Deafness Notification Report (2015)

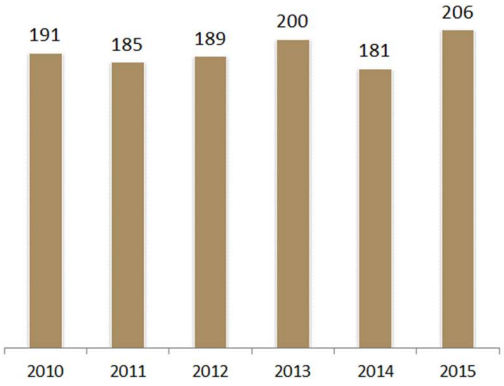
Notified cases of hearing loss (not remediable by grommets) among New Zealanders under the age of 19

Janet Digby, Levare Limited. August 2016

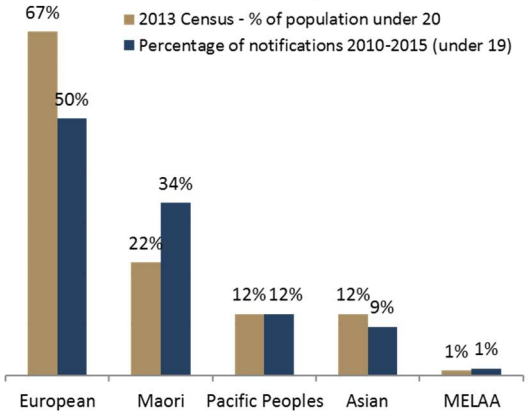
accessable
ENVIRONMENTAL HEALTH MANAGEMENT SERVICES

2010-2015 Notifications: A summary

Number of DHBs/Notifications



Ethnicity

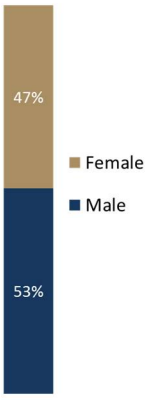


More likely to be identified later
- born overseas
- mild hearing losses
- acquired hearing losses
- unilateral hearing losses
More likely to be identified earlier
- born in New Zealand
- profound hearing loss
- losses thought to have been present at birth
- bilateral hearing losses

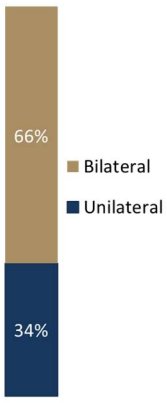
Most common reasons for delay

Audiologist had difficulties getting a confirmed diagnosis
Parents did not attend appointments
Waiting time to see hearing professional
Parents suspected something other than hearing loss
Difficulty getting a referral to audiology

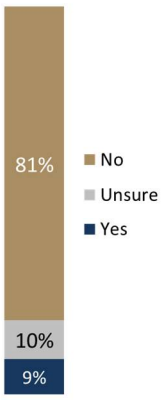
Gender



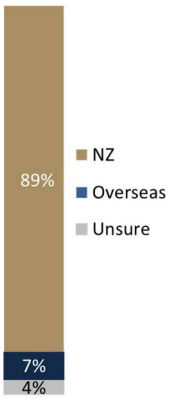
Bilateral/Unilateral



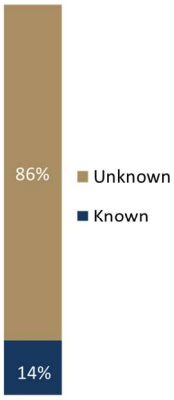
Additional disabilities



Birthplace



Aetiology



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

Making notifications to the database

The authors of this report would like to extend a big thank you to all those who have provided notifications to the database for the 2015 calendar year. We understand you are not compelled to provide this information and we know how busy you are. Thank you for contributing to our understanding of permanent hearing loss among New Zealand's children and young people.

Audiologists (including non-NZAS members) are strongly encouraged to make future notifications to the database by following this link. Audiometrists can also make notifications for newly diagnosed cases over the age of 16 years of age.

Notes for those completing notifications for the database:

1. **Consent changes:** We are delighted that, with the change of protocol in the Universal Newborn Hearing Screening and Early Intervention Programme (UNSHEIP) (and change of scripts and information provided by the screeners to babies' parent/caregivers), all babies screened by the UNSHEIP are now legally consented for entry into the Deafness Notification Database (DND), and there is no need to get the families to sign a separate consent form.

   Our thanks to Dr. Andrea Kelly (Auckland District Health Board) and to Moira McLeod (National Screening Unit) for their work on this issue.

2. **Shorter notification form:** The family history questions in the database have now been simplified and are much shorter – this will bring these questions into line with similar questions asked in overseas jurisdictions and should make the notification form easier to complete.

3. **New PDF version of form:** The new PDF version of the notification form is aligned more closely with the online form, so you can complete it and ask an administrator to enter the details into the database. It is also compact so you can print on one sheet (double sided) for your records. The new form is also available on the NZAS website.

4. **Send us your notifications as soon as possible following diagnosis:** We strongly encourage those making notifications to the database to get these in as soon as possible following diagnosis, and always before the end of the notification period in mid-March of the following year.

   This ensures these reports contain accurate information about those children notified during each calendar year. We understand that, with cases diagnosed late in the year, not all families may have consented to provide information about their child or young person to the database – this is why the deadline for notifications was extended to mid-March.

   Written consent from families is still required in the future for children whose hearing loss was not identified through newborn hearing screening in New Zealand.

5. **If you have any questions at all, please contact Janet Digby:** janet@levare.co.nz or by telephone (09) 445-6006. If in doubt about whether a case meets the criteria, please notify the DND about the case.

Contents

The 2015 Report

Introduction

Welcome to the sixth annual report describing notifications to the re-launched Deafness Notification Database (DND). This report includes diagnoses made throughout New Zealand during the 2015 calendar year.

Since the DND was re-launched in 2010, the following definition has been used to determine which cases are included in the DND, and therefore in the analysis for this report:

> Children and young people 18 years or younger, born in New Zealand or overseas, with:
> - a permanent hearing loss in one or both ears[i],
> - an average loss of 26 dB HL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0 kHz)[ii].

Historical information about the database's inclusion criteria can be found in Appendix A: History of the database, on page 45 of this report.

Steps have been taken to ensure data contained in this report are comparable with previous deafness notification data. However, 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 as a result a number of longitudinal comparisons are not possible.

Please note that, unless otherwise specified, analyses contained in this report describe characteristics of the full number of 2015 notifications for which data were provided.

A glossary of commonly used terms can be found on page 50 of this report.

Completeness of notifications

While every reasonable effort has been made to ensure the newly re-launched database improves understanding of permanent hearing losses among New Zealand children and young people, there is no way to know how many new cases which meet the criteria are *not* notified to the database.

There may be certain types of cases that are under-represented within notifications, and as a result inferences made from the data contained in this report should be taken as indicative only unless stated otherwise (see the beginning on page 8 for further information).

For example, hearing losses among Māori are more likely to be underrepresented in the DND as disparities in access to, and within, the health system exists for this group[1,2,3,4].

Despite these limitations, we can use a number of methods to provide some indication of the number of new diagnoses of hearing loss annually, among children and young people. Based on these analyses, it is likely that the database has been receiving notifications for between 50% and 70% of all cases diagnosed each year, since 2010. Further detail can be found in Appendix D which begins on page 48.

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[i] The original criteria for the database, which applied to notifications until the end of 2005, required the hearing loss to meet the audiometric criteria in both ears and for the child or young person to have been born in New Zealand. When the database was restarted in 2010, the criteria were broadened to include children with hearing loss in *one or both ears* and those born *outside* New Zealand.

[ii] Based on feedback from the audiological community, high frequency hearing losses that would not meet the original criteria, but that would exceed the 26 dB HL average based on audiometric data from 2.0, 4.0, 6.0 and 8.0 kHz, have been collected from July 2011. We will continue to include this special group within the database. A limited analysis of data from high frequency hearing losses notified in 2014 can be found in

Appendix B: High frequency hearing losses, on page 48.

As time passes, we hope that further efforts can be made to increase the proportion of notifications received, improving the ability of the database to inform the Ministry of Health, Ministry of Education, clinicians and other service providers, about the number and nature of new diagnoses of hearing loss among New Zealand children and young people.

Acknowledgements

Thank you to the 206 families who consented to share details about their child's hearing loss. As a result of their willingness to share basic information about their child's diagnosis, service providers will be better informed about current and future demand for services, including what skills are required, in various workforces, to better serve the needs of families.

The time taken by professionals around the country to make notifications is also very much appreciated, as are efforts of those who have completed the analysis for reports prior to 2006, which had unique challenges.

This report has been funded by **accessable**, through a contract with the Ministry of Health. We would like to thank the Ministry of Health for funding the database from 2012. Without this support, people working with children who are deaf or hearing impaired would not have up to date information to help them better understand the nature of new diagnoses in New Zealand.

The primary author of this report gratefully acknowledges the significant support and guidance of Professor Suzanne Purdy of the University of Auckland and Dr Andrea Kelly of Auckland District Health Board. Their input into these reports is significant and greatly appreciated.

Dr David Welch, Mr Colin Brown and Professor Peter Thorne are also acknowledged for their contributions to, and interest in, the DND over the years.

Contact details

The authors of the report hope that ongoing changes made to the way information is analysed and presented will improve the value of these reports over time. We ask that readers get in touch to provide us with feedback to help guide the development of future reports.

Feedback on this report and any questions about the DND should be directed to its primary author, Janet Digby. Janet can be contacted at: janet@levare.co.nz or by telephone, (09) 445-6006.

Notifications

General information

Two hundred and six notifications pertaining to cases first diagnosed during the 2015 calendar year, and meeting the criteria for inclusion, were received by 15 March 2016, this year's cut off for 2015 notifications[i, ii]. These notifications were received from a total of 58 audiologists, with notifications from 17 of the 20 district health boards (DHBs). This number is up on the 181 notifications received in 2014, which were received from 16 district health boards and 54 audiologists.

The increase this year in the number of notifications is likely the result of changes in consenting processes, allowing automatic consent for children screened by the UNHSEIP for the DND, and also automatic referral of audiologists completing a first hearing aid application to the online notification form. Our thanks to Dr Andrea Kelly and the National Screening Unit for work on aligning consenting processes, and to **accessable** for their work to direct audiologists to complete a notification form for children and young people receiving their first hearing aid(s).

An additional 21 high frequency hearing losses were also notified to the database for the 2015 year. These are described in Appendix B on page 47. (A significant number of notified cases that were listed by audiologists at the time of notification as 'high frequency losses' met the criteria for the main database, and so were not included in this special category.)

Among those children and young people whose hearing loss was notified to the database, notifications peaked at the end of the notification period (November to December), with a smaller peak in August. This may be the result of the general shortage of audiologists nationwide and the timing of their holidays, or owing to other reporting pressures, which are considerable.

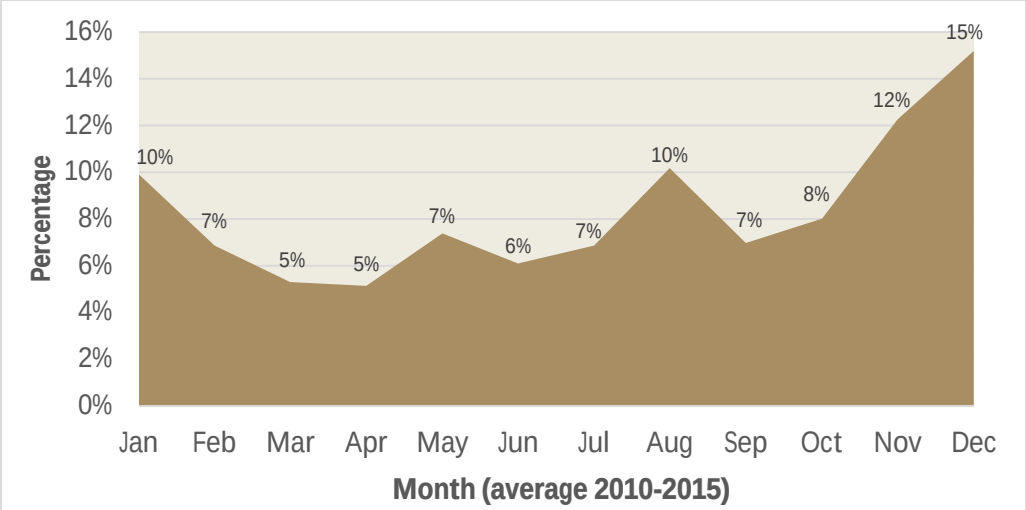

FIGURE 1: STACKED AREA CHART SHOWING AVERAGE NOTIFICATIONS BY MONTH (2010-2015)

[i] 13 March 2015 was the deadline for notifications for the 2014 calendar year. Reports prior to 2006 contained information about diagnoses notified in a given year, rather than diagnosed within that year. As a result, the number of notifications varied, increasing in years where there were greater efforts made to encourage audiologists to send in notifications. For example, in 2004 there were an additional 288 retrospective notifications received from the Children's Hearing Aid Fund (CHAF) audit.

[ii] It is not possible 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 less than five minutes to enter using the online form, as was the case in previous years. The removal of the extended family history questions in the middle of 2014, to align the form with developing best practice internationally, has reduced the time it takes for audiologists to collect notification information and submit cases.

Number of notifications

Notifications are collected through an online survey form to reduce data entry errors and make it as easy as possible to notify cases. Efforts have been made to publicise the database to all audiologists working with children and young people, in an attempt to collect as many notifications as possible.

Please note the following points regarding longitudinal data from the DND:

- notifications have been reported based on calendar years throughout the period of operation of the database, i.e. 1982-2005 and 2010-2015;
- the period from 1982 to 2005 contains notifications to the original National Audiology Centre/ Auckland District Health Board (ADHB) administered database;
- no data are provided for 2006 to 2009 as the database was not operating during this period; and
- data for 2010 to 2015 relate to notifications provided to the newly re-launched database.

Figure 2 shows the number of notifications meeting the criteria for the main dataset, in each year[i]. Information about how the inclusion criteria have changed over time is included in Appendix A which begins on page 45.

Please note that the 2001 to 2005 figures, included in previous reports, were later revised by the database's contracted provider at the time, ADHB. Figure 2 shows the number of notifications which met all inclusion criteria at the time and were included in the database's annual reports[ii] rather than the revised figures. Figure 2 illustrates the variability in the number of valid notifications provided to the original database, particularly in the last six years of its operation[iii].

2010-2014 data have also been revised slightly from previously reported figures, as further information about existing notifications is received, and as new retrospective notifications are provided to the database. Some of these changes have been described in greater detail in previous reports.

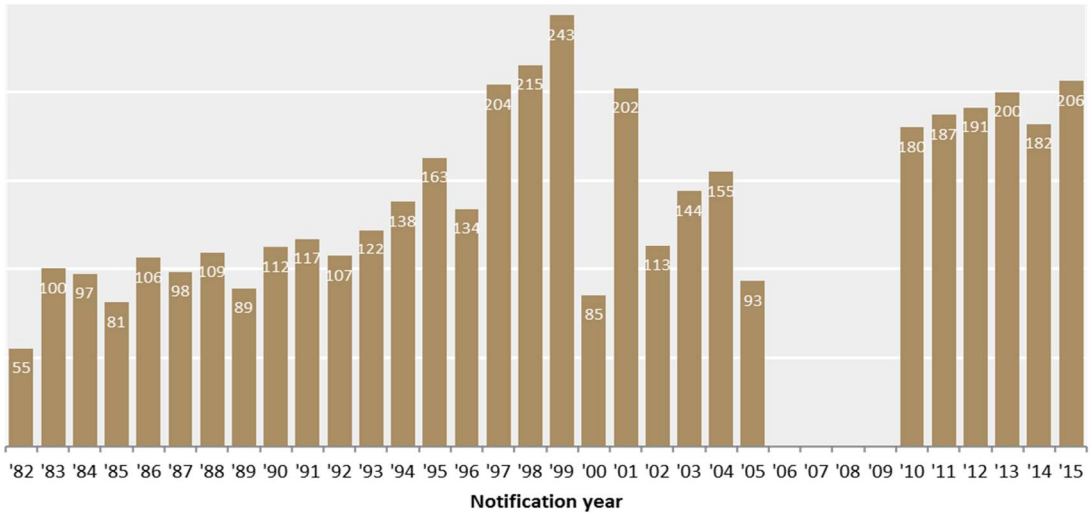

FIGURE 2: NOTIFICATIONS BY YEAR 1982-2005 AND 2010-2015

The following types of notifications are *not* accepted into the dataset based on the current inclusion criteria:

- slight losses (those not meeting the 26 dB HL average across four frequencies in at least one ear)[iv];

[i] High frequency hearing losses, which have been collected since July 2011, are not included in this main dataset.

