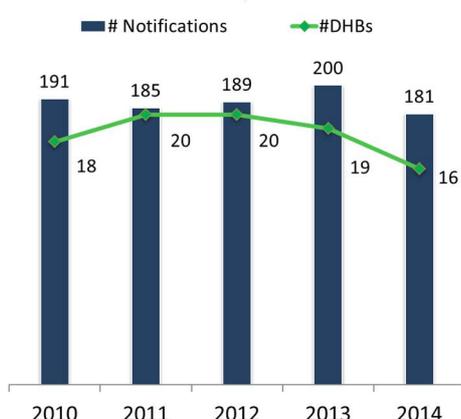
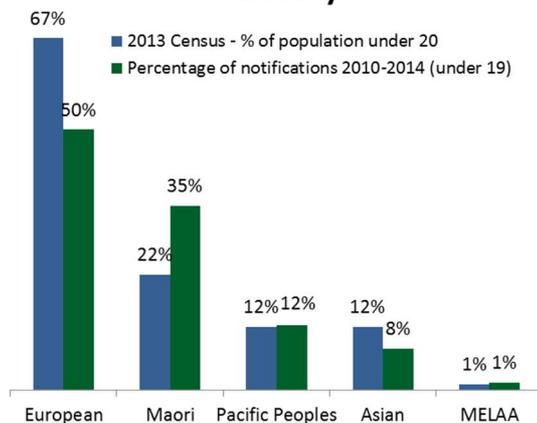


2010-2014 Notifications: A summary

Number of DHBs/Notifications



Ethnicity



More likely to be identified later

- born overseas
- mild hearing losses
- acquired hearing losses
- unilateral hearing losses

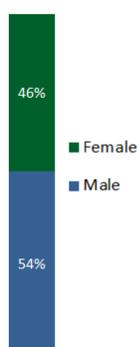
More likely to be identified earlier

- born in New Zealand
- profound hearing loss
- losses thought to have been present at birth
- bilateral hearing losses

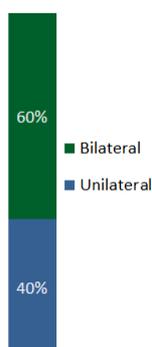
Most common reasons for delay (see report for more details)

Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)	40%
Parents did not attend appointments	28%
Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, get GA ABR, no audiology staff at the DHB, limited staff resource)	27%
Difficulty getting a referral to audiology (e.g. VHT referral not accepted by DHB, GP or other health professional dismissed parent concern and no referral was made)	12%
Parents suspected something other than hearing loss (e.g. speech delay, developmental delay, selective hearing)	11%

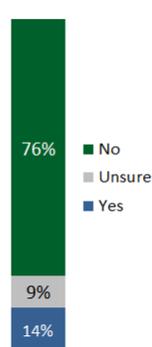
Gender



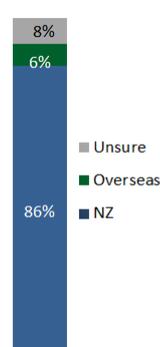
Bilateral/Unilateral



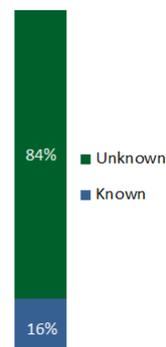
Additional disabilities



Birthplace



Aetiology



Citation: Digby JE, Purdy SC, Kelly AS (2015) *Deafness Notification Report (2014) Hearing loss (not remediable by grommets) in New Zealanders under the age of 19*. **accessible**. Auckland, New Zealand.

This and previous reports are available on the New Zealand Audiological Society website:

<http://www.audiology.org.nz>

This report can be freely quoted, copied and circulated with appropriate acknowledgement.

For notifying audiologists

The authors of this report would like to extend a big thank you to all audiologists who have provided notifications to the database for the 2014 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](#).

Notes for audiologists completing notifications for the database:

- 1. Consent changes:** We are delighted to announce 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 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 in line with similar questions asked in overseas jurisdictions.
- 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 all audiologists to get their notifications into the database as soon as possible following diagnosis, and always before the end of the notification period at the beginning 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 has been extended to mid-March the following year. Written consent from families will still be 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@leware.co.nz or by telephone (09) 445-6006. If in doubt about whether a case meets the criteria, please notify the case.

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The 2014 Report

Introduction

Welcome to the fifth annual report describing notifications to the re-launched Deafness Notification Database (DND). This report includes diagnoses made throughout New Zealand during the 2014 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 earsⁱ,
- 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 42 of this report.

Steps have been taken to ensure data contained within 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 within this report describe characteristics of the full number of 2014 notifications for which data were provided.

A glossary of commonly used terms can be found on page 47 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, which 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 exist for this group¹.

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 45.

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 which would not meet the original criteria, but which 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 40.

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 181 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 individual audiologists 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 its own unique challenges.

This report has been funded by [accessible](#), 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 nature of new diagnoses in New Zealand.

The primary author of this report gratefully acknowledges the significant support and guidance of Prof Suzanne Purdy of the University of Auckland and Dr Andrea Kelly of ADHB.

Dr David Welch, Mr Colin Brown and Prof 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@leware.co.nz or by telephone, (09) 445-6006.

Notifications

General information

One hundred and eighty one notifications pertaining to cases first diagnosed during the 2014 calendar year, and meeting the criteria for inclusion, were received by the 13th of March 2015, this year's cut off for 2014 notificationsⁱ. These notifications were received from a total of 54 audiologists, with notifications from 16 of the 20 district health board areas. This number is down from the 200 notifications received in 2013, which were received from 19 District Health Boards (DHBs) and 49 audiologists.

An additional 17 high frequency hearing losses were also notified to the database for the 2014 year. These are described in Appendix B on page 44.

It 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.

Of 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.

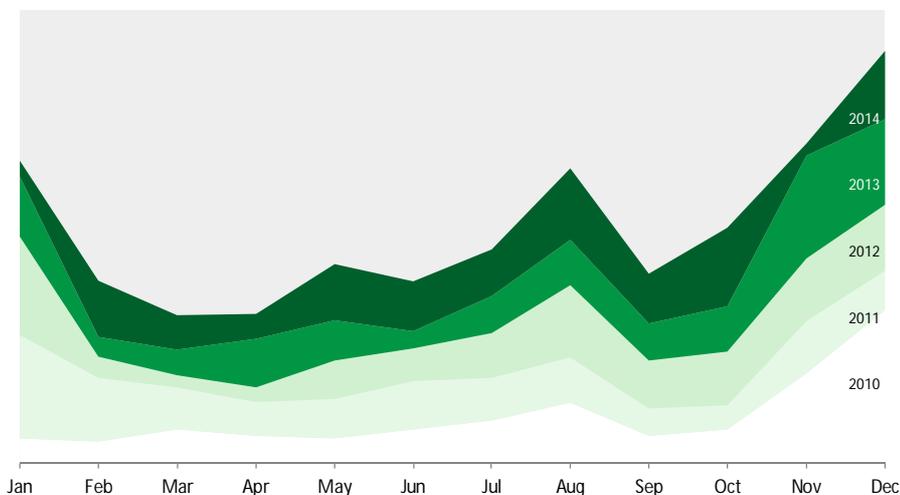


FIGURE 1: STACKED AREA CHART SHOWING NOTIFICATIONS BY MONTH (2010-2014)

Gender

Slightly more of the cases notified to the database were male (51%) than female (49%), although this difference is smaller than in previous years.

The ratio of boys to girls has ranged from a low of 1:1.06 (2014) to a high of 1:1.34 (2013), with an average of 1:1.16 during the period since the database was re-launched in 2010.

ⁱ The 13th of 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 was variable, and increased 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.

With the exception of 2014, 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% males (1:1.08 and 1:1.38) in various jurisdictions within 2011's Comprehensive Handbook of Pediatric Audiology².

A statistical 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.

Number of notifications

Notifications are collected through an online survey form to reduce data entry errors and make it as easy as possible for audiologists 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.

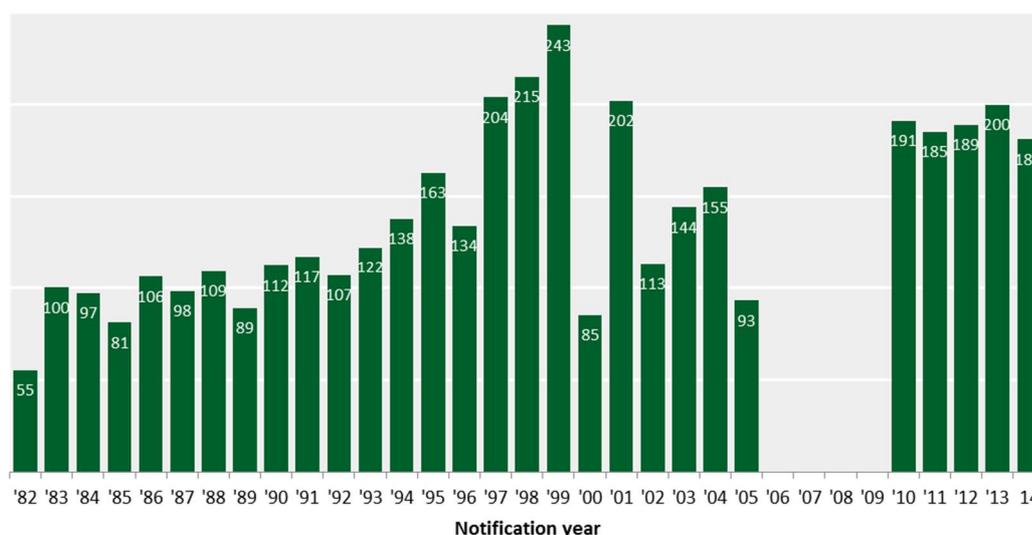


FIGURE 2: NOTIFICATIONS BY YEAR 1982-2005 AND 2010-2014

Figure 2 shows the number of notifications meeting the criteria in each year. Information about how the inclusion criteria have changed over time is included in Appendix A which begins on page 42. Please note that the 2001 to 2005 figures, included in previous reports, were later revised by the manager of the database at the time, Auckland District Health Board (ADHB). Figure 2 shows the number of notifications which met all inclusion criteria at the time and were included in the database's annual reportsⁱ rather than including the revised figures.

