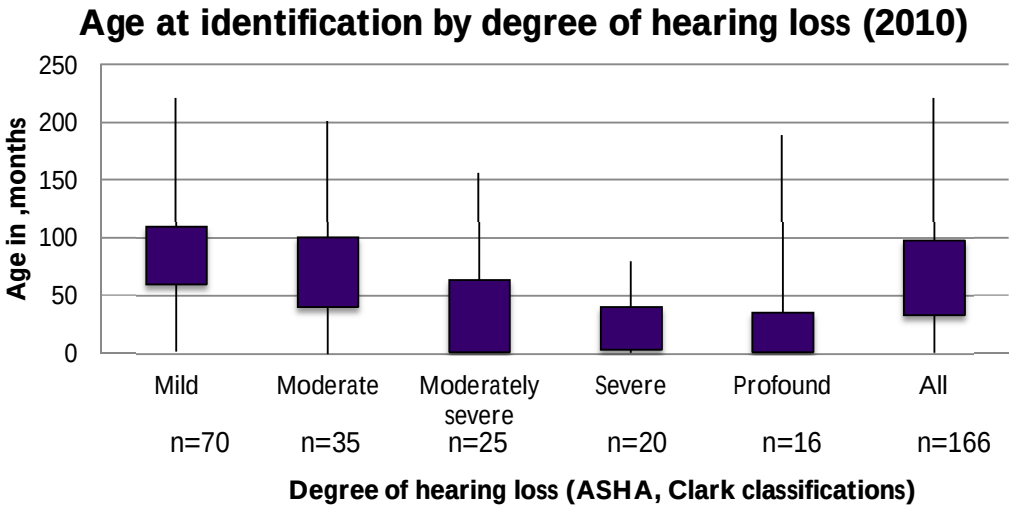


FIGURE 14: AGE AT CONFIRMATION BY DEGREE OF HEARING LOSS (2010)

Figure 15, below, shows the spread of ages of identification by ethnic group for those children and young people born in New Zealand.

A number of previous DND reports noted that Māori and/or Pacific children were identified later than European children although this difference was not reported in each DND report[27]. Milder hearing losses were more prevalent among Māori children in those years and the difficulties in identifying these losses may have resulted in later average ages of identification. It will be interesting to see whether 2011 data shows a difference between average ages of notification by ethnicity and whether a different severity profile is found with Maori having a greater proportion of milder hearing losses than other groups.

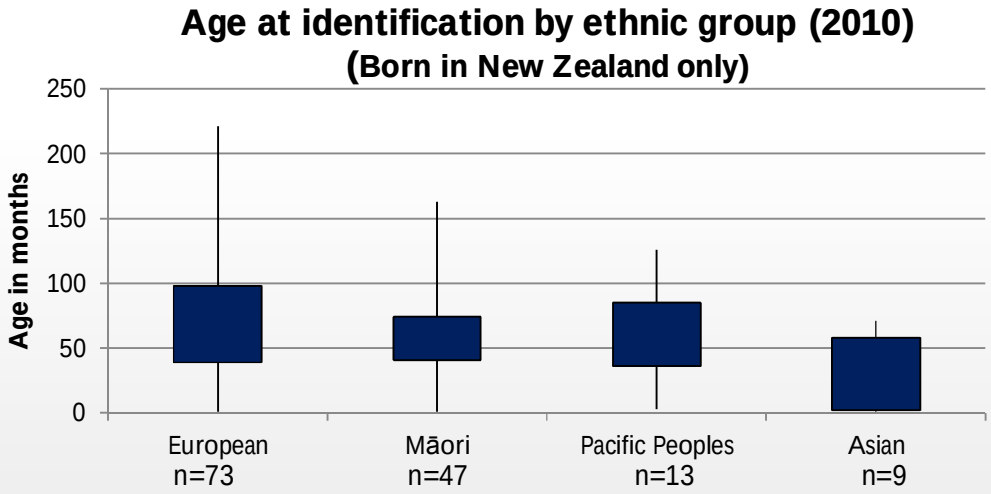


FIGURE 15: AGE AT IDENTIFICATION BY ETHNIC GROUP (2010)

[27] For example, the 1997 DND report noted a similar age of identification between Māori and non-Māori while the 2002 – 2004 reports noted a difference with European children being identified on average, earlier than Maori and Pacific children.

Introduction of newborn hearing screening

Implementation of New Zealand's Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP) began in 2007 and district health boards were included within the rollout in three stages.

From July 2007 the first stage DHBs, which were already screening newborns under their regional programmes were included under the auspices of the national programme (Waikato, Tairawhiti and Hawke's Bay). The second stage DHBs; Whanganui, Hutt Valley and Capital and Coast, began screening in June 2009 and the remaining DHBs (Northland, Waitemata, Auckland, Counties Manukau, Wairarapa, Nelson/Marlborough, Otago/Southland and West Coast) began screening at various times between July 2009 and June 2010.