[ii] Previous figures were sometimes split by those that were removed for audiometric or other reasons. The figures now show the total number of notifications that met criteria for inclusion, which were in place at that time.

[iii] Greville completed an analysis of the data in 2005, and noted that data reported in previous reports contained a number of duplicates, presumably from previous year's notifications; these have been removed for this analysis.

[iv] High frequency hearing losses are included separately in Appendix B.

- cases where the child or young person was reported as having mild hearing loss with normal bone conduction thresholds[i] (assumed to be a transient conductive hearing loss unless a permanent conductive hearing loss was specifically stated, e.g. due to ossicular fixation);
- notifications with significant missing information where no further information was provided on request; and
- notifications that didn't report that consent had been provided by the parent/caregiver.

Gender

Slightly more of 2015's notified cases to the database were male (52%) than female (48%), similar to the 2010-2015 average, in which males account for 53% of notifications and females 47%.

With the exception of 2014 notifications, DND figures are aligned with overseas research; boys are commonly found to have higher rates of hearing loss than girls. These figures range between 52% and 58% for males (1:1.08 and 1:1.38) in various jurisdictions, as reported in the 2011 Comprehensive Handbook of Pediatric Audiology[5].

Australian Hearing's data shows a similar pattern[6], with higher rates of hearing loss among males under the age of 21 who have hearing aids or cochlear implants[ii]. In this report 52.6% of children are male while 47.4% are female[iii]. This pattern is seen in all states, with the exception of South Australia and ACT, in which the ratios of male to females is almost 1:1 and in those aged 21-25 years of age, where 48.4% of those with hearing impairment were male.

An analysis was conducted in 2013 to find any differences in our database between the severity profile, type and distribution of hearing loss by gender. This analysis did not discover any gender differences in severity profile or type of hearing loss.

Birthplace

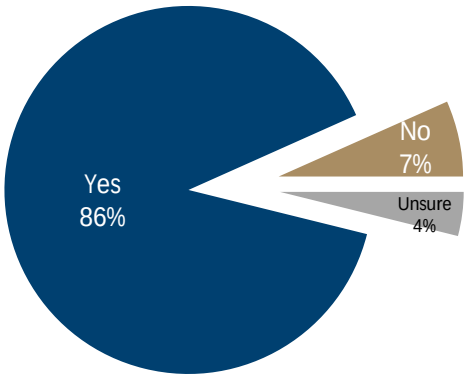

FIGURE 3: PROPORTION OF 2015 CASES BORN IN NEW ZEALAND

This is the sixth year in which children and young people born outside of New Zealand have been formally included in the database and its main analysis.

As shown in Figure 3, of the 206 notifications to the database in 2015, 7% were known to be born outside New Zealand. Birthplace was uncertain in a further 4% of cases reported to the database.

Between 3% and 9% of cases in the database were known to be born outside New Zealand between 2010 and 2015.

[i] Hearing losses meeting the criteria listed on page 6 were included within the dataset. This included a number of cases of permanent conductive loss.

[ii] This source reports on children and young people, under the age of 26 who received services from Australian Hearing in 2014.

[iii] 0.1% of cases were of unknown gender.

DHB representation

Table 1 contains the percentage of 2015 notifications from each DHB area and compares these with the percentage of the population under the age of 20 from the 2013 Census[i].

In addition to natural fluctuations in the number of hearing losses diagnosed among children and young people in a given year, other factors influencing notification levels are likely to include:

- the size of the population within the age range for the database;
- the prevalence of hearing losses within that population;
- the date the child or young person was diagnosed, and whether it is appropriate to ask for consent for the database at the time of diagnosis, or whether this is best done at a later appointment which may be after the cut-off date for notifications;
- the number of hearing professionals employed by each DHB;
- the workload of these hearing professionals; and
- the level of commitment among staff to making notifications to the database.

It is worth noting that, historically, many clinicians believe there is a preponderance of deafness in Auckland and Christchurch as many families have traditionally moved to these places from the regions so their children could be schooled at Kelston Deaf Education Centre (KDEC) (Auckland) or van Asch Deaf Education Centre (VADEC) (Christchurch).

In addition, it is interesting to note that some of the DHBs reporting higher numbers of notifications than anticipated based on their population, are often those DHBs with a higher proportion of Māori and/or Pacific populations (e.g. Tairawhiti, Waikato, Hutt, Northland, Bay of Plenty).

	Percentage of notifications received 2015 (under 19 years)	Percentage of population under the age of 20 (Statistics New Zealand, 2013 Census)
Auckland	6%	11%
Bay of Plenty	7%	4%
Canterbury	11%	11%
Capital and Coast	10%	7%
Counties Manukau	10%	13%
Hawke's Bay	2%	3%
Hutt	4%	3%
Lakes	0%	2%
Midcentral	3%	4%
Nelson Marlborough	4%	3%
Northland	5%	3%
South Canterbury	2%	1%
Southern	3%	7%
Tairawhiti	2%	1%
Taranaki	7%	2%
Waikato	11%	9%
Wairarapa	0%	1%
Waitemata	10%	13%
West Coast	0%	1%
Whanganui	2%	1%

TABLE 1: PERCENTAGE OF NOTIFICATIONS (2015) COMPARED WITH THE ESTIMATED PERCENTAGE OF POPULATION UNDER 20 YEARS (2013 CENSUS) OF AGE BY DISTRICT HEALTH BOARD

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

DHBs underrepresented in the 2015 data include:

- Auckland and Waitemata[i] DHBs – notifications are lower than would be expected due to consenting issues with previous cases. The number of notifications for children and young people living in these DHBs, and diagnosed in 2015, has risen this year due to changes in consenting processes which applied from the middle of the reporting year, allowing children screened by the UNHSEIP to automatically be consented for the DND.
- Wairarapa, Lakes and West Coast – these are relatively small DHBs, and may not have diagnosed any children or young people during the 2015 year;

Other disabilities

The presence of one or more additional disabilities can have a significant impact on both outcomes for children and young people with hearing loss, and also on the level of support they may require, particularly from special education services.

Cupples *et al.* (2009) found that there were differences in outcomes achieved by the 119 children included in the study based on the type of additional disability. Children with autism, cerebral palsy, and/or developmental delay showed poorer outcomes compared with children who had vision or speech output impairments, syndromes not entailing developmental delay, or medical disorders[7].

Children with hearing loss are thought to have a high rate of additional disabilities because many risk factors for hearing loss also involve other developmental conditions. Rates of additional disabilities among children with hearing loss are particularly high among those who have a syndrome.

Of 2015 notifications, 9% were thought to have disabilities *in addition* to hearing loss at the time the notification was made. This is lower than the proportion reported for any previous year. In a further 10% of 2015 of cases there was uncertainty regarding whether the child or young person had an additional disability[ii].

The most commonly reported conditions specified were those related to a specific syndrome (n=6), general or global developmental delays or intellectual disability (n=5) and vision problems[8, iii] (n=2).

The proportion of children notified with additional disabilities is not directly comparable to data reported prior to re-launch of the database in 2010, as an 'unsure' category has been added to allow for cases where an additional disability may be suspected but has not yet been confirmed. However, when the 'unsure' figure is added to the proportion of cases with an additional disability, as in Table 2 the figure is more consistent with those reported before the database's re-launch.

[i] Waitemata DHB's audiology for children is undertaken by audiologists at Auckland District Health Board

[ii] The proportion of New Zealand children with a hearing impairment (diagnosed at any time) who also have an additional disability which affects their learning is not known.

[iii] No local data is available on the rates of vision problems among deaf and hearing impaired populations in New Zealand, but some professionals recommend routine referral for ophthalmological assessment for children diagnosed with significant bilateral hearing impairment.

Notification Year	Proportion of cases with a known additional disability	Proportion of cases with a possible additional disability	Proportion of cases with additional disability (2002-2005). Total confirmed and possible (2010-2014)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	12%	10%	**22%**
2011	14%	5%	**19%**
2012	16%	11%	**27%**
2013	12%	12%	**24%**
2014	16%	8%	**23%**
2015	9%	10%	**19%**

TABLE 2: PROPORTION OF CASES WITH A KNOWN ADDITIONAL DISABILITY

Earlier identification of children with hearing loss is likely to result in lower levels of reported additional disabilities, which are reported at the time of diagnosis of the hearing loss. This is because children may have not yet been diagnosed with these conditions, or they have conditions that have not yet developed (e.g. diagnoses of autism spectrum disorder are typically not made in the first year of life and vision impairments are more common in older children).

Other possible reasons for the downward trend in the proportion of children reported with additional disabilities include:

- children with hearing loss in New Zealand may not be routinely assessed by a paediatrician, meaning additional disabilities may be under-diagnosed; and
- immunisation coverage in New Zealand has risen significantly since vaccination for children became a Primary Health Organisation (PHO) Performance Programme indicator in January 2006 (and a funded indicator from July, 2008). Achievement rates for the indicator 'age-appropriate immunisations completed by age two years' have doubled from approximately 45% in 2007 to 91% in September 2013[9]. Such improvements have reduced rates of meningitis in New Zealand and this may have an impact on the proportion of children with hearing loss with additional disabilities, although the numbers are likely to be small[i].

[i] It is difficult to compare the number of cases of meningitis over time as this information was not collected prior to the re-launch of the database in 2010 and as there is no specific question related to meningitis in the current database. Further information on meningitis cases can be found on page 17.

Overseas additional disability data

While it is difficult to compare reported rates of additional disabilities among hearing impaired children, as the definition for hearing loss and for disabilities differ and are not always described in journal papers, a selection of rates from various jurisdictions are described below. The first paper listed shows the huge variability in rates, presumably the result of definitional differences.

New Zealand DND figures are similar to Australian estimates of the proportion of deaf children with an additional educational need, although this is unlikely to be a fair comparison owing to differences in how additional disabilities are defined.

Source	Date	Location	Details	Rates
Ear Foundation for National Deaf Children's Society[10]	2012 (review date)	United Kingdom Review of international data	Review of 12 papers from 2002-2012 containing prevalence rates thought to be relevant to the UK, US, Australia, New Zealand	Most common additional disabilities: • visual impairment (4-57% depending on the definition) • neurodevelopmental disorders (2-14%) • speech language disorders (61-88%)
Fortnum *et a.l*[11]	2002	UK	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis[12]	1997	UK	Trent region study of permanent congenital hearing impairment	38.7% of children found to have one or more additional clinical or developmental problems, although this study used a wide definition of additional needs.
Holden-Pitt and Diaz[13]	1998	United States	60% of deaf and hearing impaired children in the United States in the 1996/97 year	20-40% of all US children with a hearing loss had an additional disability
LOCHI[14]	2013	Australia	Study examining 260 children in Australia born with hearing impairment	18% of children within their sample have one additional disability, 10% with two and 9% with three or more
The Consortium for Research into Deaf Education[15]	2011/12	UK	Annual national survey of educational staff	21% of deaf children (including unilateral and bilateral and mild to profound losses) had an additional special educational need in addition to their hearing impairment

TABLE 3: ADDITIONAL DISABILITIES, SELECTED OVERSEAS RATES FOR COMPARISON

Bilateral and unilateral loss

Background

Binaural hearing allows the auditory system to process and integrate input from both ears, making it necessary for improved understanding of speech in noisy situations and for sound localisation[16,17].

As a result of a series of studies in the early 1980s (in the United States) the significance of unilateral hearing losses was re-evaluated by professionals, who had often previously underestimated the significance of unilateral hearing loss in children[18,19,20].

Children with unilateral hearing losses are known have reduced educational performance, language delays and higher rates of behavioural issues, which are reported as significant in about a third of all cases, and a significant proportion of these hearing losses progress over time to become more severe or to affect both ears[21,22,23,24,25,26].

The Joint Committee on Infant Hearing (JCIH) noted in its 2007 statement that 'All families of infants with any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.'[27] This statement also recommended that developmental monitoring should also occur at regular six month intervals for those with permanent unilateral hearing loss because these children are at risk of having speech and language delay.

This statement now has a supplement (2013) which states:

"Goal 7: All Children Who Are Identified With Hearing Loss of Any Degree, Including Those With Unilateral or Slight Hearing Loss, Those With Auditory Neural Hearing Loss (Auditory Neuropathy), and Those With Progressive or Fluctuating Hearing Loss, Receive Appropriate Monitoring and Immediate Follow-up Intervention Services Where Appropriate." [28]

This supplement goes on to state that all children with unilateral or bilateral hearing loss should be referred to early intervention services for evaluation and consideration of enrolment. Most infants and children with bilateral hearing loss and many with unilateral hearing loss benefit from some form of personal amplification device[29].

In New Zealand, Project HIEDI recommended in 2010 that families of children with unilateral hearing loss be offered advisory services and that such children be regularly assessed to quickly determine if they are beginning to fall behind and to determine appropriate support[30].

To further investigate the impact of unilateral hearing loss on young children, The Children with Unilateral Hearing Loss (CUHL) study is being conducted by the National Acoustic Laboratories (NAL), Australia. There is currently no high quality evidence on how to best manage unilateral hearing loss in young children.

Inclusion in the DND

To reflect the importance of unilateral loss, cases where these average more than 26 dB HL in the hearing impaired ear[i], have been included in the DND since its re-launch in 2010.

Although unilateral hearing losses were not included in the DND before 2006, a number of these cases were notified to the database each year and these numbers were provided in the annual reports at that time. Comparing the proportion of unilateral/bilateral notifications with previous DND data (prior to 2005) is not possible because although a number were reported prior to 2006, these were not well reported and so are incomplete in this older dataset.

In DND reports between 2010 and 2014, the proportion of bilateral and unilateral losses was calculated based only on cases with full audiometric data and in 2014 also on those that could have data interpolated.

From now on, we will also present the proportion of bilateral and unilateral hearing losses based on cases with and without all data-points and also on interpolated figures[ii] using manual checks for those records that cannot have data interpolated. This means we can report on the number of ears affected by hearing loss in more cases within the database.

Bilateral and unilateral hearing losses:

- the proportion of 2010-2015 cases that were bilateral/unilateral using only cases with full audiometric data was 63:37, and this became 66:34 when cases without all data-points were included; and
- the proportion of 2010-2015 cases that were bilateral/unilateral *using interpolated data* and manual checks was 66:34 and this became 67:33 when cases without all data-points were included.

[i] Averaged over four frequencies – 0.5, 1.0, 2.0 and 4.0 kHz.

[ii] While only cases where all 8 audiometric data-points are present are able to be included in most severity calculations, interpolation of data has been used in some instances in this report to provide a more complete picture of the severity of hearing losses notified. Interpolation is only used where three of the four data-points are provided for one ear, and where both surrounding data-points are provided.

