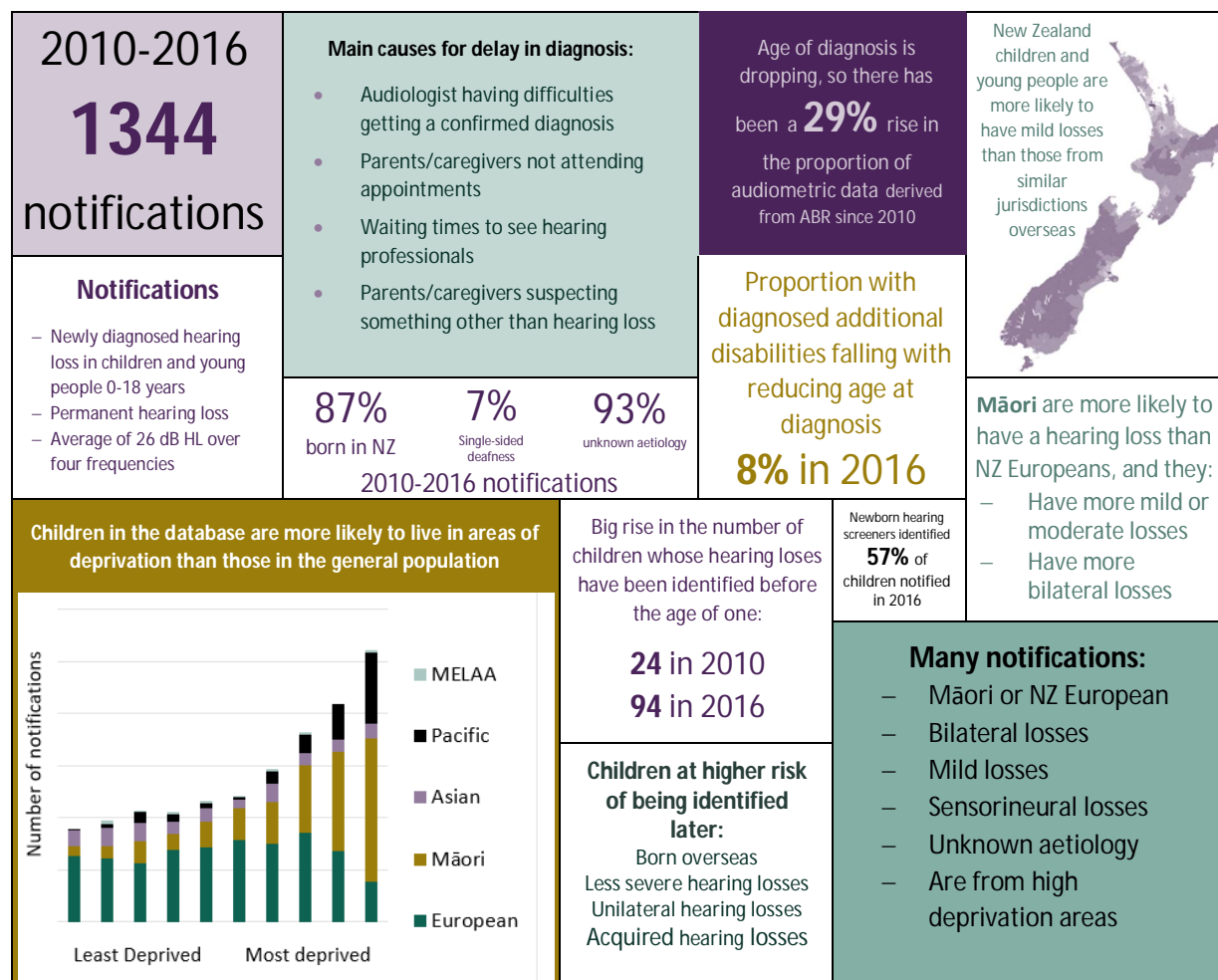


Deafness Notification Report 2016

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

*Janet Digby, Levaré Limited
December 2017*

Summary



Making notifications to the database

The authors of this report would like to extend their sincere thanks to all those who have provided notifications to the database. Your contribution to our understanding of permanent hearing loss among New Zealand's children and young people is greatly appreciated.