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 2010-2013;
- the period from 1982 to 2005 contains notifications to the original National Audiology Centre/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 2014 relate to notifications provided to the newly re-launched database.

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

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

- slight losses (those not meeting the 26 dB HL average across four frequencies in at least one ear)ⁱ;
- cases where the child or young person was reported as having mild hearing loss with normal bone conduction thresholdsⁱⁱ (assumed to be a transient conductive hearing loss unless a permanent conductive hearing loss was specifically stated, e.g. ossicular fixation);
- notifications with significant missing information where no further information was provided by the audiologist on request; and
- notifications which didn't indicate consent had been provided by the parent/caregiver.

Figure 2 also illustrates the variability in the number of valid notifications provided to the original database, particularly in the last six years of its operationⁱⁱⁱ.

There have been small changes in the number of notifications included in the database since 2010. The reasons for these changes are described below:

- The ADHB kindly allowed access to the original DND dataset from 2012^{iv} (1982-2005), so that new notifications could be checked against those received previously to ensure no duplicates were being included in the current analyses. Duplicates were identified based on National Health Index (NHI) and by name, using fuzzy matches to detect potential duplicates which couldn't be identified based on the NHI, owing to missing information or data entry errors in the ADHB dataset. A small number of small changes to pre-2012 data appear in this year's report (compared with 2010 and 2011 reports) as a result of access to this information.
- Eleven additional notifications which were submitted late for the 2010 year have been included in the dataset, as they met the criteria for inclusion and some allowance was made for audiologists being slow to respond to the 2010 re-launch of the DND.
- Occasionally, an audiologist will report to us that a diagnosis previously notified to the database and which at that time met the criteria for inclusion has been revised and no longer meets the criteria for the database. These cases are removed from the database.

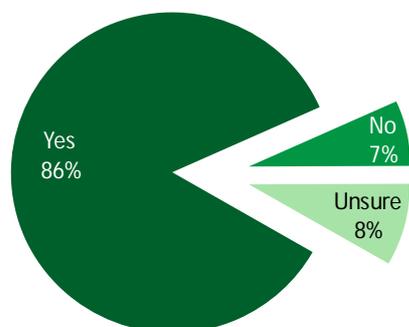


FIGURE 3: PROPORTION OF 2014 CASES BORN IN NEW ZEALAND

Birthplace

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

As shown in Figure 3, of the 181 cases included in the main analysis in 2014, 7% were known to be born outside New Zealand. Birthplace was uncertain in a further 8% of cases reported to the database.

Between 3% and 9% of cases in the database were born outside New Zealand between 2010 and 2014.

ⁱ High frequency hearing losses are included separately in Appendix B.

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

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} Access to this dataset allowed the authors to confirm that the notifications included in the database prior to 2005 included a number of duplicates, and we can confirm that the number of notifications reported before 2005 was artificially inflated as a result. There is no way of understanding which cases were included in each of the previous database's annual datasets, so the number of notifications listed in Figure 2 includes those figures reported in the original annual reports at the time of their publication.

DHB representation

Table 1 contains the percentage of 2014 notifications from each DHB area and compares these with the percentage of the population under the age of 20 from the 2013 Censusⁱ.

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 FTE audiologists employed by each DHB;
- the workload of these audiologists; and
- the level of commitment among staff to making notifications to the database.

It is worth noting that, historically, 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 the DHBs reporting higher numbers of notifications than anticipated based on their population are almost all (with the exception being Canterbury) those DHBs with a higher proportion of Māori and/or Pacific populations (e.g. Counties Manukau, Northland, Bay of Plenty).

	Percentage of notifications received 2014 (under 19 years)	Percentage of population under the age of 20 (Statistics New Zealand, 2013 Census)
Auckland	5%	11%
Bay of Plenty	7%	4%
Canterbury	17%	11%
Capital and Coast	3%	7%
Counties Manukau	15%	13%
Hawke's Bay	0%	3%
Hutt	5%	3%
Lakes	4%	2%
Midcentral	5%	4%
Nelson Marlborough	5%	3%
Northland	9%	3%
South Canterbury	0%	1%
Southern	8%	7%
Tairāwhiti	2%	1%
Taranaki	6%	2%
Waikato	7%	9%
Wairarapa	0%	1%
Waitemata	3%	13%
West Coast	1%	1%
Whanganui	0%	1%

TABLE 1: PERCENTAGE OF NOTIFICATIONS (2014) 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 2014 data include:

- South Canterbury DHB (a small DHB) which reported that they did not diagnose any children or young people under the age of 19 in 2014;
- Whanganui, Wairarapa and West Coast are all relatively small DHBs, and may not have had any diagnoses during the 2014 year;
- Auckland and Waitemataⁱ DHBs' notifications are lower than would be expected owing to consenting issues with previous cases. They hope this issue will be resolved with the change in consenting processes which will allow children screened by the UNHSEIP to automatically be consented for the DND.

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.

Of 2014 notifications, 16% were thought to have disabilities *in addition* to hearing loss at the time the notification was made. In a further 8% of cases there was uncertainty regarding whether the child or young person had an additional disabilityⁱⁱ.

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^{3,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.

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%

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

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.

because children may have not yet been diagnosed with these conditions, or they have conditions that have not yet developed (e.g. vision impairments are more common in older children and diagnoses of autism spectrum disorder are typically not made in the first year of life).

Other possible reasons for 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 2013⁴. 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 smallⁱ.

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 within 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 ⁵	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: <ul style="list-style-type: none"> • visual impairment (4-57% depending on the definition) • neurodevelopmental disorders (2-14%) • speech language disorders (61-88%)
Fortnum et al ⁶	2002	UK	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis ⁷	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 ⁸	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 ⁹	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 impairment
The Consortium for Research into Deaf Education ¹⁰	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

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.

Bilateral and unilateral loss

Unilateral hearing losses are known to impact on educational performance, and a significant proportion of these hearing losses progress over time¹¹. As a result, cases of unilateral loss, where these average more than 26 dB HL in the hearing impaired earⁱ, 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. Comparing the proportion of unilateral/bilateral notifications with previous DND data (prior to 2005) is not possible - although a number were reported prior to 2006, these were not well reported and so are incomplete within this older dataset.

In DND reports since 2010, the proportion of bilateral and unilateral losses was calculated based on cases with full audiometric data. These data downplayed the true proportion of hearing losses which were bilateral, and so, from now on, we will also present the proportion of bilateral and unilateral hearing losses based on interpolated figuresⁱⁱ and using manual checks for those records which cannot have data interpolated:

- the proportion of 2010-2014 cases which were bilateral/unilateral using only cases with full audiometric data was 60:40 and 57% of 2014 DND cases were bilateral in nature; and
- the proportion of 2010-2014 cases which were bilateral/unilateral using *interpolated* data and manual checks was 66:34 and 67% of 2014 DND cases were bilateral in nature.

While immunisation coverage (including for conditions such as mumps) in New Zealand has risen significantly from 45% in 2007 to 92% in 2012¹² 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 loss¹³.

Differences between the proportions of bilateral and unilateral notifications within each severity category are shown on page 34 .

Types of hearing loss

Information on the types of hearing loss notified is now being collected for each hearing impaired earⁱⁱⁱ.

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, i.e. this group is effectively a subgroup of the sensorineural category.

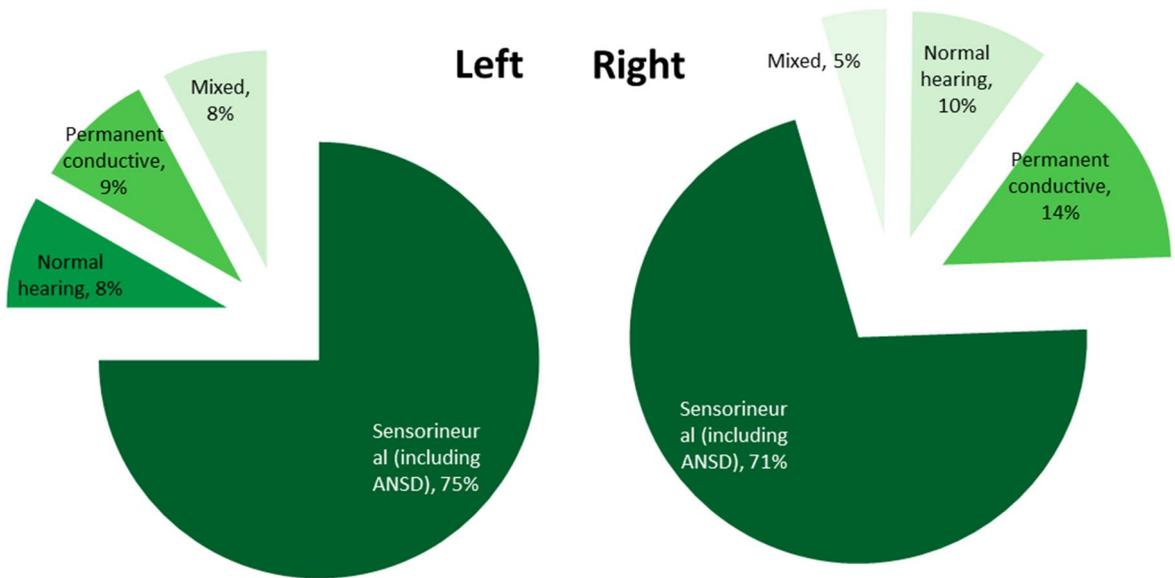
The most commonly reported type of hearing loss within notifications to the DND which included this information, was sensorineural (71% in the right ear and 75% in the left), followed by permanent conductive losses (9% in left ears and 14% in right ears – some of which were atresia) and mixed losses (11% in left ears and 9% in right ears). Two percent of all cases were recorded as ANSD (3% of the sensorineural category), while 1 to 2% of cases were listed as uncertain type.

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 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 surrounding data-points are provided.

ⁱⁱⁱ 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.

We hope to be able to include further information on hearing loss types in future reports, now that these data are regularly being collected.



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.

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

The previous question asked whether there was a family history of hearing loss. The results from this question are below. *Please keep in mind that data from 2014 contain information from approximately half the notifications for that period, as the question was changed in the middle of the year – the smaller sample here may explain the variation for this year's result.*

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)."