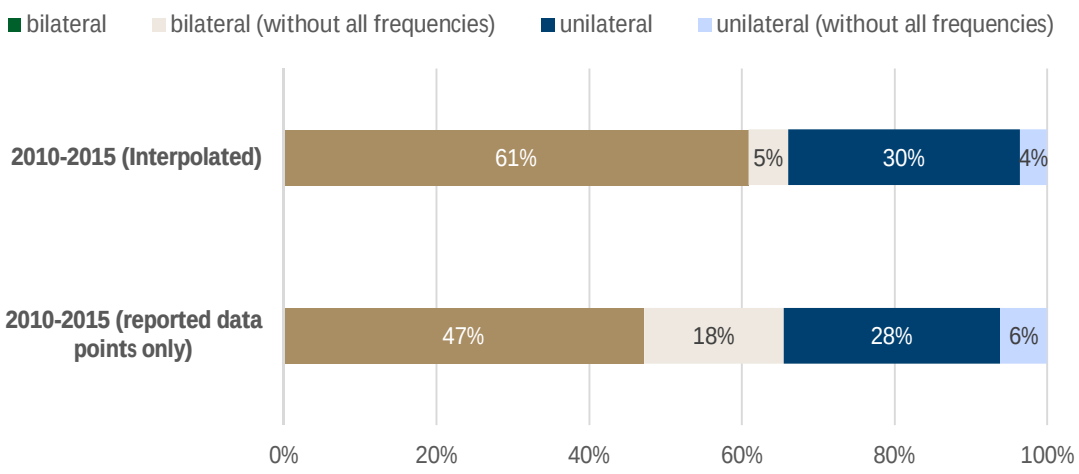

FIGURE 4: BILATERAL AND UNILATERAL CASES BY GROUPING (2010-2015)

While immunisation coverage (including for conditions such as mumps) in New Zealand has risen significantly from 45% in 2007 to 92% in 2012[31] there is no obvious reduction in the proportion of newly diagnosed unilateral hearing losses over time – perhaps not surprising given the number of cases of these conditions is likely to be very small.

Genetic or epigenetic factors are thought to play a role in some cases of unilateral hearing loss. Further research is required to establish the aetiological patterns of unilateral hearing loss[32].

Differences between the proportions of bilateral and unilateral notifications within each severity category are shown on page 36.

Single sided deafness

Severe or profound unilateral hearing loss is sometimes referred to as single-sided deafness (SSD). Unilateral hearing loss impairs the listening which is possible with two ears – including speech perception in noisy situations and the ability to localise sound[33].

A number of requests have been received in recent months, asking us to describe cases of SSD in this report. Different case definitions of SSD are used internationally; for example, some definitions include only those with severe loss in the worse ear and others only those with profound hearing loss[34,35]. The boundaries for these degrees of loss also differ depending on the jurisdiction. One reason for examining the proportion of unilateral losses which are SSD is that there are differences in the types of hearing technology that may benefit children with SSD (e.g. cochlear implant) versus less severe degrees of hearing loss (e.g. bone conduction hearing aid).

Cases of SSD within our data have been defined as children and young people in the main dataset who have a hearing loss of more than 70 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the worse ear, and a hearing loss of less than 26 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the better ear. These average thresholds have been chosen considering the ASHA codeframe for severity, and because 26 dB HL is the lower limit for average notifications to be accepted into the database and as a 70 dB HL[i] average is the boundary between moderately severe and severe losses.

The data contained in Table 4, below, show the proportion of notifications received in each notification period that meet the criteria (above) for SSD. These cases have been identified from data containing all threshold information in addition to those that have had one missing data-point completed by interpolation.

[i] This 70 dB HL average for the lower limit will eliminate most cases of atresia, as these are mostly conductive, and therefore not severe enough to meet this threshold criterion. Such children will benefit from a bone conduction hearing aid and are, as a result, a different group to those we wish to identify as having SSD.

Notification Year	Proportion of cases with single sided deafness (SSD)
2010	6%
2011	4%
2012	8%
2013	10%
2014	7%
2015	5%
Average 2010-2015	7%

TABLE 4 SINGLE SIDED DEAFNESS CASES BY YEAR

Types of hearing loss

Information on the types of hearing loss notified is now being collected for each hearing impaired ear[i]. Options provided were; 'sensorineural', 'mixed', 'permanent conductive', 'ANSD' (Auditory Neuropathy Spectrum Disorder), 'other' and 'don't know'. Please note that the ANSD group have sensorineural hearing losses; that is, this group is effectively a subgroup of the sensorineural category. Those notifying cases could also select normal hearing for the hearing ear in children and young people with unilateral hearing loss.

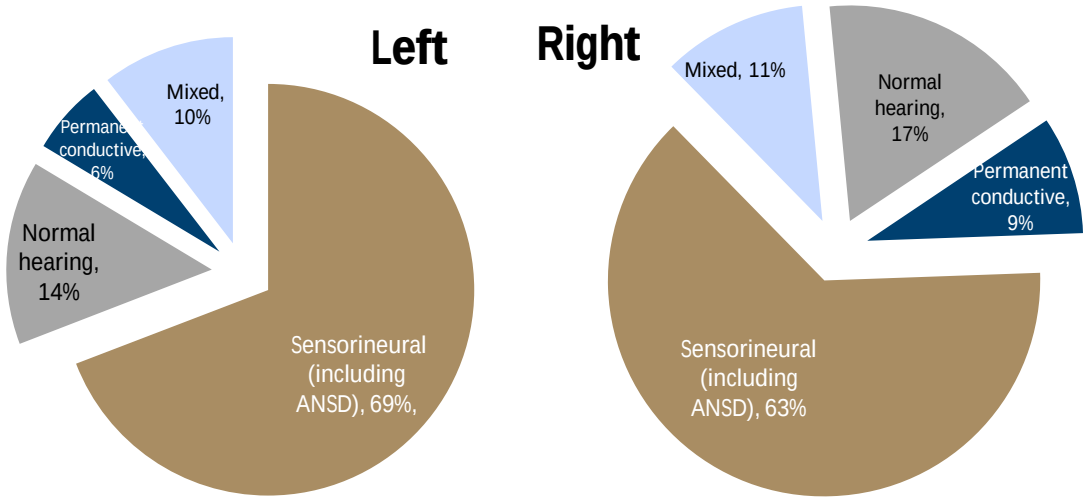

FIGURE 5: TYPE OF HEARING LOSS (2015)

The most commonly reported type of hearing loss contained in notifications to the DND that included information on the type of hearing loss, was sensorineural (63% in the right ear and 71% in the left), followed by mixed losses (10% in left ears and 11% in right ears) and then permanent conductive losses (6% in left ears and 9% in right ears – some of which were atresia). Three percent of right ears and 5% of left ears were recorded as ANSD.

Prevalence of ANSD among those with permanent hearing loss is likely to be 10%, according to a 2015 review by Rance [36].

[i] Part way through the 2013 year, we began asking audiologists "Bearing in mind the maximum thresholds of BC testing… Do you think it is most likely that this hearing loss is…", for each ear, to ascertain the type of hearing loss.

Family History

The DND reports prior to 2005 note that a relatively high proportion of cases recorded '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).

In 2010, when the database was re-launched, changes were made to this question to try to gain more specific responses about the nature of the family history.

Questions on this topic began with a general question asking whether there was a family history of hearing loss. More specific questions were then asked about whether the relative was a parent, sibling or grandparent, and then about each specific relative. Between 13% and 24% of cases reported a 'family history of hearing loss' between 2010 and 2013.

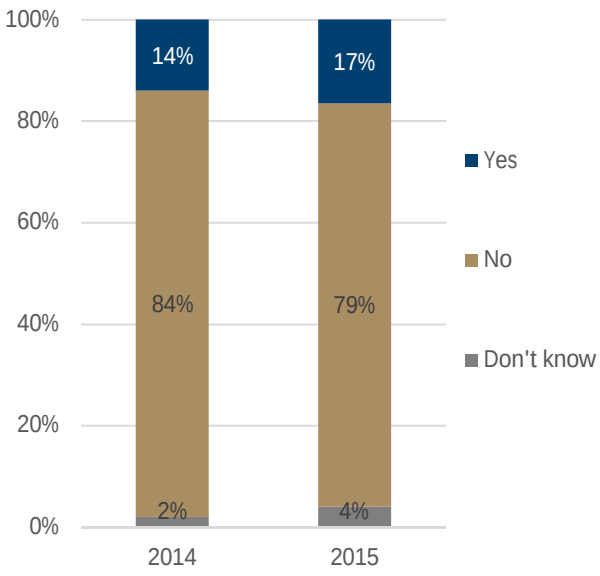

FIGURE 6: IMMEDIATE FAMILY MEMBER WITH HEARING LOSS (2014 AND 2015)

During 2014, the questions in this section of the notification form were changed, in part to make them easier to complete (this section was not well completed previously) and also to bring the questions into line with developing international practice.

The new question in the DND relating to family history is 'Does an immediate family member (only a mother, father or sibling) have a permanent hearing loss? (or had a permanent hearing loss if they have died).' The results for this question are shown in Figure 6.

Please keep in mind that data from 2014 contains information from approximately half the notifications for that period, as the question was changed in the middle of the year. 2015 data contains completed information based on the new question in the notification form.

Ethnicity

Representation

All but one of the 2015 notifications contained one or more ethnicity codes, which is pleasing. However, a smaller proportion of 2015 notifications received specified more than one ethnic group when compared with 2013 Census data. This may mean that coding for ethnicity is less complete for deafness notifications than for the Census data, and this may affect the validity of this comparison.

The majority of notifications provided to the database since its re-launch in 2010 relate to children and young people of New Zealand European and/or Māori[i] ethnicity.

The MELAA category included in this and other sections of this report relates to children and young people of Middle Eastern, Latin American or African ethnicity.

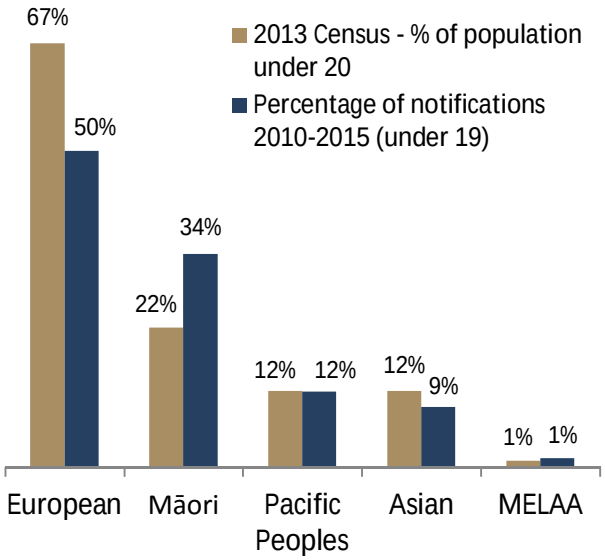

FIGURE 7: COMPARISON ETHNICITY IN NOTIFICATIONS (2010-2015 UNDER 19 YEARS OF AGE) TO CENSUS DATA (2013 – UNDER 20 YEARS OF AGE)

Multi-coded 2013 Census data was included in Figure 7 for the first time in the 2014 report. This figure shows the total response count (as individuals can identify with more than one ethnicity, the totals sum to more than 100%) for ethnicity from the 2013 Census (for those under the age of 20) and compares this to the ethnicity breakdown for deafness notifications from 2010-2015 (which includes those under the age of 19[ii]).

Ethnicity and prevalence

Compared to the general population, the proportion of notifications from those of European ethnicities are lower than one would expect based on the size of their population under 20 years, and notifications from those of Māori ethnicity are higher than expected.

Nevertheless, Māori may still be underrepresented in DND statistics because of their greater chance of having a less severe (mild or moderate) hearing loss, which is more likely to go unidentified. It may also be that disparities in 'access to, and within, the health system' for Māori may mean fewer cases are found or notified when compared with the New Zealand European population.

A number of sources support the difference in rates of hearing loss between Māori and New Zealand Europeans which is illustrated above:

- Reported diagnoses from the UNHSEIP which show that Māori have higher rates of hearing loss at diagnosis from this programme[37].
- The Household Disability Surveys (1991-2006[38]) – these surveys suggest Māori may have higher rates of hearing disability (children and adults) and higher rates of unmet need for technology and equipment when compared with non-Māori[39]. (For information about the limitations of this data please see the 2011 DND Report[40].)
- Findings from Digby *et al.* (2014) indicated young Māori have higher rates of permanent hearing loss than their European peers, based on the previous DND dataset (1982-2005)[41].

[i] In this report New Zealand Māori ethnic group is referred to as Māori.

[ii] Individual year age data for ethnicity is not freely available from Statistics New Zealand.

- Referral rates from the B4 School Check[i] (2011) analysed by Searchfield *et al.*, show higher rates of referral from hearing screening for Māori children (9%) compared with non-Māori (5%)[42]. It is important to note that high referral rates for Māori may relate to higher rates of ear disease, as these figures do not just relate to permanent hearing loss.

For further information on ethnicity coding within the database, please refer to Appendix C: Notifications and ethnicity, on page 48.

Unilateral and bilateral hearing losses

Ninety-eight per cent of cases notified to the database during the 2010 to 2015 period had one or more ethnicity codes recorded. Please keep in mind that some records (less than 10%) contained multiple codes for ethnicity, and so appear in more than one group.

Considering all 2010-2015 cases and using interpolated audiometric data and manual determinations of bilateral/unilateral status, 66% of cases notified are recorded as having bilateral hearing losses, while the remaining 34% have unilateral hearing losses. Figure 8, below, shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group during the 2010-2015 period. This year, these data include not only interpolated cases, but also those with one or more frequencies missing. (As a result, these data contain more cases compared with data presented in previous reports although they are still comparable with 2010-2014 report data.)

Please note that MELAA figures relate only to a very small number of cases.

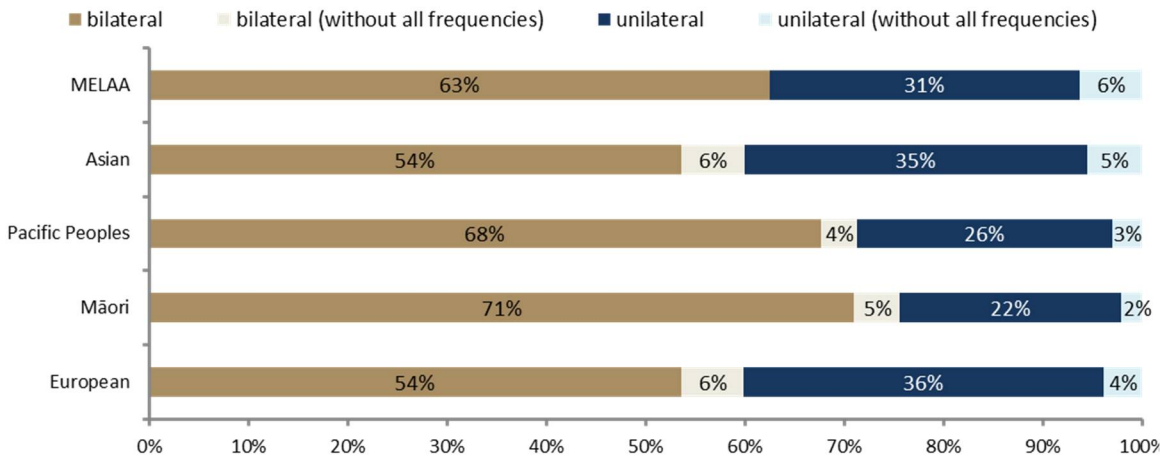

FIGURE 8: PROPORTION OF UNILATERAL AND BILATERAL HEARING LOSSES BY ETHNICITY (2010-2015) BASED ON INTERPOLATED DATA AND MANUAL CHECKS TO DETERMINE BILATERAL/UNILATERAL STATUS

The significant difference between Māori and New Zealand European rates of bilateral loss (found on analysis of the now larger sample in 2015) supports the 2014 paper by Digby *et al*. which found a larger proportion of bilateral hearing losses among young Māori when compared with their New Zealand European counterparts[41].

These data suggest that Pacific Peoples may also have higher rates of bilateral hearing loss than their New Zealand European Counterparts, although further analysis will be required to confirm this difference.

This difference can also be seen by comparing bilateral losses among children and young people listed as having 'Māori' ethnicity (78%), with those listed as 'New Zealand European' (59%) and those with whose ethnicity is described using both 'Māori' and 'European' ethnicity codes (63%).

[i] For more information on the B4 School Check, please click here or view the glossary on page 51.

Aetiology

Causes of deafness

The aetiology of hearing loss is either genetic (syndromic or non-syndromic) or non-genetic, and may be known or unknown depending on whether testing has been completed and whether a cause is able to be identified.