It is worth noting that the large Auckland DHBs (Counties Manukau, Waitakere and Auckland) had all begun their screening programmes by April 2010 and Northland and Southern DHBs were the last to begin screening in July 2010.

Referrals from the UNHSEIP

Overseas, a number of comparable newborn hearing screening programmes (such as those in the UK and Australia) seem to be converging at a birth prevalence of approximately 1.10 per thousand for bilateral hearing losses, and approximately an additional 0.5 per thousand unilateral hearing losses. This suggests that if and when the UNHSEIP achieves high coverage and low loss to follow up we may expect up to 90 diagnoses directly from the programme each year, based on 62,000 births per annum[28].

A total of 27 of the 2010 notifications related to children born in New Zealand in 2008, 2009 or 2010 who were diagnosed as a direct result of newborn hearing screening. It is not known how many cases of hearing loss are missed as these children were either not screened by the UNHSEIP or are being lost to follow-up.

While some programme staff are reporting the number of diagnoses resulting from newborn hearing screening is smaller than they expected, very little can be inferred from 2010 notification data, primarily as the:

- completeness of the DNDs dataset for 2010 is not known;
- coverage rates at district health board UNHS programmes have not been reported; and as
- number of babies screened, particularly within specific DHBs is still relatively small and the number of diagnoses may be 'lumpy' as a result.

Key screening goals – age at diagnosis

The UNHSEIP was implemented in New Zealand to reduce the age of intervention for children born with hearing loss, as this approach has been successful overseas in improving outcomes. Screening programmes achieve this by significantly reducing the age at diagnosis for hearing losses present at birth compared with approaches reliant on risk factors. Key aims of newborn screening programmes include the screening of children by one month of age, diagnosis of hearing loss by three months and the start of intervention by six months of age. These are known as the 1-3-6 goals, and are commonly used in newborn hearing screening programmes internationally.

Measuring the proportion of children with hearing losses identified before the benchmark of three months of age as a result of a referral from newborn hearing screening will be an important measure

[28] This is an approximation of the number of births reported in 2010.

of the success of the newborn hearing screening in future. The Deafness Notification Database will provide useful data to show how the overall age at identification changes over time.

27 babies born in 2008, 2009 and 2010 were reported to the database as being diagnosed as a direct referral from newborn hearing screening diagnosed on average at 3.9 months of age.

17 of these babies were identified before the three month goal. Three of these cases were identified much later, at 18, 19 and 21 months of age. Reasons were given for the delay in diagnosis for two of these cases. One was attributed the audiologist having difficulties getting a confirmed diagnosis, parents not bringing the child to appointments and a transfer between DHBs while the other was attributed to the audiologist having difficulties getting a confirmed diagnosis and parents not bringing the child to appointments.

Other key points related to referrals and UNHS include:

- Six children diagnosed with a hearing loss in 2010 were not screened as screening was not offered in their area at the time of the child's birth
- In three cases it was unclear whether screening had been offered by the family
- One child was living in an area where screening was offered but was not screened as the baby was unwell. A decision was made by audiology, SCBU and screening staff that a direct referral should be made to audiology, given the number of risk factors involved, and the diagnoses resulted from this referral
- A total of four diagnoses were reported as delayed according to the audiologist as a result of transfers from one area to another (two of these were those cases identified between 18 and 21 months – previously mentioned)
- One child was referred directly from the screening programme to audiology due to atresia.

Identification of false negatives

The DND provides probably the only method for identifying potential false negatives from the newborn hearing screening programme.

Two children diagnosed with hearing loss in 2010 had been screened previously (under regional screening programmes) and passed this screening programme. As these children were born in 2003 and 2005 this leaves a significant time period between screening and diagnosis for progressive or acquired hearing losses to present.

Delays in diagnosing hearing loss

Not all cases within the database included the age at which the hearing loss was first suspected. This question has now been made compulsory within the online notification form.

The average delay between first suspicion of the hearing loss and confirmation of the loss, for all cases, *including* those born overseas and those with acquired hearing losses, was 20 months. This represents a significant average delay between first suspicion of a hearing loss and confirmation of this loss among children notified to the database.

Comparisons with the length of delay in previous years requires the removal of cases of acquired hearing loss, those born overseas, those with unilateral hearing loss and those with mild hearing losses. When this is completed, the average age drops slightly to 19 months as shown in Table 11, below.