Of the 49 notifications where this question was completed, 86% recorded 'no', 2% recorded 'don't know' and 12% recorded 'yes'. 2015 data will span the full notification period and will be included in next year's report.

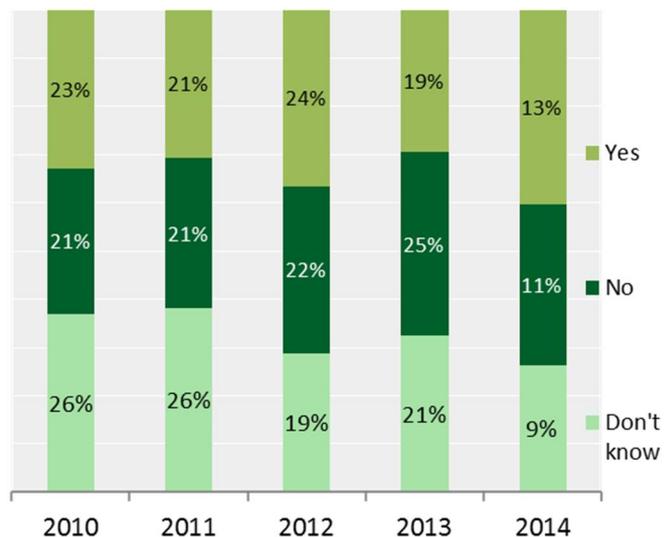


FIGURE 4: FAMILY HISTORY OF HEARING LOSS

Ethnicity

Representation

All but four of the 2014 notifications contained one or more ethnicity codes, although only a fairly small proportion of notifications specified more than one ethnic group. Of those with one or more codes, 91% of respondents selected one code for their child's ethnicity, while 6% selected two

codes and 0.5% selected three.

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 ethnicity.

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

Multi-coded 2013 Census data was not available at the time the last report was published, so it is

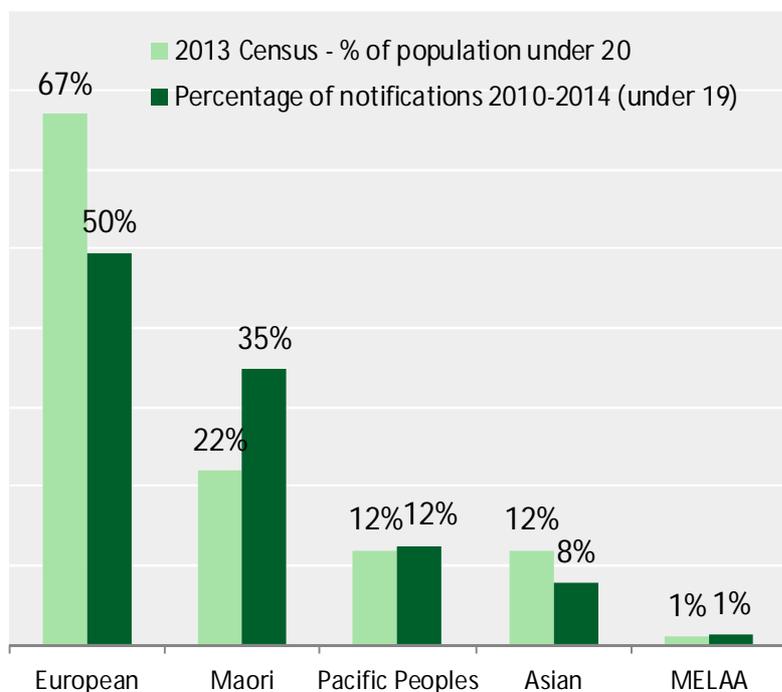


FIGURE 5: COMPARISON OF TOTAL ETHNICITY HEARING LOSS NOTIFICATIONS (2010-2014 UNDER 19 YEARS OF AGE) TO CENSUS DATA (2013 – UNDER 20 YEARS OF AGE).

included in Figure 5 for the first time. 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 2010-2014 (which includes those under the age of 19ⁱⁱ).

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. This is aligned with findings from Digby *et al* (2014)¹⁴ which indicated young Māori have higher rates of permanent hearing loss than their European counterparts.

However, please keep in mind that generally fewer children and young people in the DND dataset have more than one ethnicity code than in the Census data. This may mean that coding for ethnicity is less complete for deafness notifications than for the Census data, which would affect this comparison.

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.

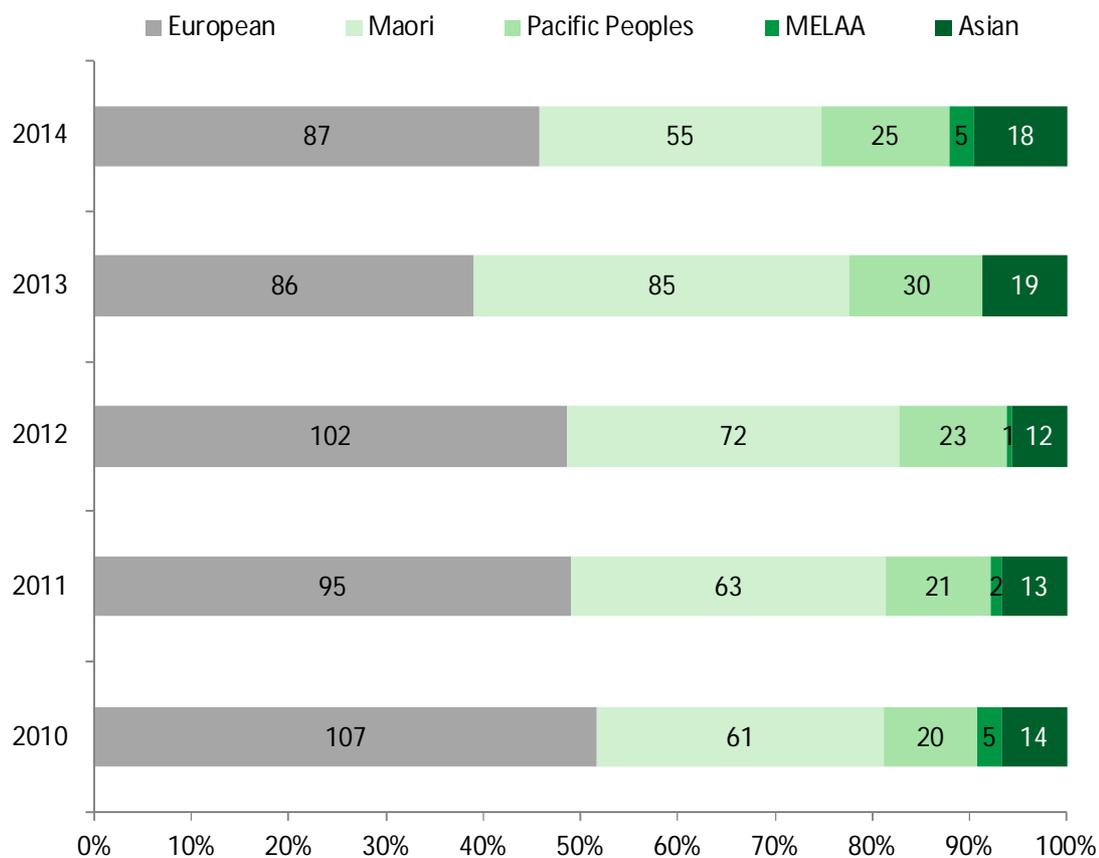


FIGURE 6: ETHNICITY OF 2010-14 CASES

Ethnicity differences

A number of sources suggest possible differences in prevalence of hearing loss between Māori and New Zealand Europeans, although no difference has ever been confirmed:

- The Household Disability Surveys (1991-2006¹⁵) – these 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-Māori¹⁶. *For more about the limitations of this data please see the 2011 DND Report¹⁷.*
- Referral rates from the [B4 School Check¹](#) (2011) analysed by Searchfield *et al.*, show higher rates of referral from hearing screening for Māori children (9%) compared with non-Māori (5%)¹⁸. It is important to note that high referral rates for Māori may indicate higher rates of ear disease as these figures do not just relate to permanent hearing loss.
- Universal newborn hearing screening: While only limited programme data is available to describe diagnoses resulting from newborn hearing screening, Māori children were referred at higher rates (2.4% compared with their New Zealand European counterparts 1.2%) and diagnosed at higher rates (13.8% compared with 10.8% for their New Zealand European counterparts) between April 2012 and December 2012¹⁹.

Historically, DND reports have shown that the greatest number of notifications pertain to Māori and New Zealand European children and young people, and that milder degrees of hearing loss are more commonly reported among Māori^{20,21}.

ⁱ For more information on the B4 School Check, please click [here](#) or view the glossary on page 52.

A recent analysis of DND data²² indicates significant differences exist in both the prevalence of hearing loss and severity profile, and that young Māori are less likely to be reported as having severe or profound losses than their New Zealand European peers. This information can be used to inform policy and practice in both screening and clinic settings, to identify hearing loss early and allow engagement with intervention services. This is particularly important as Māori health is 'characterised by systematic disparities in health outcomes, exposure to the determinants of health and health system responsiveness'²³.

Although Māori are well represented among hearing loss notifications, this group may still be underrepresented in DND statistics because of their greater chance of having a less severe (mild or moderate) hearing loss. It may also be that disparities in 'access to, and within, the health system'²⁴ for Māori may mean not all cases are found or notified.

For further information on ethnicity coding within the database, please refer to Appendix C: Notifications and ethnicity, on page 45.

Unilateral and bilateral losses

A total of 926 children and young people with completed ethnicity information were notified and met the criteria during the 2010 to 2014 period. These data contain some records (less than 10%) which had multiple codes for ethnicity, and so appear in more than one group.

Considering 2010-2014 cases with at least one ethnicity code 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.