The American College of Medical Genetics and Genomics estimated in 2014 that 30% of genetic deafness is syndromic[43]. In non-syndromic deafness with a genetic cause, the most common genetic mutations found are in the GJB2 and Pendrin genes. The Otoferlin gene has been implicated in cases of ANSD[44].

The proportion of hearing losses with a confirmed genetic cause is increasing over time[45,46], as more hearing losses are better understood in terms of their aetiology, and as genetic testing becomes cheaper and more widely available.

Non-genetic causes of hearing loss among children and young people include: prematurity, infections during pregnancy (such as cytomegalovirus, toxoplasmosis and rubella), and diseases like meningitis and mumps.

In children, mumps is thought to be the commonest cause of unilateral acquired sensorineural deafness and is usually sudden in onset and profound in severity[47].

Cytomegalovirus (CMV) is a significant cause of deafness among children and young people in overseas studies, causing 10-20% of cases in those under the age of 5[48]. In New Zealand, CMV seroprevalence was assessed from 9343 first-time New Zealand blood donors. The highest prevalence was found among Pacific Islanders (93.2%) and the lowest in Caucasians (54.8%)[49].

Internationally, as reported by Davis and Davis[5] , it is common for a high proportion of cases (between 15% and 57%) of hearing loss to be of unknown aetiology. Aetiology is reported as more likely to be investigated in cases of bilateral hearing loss, and where the hearing loss is more severe in nature, compared with unilateral cases or those which are less severe[50].

It is worth noting that identification of one aetiology does not exclude an underlying genetic predisposition. For example, the A1555G mitochondrial mutations may predispose a patient to hearing loss, and this hearing loss is expressed when certain antibiotics are used[51].

New Zealand data

All but three of the 206 cases that met the inclusion criteria for the 2015 period, contained information relating to whether the cause of the hearing loss was known or unknown.

As seen in Figure 9 (below), the proportion of hearing losses where the cause was thought to be known has decreased significantly 2010 - 2015, when compared with figures from before 2006. At least some of this difference is thought to be the result of changes in the cause information requested, as the notification form has been made more specific, asking for confirmed, and not suspected cause.

Another reason for the increasing proportion of cases without a known cause recorded, is that more children in more recent times are being diagnosed with hearing loss earlier, owing to the introduction and roll-out of newborn hearing screening. For example, now that more babies are being diagnosed with hearing loss, genetic testing is less likely to have been performed at the time the hearing loss is diagnosed. In addition, hearing losses may now be identified before a full picture of possible other issues is established, perhaps reducing the likelihood that hearing losses that are part of a syndrome being identified at the time of notification.

As mentioned earlier in this report, immunisation coverage in New Zealand has risen significantly since vaccination for children became a Performance Programme indicator in 2006. This is likely to be resulting in

reduced rates of hearing loss from diseases such as meningitis (which presents with additional disabilities, although the numbers are likely to be small[i]), measles, mumps and rubella.

The importance of cytomegalovirus (CMV) in causing deafness among children and young people in New Zealand is not yet understood. There may be differences in rates of exposure to CMV among different ethnic groups in New Zealand, with serologic information from three-year-olds indicating higher levels of exposure among Māori and Pacific Island groups when compared with their European counterparts[52].

In New Zealand, during the 2010-2015 period, bilateral hearing losses and those which were recorded as severe or profound in severity were more likely to have a known aetiology than those categorised as mild and/or unilateral in nature.

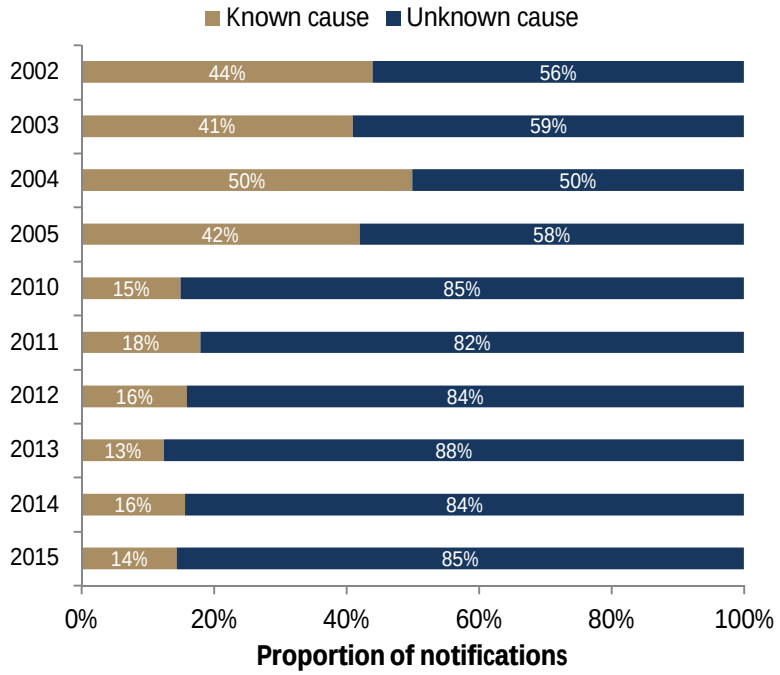

FIGURE 9: PROPORTION OF HEARING LOSSES OF KNOWN AND UNKNOWN CAUSE NOTIFIED TO THE DND BY YEAR DIAGNOSED (2002-2005 AND 2010-2015)

Testing for aetiology in New Zealand

There has been a drive among the New Zealand based ENT specialist community during the last few years to increase the proportion of hearing losses which undergo aetiological investigations, such as genetic testing, MRI and CT scans[53].

Although practice varies, ENT specialists generally refer young people/families of children with hearing loss for genetic testing where there is no clear explanation of the cause of the hearing loss.

Over time, more genes and mutations are being added to those for which testing is available in New Zealand. ENT specialists request the tests and counsel patients about the results. If multiple or unusual mutations exist, ENT specialists refer to genetic services[54].

[i] It is difficult to compare the number of cases of meningitis over time as this information was not collected prior to the re-launch of the database in 2010 and as there is no specific question related to meningitis in the current notification form.

Identification of hearing losses

Who first suspected the hearing loss?

Information on who first suspected the child's or young person's hearing loss was recorded for all children and young people confirmed as being born in New Zealand and who were diagnosed in 2015.

Table 5 shows the top three groups first suspecting the hearing loss among notified cases since the re-launch of the database in 2010.

Parents/caregivers have gone from first suspecting hearing loss in almost half of all cases to being first in about 1 in 5 cases. Newborn hearing screeners were not in the top three groups to suspect a hearing loss in 2010 or 2011[i] and yet they are now first to suspect almost half of all cases of hearing loss notified to the database. For the first time, Vision Hearing Technicians (VHTs) are not listed in the top three groups most likely to first suspect a hearing loss. This is likely to be because many hearing losses previously identified first by the VHTs are now being identified through newborn hearing screening.

2010	2011	2012	2013	2014	2015
Parent or caregiver (49%)	Parent or caregiver (42%)	Parent or caregiver (33%)	Newborn hearing screener (34%)	Newborn hearing screener (40%)	Newborn hearing screener (48%)
VHT (22%)	Medical Professional (21%)	VHT (23%)	Parent or caregiver (20%)	Parent or caregiver (22%)	Parent or caregiver (18%)
Medical Professional (12%)	**VHT (15%)**	Newborn hearing screener (23%)	**VHT (16%)**	**VHT (13%)**	Medical professional (other than GP) (10%)

TABLE 5: THREE GROUPS MOST LIKELY TO FIRST SUSPECT A HEARING LOSS 2010-2015 (BORN IN NEW ZEALAND)

Strong evidence exists that behavioural methods for identifying a hearing loss, even those used by paediatric audiologists or hearing screeners, are not an accurate method of screening for hearing loss in young children[55,56]. In addition, the challenges parents face in trying to identify a hearing loss in their young child are considerable, particularly when their hearing loss is not so severe as to prevent speech and language development.

Therefore, it is very pleasing to see that there has been a noticeable change in the groups most likely to first suspect a hearing loss among children and young people, over the last three years, towards those using objective methods, particularly those using these measures in newborn hearing screening.

The proportion of 2015 cases first suspected by parents or caregivers is again lower than at any time since the database was re-launched in 2010. It is also below historic levels in the original DND database, which reported between 34% and 52% of cases first suspected by parents in the 2000 to 2005 period.

[i] Further information was added to the notification form in 2012 to ensure audiologists were clear about how to code the answer to this question, should the child have been identified through newborn hearing screening. This change may be partially responsible for the reported increase in the role of newborn hearing screeners in first suspecting the hearing loss from 2012, given that the UNHSEIP coverage rates had not at that time increased significantly from 2011 levels. However, the growing role of newborn hearing screeners is undeniable.

Age at diagnosis

Figure 10, below, shows the number of cases identified by the age of the child[i]. There is a notable peak in the number of notifications during the first year of life – this is undoubtedly the effect of the universal newborn hearing screening programme. The peak for diagnosis in the first year of life is now almost three times as high as it was in 2010, when the database was re-launched. This is a positive trend, as it indicates more and more children are being diagnosed early.

A further, smaller peak can be seen for four, five and six year olds; this is likely to correspond to the B4 School Check[ii, 57]. The number of children being identified at this time has fallen by almost half since 2010, although screening coverage for the hearing portion of the B4 School Check has been rising during this time. See page 30 for more details about the B4 School Check.

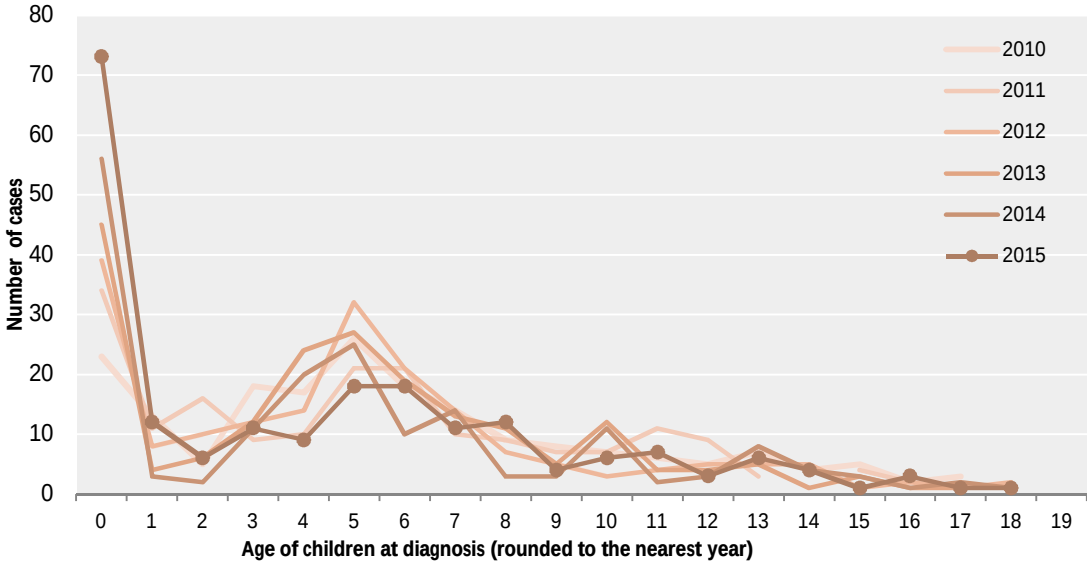

FIGURE 10: NUMBER OF CHILDREN DIAGNOSED BY AGE (2010-2015)

Overall age at identification

Caution: there are a number of issues with reporting the average age at identification (diagnosis) for all groups of children. While this may have some meaning, as it describes the average age at which providers will begin working with children to provide interventions of some type, the average relates to all newly diagnosed children, as it is not possible to separate out children with hearing losses that are late onset (such as progressive and acquired hearing losses).

It is important to remember this average age includes all children diagnosed in the notification period, for whom specific confirmation age data was available[iii], *including those born before newborn hearing screening was implemented* and, as mentioned above, those with acquired or progressive hearing losses.

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[i] Please note that the majority of children having their B4 School Check since the end of 2013 will have been screened for hearing loss soon after birth.

[ii] The B4 School Check aims to screen all children before they reach school, and to identify and provide intervention to those children identified with targeted conditions. Part of this Check involves screening children for hearing loss. This screening should be completed on all children not already under the care of an ENT specialist or audiologist following their fourth birthday. Those not screened before they reach school should be screened after their arrival at school. This screening involves audiometry, usually conducted by a Vision Hearing Technician. If the child passes this test, no further referrals are required. Should the child refer on audiometry, tympanometry should be conducted.

[iii] Confirmation age data is now being requested as a date of diagnosis, rather than an age at diagnosis to improve the quality of this data. This information is also being requested at the same time as suspicion age, to emphasise the differences between these two pieces of information and reduce data entry errors.

Keeping this in mind, the average ages at diagnosis for children diagnosed and notified to the database are described in Table 6. This table shows that, although there has been a fall in the overall average age of confirmation, the reduction is quite slow and seems to have been influenced by the increase in the number of notifications around five years of age for 2012 and 2013 as well as the increases at ten years of age for 2013 and at 10-11 years for 2011. Those born in New Zealand have a more marked drop in the average age than the full sample, which includes those born overseas and a small number where the place of birth is unknown.

Please note that the data in the table below have been slightly revised compared to those reported previously, to account for some notifications that were later removed from the database, and others that have been added retrospectively. These changes are small.

	2010	2011	2012	2013	2014	2015
Average all cases	67	59	61	60	60	53
Average born in New Zealand	64	54	56	54	52	48

TABLE 6: AVERAGE AGES OF DIAGNOSIS FOR ALL CASES IN MONTHS (2010-2015)

For the purpose of comparison with previous data, the average age at diagnosis is presented, but those groups who are more and less likely to be identified later can be found in Table 7 below.

Groups more likely to be identified later	*Groups more likely to be identified earlier*
born overseas	born in New Zealand
mild hearing losses	profound hearing loss
acquired hearing losses, e.g. late onset, progressive and trauma related	hearing loss suspected to have been present at birth
unilateral hearing losses	bilateral hearing losses

TABLE 7: EARLY AND LATE AVERAGE AGES OF IDENTIFICATION (2010-2015)

Age at diagnosis by severity of hearing loss

Table 8 shows the average age at diagnosis (confirmation of hearing loss) for children and young people with *bilateral* hearing loss in each of the American Speech-Language-Hearing Association (ASHA) severity categories. As expected, mild and moderate hearing losses are identified later than more severe losses.

Please note that, as a number of records in the database contain incomplete severity information, we have included those determined to be bilateral using both data by the audiologist and interpolated data-points. Also, please note that the table below only includes cases of bilateral hearing loss.

Additionally, 'moderately severe', 'severe' and 'profound' categories contain small samples so we have grouped these together this year. Younger children are more likely to be missing some severity data, meaning they could not be classified for the table below and that may be the reason why reductions in average age of diagnosis are not as clear within this data.

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2010-2015)	Total number of cases
mild	72	368
moderate	47	202
moderately severe	28	50
severe	26	22
profound	15	41

TABLE 8: AVERAGE AGE AT DIAGNOSIS, IN MONTHS, FOR *BILATERAL* HEARING LOSSES BY DEGREE (ASHA CODEFRAME) USING INTERPOLATED DATA WITH MANUAL CHECKS (2011-2015)[i]

The greatest variability in the age at diagnosis is for mild and moderate hearing losses – understandable given that these losses can be difficult to identify. The database does not include information about the proportion of losses which are thought to be progressive in nature.

Age at diagnosis and ethnicity

Figure 11 shows the average identification ages for each ethnic group, for all children and young people notified, where ethnicity information was provided. Keep in mind that these data are not priority coded, hence a small number of cases are in two or more ethnicity groups at one time. MELAA data are contained in this graph but keep in mind that this group is historically very small, hence its variation in the average is more visible.

While Māori are more likely to have bilateral hearing losses (which are on average identified earlier than unilateral losses), they are also more likely to have mild and moderate hearing losses than their New Zealand European peers, and mild and moderate losses are on average identified later than severe and profound losses[41]. These opposing effects make it difficult to understand how effectively the system is working to detect hearing losses early among Māori children and young people.