Audiologists (including non-NZAS members) are encouraged to make future notifications to the database by following [this link](#). Audiometrists are encouraged to make notifications for cases they have diagnosed in over sixteen-year olds.

Notes for those completing notifications for the database:

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

This ensures these reports contain accurate information about those children and young people notified during each calendar year.

Resources for clinicians making notifications can be found [here](#) – these include a PDF version of the notification form, background information about the database and previous database reports.

2. **Consent:** babies screened by the UNSHEIP are legally consented for entry into the Deafness Notification Database (DND), and there is no need to get the families to sign a separate consent form.

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

The families of other babies and children being notified to the database will still be required to sign the [consent form](#) which clinics should keep on file.

3. **Questions:** If you are in doubt about whether a case meets the criteria, please notify the case. For answers to any questions at all, please email [Janet Digby](#): or telephone (09) 445-6006.

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The Deafness Notification Database

Introduction

Nau mai, afio mai - welcome to the seventh annual report describing notifications to the New Zealand Deafness Notification database. This report includes data on diagnoses made throughout New Zealand during the 2016 calendar year.

The New Zealand Deafness Notification Database was established in 1982 and contains information on new diagnoses of permanent hearing loss among children and young people under the age of 19.

Audiologists from around the country send notifications electronically when they diagnose a child with hearing loss. These notifications contain information about the configuration, type and severity of the hearing loss, as well as demographic information, assuming the parents provide consent for this information to be shared.

The analyses contained in this report generally pertain to 1344 children and young people notified to the database with a hearing loss diagnosed between the start of 2010 and the end of 2016ⁱ.

“Ka mua, ka muri”

Māori proverb - ‘walk backwards into the future’ about learning from the those who have gone before us.

ⁱ There are additional notifications which have been included in the main dataset from years 2003-2009 and from early in 2017, which brings the total number in the dataset at the time of writing to 1476 children and young people who were initially diagnosed with a hearing loss between 2003 and 2017.

ⁱⁱ The original criteria for the database, which applied to notifications until the end of 2005, required the hearing loss

The information contained within the Database has enabled us to shed light on the nature of hearing losses among children and young people, including by helping us understand that Māori are more likely than their New Zealand European counterparts to be diagnosed with a hearing loss, and also that Māori are more likely to have mild and bilateral hearing losses.

Recent database notifications have also shown that there is a growing number of children being identified under the age of one year. This is pleasing as the earlier a child’s hearing loss is identified the earlier intervention can be provided. This shift is almost certainly the result of implementation of nationwide newborn hearing screening.

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

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.

ⁱⁱⁱ Based on feedback from the audiological community, high frequency hearing losses that would not meet the original criteria, but that would exceed the 26 dB HL average based

Since the database was relaunched in 2010, in addition to children with bilateral hearing losses, those with unilateral hearing losses that are mild or greater in severity, have been included.

Further historical information about the database's inclusion criteria can be found in Appendix A: History of the database, on page 59 of this report.

Steps have been taken to allow data contained in this report to be compared 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 specific area (such as family history), and as a result some longitudinal comparisons are not possible.

Please note the following regarding longitudinal data from the DND:

- notifications have been reported for each calendar year throughout the period of operation of the database, i.e. 1982-2005 and 2010-2016;
- the period from 1982 to 2005 contains notifications to the original National Audiology Centre/ Auckland District Health Board (ADHB) administered database;
- no annual reports were completed for the years 2006 to 2009 as the database was not operating during this period; and
- data for 2010 to 2016 relate to notifications provided to the newly re-launched database.

A glossary of commonly used terms can be found on page 63 of this report.

Terminology

There are several terms used by young people with a hearing loss and their families/whānau. Those whose information is included within this

report range from those which are unilateral and mild in severity, through to those whose hearing losses are bilateral and profound in nature. The terms commonly used differ within these groups as well as between them.