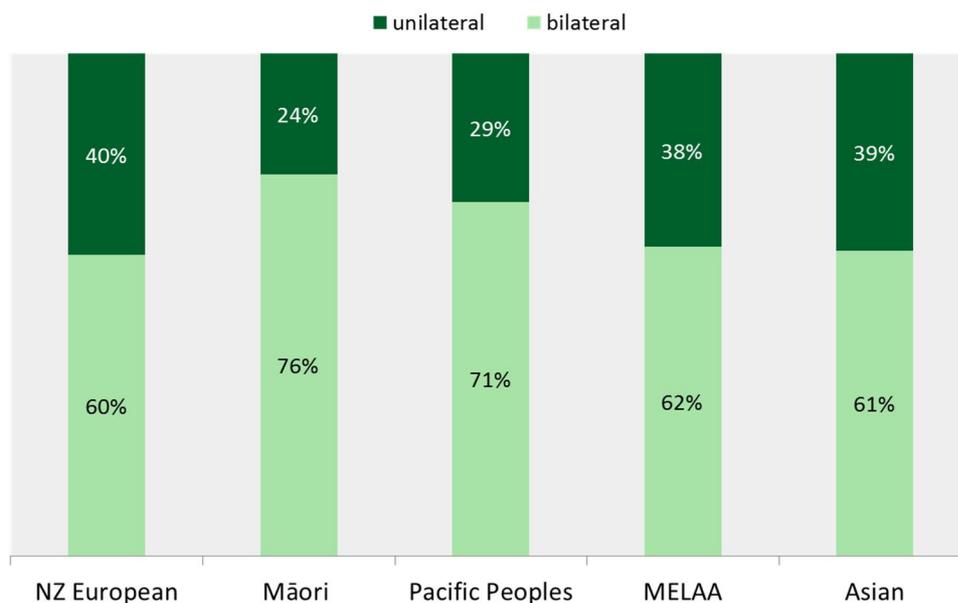


Figure 7 below shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group. Please note that MELAA figures relate only to very small sample sizes for the 2010 to 2014 period.

The significant difference between Māori and New Zealand European rates of bilateral loss supports the 2014 paper by Digby *et al.* which confirms a larger proportion of bilateral hearing losses among young Māori when compared with New Zealand European counterparts.

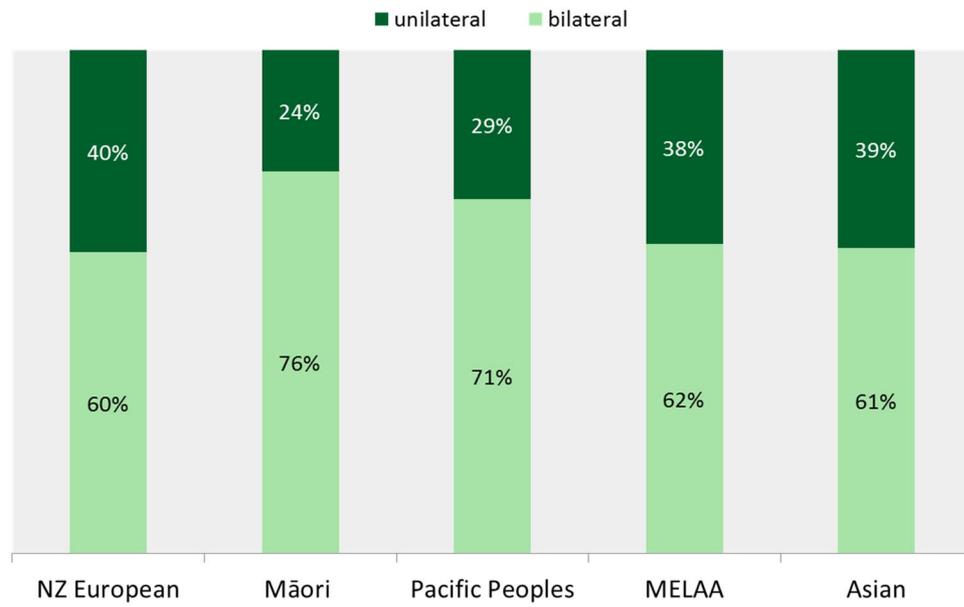


FIGURE 7: PROPORTION OF UNILATERAL AND BILATERAL HEARING LOSSES BY ETHNICITY (2014) FOR CASES WHICH HAVE AT LEAST ONE ETHNICITY CODE, BASED ON INTERPOLATED DATA AND MANUAL CHECKS TO DETERMINE BILATERAL/UNILATERAL STATUS

Aetiology

All but two of the 181 cases which met the [inclusion criteria](#) for the 2014 period, contained information relating to the cause of the hearing loss.

As seen in Figure 8 below, the proportion of hearing losses where the cause was thought to be known has decreased significantly through the period 2010 to 2014, when compared with figures from before 2010. 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 earlier with hearing loss, owing to the introduction and roll-out of newborn hearing screening. 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 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.

Additional genetic causes of hearing loss are being identified as genetic research progresses^{25,26}, however genetic testing is typically not occurring early, or in all cases of identified hearing loss.

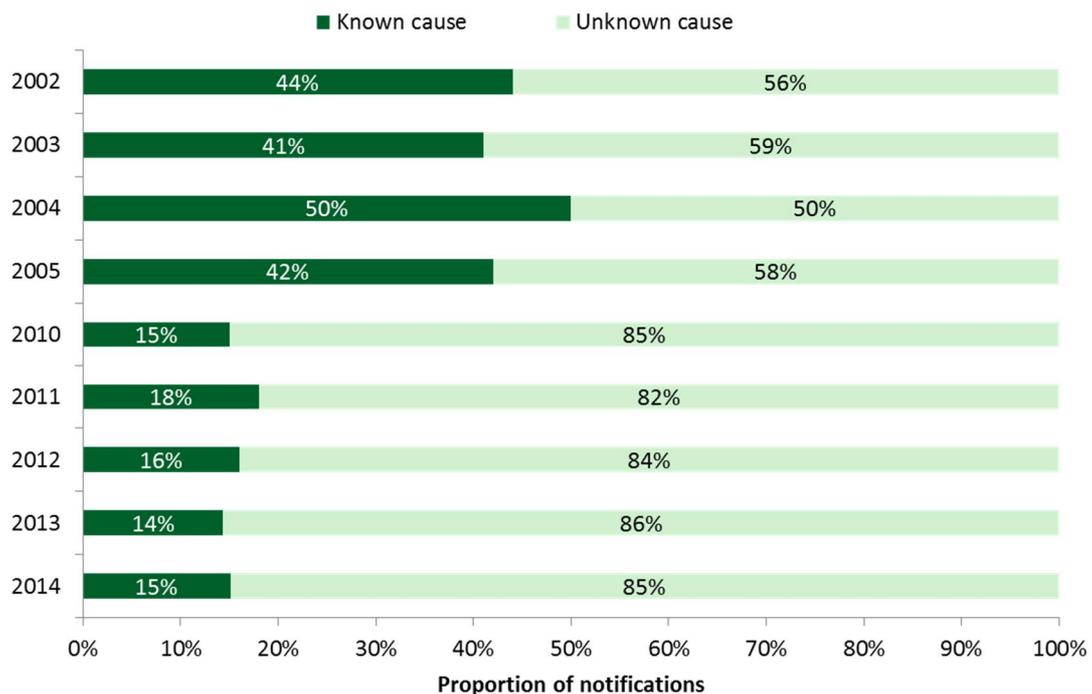


FIGURE 8: PROPORTION OF HEARING LOSSES OF KNOWN AND UNKNOWN CAUSE NOTIFIED TO THE DND BY YEAR DIAGNOSED

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 scans²⁷.

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. The most common mutations found are in the GJB2 and Pendrin genes. The Otoferlin gene has been implicated in cases of ANSD²⁸. ENT specialists request the tests and

counsel patients about the results. If multiple or unusual mutations exist, ENT specialists refer to genetic services²⁹.

Table 4, below, shows the aetiological breakdown where the aetiology was recorded as 'known'.

Aetiology breakdown	2010 (n=)	2011 (n=)	2012 (n=)	2013 (n=)	2014 (n=)
Acquired hearing loss	11	16	17	11	11
Genetic cause (Non-syndromic)	3	2	3	1	4
Syndromic	3	4	6	6	9
Other	6	6	3	2	3
Not listed	2	5	4	8	0

TABLE 4: KNOWN AETIOLOGY BY TYPE – FIGURES SHOWS ARE THE NUMBER OF CASES (2010-2014)

Of the cases of hearing loss diagnosed in 2014 and notified to the DND, none were listed as being the result of meningitis, while one was listed as being caused by mumps. This compares with three cases in 2012, four cases in 2011, and six cases in 2010 listed as being the result of meningitis³⁰. However, it is worth noting that this information is not specifically requested and so may be incomplete.

Overseas, [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 cases which are milder in terms of their severity³¹.

In New Zealand, during the 2010-2014 period, bilateral hearing losses and more severe losses were more likely to have a known aetiology than those which are unilateral.

Internationally, as reported by Davis and Davis (2011), it is common for a high proportion of cases (between 15% and 57%) of hearing loss to be of unknown aetiology. The proportion of hearing losses with a confirmed genetic cause is likely to increase over time, as more hearing losses become understood in terms of their aetiology, and as genetic testing becomes cheaper and more widely available.

It is worth noting that identification of one aetiology does not exclude another aetiology. For example, the A1555G mitochondrial mutations may predispose a patient to hearing loss, and this hearing loss is expressed when certain antibiotics are used³².

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 150 children and young people confirmed as being born in NZ and who were diagnosed in 2014.

Table 5 shows the top three groups which first suspected hearing losses among notified cases since the re-launch of the database in 2010. Other groups who commonly suspected hearing losses first in 2014 included: medical professionals (other than the GP) – 9%, and educators and teachers – 3%.

2010	2011	2012	2013	2014
Parent or caregiver (49%)	Parent or caregiver (42%)	Parent or caregiver (33%)	Newborn hearing screener (34%)	Newborn hearing screener (40%)
VHT (22%)	Medical Professional (21%)	VHT (23%)	Parent or caregiver (20%)	Parent or caregiver (22%)
Medical Professional (12%)	Vision hearing technician (15%)	Newborn hearing screener (23%)	Vision hearing technician (16%)	Vision hearing technician (13%)

TABLE 5: THREE GROUPS MOST LIKELY TO FIRST SUSPECT A HEARING LOSS 2010-2014 (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^{33,34}. 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. For the first time in 2013, newborn hearing screeners, not parents, are most commonly the first to suspect hearing lossⁱ.

The proportion of 2014 cases first suspected by parents or caregivers is now 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.