In addition, the proportion of cases reported as Māori in the database has grown since 2010 – this could be due to a greater focus on accurately coding of ethnicity in some areas, although we have no data to confirm this suggestion.

Pacific Peoples consistently have the highest average age at detection of losses when compared with the other groups in the sample. We hope future analyses will shed light on the types of losses among Pacific Peoples so we can better understand the reasons for their later average diagnoses.

A number of previous DND reports (1995-2005) noted that Māori and/or Pacific children were identified later than New Zealand European children, although this difference was not reported in every DND report[ii].

[i] Some 2011 and 2012 figures contained in this table differ from those reported previously, owing to small differences in the way these data were calculated, and also small reductions in the number of notifications included in the database since the original dataset was provided to allow checks for duplicates.

[ii] 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 New Zealand European children being identified, on average, earlier than Māori and Pacific children.

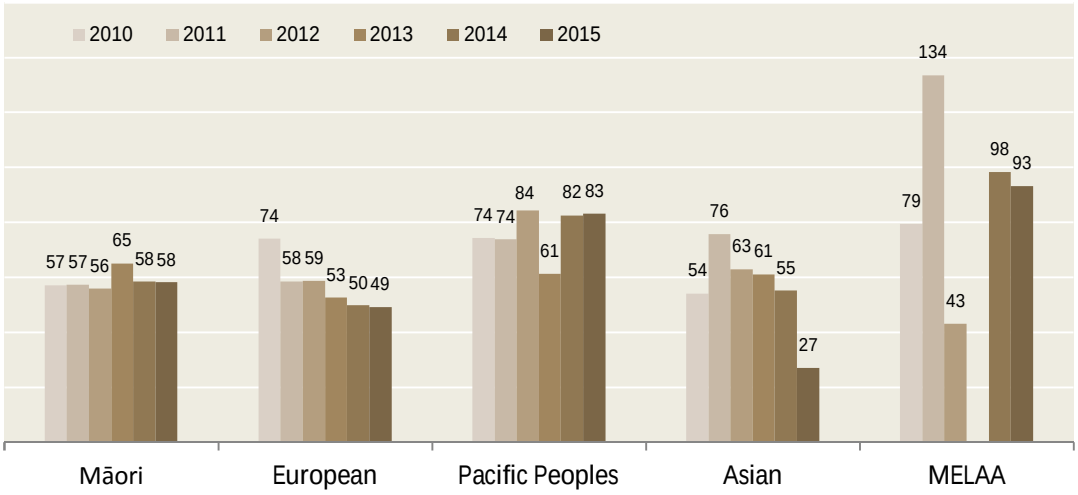

FIGURE 11: AVERAGE AGE OF DIAGNOSIS BY ETHNICITY IN MONTHS (2010-2015)

Newborn hearing screening

All district health boards have been screening babies for the full notification period (calendar years) since 2011[i]. Data in this section of the report relate only to those children born in New Zealand.

Screening status

Table 9 shows the screening status of New Zealand-born children notified to the database (and therefore diagnosed) in the period 2010 to 2015.

Please note that this table shows children diagnosed at varying ages, so just over a quarter (26%, n=47) of the cases notified for 2015 were not screened as no UNHSEIP service was available in their area at the time of their birth. As expected, the proportion of children being diagnosed as a result of a referral from the UNHSEIP is increasing, and the proportion of notifications not offered screening is falling.

Loss to follow-up is a significant issue for newborn hearing screening programmes internationally. As audiological assessment data from the UNSHEIP is still incomplete, the true extent of loss to follow-up in the UNHSEIP cannot be ascertained. However, we can look for differences in loss to follow-up within the DND.

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 1.0 to 1.1 per thousand babies for bilateral hearing losses, and approximately an additional 0.5 per thousand unilateral hearing losses[58, 59, 60, 61].

Because overall population prevalence in New Zealand for these targeted types of permanent hearing loss is not known, we can only use these rates as a guide to the number of cases which may be found in New Zealand when the UNHSEIP achieves high coverage and low loss to follow-up in all regions.

[i] Implementation of New Zealand's Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP) began in 2007, and the last eight district health boards to be included in the roll-out began screening between July 2009 and July 2010. It is worth noting that the large Auckland DHBs (Counties Manukau, Waitemata and Auckland) had all begun screening by April 2010.

Was universal newborn hearing screening (using aABR or aOAE) offered to this family after this child or young person's birth?		2010	2011	2012	2013	2014	2015
No	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	4%	2%	4%	1%
	No, this service was not available at the time (at the time of diagnosis)	69%	53%	55%	49%	38%	26%
Unsure	Unsure whether screening was offered to this family	7%	4%	6%	6%	5%	6%
Yes	Yes, a screening programme was in place, but the child was directly referred to audiology due to atresia	0%	0%	0%	0%	1%	1%
	Yes, screening was offered but this child was not screened	1%	1%	1%	1%	1%	3%
	Yes, the child was screened and referred but follow-up did not occur at the time, and so this is a delayed diagnosis	0%	4%	2%	5%	2%	2%
	Yes, this child was screened and passed	1%	7%	5%	6%	12%	15%
	Yes, this diagnosis is a result of a refer on the screening test	19%	27%	28%	32%	38%	45%

TABLE 9: SCREENING STATUS OF CHILDREN BORN IN NEW ZEALAND AND DIAGNOSED 2010-2015[i]

Using these overseas data, we might expect approximately 90 diagnoses directly from the newborn screening programme each year, based on a figure of 62,000 births per year[ii]. Current National Screening Unit (NSU) data are aligned with these overseas figures.

However, there is now good local data suggesting that young Māori may have higher rates of hearing loss than is found in the New Zealand European population. This suggests that New Zealand's UNHSEIP may expect to identify more cases of hearing loss than other programmes.

A total of 81 of the 2015 notifications (45%) related to children born in New Zealand who were diagnosed as a direct result of newborn hearing screening. This percentage has almost tripled since the database was re-launched in 2010.

It is not known how many cases of hearing loss are currently missed by the newborn hearing screening programme, as these children were either not screened by the UNHSEIP or they were lost to follow-up.

	2010	2011	2012	2013	2014	2015
Number of diagnoses resulting from universal newborn hearing screening	28	44	45	53	57	81
Diagnoses as a proportion of total notifications	16%	27%	27%	32%	38%	45%

TABLE 10: DIAGNOSES RESULTING FROM NEWBORN HEARING SCREENING IN NEW ZEALAND[iii], 2010-2015

[i] Please note that some figures in this table have been rounded and so not all sum to 100%. These figures are slightly different from those reported in previous years, due to small numbers of retrospective notifications and a small change in the codeframe.

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

[iii] Please note that the table shown in the 2011 report contained data for all cases, whereas this table contains data only for children born in New Zealand.

The most recent NSU monitoring report[37], includes screening data from 1 January to 31 December 2014, and audiology data to 11 February 2016 for children screened in 2014, reports that[i]:

- 93.3% of babies born during 2014 completed screening. Of those babies who completed screening, approximately 85.5% did this by the target of one month (corrected age) – figures for individual DHBs ranged from 51% to 102%[ii];
- 2.2% of babies screened during the period were referred to audiology;
- 67.9% of the 1,196 babies referred to audiology had audiology assessment data reported to NSU by the date of extraction for data contained in the report (Feb 2016). 58% of babies referred had their assessment completed by the target time of three months of age, well below the target of 90%;
- Māori and Pacific babies were less likely to have screening and audiological assessments completed. Rates of permanent congenital hearing loss were higher for Māori babies (2.5 per 1,000) than their European counterparts (1.5 per 1,000 babies), suggesting Māori babies have higher rates of PCHL; and
- 105 babies in total were identified with some type of hearing loss during the period.

Notifications from two large DHBs (Auckland and Waitemata) are thought to be underrepresented in the 2015 database (see page 9 for further information). This is likely to have reduced the number of potential DND notifications resulting from UNHS that were notified in 2015.

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 identification 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 of the success of the New Zealand newborn hearing screening programme. The annual DND reports should provide useful data to show how the overall age at identification changes over time.

There has been a pleasing reduction in the average age at diagnosis of cases referred from newborn hearing screening in New Zealand (therefore born in New Zealand), from ten months in 2010 to eight months in 2011 and five months in 2012, six months in 2013 and now to five months in 2014.

Of the 81 cases notified in 2015 which were identified as a result of newborn hearing screening within New Zealand, 63 (75%) were diagnosed by the internationally recommended age of three months. This is a pleasing improvement on 2011 and 2012 levels, and the same as 2013, although the target age of three months for diagnosis has still not been met across all cases.

[i] Please note that the most recent report contains data from 2014, and so doesn't align with the DND reporting period (calendar year 2015). Also, the 2014 data covers the period during which the previous AOAE/AABR protocol was in place for the UNSHEIP.

[ii] Values over 100% are calculated due to differences between the number of babies screened and the number of births.

	2010	2011	2012	2013	2014	2015
Number of diagnoses resulting from universal newborn hearing screening	28	44	45	53	57	81
Average months at diagnosis	10	8	5	7	5	6

TABLE 11: NEWBORN HEARING SCREENING DIAGNOSES AND AGE AT DIAGNOSIS

Identification of false negatives

The DND probably provides the only method for identifying potential false negatives from the newborn hearing screening programme[62].

In 2012, there was a Ministry of Health initiated recall of 3,422 babies, 2,064 of whom had potentially been incorrectly screened; 901 children of these had been rescreened by 28 November, 2012[63]. In 2015, no cases notified to the database were explicitly identified as having wrongly passed their screening, meaning we have no confirmed false negative cases for this year. This isn't to say that one or more babies weren't incorrectly passed at their newborn hearing screening, just that none were recorded as such in the notifications.

Cases included in the potential false negative category (below) may be due to deviation from the protocol on the part of the screener, hearing losses being progressive or acquired, or because the screening technology and/or protocol did not identify a child with a milder hearing loss or one with an unusual configuration.

Twenty-seven of the children identified with hearing loss during 2015 had been screened previously and passed this screening. This figure, and the fact that it is rising, is not necessarily a concern, as many children develop hearing losses after their initial diagnosis, and over time more children are being screened.

Of these 27 cases, there are two groups that may be useful to remove to help us identify potential false negatives. The first of these have known acquired hearing loss, while the second is those with hearing losses where there is some uncertainty – they were either suspected to have been present at birth, or the diagnosing professional was unsure whether the hearing loss was likely to have been present at birth[i]. As the second of these groups is based on a relatively subjective assessment by the clinician, these cases may or may not provide cause for concern.

	2010	2011	2012	2013	2014	2015
Total cases identified by year who were screened previously (i.e. are not currently referrals from the UNHSEIP) and who passed this screening	3	11	10	11	18	27
Number of cases from regional screening programmes, or from the UNHSEIP, which passed screening, which were not thought to be acquired loss, and where the notifying professional answered 'yes' or 'unsure' to the question about whether the loss was thought to have been present at birth and who were born in NZ	3	5	5	6	10	18

TABLE 12: POTENTIAL FALSE NEGATIVES AND CASES PREVIOUSLY REFERRED FROM HEARING SCREENING, 2010-2015, BORN IN NEW ZEALAND ONLY

Of the 18 cases listed in Table 12, one looks to have been miscoded by the audiologist as a pass from newborn hearing screening as they were identified before one month of age. The remaining children in this group had their hearing losses identified between the age of eight months and six years three months of age.

[i] Audiologists completing the notification form were asked to answer 'yes', 'no' or 'unsure' to the question 'Was the hearing loss thought to have been present at birth?' However, the answer to this question provides only a rough indication, as we cannot know whether the hearing loss was indeed present at birth.

Further details were provided about five:

- "family reassured by pass hearing screen, third child, exposed to two languages so put speech and language delay down to other factors than HL";
- "child had glue ear overlay which was clouding the picture of her hearing ability";
- "child passed her newborn hearing screening test and hearing loss was not suspected. She had the hearing test as she has an appointment to see an ENT specialist";
- "child was initially referred to Audiology as targeted follow-up from newborn hearing screening. Ongoing audiological testing over a long period of time has been required to completely confirm his loss - however this has been suspected since November 2011";
- "passed his BC results at the time but results on follow up were not clear."

Of the 18 cases listed, two of these were coded as 'yes' to the question about whether the loss was thought to have been present at birth. This is the category that is most likely to include false negatives, although false negatives could exist within the full group of 18. One of these two children had a hearing loss that, while unlikely, could possibly been missed by the screening protocol (the protocol will not identify all children with hearing loss restricted to a specific frequency region or children with milder hearing losses) while the other had a hearing loss which should have been identified by the protocol if the hearing loss was present at the time of screening.

B4 School Check

The B4 School Check is a nationwide programme offering a free health and development check for four-year-olds. The Check aims to identify and address any health, behavioural, social, or developmental concerns that could affect a child's ability to get the most benefit from school. It is the final core contact of the Well Child Tamariki Ora Schedule. Screening audiometry and tympanometry (if required) are administered by Vision Hearing Technicians around the country.

B4 School Check hearing screening data for 2010-2015 cohorts are shown below. Children who refer on the screening are given a brochure explaining the result, as are those pass who on their hearing screen. These brochures are available in a number of languages.

Outcome	Description	2010/11	2011/12	2012/13	2013/14	2014/15
Pass Bilaterally	The child was screen and passed.	58%	67%	71%	74%	79%
Referred	The child was screened and referred to a relevant service.	5%	5%	5%	5%	5%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	7%	8%	7%	6%	6%
Under care	The child is already under the care of a relevant service.	1%	3%	3%	3%	3%
Decline	The hearing check was declined by the caregiver.	4%	5%	4%	3%	1%
Not Checked	The child did not receive a hearing check.	24%	16%	11%	12%	6%
Population	**Derived from the PHO enrolled population.**	**63,585**	**65,692**	**64,911**	**65,335**	**63,730**

TABLE 13 B4 SCHOOL CHECK HEARING SCREENING DATA FOR CHILDREN BORN BETWEEN 08/07/2005 AND 07/07/2010 (THOSE CHILDREN SCREENED IN 2010-2015)[i,64]

[i] The Ministry of Health notes "The population used here is the PHO enrolled population. We use this rather than SNZ population (SNZ - Statistics New Zealand) projections due to the better inter census accuracy, and as SNZ population projections only include 5-year age groups. However, the PHO enrolled population has a coverage of 96% for the 0-4 age group."

Delays in diagnosis

Information about delays

Those notifying cases were asked to provide information about the length of delay in identifying hearing loss and reasons for the delay, where one existed. Not all 2015 cases for which there was a delay had one or more reasons for the delay listed.

The average delay in 2015, between first suspicion of the hearing loss and confirmation of the loss, *including* those born overseas, and mild, acquired or unilateral hearing losses was 11 months. Almost half the delays recorded in 2015 were one month or less in duration.

While 11 months is a significant average delay between first suspicion of a hearing loss and confirmation of this loss, average delays in the last four years are improvements on 2010 and 2011 figures. This is likely to be owing to the introduction of, and improvements in, newborn hearing screening programmes around the country.

Please keep in mind that these delay figures are not directly comparable owing to the changing composition of the notifications in terms of severity, the proportion of unilateral and bilateral notifications and the proportion of losses which were acquired or progressive in nature.

Year	Delay in months
2015	11
2014	13
2013	12
2012	9
2011	18[i]
2010	22[ii]

TABLE 14: DELAY IN MONTHS BY YEAR, 2010-2015

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.

In 2015, 41% of all cases had one or more reasons for delay listed, with 29% having one reason, and 12% having two or more reasons for the delay listed. The number of cases with no reasons listed for the delay has risen during the last four years.

Again, this year the analysis below examines the reasons for delay where one or more reasons are listed *and* where the delay was reported to be: a) greater than one month; and b) greater than six months, measured from the time the hearing loss was first suspected until the time when the hearing loss was diagnosed. For cases diagnosed in 2010-2015, Table 15 shows the most commonly cited reasons for delays in diagnosis.