Some families and young people prefer terms such as 'hearing impaired' or 'hard of hearing', while others use the term 'Deaf' or 'deaf'. For the purposes of this report, we need to choose a term or set of terms and use these consistently where possible to aid in the report's readability. In doing this it is not the authors' intention to exclude those who use or prefer other terms.

Following discussions with the Ministry of Health and consultation with Federation for Deaf Children a decision has been made to prioritise the terms 'deaf', and/or 'hard of hearing' within these reports, moving away from the term 'hearing impaired' which has been used in previous reports.

Completeness of notifications

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

There may be certain types of cases that are under-represented within notifications, and as a result inferences made from the data contained in this report should be taken as indicative unless stated otherwise.

Based on analyses described in the 2013 DND report and on discussions with the audiological community, the authors of this report believe it is likely that the database has been receiving

on audiometric data from 2.0, 4.0, 6.0 and 8.0 kHz, have been collected from July 2011.

notifications for between 55% and 80% of all new cases diagnosed each year.

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 stakeholders (including the Ministry of Health, Ministry of Education, clinicians, educators and other service providers) about newly diagnosed hearing losses among New Zealand children and young people.

Acknowledgements

Our sincere thanks to the 190 whānau/family members and tamariki (young people) who consented to share details about their own or their child's hearing loss to be included in the database. As a result of the willingness of families to share basic information about their or their child's diagnosis, service providers can be better informed about current and future demand for services, some variables most likely to result in delays in identification and other information that will help them better serve the needs of whānau families.

The time taken by professionals around the country to make notifications is also very much appreciated.

This report has been funded by **Enable New Zealand**, through a contract with the Ministry of Health. We would like to thank the Ministry of Health for funding the database from 2012.

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

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

This year, we would also like to extend a special thanks to Jessica McLay, Statistician from the University of Auckland, for assisting with us by completing additional analyses for this report, particularly regarding difference testing relating to ethnicity and deprivation.

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 readers to 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 by [email](#) or by telephone, (09) 445-6006.

Notifications

General information

One hundred and ninety notifications pertaining to cases first diagnosed during the 2016 calendar year, and meeting the criteria for inclusion, were received by 17 March 2017, this year's cut off for 2016 notifications^{i, ii}. These notifications were received from a total of 59 audiologists, with notifications from 18 of the 20 district health boards (DHBs).

A significant number of cases were listed by audiologists at the time of notification as 'high frequency losses'. However, on examination, a clear majority of these met the criteria for the main category, and so were not included in this sub-category. Because only five cases met the criteria for the high frequency category, we have not described this group in this year's report.

Among those children and young people whose hearing loss was notified to the database, notification numbers used to peak at the end of the notification period (November to December), with a smaller peak in August. Now, with recent changes to the consenting process, notifications are submitted more evenly throughout the year, again with a peak around August and September, but also with a peak before notifications close in March.

i Reports prior to 2006 contained information about diagnoses notified in each year, rather than diagnosed in that year. As a result, the number of notifications varied, increasing in years in which greater efforts were made to encourage audiologists to send in notifications. For example, in 2004 there were an additional 288 retrospective notifications received from the Children's Hearing Aid Fund (CHAF) audit.

ii It is not possible to ascertain how long, on average, audiologists took to make each individual notification, as online forms are often left open for a number of hours. However, it is clear that many individual notifications took fewer than five minutes to enter using the online form, as

Notifications are collected through an online form to reduce the risk of data entry errors and make it as easy as possible for hearing professionals to notify cases.

To maximise the number of notifications to the database, efforts have been made to publicise this mahi (work) through the New Zealand Audiological Society to reach the majority of those initially diagnosing with children and young people/tamariki with hearing loss.

Number of notifications

Figure 1 shows the number of notifications meeting the criteria for the main dataset, in each year. High frequency hearing losses, which have been collected since July 2011, are not included in this main dataset.

Information about how the inclusion criteria have changed over time is included in Appendix A which begins on page 59.