Age at diagnosis

Figure 9 shows the number of cases identified by the age of the child. The majority of children having their B4 School Check since the end of 2013 will have been screened for hearing loss soon after birth.

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.

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 more than twice as high as it was in 2010, when the database was re-launched. This is a positive trend, as more children are being diagnosed early.

A further peak can be seen for four, five and six year olds; this is likely to correspond to the B4 School Check³⁵. 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.

[Recent Ministry of Health data](#) from the B4 School Checks (2013/14 year) estimate that 91% of eligible children received a B4 School Check, although this figure is not broken down by aspect of the check.

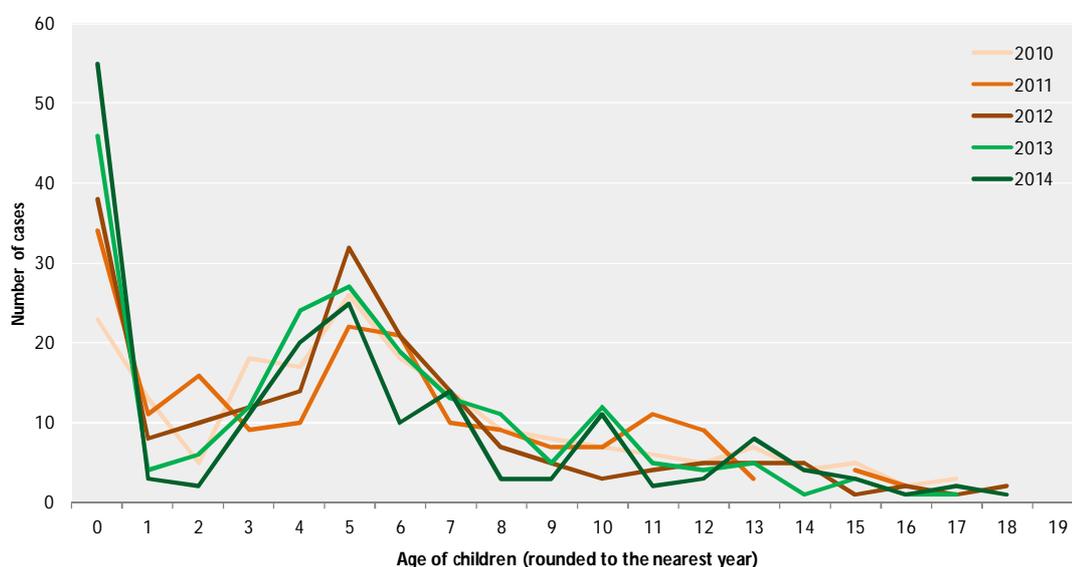


FIGURE 9: NUMBER OF CHILDREN DIAGNOSED BY AGE (2010-2014)

Overall age at identification

There are a number of issues with reporting the average age at identification (diagnosis) for all groups of children. While this may 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 which are late onset (such as progressive and acquired hearing losses).

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, while the average age at [confirmation](#) is dropping year on year, the reduction is quite slow and seems to have been influenced by the increase in the number of notifications around 5 years of age for 2012 and 2013 as well as the increases at 10 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.

	2010	2011	2012	2013	2014
Average all cases	68	62 ⁱ	61	61	60
Average born in New Zealand	65	57	56	54	53

TABLE 6: AVERAGE AGES OF DIAGNOSIS FOR ALL CASES IN MONTHS (2010-2014)

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

For the purpose of comparison with previous data, the average age at diagnosis is presented, but the average age for 2014 has also been split by further subgroups in Table 7 below, to add meaning to this measure.

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 (2014)

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 a reasonable number of records in the database contain incomplete severity data, and also that the table below only includes cases of bilateral hearing loss, so these figures do not describe the age at diagnosis for the full 2014 cohort. Those cases with incomplete severity data are more likely to have been diagnosed as babies and as a result are more likely to have an earlier diagnosis than the average found in the full sample.

Please note that 'moderately severe', 'severe' and 'profound' categories contain small samples. Younger children are more likely to be missing some severity data, meaning they could not be classified for the table below.

ⁱ This figure has been revised from 57 as previously reported to take into account the fact that a number of entries had missing confirmation dates (this data is now required as part of the form) and also that a small number of records have been reviewed against the original DND database, enabling a small number of duplicates from 2011 to be removed.

ⁱⁱ 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.

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2011)	Average months at diagnosis (2012)	Average months at diagnosis (2013)	Average months at diagnosis (2014)
Mild	81	78	75	81
Moderate	79	68	65	58
Moderately severe	53	42	NA (not enough cases)	70
Severe	48	NA (not enough cases)	NA (not enough cases)	29
Profound	39	22	18	NA (not enough cases, n=1)

TABLE 8: AVERAGE AGE AT DIAGNOSIS, IN MONTHS, FOR *BILATERAL* HEARING LOSSES BY DEGREE CONTAINING SEVERITY DATA (2014)ⁱ

The greatest variability in the age at diagnosis is for mild and moderate hearing losses – understandable given that these are the largest groups. 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 10 shows the average identification ages by 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 can be 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, which are on average identified later than severe and profound losses³⁶. This makes 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 within 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.

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.

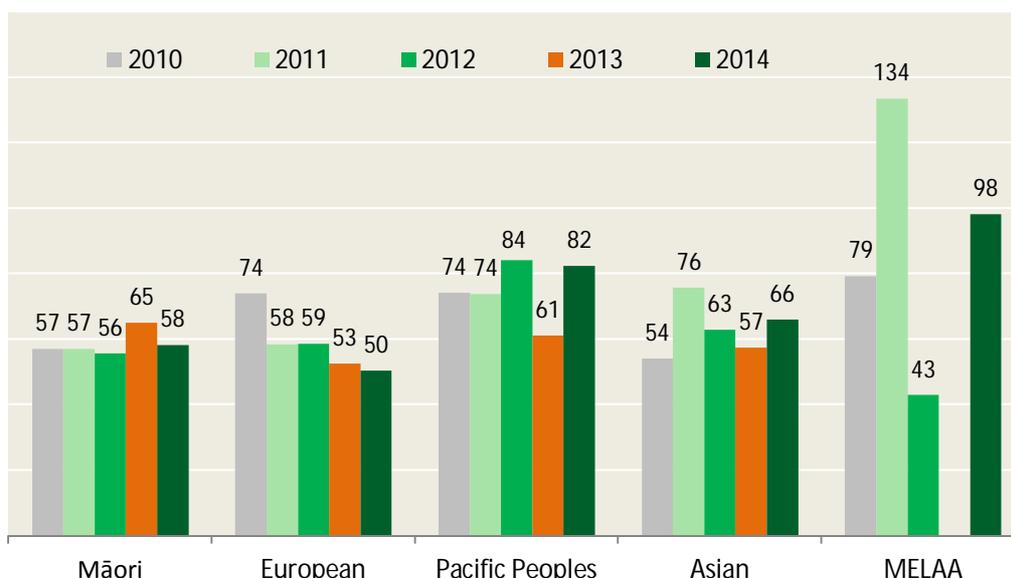


FIGURE 10: AVERAGE AGE OF DIAGNOSIS BY ETHNICITY IN MONTHS (2010-2014)

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 reportⁱ.

Newborn hearing screening

All district health boards have been screening babies for the full notification period (calendar years) since 2011ⁱⁱ. 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 NZ-born children notified (and therefore diagnosed) in the period 2010 to 2014.

Please note that this table shows children diagnosed at varying ages, so just over a third (38%, n=56) of the cases notified for 2014 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.

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
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%
	No, this service was not available at the time (at the time of diagnosis)	69%	54%	55%	48%	38%
Unsure	Unsure whether screening was offered to this family	7%	4%	6%	6%	5%

ⁱ 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 NZ European children being identified, on average, earlier than Māori and Pacific children.

ⁱⁱ 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 within 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
Yes	Yes screening was offered but this child was not screened	1%	1%	1%	0%	0%
	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%
	Yes, this child was screened and passed	1%	7%	5%	7%	12%
	Yes, this diagnosis is a result of a refer on the screening test	19%	27%	27%	32%	38%

TABLE 9: SCREENING STATUS OF CHILDREN BORN IN NEW ZEALAND AND DIAGNOSED 2010-2014ⁱ

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^{37, 38, 39, 40}.

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

A total of 57 of the 2014 notifications (38%) related to children born in New Zealand who were diagnosed as a direct result of newborn hearing screening. This percentage has doubled since the database was re-launched in 2010.

It is not known how many cases of hearing loss are currently missed from 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
Number of diagnoses resulting from universal newborn hearing screening	28	44	45	53	57
Diagnoses as a proportion of total notifications	16%	27%	27%	32%	38%

TABLE 10: DIAGNOSES RESULTING FROM NEWBORN HEARING SCREENING IN NEW ZEALANDⁱⁱⁱ, 2010-2014

The most recent NSU monitoring report, including screening data from 1 January to 30th June 2013 and audiology data to the end of January 2014, reports that:

- approximately 87.5% of babies born across the country during this period completed their newborn hearing screening;
- of those babies who completed screening, approximately 93% did this by the target of one month (corrected age) – figures for individual DHBs ranged from 44% to almost 100%;

ⁱ Please note that some figures in this table have been rounded and so not all sum to 100%.

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 NZ.

- 82.8% of those babies completing audiology had their audiology assessment completed by the target of three months of age;
- audiological data was provided to NSU for 92% of referred babies, a significant improvement in recent years; and
- 153 babies in total were identified with some type of hearing loss during the period, including those with temporary losses⁴¹; 39 of these were cases of permanent congenital hearing loss.

Notifications from two large DHBs (Auckland and Waitemata) are thought to be underrepresented in the 2014 database. (See page 10 for further information.) This is likely to have reduced the number of potential DND notifications resulting from UNHS that were notified in 2014.

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 10 months in 2010 to 8 months in 2011 and 5 months in 2012, 6 months in 2013 and now to 5 months in 2014.

Of the 57 cases notified in 2014 which were identified as a result of newborn hearing screening within NZ, 44 (77%) 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 3 months for diagnosis has still not been met across all cases.

Identification of false negatives

The DND probably provides the only method for identifying potential [false negatives](#) from the newborn hearing screening programme⁴².