Table 15 shows the reasons for delay for cases with delays of six months or more. It also contains some possible approaches to reducing the various types of delay.

[i] Revised from the 20 months reported in 2011.

[ii] Revised from the 20 months reported in 2010.

Rank (most frequently mentioned)	Reasons for delay	Possible ways to reduce type of delay
1st	Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)	• efficient clinical practice to complete assessments over fewer appointments (Following 2016's Diagnostic and amplification protocols[65] which can be found on the National Screening Unit website and which used to be referred to as Appendix F) • prompt referral from newborn hearing screening
2nd	Parents did not attend appointments (for any reason)	• better communication with parents • flexible appointments for families • assistance with travel costs • more attempts to contact families before discharging from service • audiology services closer to home for families (e.g. community based clinics or outreach) • reduced waiting times
3rd	Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)	• better funding for audiology resources/DHBs to prioritise newborn hearing screening referrals and other paediatric cases
4th	Parents suspected something other than hearing loss (e.g. speech delay, developmental delay, selective hearing)	• better education of parents so they can identify signs of a possible hearing loss (including before baby is born through newborn hearing screening materials and using these as an opportunity for discussion) • clear guidance on pathways for assessment for parents
5th	Difficulty getting a referral to audiology (e.g. GP or other health professional dismissed parent concern and no referral was made)	• education for non-hearing professionals such as doctors, well-child nurses, preschool teachers, childcare workers, speech language therapists (SLTs) and specialists
6th	Follow-up lost in the system and did not occur as scheduled (between professionals or annual review or follow up appointment not made) OR Referral not made between professionals	• better systems and processes for scheduling and seeing follow-up occurs
7th	Child was born or lived overseas and hearing loss not diagnosed there	• audiologists and others (including the managers of the DND) could engage with refugee and new immigrant agencies to advise them of the existence of free hearing services and the pathway for early referral

TABLE 15: TOP REASONS FOR DELAY FOR THOSE WITH DIAGNOSIS DELAYS OF MORE THAN SIX MONTHS AND POSSIBLE REMEDIES (2010-2015)

Comments received regarding 2015 diagnoses sometimes contained information about the reason for delays, and some of these are reproduced below. The first comment highlights the importance of following the protocol and obtaining separate ear information at the beginning of testing.

"This child was tested via soundfield VRA at age 9 months which she passed (child's left ear is essentially normal). Subsequent audiology visits showed conductive overlays, but again testing was done via soundfield, so the unilateral hearing loss (which may have been present at the time) was not picked up. The child was subsequently referred to ENT for grommets. Separate ear information was only achieved recently."

"Born in Brisbane, Australia. Moved back to NZ for a short period of time before moving back after the Christchurch earthquakes. Once she came back, they thought she had glue ear and waited for ventilation tubes, once these were in, it has been found that her hearing has not improved and she has a permanent conductive hearing loss bilaterally."

"Child was initially referred to Audiology as targeted follow-up from newborn hearing screening. Ongoing audiological testing over a long period of time has been required to completely confirm his loss - however this has been suspected since November 2011."

"Referral from newborn screening not received by audiology despite being entered as a refer result in local database. This was detected in a database audit. Child was 7 months old at time of audit/subsequent resending of referral. Family did not attend first appointment offered at 9 months of age, but attended subsequent appointment after this."

Delays attributed to newborn hearing screening

Of the nine children whose diagnosis was a direct result of referral from the UNHSEIP and whose diagnosis was later than three months of age, one or more reasons for the delay were reported in six cases. Four of these cases had the diagnostic delay attributed to parents not attending appointments:

- audiologist having difficulties getting a confirmed diagnosis (n=2);
- parents not attending appointments (n=4);
- other – other medical issues took precedence (n=1)[i].

More information about the causes of delays in all groups can be found in the section on *Delay causes,* beginning on page 31.

One important consideration for newborn hearing screening referrals is the importance of prompt referral from the UNHSEIP to audiology, and the high priority of these cases by the DHB, to enable auditory brainstem response (ABR) to be completed before the approximate age of four months, by which time ABR becomes more difficult due to babies being less likely to sleep without sedation or anaesthesia. Without early ABR on these children it can be more difficult to obtain a diagnosis for this group until they can be tested using Visual Reinforcement Audiometry (VRA) at six months to two years of age. (Some children may not be testable using VRA until after six months due to other developmental difficulties.)

[i] These six cases contained seven reasons for the delay.

Severity

Audiometric data

Audiometric data are requested for both the right and left ears of all children and young people notified to the DND. Those notifying cases to the database were asked to provide air and bone conduction thresholds from the pure tone audiogram. In cases where the young age of the child meant the audiologist was unable to obtain audiometric data from pure tone audiometry, audiologists were asked to estimate thresholds from the ABR using correction factors from the National Screening Unit's 2009 Universal Newborn Hearing Screening and Early Intervention Programme National Policy and Quality Standards[i].

Examining the four data-points for each ear shows that these data points were provided for 128 and 122 of the 206 cases notified to the database, for right and left ears respectively. Notifying audiologists/ audiometrists are encouraged to provide as much audiometric data as possible for cases being notified.

Those notifying cases were approached about a number of cases, and were able to provide some missing information. Of the cases that still contained missing data, data are more commonly reported for 0.5 kHz and 2.0 kHz and less likely to be reported for 4.0 kHz and 1.0 kHz frequencies. This demonstrates that frequencies which are typically tested at the end of the protocol for testing young children are less likely to be complete (i.e. 4.0 kHz and 1.0 kHz).

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

As shown in Figure 12, below, the proportion of cases for which the thresholds were determined through ABR is rising, from 21% in 2010 to 41% in 2015. This is thought to indicate that fewer children are old enough to have their hearing assessed behaviourally. We hope to see this figure drop further in future years as newborn hearing screening programme coverage rates continue to increase and hearing loss is diagnosed at younger ages.

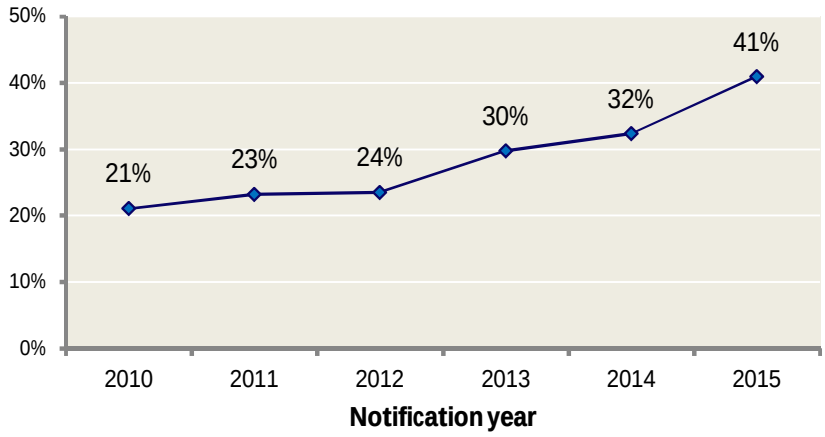

FIGURE 12: PROPORTION OF THRESHOLDS FROM ABR VS PTA, BY NOTIFICATION YEAR 2010-2015

[i] Correction factors: 5, 5, 0, and -5 dB for 0.5, 1.0, 2.0 and 4.0 kHz respectively as contained in 2016's Diagnostic and amplification protocols which can be found on the National Screening Unit website and which used to be referred to as Appendix F.

[ii] Correction factors for ABR and bone conduction were provided within the online notification form. These are from National Screening Unit (2016) Amplification protocols as noted above.

Classifications

In New Zealand, the Clark (ASHA) codeframe is the one used most commonly by clinicians. Therefore, this is the codeframe chosen for the majority of analyses in this report. Further information about severity classifications can be found in Appendix E on page 49.

Degree of loss	Clark 1981 (ASHA)[66]
Normal	-10-15 dB HL
Slight	16-25 dB HL
Mild	26-40 dB HL
Moderate	41-55 dB HL
Moderately Severe	56-70 dB HL
Severe	71-90 dB HL
Profound	≥91 dB HL

TABLE 16: CLARK'S 1981 ASHA SEVERITY CODEFRAME

Calculating severity for notifications

While the New Zealand DND collected some audiometric data for a number of years until the end of 2005, this information was insufficient to allow comparisons to be made easily with data from other jurisdictions. From 2010, the re-launched database has requested full audiometric data from those notifying cases, in the hope that more meaningful comparisons can now be made with overseas data.

As the original database (1982-2005) did not keep detailed records of how the analysis was conducted, it may not be possible to exactly replicate the inclusions made to calculate these figures. For example, we are unsure whether some or all database analysis prior to 2005 excluded cases which did not contain all eight audiometric data-points, or whether interpolation or averaging was used for records with fewer tested frequencies.

Interpolation

Table 17 shows the severity of hearing losses diagnosed for the first time in 2014, which is calculated in two ways. The first of these is using data containing all eight data-points, while the second includes interpolation.

While only cases where all eight audiometric data-points are present can be included in most severity calculations, interpolation of data has been used in some cases to provide a more complete picture of the severity of hearing losses notified. Interpolation is only used where three of the four data-points are provided for one ear, and where both data points surrounding the interpolated point are provided. This technique is becoming increasingly useful as more children are being diagnosed earlier, meaning they cannot have their hearing assessed behaviourally.

Please note that the severity analyses include either unilateral or bilateral losses, and are based on the hearing impaired ear in the case of unilateral losses, and on the better ear in the case of bilateral losses.

Key comments on these data include:

- that the proportion of less severe hearing losses is higher among bilateral cases when compared with those pertaining to only one ear;
- the number of bilateral hearing losses for which severity can be calculated rises when interpolation is used;
- that the proportion of mild bilateral losses drops when these cases are removed, increasing the proportion of moderate and greater hearing losses; and
- the proportion of moderate and moderately severe losses rises for unilateral cases.

The table below compares the proportion of bilateral/unilateral cases, comparing those which have not been interpolated and not had manual checks to those which have. Please note that this includes all notifications from 2010-2015 while last year's report only contained 2014 data.

Degree of loss using ASHA severity codeframe	Bilateral 2010-2015	Bilateral 2010-2015 (interpolated and manual checks)	Unilateral 2010-2015	Unilateral 2010-2015 (interpolated and manual checks)
Mild	61%	54%	47%	47%
Moderate	28%	29%	18%	17%
Moderately severe	5%	7%	11%	11%
Severe	2%	3%	5%	4%
Profound	4%	6%	19%	21%
Sample size	N=537	n=691	n=322	n=346

TABLE 17: COMPARISON OF SEVERITY CLASSIFICATIONS BY METHODOLOGY, 2010-2015

Severity profile differences between bilateral and unilateral hearing losses

Last year's report contained a graph showing the severity profile for children and young people notified to the database whose losses were bilateral, and compared these with children and young people whose losses were unilateral. Cases selected required all four data-points to be completed for each hearing impaired ear.

This year, we have included a similar graph, but this time we have included the severity profiles for bilateral and unilateral hearing losses for cases in which missing audiometric data could be interpolated (meaning more cases can be classified by their severity) and where a manual determination of whether the loss was bilateral or unilateral could be made based on available data. The authors of this report believe this shows a more accurate picture, and we will be using this method of analysis in future.

Figure 13 shows a difference can be seen between the severity profile of bilateral hearing losses (less severe and profound losses) and those with unilateral hearing losses (more severe and profound losses).

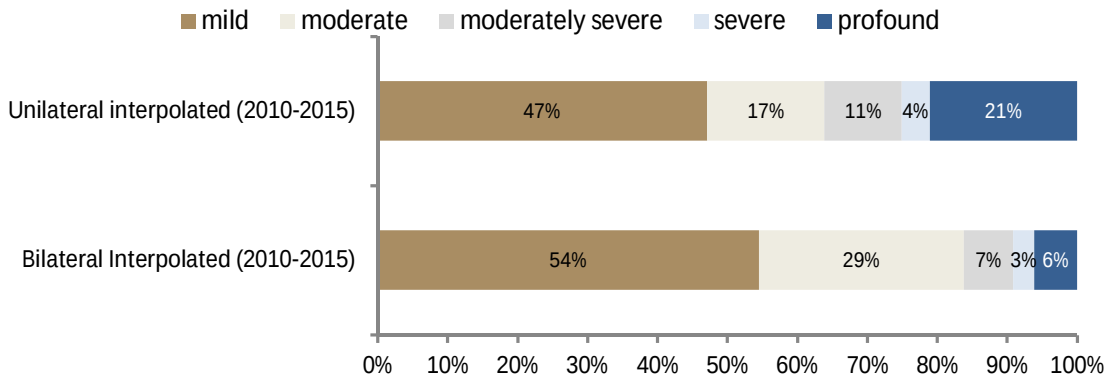

FIGURE 13: UNILATERAL AND BILATERAL HEARING LOSSES BY DEGREE (2010 - 2015)

This is particularly the case when the comparison is made between the ear with hearing loss in unilateral cases and the better ear in cases of bilateral loss. Clearly, these differences lessen when comparison is made with the worse ear in bilateral cases.

Other reasons for these differences may relate to:

- unilateral hearing losses within the database, which are, on average, found later than bilateral hearing losses and may have had more time to become more severe where these are progressive

losses. Bilateral hearing losses are more likely to be identified more quickly and therefore have less time to progress;
- low and mid frequency congenital hearing losses, which are more likely to be bilateral in nature and are more likely to be mild or moderate; and
- differences in genetic and other causes of unilateral versus bilateral hearing losses.

Comparisons with previous data

By categorising the notifications using the DND severity codeframe (1996-2005), a longitudinal comparison of the proportion of children in each group is possible using data reported between 2001 and 2005 and more recent data.

The database at that time excluded cases of unilateral hearing losses, children born overseas and those with acquired hearing losses. The 2010 to 2015 figures shown here match those exclusions from the earlier database.

Table 18 shows the average proportion of hearing loss notifications in each category between 2010 and 2015 and compares this with data from 2001 to 2004[i].

Proportion of cases notified by degree of hearing loss	Average 2001-2004	Average 2010-2015
Mild	48%	55%
Moderate	35%	34%
Severe	10%	6%
Profound	6%	6%

TABLE 18: 2001-2004 DND DATA COMPARED WITH INTERPOLATED 2010-2015 NOTIFICATION DATA, SELECTED CASES ONLY, 1996-2005 DND SEVERITY CODEFRAME

Previously we found that the severity profile of cases seemed to be different from previous years – we noted that we would be watching future data to see whether the severity profile returned to a pattern that more closely matched that seen before 2005. A return to historical patterns with fewer mild losses is not evident, either when cases containing full audiometric thresholds are considered, or when comparing data in Table 17 which includes more cases by using interpolated and manually checked thresholds.

Findings during the last two years show a very small proportion of severe and profound hearing losses and the highest proportion of mild cases since the database was re-launched. Factors which may be contributing to the generally small proportion of more severe hearing losses are listed below:

- information about individual children and young people are included in the dataset at the time of first diagnoses. A greater proportion of hearing losses are now being identified earlier, thanks to the introduction of newborn hearing screening. As a result, progressive hearing losses have not yet had the time to worsen, meaning the proportion of more severe losses may be less;
- some cases with audiometric data points in the severe and profound range did not contain complete audiometric data and these have not been included in this table, meaning severe losses (and other degrees too) may be under-represented[ii];
- often children diagnosed with hearing loss have a sloping hearing loss and the better thresholds reduce the average degree of hearing loss; and

[i] Data up to and including 2004 is used as it is unclear from the 2005 report which figures relate to which of the ASHA categories.

[ii] We have not been able to determine the criteria for calculating severity before 2006 making it difficult to attempt replication of the methods used.

- as noted previously, vaccination programmes have reduced rates of meningitis in New Zealand and this reduction is expected to have led to a reduction in rates of (more severe) hearing loss[67]. However, the reduction in the number of more severe cases due to meningitis is likely to be small.