Figure 1 shows the number of notifications which met all inclusion criteria at the time and were included in each of the database's annual reportsⁱⁱⁱ.

was the case in previous years. The removal of the extended family history questions in 2014 has reduced the time it takes for audiologists to collect notification information and submit cases.

iii Please note that the 2001-2005 figures, included in previous DND reports, were later revised by the database's contracted provider at the time, ADHB. Previous figures were sometimes split by those that were removed for audiometric or other reasons. The figures now show the total number of notifications that met criteria for inclusion, which were in place at that time.

This illustrates the variability in the number of notifications provided to the original database, particularly in the last six years of its operationⁱ.

Data from 2010 to 2016 have been revised slightly from previously reported figures, as further information about existing notifications is received, and as retrospective notifications are provided to the database. Specific changes are described in detail in the reports in which these were first made. Previous reports can be found on the New Zealand Audiological Society website.

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

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

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

ⁱ 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 are excluded from the data reported here.

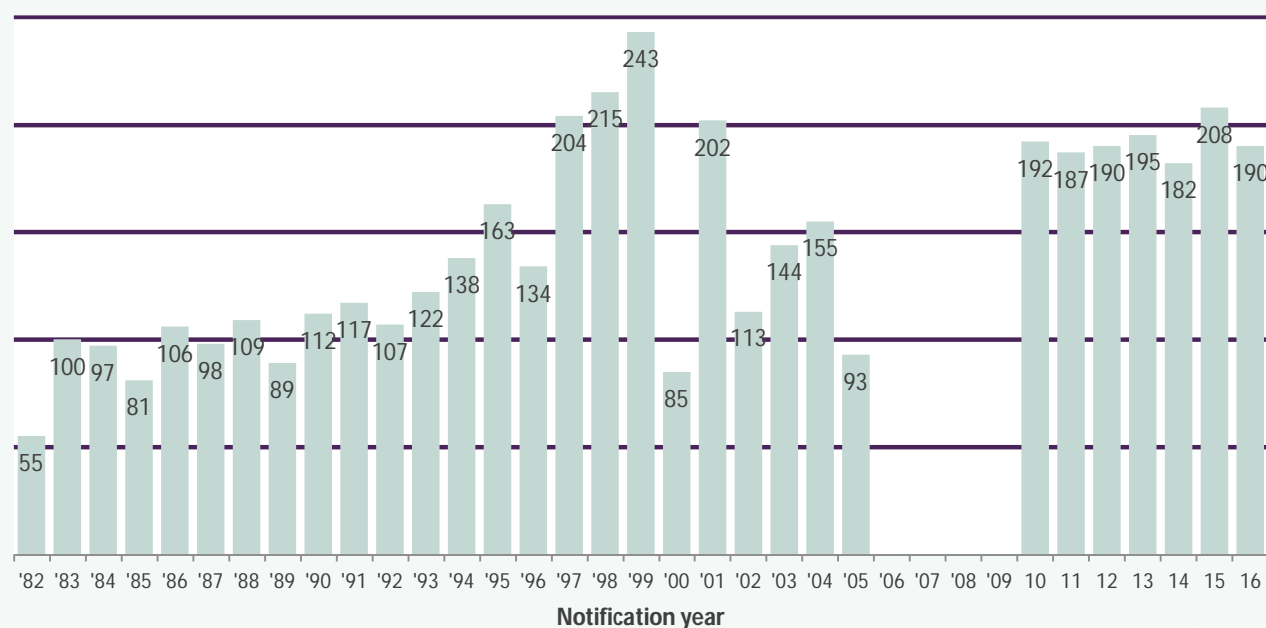


Figure 1: Notifications by year 1982-2005 and 2010-2016

Gender

Of the 1344 cases (2010-2016) contained in the main dataset, 45% of these are listed as female (n=611) and 55% male (n=733). This represents a ratio of 1: 1:22. This gender difference is particularly noticeable in 2016 with only 38% of notifications recorded as female, and 62% male. It will be interesting to see if next year's data are more typical of the long-term average.