Cases included in the potential false negative category (below) may be the result of 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.

Eighteen of the children identified with hearing loss during 2014 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 birth – these losses account for approximately half of all cases of hearing loss.

Of these 18 cases, there are two groups which 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 audiologist was unsure whether the hearing loss was likely

to have been present at birthⁱ. As the second of these groups is based on a relatively subjective assessment by the clinician, these cases may or may not be cause for concern.

	2010	2011	2012	2013	2014
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
Number of cases from regional screening programmes, or from the UNHSEIP, which were not acquired loss, and cases where the audiologist answered 'yes' or 'unsure' to the question about whether the loss was thought to have been present at birth	3	5	5 ⁱⁱ	6	10

TABLE 11: POTENTIAL FALSE NEGATIVES AND CASES PREVIOUSLY REFERRED FROM HEARING SCREENING, 2010-2014, BORN IN NEW ZEALAND ONLY

To narrow our focus further, we can examine cases where the hearing loss was thought to have been present at birth (as opposed to those that could have been or were) and *not* an acquired hearing loss. The cases in this group were most likely to be false negatives. This year there were ten children in this group, but this figure may not represent all false negatives from the UNHSEIP who were diagnosed during 2014.

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 November 28, 2012⁴³.

In 2014, two cases notified to the database were explicitly identified as having wrongly passed their screening (both list in the comments that the screener had screened their own ears), meaning they are confirmed false negative cases. These two children had their hearing loss identified at 4.3 and 2.8 years of age, rather than the five or six month average which would have been expected had they been referred from screening soon after birth and seen for an audiological assessment.

The audiologist notifying one further case states they were recalled by their district health board following concerns regarding screener practice. This child's hearing loss was diagnosed at 3.7 years 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.

ⁱⁱ Please note this label was incorrect in the 2012 report.

Delays in diagnosis

Information about delays

Audiologists were asked to provide information about the length of delay in identifying hearing loss and reasons for the delay, where one existed. Not all 2014 cases for which there was a delay had one or more reasons for the delay listed.

The average delay in 2014, between first suspicion of the hearing loss and confirmation of the loss, *including* those born overseas and mild, acquired or unilateral hearing losses was 13 months.

While this is a significant average delay between first suspicion of a hearing loss and confirmation of this loss, average delays in the last three years are improvements on 2010 and 2011 figures. This is likely to be owing to the introduction of, and improvements within, newborn hearing screening programmes around the country. Please keep in mind that these delay figures are not directly comparable because of the changing composition of the notifications in terms of severity, and the proportion of unilateral and bilateral notifications and the proportion of notifications which were from the UNHSEIP.

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

TABLE 12: DELAY IN MONTHS BY YEAR, 2010-2014

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 2014, 45% of all cases had one or more reasons for delay listed, with 29% having one reason, and 15% having two or more reasons for the delay listed.

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 1 month and b) greater than 6 months, measured from the time the hearing loss was first suspected, until the time when the hearing loss was diagnosed. For cases diagnosed in 2010-2014, Table 13 shows the most commonly cited reasons for delays in diagnosis.

Of particular concern are delays which are system related, such as follow up being lost in the system, waiting times to see professionals and difficulties getting a referral to audiology.

"No referral ever made to audiology despite concerns for speech development and having SLT [speech language therapy] throughout primary school."

"Referred from NBHS however did not complete follow-up due to family circumstances, then referred from VHT as a 4 year old."

"First ABR showed likely bilateral conductive HL, then would not sleep for repeat ABR, subsequent VRA appts [appointments] - distressed with things in ears, then contracted [medical condition] and seen as ward [patient] again unhappy with things in ears. Finally diagnosed with Play audiometry."

ⁱ Revised from the 20 months reported in 2011.

ⁱⁱ Revised from the 20 months reported in 2010.

Table 13 shows the reasons for delay and compares these for groups with longer (more than 6 months) delays and shorter delays (1-6 months). It also contains some possible methods for reducing the various types of delay.

Reasons for delay	Cases with one or more reason for delay and delay of more than 1 month 2010-2014	Cases with one or more reason for delay and delay of more than 6 months 2010-2014	Possible ways to reduce type of delay
Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)	40%	46%	<ul style="list-style-type: none"> efficient clinical practice to complete assessments over fewer appointments (Following Appendix F Guidelines) prompt referral from newborn hearing screening
Parents did not attend appointments (for any reason)	28%	27%	<ul style="list-style-type: none"> 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
Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)	27%	24%	<ul style="list-style-type: none"> better funding for audiology resources/DHBs to prioritise newborn hearing screening referrals and other paediatric cases
Difficulty getting a referral to audiology (e.g. GP or other health professional dismissed parent concern and no referral was made)	12%	13%	<ul style="list-style-type: none"> education for non-hearing professionals such as doctors, Speech Language Therapists (SLTs) and specialists
Parents suspected something other than hearing loss (e.g. speech delay, developmental delay, selective hearing)	11%	10%	<ul style="list-style-type: none"> 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
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	8%	7%	<ul style="list-style-type: none"> better systems and processes for scheduling and seeing follow-up occurs
Child was born or lived overseas and hearing loss not diagnosed there	4%	1%	<ul style="list-style-type: none"> 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 13: TOP REASONS FOR DELAY FOR TWO KEY GROUPS AND POSSIBLE REMEDIES (2010-2014)

Delays attributed to newborn hearing screening

Of the 9 children whose diagnosis was a direct result of referral from the UNHSEIP and whose diagnosis was later than 3 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 precedent (n=1)ⁱ;

ⁱ These six cases contained seven reasons for the delay.

More information about the causes of delays in all groups can be found in the section on *Delay causes*, beginning on page 29.

Three children whose hearing loss was identified in 2014 had been wrongly passed by the newborn hearing screening programme after the screening was performed in the screeners' ear(s). Disappointingly, these children did not have their hearing losses detected until 2.8 years, 3.7 years and 4.3 years of age, respectively.

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 as babies are 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 18 months to 2 years of age.

Severity

Audiometric data

Audiometric data are requested for both the right and left ears of all children and young people notified to the DND. Audiologists 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 Standardsⁱ.

Examining the four data points for each ear shows that these data-points were provided for 136 and 133 of the 181 cases notified to the database, for right and left ears respectively. Notifying audiologists are encouraged to provide as much audiometric data as possible for cases being notified.

Audiologists were approached about a number of cases, and were able to provide some missing information. Of the cases which 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 to audiologists in the online notification formⁱⁱ.

As shown in Figure 11, below, the proportion of cases with pure tone audiometry data is dropping slowly, from 79% in 2010 to 68% in 2013. 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.

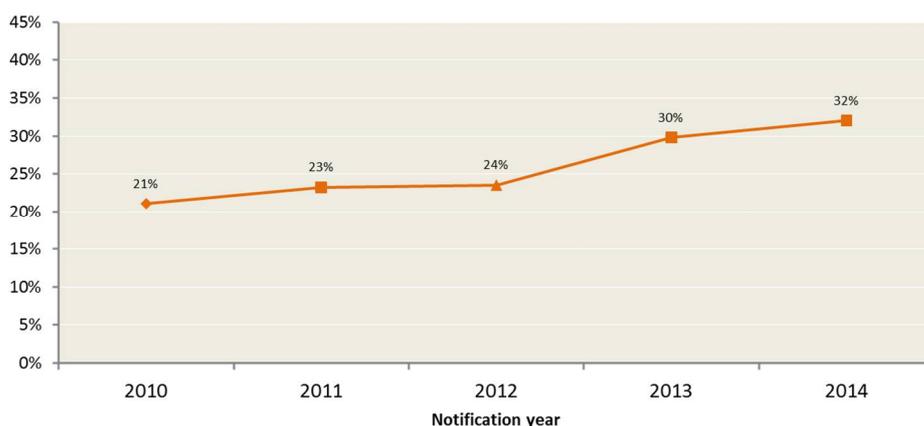


FIGURE 11: PROPORTION OF THRESHOLDS FROM ABR VS PTA, BY NOTIFICATION YEAR 2010-2014

ⁱ Correction factors: 5, 5, 0, and -5 dB for 0.5, 1.0, 2.0 and 4.0 kHz respectively as contained in Appendix F Diagnostic and Amplification Protocols

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

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. You can find further information about severity classifications in Appendix E on page 46.

Degree of loss	Clark 1981 (ASHA) ⁴⁴
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 14: 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 audiologists 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 15 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 8 data points, while the second includes interpolation. Remaining graphs in this report contain severity data from records containing all eight data points only.

While only cases where all 8 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.

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.

This analysis categorises severity based on the [ASHA](#) Clark codeframe in common use by New Zealand audiologists. Key comments on these data include the:

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

- 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.

Degree of loss using ASHA severity codeframe	Bilateral 2014	Bilateral 2014 (interpolated and manual checks)	Unilateral 2014	Unilateral 2014 (interpolated and manual checks)
Mild	64%	53%	38%	40%
Moderate	29%	28%	15%	16%
Moderately severe	1%	8%	10%	11%
Severe	5%	6%	3%	4%
Profound	1%	5%	33%	30%
Sample size	n=80	n=116	n=60	n=57

TABLE 15: COMPARISON OF SEVERITY CLASSIFICATIONS BASED ON METHODOLOGY, 2014

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 where 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.

This year’s graph, Figure 12, 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).

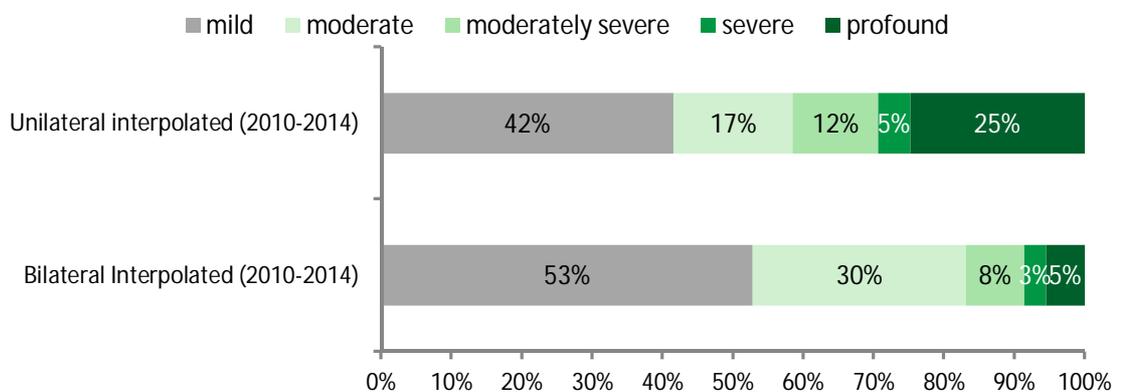


FIGURE 12: UNILATERAL AND BILATERAL HEARING LOSSES BY DEGREE (2010 - 2014)

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.