Year	Age in months at confirmation of hearing loss	Delay in months
2010	50	19
2005	33	10
2004	45	14
2003	46	11
2002	35	11
2001	34	7

TABLE 11: DELAY COMPARISONS WITH PREVIOUS DATA (EXCLUDING ACQUIRED, UNILATERAL, BORN OVERSEAS AND MILD LOSSES)

Children with mild or moderate hearing losses are more likely to experience a long delay (24 and 22 months respectively) when compared with other degrees of hearing loss with severe and profound hearing losses (7 and 14 months respectively).

European children notified to the database for 2010 have the longest average delay between suspicion and confirmation at 25 months, followed by Pacific Peoples and Māori and 16 and 15 months. Please note – not all cases contained age at first suspicion data so these samples are particularly small for MELAA and Asian children and young people.

Ethnic group	Average months between suspicion and confirmation
European	25
Maori	15
Pacific Peoples	16
MELAA	7
Asian	6

TABLE 12: AVERAGE TIME BETWEEN SUSPICION AND CONFIRMATION BY ETHNICITY (THOSE BORN IN NZ AND NOTIFIED IN 2010)

Some previous notification reports have calculated the proportion of cases with a significant delay of six months or more between first suspicion and confirmation of the hearing loss. As the number of newborns identified with hearing loss grows, and as the goal for identifying these losses is before 3 months of age, applying this six month threshold for determining whether a delay exists no longer seems appropriate.

Delay causes

The notification form also requests information on the reasons for a delay between suspicion of a hearing loss and confirmation of the loss through diagnosis.

The most frequently mentioned cause of delay related to difficulties the audiologist had getting a confirmed diagnosis. This was followed by delays as a result of parents not attending appointments, difficulties the family had getting a referral to audiology and waiting times to see hearing professionals.

Reasons for delay	Number of cases where option selected
Audiologist had difficulties getting a confirmed diagnosis (e.g. child unwell)	28
Parents did not attend appointments (for any reason)	12
Difficulty getting a referral to audiology (e.g. GP or other health professional dismissed parent concern and no referral was made)	8
Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)	11

TABLE 13: REASONS FOR DELAY BETWEEN SUSPICION AND CONFIRMATION OF HEARING LOSS

Further information regarding causes of delays

There were a number of themes underlying comments made about the cause of delays, including those grouped by the categories in Table 13:

- As seen in the table, audiologists commonly reported difficulties obtaining a confirmed diagnosis. In two cases these delays were reported to be the result of conductive overlay/ORL referral or pathway delays, and this area was also mentioned 7 times in the 'other' category.

  One child was reported to have had a delayed diagnosis as a result of being under the care of an Ear Nose and Throat Specialist for 10 sets of grommets.

- Some audiologists notifying cases in which delays were recorded as being the result of parents not attending appointments provided further information about the cause of these delay:

  "Family had transport problems and lives in [an area] approximately 3 hours away."

  "Parents declined ABR and VRA appointments. Moved without forwarding address and were out of contact for 2 years. Failed to attend 4 appointments in 2010 prior to diagnosis."

- Some delays seemed to be the result of issues with systems and processes, including those which ensure a child whose family moves between DHB areas is tracked and follow up appointments are scheduled:

  "Audiology appointment requested following grommets but appointment was not made. Lost in system."

  I think [child] has a progressive hearing loss similar to her Father's. It appears to have started sometime in 2009. Previously she had been seen for hearing tests up until 2004 which showed normal hearing.

Annual follow-up did not take place for some reason, so [child]
was only seen when she and her mother really began to notice her hearing loss.

"This child was not screened as she was transferred to [another hospital].
She was then inpatient for a long time and did not attend appointments
however Audiology was unaware of this.
Ward staff noticed that [the child] was not talking and got mum
to come and arrange a hearing test."

- Delays were reported in three cases to have been caused by delays seeing non-hearing professionals such as anaesthetists whose services are required when ABR is conducted under a general anaesthetic
- In one case a parent was reassured wrongly about their child's hearing status:

"Teachers at preschool advised mother not to worry
about her poor speech back in Oct 2009."

2009 Notifications to the database

Tables below provide some very basic information on those retrospective notifications from 2009 which were collected through the re-launched DND during 2010. 91 cases from 2009 met the inclusion criteria and are described below:

- 86% of children and young people notified were born in New Zealand, with 5% born overseas and uncertainty about the birthplace of the remaining 9%
- The average age at confirmation for the complete sample was 57 months
- Parents and caregivers were the most likely to identify a hearing loss

Ethnic group (grouped total responses)	Number of cases 2009 DND[29]	Percentage of cases associated with each ethnic group
European	53	54%
Māori	25	26%
Pacific Peoples	12	12%
Asian	7	7%
Middle Eastern / Latin American / African[5]	1	1%

Degree of hearing loss (ASHA Clarke)	Number of cases
mild	35
moderate	23
moderately severe	18
severe	5
profound	10

[29] Please note due to the methods used the sum of the number of cases in each ethnicity does not equate to the total number of notifications

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