Ethnicity and severity profiles

Historically, DND reports have shown that the greatest number of notifications pertain to New Zealand European and Māori and children and young people, and that milder degrees of hearing loss are more commonly reported among Māori[40, 68]. A recent analysis conducted on the previous DND dataset (1982-2005)[41] found young Māori were more likely to have mild or moderate hearing losses when compared with their New Zealand European peers.

This pattern is repeated with recent Deafness Notification data. Last year's analysis showed the proportion of cases in each of the severity categories, split by ethnicity grouping – which showed Māori had a higher proportion of mild and moderate cases than their New Zealand European peers.

This year an analysis was completed which also included interpolated data and this showed the same pattern of results. In addition, an analysis of those cases that were coded only as Māori, or New Zealand European was also completed, showing the proportion of cases of moderately severe or greater severity was 8% among those recorded as Māori and not New Zealand European, compared with 14% among those classified as New Zealand European and not Māori. It was 7% among those listed as with both Māori and New Zealand European ethnicities.

Comparisons with international data

The 2012 DND report contained a comparison of moderate and greater hearing losses in our database with those from the UK, Finland and the USA. This showed a greater proportion of moderate hearing losses, and fewer severe and profound losses than in the other countries' analyses. This could in some part be owing to the fact that our local data contains some records with only limited information. In addition, the overseas data excludes cases of mild hearing losses.

Severity codeframe used in Colorado	Bilaterally hearing impaired children in Colorado who received early intervention services between birth and 3yrs (2006-2012)		2010-2015 bilateral Deafness Notifications, born in New Zealand, under the age of 18 *Interpolated*		2010-2015 bilateral Deafness Notifications, born in New Zealand, under the age of 18	
	n=	%	n=	%	n=	%
Mild (26-40 dB HL)	99	37%	342	49%	304	57%
Moderate (41-70 dB HL)	102	38%	283	41%	195	36%
Severe and profound (>70 dB HL)	67	25%	66	10%	38	7%

TABLE 19: SEVERITY COMPARISON COLORADO AND NEW ZEALAND

It is interesting to note that some overseas data (including those from Finland and the US described by Davis and Davis in 2011[5]) also show lower numbers of children/young people in the severe category when compared with the profound category. This includes data with codeframes where the intervals between the average thresholds are evenly spaced. One caution relating to this comparison is that the data used from Finland and the UK are older, meaning that they are likely to relate to children identified prior to newborn hearing screening. As a result, some hearing losses may not have had time to progress in their severity. The significance of this effect in these samples is not known.

The 2013 report included a table which compared our bilateral local data (2010-2014) with data from Colorado[69], which covered the 2006-2012 period and was from a largely European population. Local DND data have been coded to match the severity codeframe used in Colorado. Of the Colorado sample, almost

half the children are under the age of three so, like our New Zealand data, it is similarly likely to underestimate the proportion of severe and profound loss that may be found in later childhood.

The New Zealand data has been extended to include children notified from 2010 to 2015 and is included again in Table 19. It shows the New Zealand sample contains a relatively high proportion of mild hearing losses and fewer with severe and profound losses. This year we have included records with the interpolated severity data, as we did last year. The basic pattern holds – there are still considerably fewer severe and profound losses in the New Zealand sample when compared to Colorado.

A further analysis has also been completed this year, looking at differences between New Zealand and Australian figures[6]. Severity information for cases in the DND were recoded, using the codeframe used by Australian Hearing (0-40dBHL, 41-60 dBHL, 61-90dBHL, 91dBHL+). Cases in which data-points have been interpolated have been included in the New Zealand data, as without these more severe losses are under-reported. Newborn hearing screening is nationwide in Australia, and so again the issue of comparing largely later identified children with our sample in New Zealand doesn't apply.

Severity codeframe used by Australian Hearing	Children and young people with aids and implants under the age of 26 (Australian Hearing Data)	2010-2015 bilateral Deafness Notifications under the age of 18 (*Interpolated data included)*
0-40 dBHL	62%	66%
41-60 dBHL	19%	25%
61-90 dBHL	10%	6%
91+ dBHL	9%	4%

TABLE 20: SEVERITY PROFILE COMPARISON AUSTRALIAN HEARING (2014) AND NEW ZEALAND DATA (2010-2015)

With the mounting evidence described above, it seems clear that New Zealand has a smaller proportion of severe and profound hearing losses than other similar countries. This may be in part due to the fact that Māori have a different severity profile to other groups.

Intervention

Ministry of Education

The figures below relate to the period 1 January 2014 to 31 December 2014 and were presented by the Ministry of Education to the UNHSEIP Advisory Group meeting on 2 November 2015[70].

During this period the Ministry of Education Special Education received a total of 129 referrals nationally from the newborn hearing screening programme. This figure is not comparable to the figures shown in the Newborn hearing screening section of this report, which begins on page 26. This is because the number of children *diagnosed*, as provided by the National Screening Unit, does not relate to the same period of time.

It is worth noting that children who have a bilateral hearing loss that meets the UNHSEIP threshold and are referred from the programme are automatically accepted for Advisors on Deaf Children (AODC) services.

Key points from this report:

- 42% of families/whanau were contacted within two working days following receipt of referral, 60% within five working days, 74% within ten working days and 85% within 20 working days. The target for first contact is 95% within ten working days.
- Reasons for delays for the first contact with families included that the AODC was unable to make contact with the family, that the referral was incorrectly coded, that an AODC was on leave or their position was vacant.
- The majority of referrals were from children listed as having New Zealand European or Māori ethnicities. Twenty-one children had no ethnicity listed. Reasons for absent ethnicity information are thought to include that this information was missing on the audiology referrals, or that parents did not provide this information.
- The proportion of children eligible for early intervention services who began receiving service one within a month of receipt of the referral was 92.25%. The target for the proportion is 90%.
- 36.4% of referrals to the Ministry of Education (n=47) were received for children over the age of six months, with 11 of the 129 referrals not received until more than 36 months. Reasons for the delayed referral were:
    - information not held about why the referral was delayed (n=15),
    - audiology waiting to confirm hearing loss (n=9),
    - case closed on the Ministry of Education's database (Special Education Case Management System) then re-opened (new case worker, change of district, returned from overseas), presents as a late referral (n=6),
    - family initially declined audiology follow up, then engaged with audiology (n=4),
    - referral late from audiology rationale not known (n=3),
    - children with complex needs, fragile health, other issues a priority (n=3),
    - unilateral hearing loss, monitored by Audiology and Family then referred to the Ministry of Education (n=3),
    - incorrect coding on Ministry's CMS then coding corrected, presents as a late referral (n=2),
    - family did not want Ministry involvement, and then requested referral to Ministry (n=1),
    - screening incident, identified in the re-screen process (n=1).

Deaf Education Centres

Kelston Deaf Education Centre (KDEC) and van Asch Deaf Education Centre (vADEC) are responsible for Deaf and hard of hearing students enrolled at or receiving services from the Combined Board of Trustees.

As at 31 December 2015, Deaf Education Centres provided services to the following groups[71]:

- residential placements for enrolled students in years 7 – 15 (19 students out of a maximum of 23 at KDEC and eight of a maximum of 20 at vADEC[i]),
- enrolled students attending the School Provisions in partner schools as day pupils (109 at KDEC and 28 at vADEC),
- specialist preschools had 29 Deaf enrolled students (KDEC n= 14 and vADEC n=15),
- Regional Services through Resource Teachers of Deaf visiting regular schools (n=526 at KDEC and n=314 at vADEC).

The schools offer flexible programmes to meet the specific needs of their students. NZSL and spoken language are equally respected and valued.

Of those children who are receiving these services from KDEC or vADEC:

- 39% were recorded as Māori, 38% as New Zealand European, 12% Pasifika, 8% Asian and 3% MELAA; and
- 1650 children were over the age of five: 77% of these were not Ongoing Resource Scheme (ORS) verified, 15% were verified as 'high needs', and the remaining 7% were verified as 'very high needs'.

In addition, 820 children have begun to receive services from the Deaf Education Centres over the last 18 months (through their Regional Services, provided by Resource Teachers Deaf) during stage I and II of implementation of the Wilson report. This figure has increased significantly from 299 in last year's report, due to stage III implementation of the Wilson report (remaining areas of New Zealand) beginning in July 2015. A new system is being implemented to track device usage among the 820 children mentioned above. Once this is implemented we hope to include further information about this group.

Since 2012, a single combined Board of Trustees has governed the Deaf Education Centres (DECs). The 2016 Combined Charter[72] contains useful information about the numbers of children who receive services from the schools and their staff, nationwide. The combined Board has prioritised the development of an accurate national picture of the deaf student population. This Charter contains information about student performance against National Standards.

[i] Some of the unused beds for van Asch are for immersion courses where up to eight regional students attend six times a year for a week.

Hearing aids

All but three cases notified to the database contained information about whether hearing aids were to be fitted[i].

As has been the case with data since 2010, the majority of children and young people whose cases were notified to the database and whose loss was first diagnosed in 2015 are to be fitted with two hearing aids (54%). Figure 14 shows this proportion is dropping, likely because the average age of diagnosis is falling. The proportion of cases in which the professional notifying the case is unsure whether hearing aids will be provided is rising, for the same reason.

Since the database was re-launched in 2010, the number of children who are receiving hearing aids before six months of age has almost tripled, moving from 23 to 73. This is almost certainly the result of the introduction of newborn hearing screening.

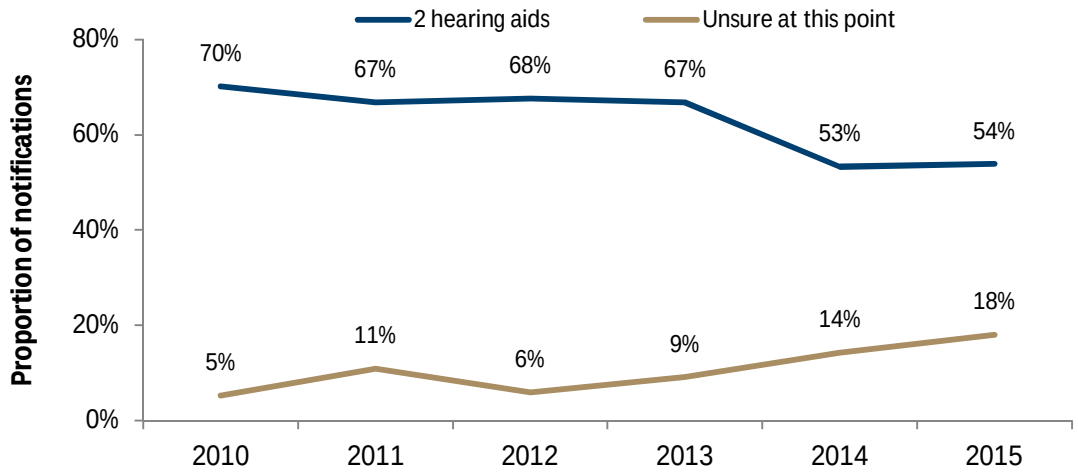

FIGURE 14: PROPORTION OF NOTIFICATIONS TO RECEIVE TWO HEARING AIDS COMPARED WITH THOSE FOR WHICH THE HEARING PROFESSIONAL IS UNSURE WHETHER HEARING AIDS WILL BE PROVIDED (2010-2015)

When data for all children notified from 2010 to 2015 is considered, audiologists reported that the majority of children with bilateral hearing losses (85%) would be fitted with two hearing aids, with 4% receiving one and 4% receiving no aids. The audiologist was unsure whether the child would receive hearing aid(s) in 8% of these cases.

For unilateral losses a higher proportion audiologists reported that 41% would be fitted with one hearing aid, with 24% would receive two aids (keep in mind that some have hearing loss in the other ear even if it didn't meet the 26dB pure tone average required to be considered a loss in the database) and 19% would receive no aids. The audiologist was unsure whether the child would receive hearing aid(s) in 17% of these cases.

Funding for hearing aids

In an attempt to provide some context for these figures, data provided by accessable, the Ministry of Health's provider for Hearing Aid Services during the period covered by this report, are shown below. Please note, these data pertain to all children receiving hearing aids and not just to those receiving hearing aids for the first time.

[i] It is worth noting that some children with unilateral hearing losses were reported to be receiving more than one hearing aid. In these cases, we can confirm that is because, although the average threshold for the better ear does not meet the 26 dB HL average required for inclusion in the database, one or more hearing thresholds are seen as sufficiently poor to warrant amplification in the better ear. This is indicative of one of the limitations related to classification systems that average hearing thresholds across four frequencies and categorise children into broad severity groups.

This shows MOH funded hearing aids for children and young people under the age of 21 who are in primary, secondary or tertiary education[73] during the 2014 calendar year. A total of 2494 service users (children and young people) received hearing aid(s) during this period, down slightly on the 2575 in 2014 and the 2659 in the year ending December 2013[i].

Of those children receiving aids as below, 60% were receiving aids in both ears, while the remaining 40% were receiving aids in only one ear.

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
NZ Māori	109	118	513	54	794
European	149	92	598	137	976
Pacific	120	57	227	61	465
Other	47	22	163	27	259
Total	425	289	1501	279	2494

TABLE 21: MOH FUNDING OF CHILDREN'S HEARING AIDS CALENDAR YEAR ENDING 31 DECEMBER 2015, ACCESSABLE

These data make for an interesting comparison with the 2010-2015 DND data. Figure 14 shows that 54-70% of children and young people were to receive two aids between 2010 and 2015, and 15-19% one hearing aid. The difference between these figures and those from **accessable** are likely to be at least in part owing to the fact that the DND data contains information about the number of aids to be provided *at the time of diagnosis*. These figures do not capture which of these cases then go on to receive more than the intended number of aids (as stated on the notification form) as they get older, either because their losses progress, or because the initial amplification strategy changes.

Cochlear implants

Although the notification form does request specific information about cochlear implants, it is useful to provide some information about the number of cochlear implants provided to children and young people in New Zealand, and some background on the funding for these implants.

Funding from the Ministry of Health 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 the area south of this line.

The majority of children receiving cochlear implants have severe or profound hearing losses, or progressive hearing losses which are becoming more severe. Some children have high frequency losses that are severe to profound in the higher frequencies and normal or near normal in the lower frequencies.

During the 2015 calendar year there were 46 publicly funded cochlear implants provided in the Northern Region and 47 in the Southern Region, to children and young people under the age of 19. These implants are provided based on Ministry of Health candidacy criteria for children and young people who are assessed by the cochlear implant teams[ii].

[i] Data provided in 2014 for the 2013 year was incorrect as these covered only a six-month period.

[ii] Since 1 July 2014, the Ministry of Health has funded bilateral cochlear implants (where this is clinically appropriate) for New Zealand children who are newly implanted. Children under the age of six at that time qualified for a retrospective second public implant.

Children receiving cochlear implants during the 2015 calendar year	Southern Cochlear Implant Programme[74]		Northern Cochlear Implant Programme[75]	
	Ears	Children	Ears	Children
ACC cases	0	0	1	1
Retrospective second cochlear implants (second ear for those under 6 already with one publicly funded ear) (1 Jul to 31 Dec)	3	3	2	2
Public Funding - (1 Jan to 30 Jun – before bilateral public funding allowed)	36	18	39	23
Private procedures	1	1	2	2
Re-implants – recalled devices, failed integrity tests, or soft failures	7	7	4	3
	47	29	46	29

TABLE 22: PUBLICLY FUNDED COCHLEAR IMPLANTS IN NEW ZEALAND (2015)[i]

[i] In some years the number of cochlear implants provided exceeds the number of profound or severe cases notified to the database. While the DND may be missing some notifications for children in the severe and profound categories, there are a number of other reasons why this figure is low compared with the number of children implanted during the same period. One is that some children who are notified to the database as having less severe hearing losses develop more significant losses over time, something which is not tracked by the database.

Appendices

Appendix A: History of the database

History of the DND

The 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 released.

The data presented in reports before 2006 contains notifications provided to the database within a specific year; that is, they pertained to children notified to the database in a calendar year, rather than those who are identified in that year[68]. During most of that time 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 among children and young people could be estimated, and from which the characteristics of new cases of hearing loss among children and young people could be understood.