In overseas research, males are commonly found to have higher rates of hearing loss than females. These figures range between 52% and 58% for males (1:1.08 and 1:1.38) in various jurisdictions, as reported in the 2011 *Comprehensive Handbook of Pediatric Audiology*ⁱ.

Australian Hearing's data on those under the age of 21 who have hearing aids or cochlear implantsⁱ shows a similar pattern², with higher numbers of

hearing loss among males 52.6% than females (47.4%)ⁱⁱ. This pattern is seen in all Australian states, except for South Australia and ACT, in which the ratios of male to females is almost 1:1, and for those aged 21-25 years of age, where less than half of cases were male (48.4%).

Birthplace

Tamariki born outside New Zealand have been formally included in the database and its main analysis since 2010. During that time, an average of 6% of children and young people notified have been born overseas, with the birthplace of an additional 7% being uncertain.

Of the 190 notifications to the database in 2016, 6% were known to be born outside New Zealand. Birthplace was uncertain in a further 6% of 2016 cases reported to the database. Figure 2 shows the proportion of cases notified by birthplace for the 2010-2016 period.

i This source reports on tamariki, under the age of 26 who received services from Australian Hearing in 2014.

ii 0.1% of cases were of unknown gender.

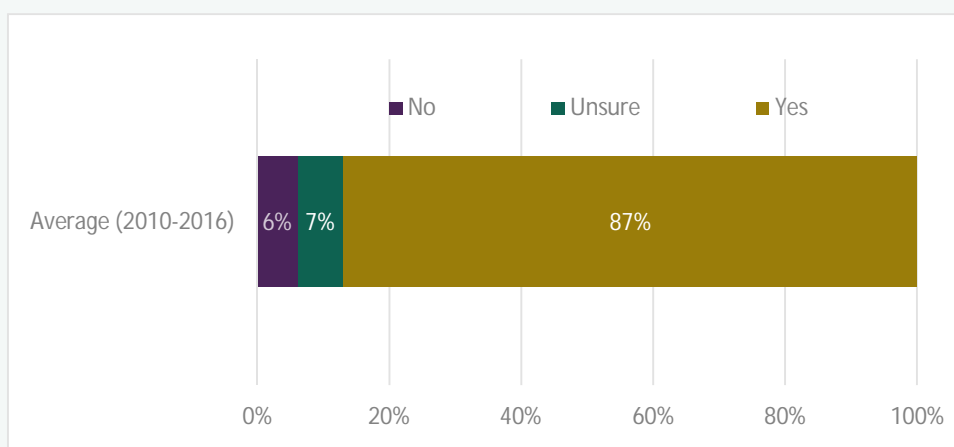


Figure 2: Proportion of cases born in New Zealand (2010-2016)

DHB representation

Table 1 contains the percentage of 2016 notifications from each DHB area and compares these with the percentage of the population under the age of 20 from the 2013 Censusⁱ. This year, we have added a third column, showing the percentage of notifications received for 2010-2016 from each district health board – this can usefully be compared with the percentage of the population under the age of 20. Please note, these percentages are rounded.

Tamariki in the database are more likely to be of Māori ethnicity than their proportion in the general population would suggest. DHBs with more than 20% of their population identified as Māori are listed in a different colour in Table 1³. It is interesting to note that several of the DHBs reporting higher numbers of notifications than anticipated based on their population, are those DHBs with a higher proportion of Māori (e.g. Tairāwhiti, Waikato, Hutt, Northland, Bay of Plenty).