Table 16 shows the proportion of hearing loss notifications in each category between 2010 and 2014 and compares this with data from 2001 to 2004ⁱ. The 2010 to 2014 figures shown here exclude those children born overseas, unilateral hearing losses and those with acquired hearing losses, as reports prior to 2005 excluded these cases.

Proportion of cases notified by degree of hearing loss	Average 2001-2004	2010	2011	2012	2013	2014	Average 2010-2014
Mild	48%	54%	55%	47%	55%	54%	53%
Moderate	35%	35%	33%	44%	34%	33%	36%
Severe	10%	6%	7%	4%	5%	8%	6%
Profound	6%	5%	6%	5%	6%	4%	5%

TABLE 16: INTERPOLATED CASES BY DEGREE OF HEARING LOSS USING 1996-2005 CLASSIFICATION SYSTEM, SELECTED CASES ONLY

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 which more closely matched that seen before 2005. However, a return to historical patterns with fewer mild losses is not evident when only cases containing full audiometric thresholds are considered, and when compared with data in Table 15 which includes more cases by using interpolated and manually checked thresholds.

Findings this year 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;

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

- 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-representedⁱ;
- often children diagnosed with hearing loss have a sloping hearing loss and the better thresholds reduce the average degree of hearing loss; and
- 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 loss⁴⁵. However, the reduction in the number of more severe cases due to meningitis is likely to be small.

It is interesting to note that some overseas data, including those contained in Table 17 also indicate lower numbers in the severe category when compared with the profound category, including when the codeframes are standardised as they are in this case.

Ethnicity and severity profiles

The 2005 DND report noted that Māori children notified in 2005 and between 1990 and 2005 were more likely to have a mild hearing loss than other ethnic groupings. This pattern is repeated with recent data.

Figure 13, below, shows the proportion of cases in each of the various degrees of loss which were notified to the database, split by ethnicity grouping. Only bilateral hearing losses are included in this figure, as severity is categorised by the ASHA Clark classification system, and as a result, these data are not comparable to data included in the 2010 report, as that year's report included both bilateral and unilateral figures. Asian and MELAA samples have been excluded from this figure as they are particularly small samples.

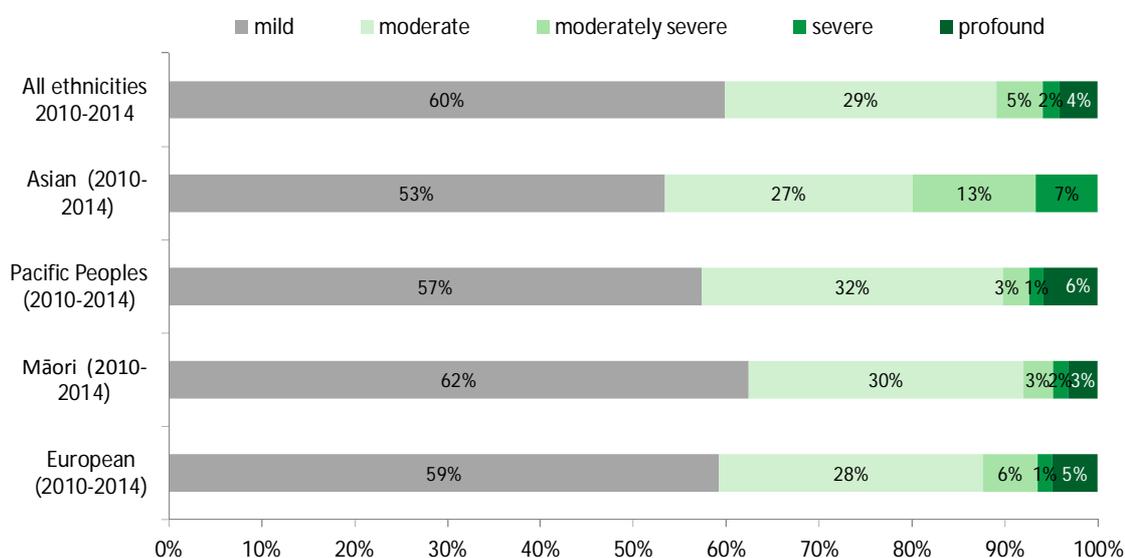


FIGURE 13: 2013 BILATERAL CASES BY DEGREE AND ETHNICITY (CASES CONTAINING COMPLETE SEVERITY DATA)

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

Comparisons with international data

Considering cases with full audiometric data only, it would seem that New Zealand may have a smaller proportion of severe and profound hearing losses than other similar countries.

The 2012 Notification report contained a comparison of moderate and greater hearing losses within 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.

The 2013 report included a table which compared our bilateral local data (2010-2014) with data from Colorado⁴⁶ 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. This data is included again in Table 17 below, and 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 added data from the slightly extended year range (2010-2014 rather than 2010-2013) and have included records with the interpolated severity data. We believe this is more accurate. The basic pattern holds, even with slightly higher numbers of moderate and greater losses, there are still considerably less severe and profound losses in the New Zealand sample.

	2010-2014 bilateral Deafness Notifications, born in New Zealand, under the age of 18 <i>Interpolated</i>		2010-2013 bilateral Deafness Notifications, born in New Zealand, under the age of 18		Bilaterally hearing impaired children in Colorado who received early intervention services between birth and 3yrs	
	n=	%	n=	%	n=	%
Mild (26-40 dB HL)	293	49%	209	55%	99	37%
Moderate (41-70 dB HL Colorado)	249	41%	147	38%	102	38%
Severe and profound (>70 dB HL)	55	9%	26	7%	67	25%

TABLE 17: SEVERITY COMPARISON COLORADO AND NEW ZEALAND

Intervention

Ministry of Education

The Ministry of Educationⁱ was approached for information on the number and characteristics of children with hearing loss being supported by [Advisors on Deaf Children](#) (AODC) during 2014. The AODC service is in transition at the current time, working to implement its response to the Wilson Report (2011)ⁱⁱ. Therefore, the Ministry representative declined to provide data for 2014, preferring instead to include information in the 2015 report by which time the shift to service provision with an Early Years focus for AODCs will be complete.

The MOE's Special Education project aimsⁱⁱⁱ:

- to clarify and differentiate between the Advisers on Deaf Children and Resource Teachers of the Deaf roles;
- to ensure that deaf education services are more consistent;
- to make a shift to service provision with an Early Years focus for AODC;
- to further clarify the AODC role with school-aged children; and
- to develop a framework of services that seamlessly links the Universal Newborn Hearing Screening Programme, early identification and the Ministry of Education's Early Years services.

Deaf Education Centres

Kelston Deaf Education Centre (KDEC) and van Asch Deaf Education Centre (vADEC) run Special Schools. As at December 31st 2014, these schools provided services to the following groups:

- residential placements for students in years 7 – 15 (a maximum of 23 at KDEC and 20 at vADEC)
- enrollments for students attending the special schools as day pupils (110 at KDEC and 27 at vADEC)
- specialist preschools for enrolled students (KDEC n= 11 and vADEC n= 7)
- Regional Services through School Provisions in partner schools (n=406 at KDEC and n=317 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 and vADEC:

- 62% have one or more hearing aids, 27% have one or more cochlear implants and 11% have a cochlear implant and a hearing aid. A further 40 children are not able to benefit from assistive technologies, such as aids or implants;
- 40% were recorded as Māori, 37% as New Zealand European, 12% Pasifika, 8% Asian and 3% MELAA; and
- 838 children were over the age of 5: 59% of these were not [Ongoing Resource Scheme \(ORS\) verified](#), 29% were verified as 'high needs', and the remaining 12% were verified as 'very high needs'.

In addition, 299 children have begun to receive services from the Deaf Education Centres (through their Regional Services, provided by [Resource Teachers Deaf](#)) over the last 18 months, during stage

ⁱ Practice Leader Deaf and Hard of Hearing, Professional Practice Unit.

ⁱⁱ Report for Review of the Role of the Advisor on Deaf Children (2011) Reviewer: Ross Wilson, Commissioned by the Ministry of Education, Special Education.

ⁱⁱⁱ Development Project 'Responding to the Wilson Report 2011 – Services to Deaf and Hard of Hearing Children Stakeholder Update (8) Wednesday 25th March 2015. Ministry of Education.

I and II of implementation of the Wilson report. This figure is expected to increase significantly as these changes are further rolled-out to include remaining areas from July 2015.

In 2012, a single combined Board of Trustees was formed to govern the Deaf Education Centres (DECs). The [2015 Combined Charterⁱ](#) 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.

Hearing aids

All but six cases notified to the database contained information about whether hearing aids were to be fitted.

As has been the case with other data since 2010, the majority of children and young people with a hearing loss which was first diagnosed in 2014 are to be fitted with two hearing aids.

Figure 14 below, shows the number of hearing aids fitted or to be fitted by notification year. The small drop in the proportion of cases to receive aids may be the result of the lower overall age of children being identified with hearing loss and/or difficulties in accurately diagnosing hearing losses among younger children in order to provide amplification. The slight rises in the proportion of cases where there is uncertainty around whether hearing aids are to be fitted may also be owing to the lower age of children being notified.