In 2006, the Auckland District Health Board discontinued its contract to provide services associated with this database. No new provider was sought by the Ministry of Health. Between 2006 and 2009, a number of groups 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 DND was seen to have even greater importance from 2007, the start of implementation 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 the only way to identify potential false negatives within the screening programme.

In 2010, the DND was re-launched, with audiologists around the country encouraged to notify diagnosed hearing losses through a new online form. This re-launched database was initiated by Janet Digby and was part funded and supported by the New Zealand Audiological Society, which allowed communication with its members.

We are delighted that the Ministry of Health began funding the DND from the start of 2012. The database is now managed through a contract with *accessable* and builds on the work done by the New Zealand Audiological Society and Janet Digby.

Inclusion criteria

The original criteria for inclusion in the DND were based on the Northern and Downs definition below, and were 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 26 dB HL in the better ear (Northern and Downs classification, 1984)[76]."

There was a strong view among audiologists consulted, that the previous definition (above), which was used before 2006, 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 may not be appropriate.

The criteria for inclusion were modified for the 2010 re-launch of the database, based on feedback from a small working group[i].

The new definition now includes children and young people 18 years or younger and is aligned with the age range used for the paediatric cochlear implant programmes.

In addition, this database now includes children:

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

Specific guidance has been provided to hearing professionals to clarify the type of cases which are included in the database, to try to increase consistency in the types of losses notified:

- included in the database; atresia, congenital ossicular fixation, meningitis, acquired hearing losses;
- excluded from the database; hearing losses which can be remediated by the use of grommets (ventilation tubes), such as hearing loss associated with otitis media.

Notifying cases

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

Notifications are collected through an online survey form, to reduce data entry errors (which can occur when transferring data from paper forms to electronic formats) and to try to make it as easy as possible for cases to be notified. A revised consent process was also implemented on re-launch to ensure all information is collected with the consent of the family. Data is backed up regularly and information is sent through a secure link. Standardised methods for data analysis are now being used.

Future renaming of the database

During 2012, feedback on the name of the database was sought from parents of deaf children, Advisors on Deaf Children (AODCs), and audiologists, on a possible change to the name of the database. This feedback did not provide a clear path for renaming the database.

Some individuals and groups felt that changing the name to a broader title, such as the Hearing Loss Notification Database, would have merit, as it would acknowledge the range of types and severity of hearing losses included. Others felt changing the name of the database could cause confusion and reduce the number of notifications in the short term.

The name of the database (Deafness Notification Database) remains open for consideration. 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 are now included in the database.

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

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

[ii] While cases of unilateral hearing loss were technically excluded from the database until 2005, there were still large numbers of notifications sent to the administrators of the database, although these were not included in the main analysis. Professionals consulted in the development of the re-launched database unanimously believed this group should be included in the database, at least in part as there is strong evidence that this group has increased risk for poorer educational and speech/language outcomes compared to children with normal hearing in both ears.

Appendix B: High frequency hearing losses

Based on feedback from the audiological community, high frequency hearing losses (averaging over 26 dB HL over 2.0, 4.0, 6.0 and 8.0 kHz) and that would not meet the original criteria (26 dB HL average over 0.5,1.0, 2.0 and 4 kHz) have been collected from July 2011.

As these cases are not included in the main analyses in this document, a limited analysis of data from high frequency hearing losses notified in 2015 is provided below. A number of notifications that were recorded as high frequency losses by those notifying the case met the criteria for the main dataset, and so were included in that dataset and analysed as such.

This year we have included the table below, which shows the proportion of cases notified that were for children and young people over the age of five years of age. In future, reporting will focus only on those over five as those under five are unlikely to be tested at higher frequencies, and therefore won't be a representative sample.

Number of notifications	2011 (July to December)	2012	2013	2014	2015
High frequency losses	n=8	n=10	n=14	n=17	n=22
High frequency losses over the age of 5	n=8	n=6	n=6	n=16	n=16

TABLE 23: HIGH FREQUENCY HEARING LOSSES (2010-2015)

Other characteristics of this group of children/young people included:

- all of the 16 cases in this category in 2015 had the aetiology listed as 'unknown';
- eleven of the children and young people notified to this category were to receive one or two hearing aids;
- years of birth for this sample ranged from 1999 to 2009;

The figures below show the audiometric data for the 22 children or young people with high frequency hearing losses in this category this year. Please note that not all children and young people in this category had hearing loss in both ears, and not all audiometric data-points were provided for all cases.

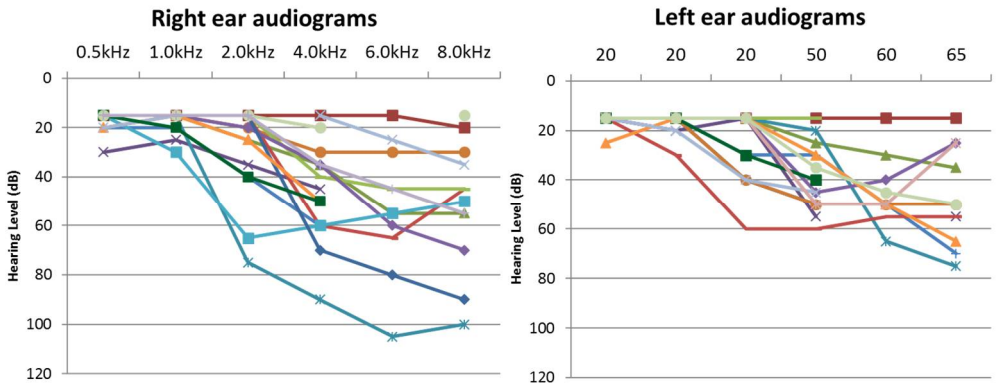

FIGURE 15: 2015 AUDIOGRAM DATA (HIGH FREQUENCY HEARING LOSSES) FOR RIGHT AND LEFT EARS

Appendix C: Notifications and ethnicity

The method used in this report to classify ethnicity is the total response method, where every person identifying with a particular ethnicity is included in that specific grouping[77]. For example, if someone considers their child to be of Samoan and Māori ethnicities 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.

Using this method provides a more detailed and realistic 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, the one with which they mostly identified, or where ethnicities are prioritised to include only one ethnic grouping per child. Using the total response 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 proportion of notifications in each ethnic group was calculated differently in DND reports before 2006, with respondents being coded initially as belonging to one 'race' and later as one 'ethnic group'. Categories used have also changed. As a result, direct comparison with ethnicity data from before the re-launch in 2010 is not possible.

The New Zealand Census (2006 and 2013) categorises respondents into five major groupings. These groups are: Māori, Pacific Peoples, Middle Eastern/Latin American/African (MELAA), New Zealand European and Asian. While it would be preferable to collect more detailed information on ethnicity, we understand this may not be available for all cases and we don't want to put audiologists off notifying cases by requesting more detail than they have available.

Appendix D: Estimating the total number of new diagnoses per year

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

However, we can use a number of methods to provide some indication of the number of new diagnoses of hearing loss annually among children and young people. It is likely that the database has been receiving notifications for between 50% and 70% of all cases diagnosed each year, since 2010. The 2013 Deafness Notification Database report contains further information on how this range was calculated on page 46.

Appendix E: Severity codeframes

A large number of classification systems are used to categorise hearing loss severity, locally and in overseas jurisdictions. Differences between these systems make it difficult for meaningful direct longitudinal and geographical comparisons of the proportion of children in a particular severity category[i]. Unfortunately, there is no clear standard internationally for classifying hearing loss, or a consistent definition for where a hearing loss begins for the purposes of epidemiological comparison.

Table 24 shows some of the differences between local and overseas severity classifications (these systems use an average of the pure-tone thresholds at 0.5 kHz, 1.0 kHz, 2.0 kHz and 4.0 kHz). Audiologists in New Zealand are commonly using Clark's 1981 (ASHA) classifications within their clinical practice, as per the New Zealand Audiological Society practice guidelines.

Australian Hearing uses the following codeframe (0-40dBHL, 41-60 dBHL, 61-90dBHL, 91dBHL+), but don't name the categories so these are not included in the table below.

	1996-2005 NZ DND	1982-1996 NZ DND	Clark 1981 (ASHA)	Jerger and Jerger (ASHA)[78]	World Health Organisation[79]	CDC[80]	Proposed code from Davis and Davis[5]
Normal			-10-15dB HL		≤25dB HL		
Slight			16-25dB HL	0-20dB HL	26-40dB HL		
Mild	26-40dB HL	30-55dB HL	26-40dB HL	20-40dB HL		21-40dB HL	30-39 dB HL
Moderate	41-65dB HL		41-55dB HL	40-60dB HL	41-60dB HL	41-70dB HL	40-69 dB HL
Moderately Severe		56-85dB HL	56-70dB HL				
Severe	66-95dB HL		71-90dB HL	60-80dB HL	61-80dB HL	71-90dB HL	70-94 dB HL
Profound	>95dB HL	≥86dB HL	≥91dB HL	≥81dB HL	≥81dB HL	≥91dB HL	95+ dB HL

TABLE 24: COMPARISON OF AUDIOMETRIC SEVERITY CLASSIFICATION SYSTEMS

[i] These systems, by and large, do not acknowledge any differences that may exist between the way hearing losses in children, young people and adults might best be categorised, i.e. there should be one system of classification for all groups.

Glossary

accessable: The Ministry of Health's contracted Services Manager which administers and manages Hearing Aid Services nationally.

Advisors on Deaf Children (AODCs): The Ministry of Education employs advisers on deaf children to help families understand their child's hearing loss and to guide parents as they consider the technology and communication options available. Advisors also provide assessments and information about a child's development and behaviour to other professionals working with the family. They work closely with teachers from the two Deaf Education Centres[81]. Implementation of changes proposed in the Wilson Report (2011) were completed in 2015, meaning AODCs are now focused on working with an Early Years focus.

Aetiology: The cause or set of causes; in the case of this report this refers to cause(s) of a child or young person's hearing loss.

Audiometric data: Audiometric data is about a person's hearing acuity given variations in sound intensity and pitch (frequency), involving thresholds and differing frequencies. The database collects information at 0.5, 1.0, 2.0 and 4.0 kHz wherever possible, and at higher frequencies for children and young people whose hearing loss meets the criteria for inclusion as a 'high frequency hearing loss'.

Audiometrist: Audiometrists conduct hearing screening, audiological assessment, including diagnostic hearing assessment, rehabilitation and hearing aid fitting, and follow-up specific to adults and young people over the age of 16 with non-complex hearing loss.

Auditory Neuropathy Spectrum Disorder (ANSD): This condition relates to issues in the transmission of sound from the inner ear through the auditory nerve that makes sound more difficult to discriminate when it reaches the brain. Someone with ANSD can have difficulty distinguishing sounds even when the audiogram indicates a mild loss, including speech, which can sound distorted.

American Speech-Language-Hearing Association (ASHA): This Association is relevant to the Deafness Notification Database in that they publish categories, which are widely used in New Zealand, to indicate the severity of hearing loss.

Bilateral hearing loss: Hearing loss affecting both ears.

B4 School Check: The B4 School Check is a Ministry of Health-funded programme that aims to screen all children before they reach school, and to identify and provide intervention to those children identified with the targeted conditions, including hearing loss. This screening takes place when the child is aged four, or five if they are not checked earlier.

Confirmation of hearing loss: For the purposes of this database, this is the date at which the hearing loss was first diagnosed. In most cases this would mean the audiologist has completed air and bone conduction testing (behaviourally or via ABR).

Cochlear implant: A cochlear implant is an implanted electronic device which provides a sense of sound to the recipient by directly stimulating the auditory nerve with current pulses, rather than via amplified sound as occurs in hearing aids. Those receiving cochlear implants usually have a hearing loss that is severe or profound in terms of its severity classification.

DHB/district health board: These are organisations established to provide health and disability services to populations within a defined geographical area. There are currently 20 district health boards in New Zealand.

False negatives: False negative is a term used to describe screened children who are incorrectly categorised as having a low risk of the target condition. In this report, this term relates to potential false negatives resulting from the newborn hearing screening programme (UNHSEIP) (i.e. a child who passed the screening test where it is possible that they had a hearing loss at the time the screening was conducted).

Full Time Equivalents or FTE: These are used to measure the number of full time equivalent positions for audiologists and generally equate to approximately one full time equivalent for every 38 hours worked per week.

Inclusion criteria: The current Deafness Notification Database contains information about children and young people 18 years or younger, born in New Zealand or overseas, with:

- a permanent hearing loss in one or both ears
- an average loss of 26 dB HL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0 kHz)[i]

[i] Based on feedback from the audiological community, high frequency hearing losses which would not meet the original criteria have been collected as a trial from July 2011. We will continue to trial inclusion of this special group in the database. A limited

Kelston Deaf Education Centre (KDEC): Kelston Deaf Education Centre provide educational programmes and services to Deaf and hard of hearing students in the northern part of New Zealand, roughly from Taupo northwards.

Notifications: Notifications contain data about an individual case of hearing loss, including demographic information, information on the hearing loss and its diagnosis. Information is provided to the DND database with the consent of the young person who has been diagnosed with a hearing impairment, or their parent in the case of babies and children. This information has been provided to the database manager via an online form since 2010.

Ongoing Resource Scheme: The Ongoing Resource Scheme (ORS) provides support for a very small number of students, with the highest level of need for special education, to help them join in and learn alongside other children at school. This funding provides Specialist Services staffing for students (who are ORS funded) including school counsellors. This scheme was previously reviewable (ORRS).

Suspicion age: For the purposes of this database, this is the age at which the hearing loss was first suspected. This may relate to the age the child was referred from the newborn hearing screening programme.

Resource Teachers: Deaf (RTDs)[i]**:** Resource Teachers of the Deaf (RTDs) provide a range of teaching and specialist services to Deaf and hearing impaired students in mainstream schools around the country. Eligibility is decided on the basis of individual need, and recognises the importance of language, communication and culture to a student's success. Caseloads are reviewed each term and measured against specific eligibility criteria.

An RTD is a trained specialist teacher who can:

- Provide specialist 1:1 teaching
- Assist classroom teachers with curriculum adaptation and delivery
- Provide specialist advice, guidance and assistance for classroom environment and management
- Assist classroom teachers with the assessment of learning outcomes involving language and literacy achievement
- Liaise with all staff, support agencies and caregivers
- Monitor and support the use of audiological equipment
- Provide improved access to the curriculum for Deaf and hearing impaired students

The ASSIST programme (Assessment Involving Specialist Teacher) has been implemented by KDEC and Van Asch Deaf Centres region by region across New Zealand since 2013. The ASSIST team consists of Resource Teachers of the Deaf who work in an ASSIST role with students who are deaf and hard of hearing and are in Years 4 to Year 13. Their work currently comprises the management of student's audiological equipment, responding to notifications via audiology and gathering assessment data on students' language development.

Unilateral hearing loss: Hearing loss affecting one ear. With regard to the DND, there may be minimal hearing loss in the other ear but it qualifies as unilateral where the hearing loss in the other ear does not meet the 26 dB HL four frequency average criterion.

Universal newborn hearing screening and early intervention programme (UNHSEIP): This New Zealand programme, managed by the National Screening Unit as part of the Ministry of Health, aims to provide early and appropriate intervention services to all children born with permanent congenital hearing impairment. Children are screened soon after birth and those who 'refer' on this screening are referred to see an audiologist who conducts a full diagnostic assessment. Children diagnosed with a hearing loss then have access to the very important early intervention services they require to allow improved outcomes.

van Asch Deaf Education Centre (vADEC): van Asch Deaf Education Centre provides educational programmes and services to Deaf and hard of hearing students, from roughly Taupo southwards.

Vision Hearing Technician (VHT): Vision Hearing Technicians are employed by district health boards to screen children around the country for hearing and vision problems. Hearing screening involves audiometry and if the child refers on this screening, tympanometry is also conducted. The work of the VHT includes vision and hearing screening done as part of the B4 School Check.

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analysis of data from high frequency hearing losses notified in 2013 can be found in

Appendix B: High frequency hearing losses, on page 47.

[i] This information is adapted from a very helpful description which can be found here on the KDEC website.

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