District Health Board	Percentage of notifications received 2016 (under 19 years)	Percentage of population under the age of 20 (Statistics New Zealand, 2013 Census)	Percentage of notifications received 2010-2016 (under 19 years)
Auckland	13%	11%	7%
Bay of Plenty	6%	4%	6%
Canterbury	14%	11%	15%
Capital and Coast	6%	7%	8%
Counties Manukau	8%	13%	13%
Hawke's Bay	3%	3%	3%
Hutt	2%	3%	3%
Lakes	3%	2%	3%
Midcentral	2%	4%	3%
Nelson Marlborough	3%	3%	3%
Northland	6%	3%	6%
South Canterbury	2%	1%	2%
Southern	5%	7%	6%
Tairāwhiti	3%	1%	2%
Taranaki	3%	2%	4%
Waikato	9%	9%	8%
Wairarapa	2%	1%	1%
Waitematā	11%	13%	6%
West Coast	0%	1%	0%
Whanganui	0%	1%	1%

Table 1: Percentage of notifications (2016) 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.

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

DHBs underrepresented in the data include:

- Auckland and Waitematāⁱ – due to consenting issues with previous cases. The number of notifications for tamariki living in these DHBs, and diagnosed since 2015, has risen due to changes in consenting processes which applied from the middle of the reporting year and were outlined in the [2015 report](#);
- Wairarapa, Lakes and West Coast – these are relatively small DHBs, and during some years they may not diagnose any tamariki with a hearing loss;
- Hutt Valley has not diagnosed as many tamariki this year as they had in previous years.

Other disabilities

The presence of one or more 'additional disabilities' can have a significant impact on both outcomes for tamariki with hearing loss, and on the level of support they may require, particularly from *Learning Support* ([previously Special Education](#)).

Cupples *et al.* (2009) found that there were differences in outcomes for the 119 children included in their study based on the type of additional disability. Children with autism, cerebral palsy, and/or developmental delay showed poorer outcomes compared with children who had vision or speech output impairments,

In addition to the comments above, and natural fluctuations in the number of hearing losses diagnosed among tamariki in each 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 the clinician decides it is appropriate to ask for consent for the database at the time of diagnosis, or whether this is best done at a later appointment, which may be after the cut-off date for notifications;
- the number of hearing professionals employed by each DHB;
- the workload of these hearing professionals; and
- the level of commitment among staff to making notifications to the database.

syndromes not entailing developmental delay, or medical disorders⁴.

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

Of 2016 notifications, 8% were thought to have disabilities *in addition* to hearing loss at the time the notification was made. This is lower than the proportion reported for any previous year. In a

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

further 10% of 2016 cases there was uncertainty regarding whether the child or young person had an additional disabilityⁱ.

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 (n=2)^{5, ii}.

The proportion of tamariki 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.

When the 'unsure' figure is added to the proportion of cases with an additional disability, as shown in Table 2, the figure is more consistent with those reported before the database's re-launch in 2010.

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 (2010-2016)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	12%	11%	22%
2011	15%	5%	20%
2012	15%	11%	27%
2013	13%	12%	24%
2014	15%	8%	23%
2015	10%	10%	20%
2016	8%	10%	18%

Table 2: Proportion of cases with a known additional disability

Earlier identification of tamariki with hearing loss is the likely reason behind the reduced proportion of tamariki with confirmed additional disabilities, which are reported at the time of diagnosis of the hearing loss. This is because tamariki may have not yet been diagnosed with these conditions, or they have conditions that have not yet developed. For example, diagnoses of autism spectrum disorder are typically not made in the first year of

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

- tamariki with hearing loss in New Zealand may not be routinely assessed by a paediatrician, meaning additional disabilities may be under-diagnosed;

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

ⁱⁱ No local data are available on the rates of vision problems among deaf and hard of hearing populations in New Zealand,

but some professionals recommend routine referral for ophthalmological assessment for children diagnosed with significant bilateral hearing loss.

- immunisation coverage in New Zealand has risen significantly in recent years^{i,6}. Such Improvements have reduced rates of meningitis in New Zealand and this may have an impact on the proportion of tamariki with hearing loss with additional disabilities, although the numbers are likely to be smallⁱⁱ.

A logistic regression was conducted to see whether a linear or non-linear relationship exists between a tamariki having other known disabilities and her/his level of deprivation. No association was found ($p=0.7801$).