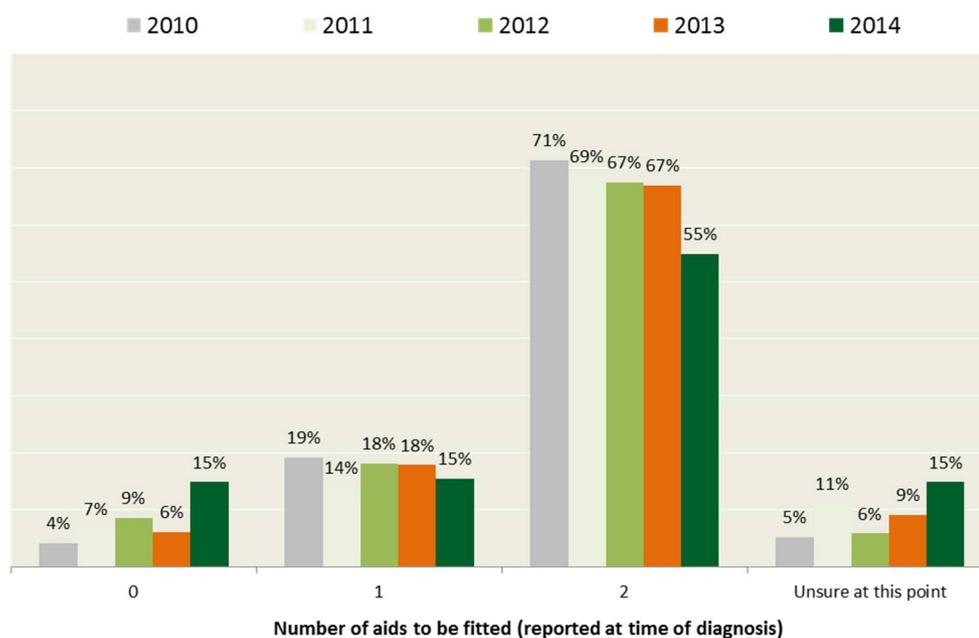


FIGURE 14: PROPORTION OF CHILDREN TO BE FITTED WITH HEARING AIDS (2010 TO 2014)

Since the database was re-launched in 2010, the number of children who are receiving hearing aids before 6 months of age has more than doubled, from 23 to 55. This is likely to be, in large part, the result of the introduction of newborn hearing screening.

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

ⁱ van Asch Deaf Education Centre and Kelston Deaf Education Centre (2015) van Asch Deaf Education Centre and Kelston Deaf Education Centre Combined Board Charter.

databaseⁱ, 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.

In such cases, the term unilateral hearing loss, as it is defined here, does indicate some level of asymmetry, but does not show whether the child requires help to improve their hearing in their better ear.

Funding for hearing aids

In an attempt to provide some context for these figures, data provided by [accessible](#) are shown below. Please note, these data pertain to all children receiving hearing aids, not those receiving hearing aids for the first time, who are included in the DND.

This shows MOH funded hearing aids for children and young people⁴⁷ during the 2014 calendar year. A total of 2575 service users (children and young people) received hearing aid(s) during this period, down slightly on the 2659 in in the year ending December 2013ⁱⁱ.

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	126	126	444	63	759
European	161	125	653	126	1065
Pacific	35	38	204	26	303
Other	98	40	255	55	448
Total	420	329	1556	270	2575

TABLE 18: MOH FUNDING OF CHILDREN'S HEARING AIDS
CALENDAR YEAR ENDING 31ST DECEMBER 2014, ACCESSABLE

These data make for an interesting comparison with the 2010-2014 DND data. Figure 14 shows that 55-71% of children and young people were to receive two aids between 2010 and 2014, and 15-19% one hearing aid. The difference between these figures and those from *accessible* 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 specified number of aids as they get older, either because their losses progress, or because the initial amplification strategy changes.

Cochlear implants

Although the notification form does not include 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

ⁱ Over the four audiometric frequencies: 0.5, 1.0, 2.0 and 4.0 kHz.

ⁱⁱ Data provided in 2014 for the 2013 year was incorrect as these covered only a six month period.

losses which are severe to profound in the higher frequencies and normal or near normal in the lower frequencies.

During the 2014 calendar year there were 61 publicly funded cochlear implants provided in the Northern Region and 51 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.

Since July 1st 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 6 at that time qualified for a retrospective second public implant.

Children receiving cochlear implants during the 2014 calendar year	Southern Cochlear Implant Programme ⁴⁸		Northern Cochlear Implant Programme ⁴⁹	
	Ears	Children	Ears	Children
ACC cases	0	0	3	3
Retrospective second cochlear implants (second ear for those under 6 already with one publicly funded ear) (1 Jul to 31 Dec)	16	16	9	9
Public Funding - (1 Jan to 30 Jun – before bilateral public funding allowed)	8	8	19	14
Public Funding - (1 Jul to 31 Dec - after bilateral public funding allowed)	19	12	28	15
Re-implants – recalled devices, failed integrity tests, or soft failures	7	6	2	2
	50	42	61	43ⁱ

TABLE 19: PUBLICLY FUNDED COCHLEAR IMPLANTS IN NEW ZEALAND

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 reason is that some children who were notified to the database as having less severe hearing losses develop more significant losses over time, something which is not tracked by the database. For example, The Northern Cochlear Implant Programme reported in 2011, that an increased and significant number of children and young people receiving cochlear implants over the last two years had progressive hearing losses. In such cases, the hearing losses would have been less severe at the time of initial identification and notification to the database.

ⁱ This total is not a sum of figures above as two children are recorded in two categories.

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 year⁵⁰. 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 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 part funded by the New Zealand Audiological Society and Janet Digby donated part of her time.

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 *accessible* and will build 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)⁵¹.”

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 groupⁱ.

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

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*ⁱ,
- who are born inside or *outside* of New Zealand.

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

- included within 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 and to try to make it as easy as possible for audiologists to notify cases. 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@leware.co.nz.

ⁱ 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) which 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.

Number of notifications	2011 (July to December)	2012	2013	2014
High frequency losses	n=8	n=10	n=14	n=17

As these cases are not included in the main analyses within this document, a limited analysis of data from high frequency hearing losses notified in 2014 is provided below. A number of notifications which were recorded as high frequency losses by the notifying audiologist actually met the criteria for the main dataset and so were included in this dataset and analysed as such.

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

- all of the 17 cases in this category in 2014 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;
- 18% of children in this category were listed as being of Māori ethnicity, compared with 43% in 2013, 63% in 2011 and 60% in 2012;
- years of birth for this sample ranged from 1999 to 2009 with seven children aged 4 to 6 years old at diagnosis;

The figures below show the audiometric data for the 17 children or young people with high frequency hearing losses contained within 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.

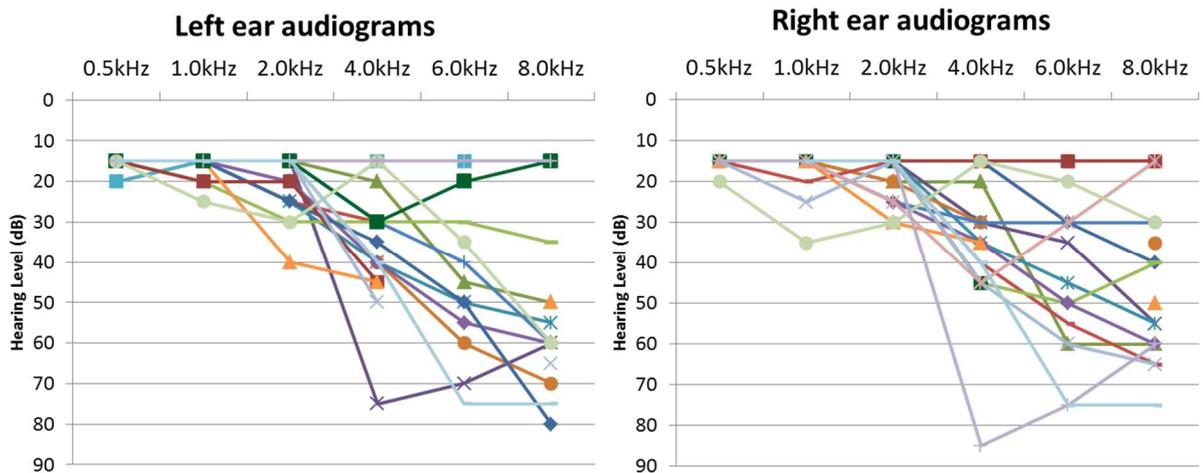


FIGURE 15: 2013 AUDIOGRAM DATA FROM HIGH FREQUENCY HEARING LOSSES FOR RIGHT AND LEFT EARS

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 grouping⁵². 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) categorises respondents into five major groupings and these groupings will continue to be used for the next Census. These groups are: Māori, Pacific Peoples, Middle Eastern/Latin American/African (MELAA), New Zealand European and Asian.

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 which 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 categoryⁱ. 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 20 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.

	1996-2005 NZ DND	1982-1996 NZ DND	Clark 1981 (ASHA) ⁵³	Jerger and Jerger (ASHA) ⁵⁴	World Health Organisation ⁵⁵	CDC ⁵⁶	Proposed code from Davis and Davis (2011)
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 20: COMPARISON OF AUDIOMETRIC SEVERITY CLASSIFICATION SYSTEMS

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

Glossary

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

Advisors on Deaf Children: The Ministry of Education employs advisors 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 Centresⁱ.

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'.

Auditory Neuropathy Spectrum Disorder (ANSD): This condition relates to issues in the transmission of sound from the inner ear through the auditory nerve which 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 which 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 which 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 NZ or overseas, with:

- a permanent hearing loss in one or both ears

ⁱ Ministry of Education (No date) [Support for child who is Deaf or hearing impaired](#). Special Education. Accessed on April 2nd 2015.

- an average loss of 26 dB HL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0 kHz)ⁱ

KDEC – Kelston Deaf Education Centre: One of two residential resource centres for deaf children, based in Auckland, providing onsite services and offsite services and support for mainstream students and their teachers.

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): These teachers observe and assess a child's needs in a one-on-one, classroom and home context. They help educators to adapt their teaching to better suit learners who are deaf or hearing impaired⁵⁷. They can team up with other specialists to provide teachers and families with education workshops. 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 eligibility criteria.

RTDs will help put in place, monitor and review a programme for a child. Resource Teachers: Deaf are employed by one of the two Deaf Education Centres.

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: One of two resource centres for deaf children, based in Christchurch, providing services onsite and services and support offsite for mainstream students and their teachers.

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](#).

ⁱBased 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 within the database. A limited analysis of data from high frequency hearing losses notified in 2013 can be found in Appendix B: High frequency hearing losses, on page 40.

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