Overseas additional disability data

While it is difficult to compare reported rates of additional disabilities between groups of tamariki who are hard of hearing, as the definition for hearing loss and for disabilities differ and are not always described in journal papers, a selection of rates from various jurisdictions are described in Table 3 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 tamariki who are hard of hearing and have an additional educational need, although this is unlikely to be a fair comparison owing to jurisdictional differences in how additional disabilities are defined.

Source	Date	Location	Details	Rates
Ear Foundation for National Deaf Children's Society⁷	2012	United Kingdom Review	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 <i>et al</i>⁸	2002	UK	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis⁹	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 Diaz¹⁰	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
LOCHI¹¹	2013	Australia	Study examining 260 children in Australia born with hearing impairment	18% of children within their sample have one additional disability, 10% with two and 9% with three or more
The Consortium for Research into Deaf Education¹²	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 These increases in rates have occurred since vaccination for children became a Primary Health Organisation (PHO) Performance Programme indicator in January 2006, and a funded indicator from July 2008. Achievement rates for the indicator 'age-appropriate immunisations completed by age two years' have doubled from approximately 45% in 2007 to 91% in September 2013.

ii 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, because 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

Background

Binaural hearing allows the auditory system to process and integrate input from both ears, making it necessary for improved understanding of speech in noisy situations and for sound localisation^{13,14}. A series of studies in the early 1980s (in the United States) caused the significance of unilateral hearing losses to be re-evaluated by professionals, who had often underestimated the significance of unilateral hearing loss in children^{15,16,17}.

Children with unilateral hearing losses are known to have reduced educational performance, language delays and higher rates of behavioural issues, which are reported as significant in about a third of all cases^{18,19,20,21,22}. A significant proportion of these hearing losses progress over time to become more severe or to affect both ears²³.

The Joint Committee on Infant Hearing (JCIH) noted in its 2007 statement that 'All families of infants with any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.'²⁴ This statement recommended that developmental monitoring should also occur at regular six-month intervals for those with permanent unilateral hearing loss because these children are at risk of speech and language delay.

This 2007 statement now has a supplement (2013) which states that all children with unilateral or bilateral hearing loss should be referred to early intervention services for evaluation and consideration of enrolment. Most infants and children with bilateral hearing loss and many with

unilateral hearing loss benefit from some form of personal amplification device²⁵.

In New Zealand, Project HIEDI recommended in 2010 that families of children with unilateral hearing loss be offered advisory services and that such children be regularly assessed to quickly determine if they are beginning to fall behind and to determine what support is appropriate²⁶.

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

Inclusion in the DND

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

Although unilateral hearing losses were not included in the DND before 2006, several of these cases were notified to the database each year and these numbers were provided in the annual reports at that time. However, comparing the proportion of unilateral/bilateral notifications with previous DND data (prior to 2005) is not possible because reporting prior to 2006 was incomplete in this older dataset.

From 2015, we describe the proportion of bilateral and unilateral hearing losses based on cases with and without all data-points and also on interpolated figuresⁱⁱⁱ using manual checks for those records that

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

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

iii While only cases where all eight-audiometric data-points recorded are able to be included in most severity calculations,

interpolation of data has been used in some instances in this report to provide a more complete picture of the severity of hearing losses notified. Interpolation is only used where three of the four data-points are provided for one ear, where both surrounding data-points are provided.

cannot have data interpolated. This change means we can now report on the number of ears affected by hearing loss in more cases.

Bilateral and unilateral hearing losses:

- the proportion of 2010-2016 cases that were bilateral/unilateral using only cases with full audiometric data was 63:37, and this became 68:32 when cases without all data-points were included; and
- the proportion of 2010-2016 cases that were bilateral/unilateral *using interpolated data* was 67:33 and this remained the same when cases without all data-points and manual checks were included.

While immunisation coverage (including for conditions such as mumps) in New Zealand has risen significantly from 45% in 2007 to 92% in 2012²⁷ 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 and/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 loss²⁸.

Differences between the proportions of bilateral and unilateral notifications in each severity category are shown in Figure 3.

Single sided deafness

Severe or profound unilateral hearing loss can be referred to as single-sided deafness (SSD).

Unilateral hearing loss impairs speech perception in noisy situations and the ability to localise sound²⁹.

While there are few studies on children in this area, a recent review focused on adult research (2016) concluded that no recommendations for the management of unilaterally deaf adults could be based on the current evidence³⁰.

Different case definitions of single sided deafness (SSD) are used internationally; for example, some definitions include only those with severe loss in the worse ear and others only those with profound loss^{31,32}. The boundaries for these degrees of loss also differ depending on the jurisdiction. One reason for examining the proportion of unilateral losses that are categorised as SSD, is that there are differences in the types of hearing technology that may benefit tamariki in this group. For example, tamariki with

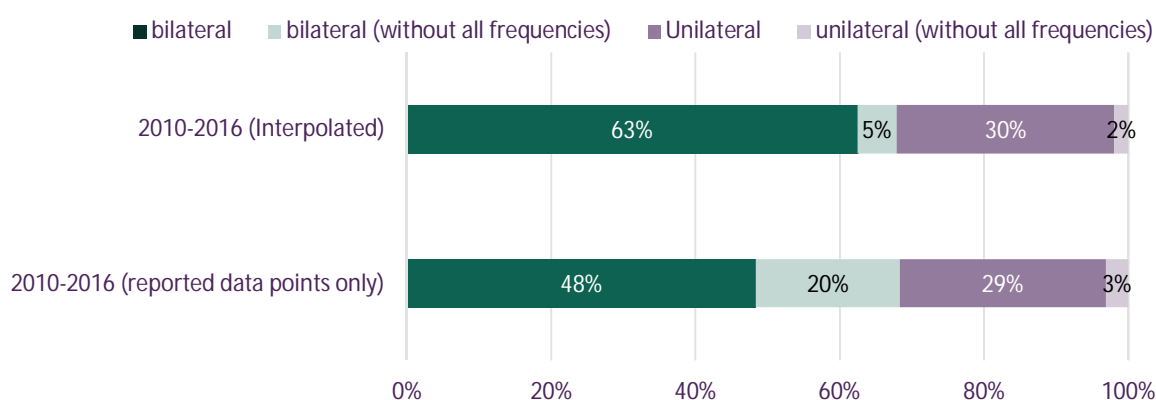


Figure 3: Proportion of bilateral and unilateral cases (2010-2016)

SSD receiving cochlear implants versus those with less severe degrees of hearing loss, who may receive a bone conduction hearing aid.

Cases of SSD in our analysis have been defined as children and young people in the main dataset who have a hearing loss of more than 70 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the worse ear, and a hearing loss of less than 26 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the better earⁱ.

This category is effectively a subgroup of the unilateral hearing category mentioned elsewhere in this report. The proportion of unilateral hearing loss casesⁱⁱ which met the DND’s criteria for SSD is 22%.

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

Notification Year	Proportion of cases with single sided deafness (SSD)
2010	6%
2011	4%
2012	8%
2013	10%
2014	7%
2015	6%
2016	6%
Average 2010-2016	7%

Table 4 Single Sided Deafness Cases by Year (2010-2016)

ⁱ These average thresholds have been chosen considering the ASHA codeframe for severity, and because 26 dB HL is the lower limit for average notifications to be accepted into the database and as a 70 dB HLⁱ average is the boundary between moderately severe and severe losses.

This 70 dB HL average for the lower limit will eliminate most cases of atresia, as these are mostly conductive, and

therefore not severe enough to meet this threshold criterion. Such children will benefit from a bone conduction hearing aid and are, as a result, a different group to those we categorise as having SSD.

ⁱⁱ Based on determinations including interpolated data.

Types of hearing loss

A question about the type of hearing loss was added to the notification form part way through 2013. This question asks audiologists to describe the type of loss in each earⁱ. Options provided are; 'sensorineural', 'mixed', 'permanent conductive', '**ANSD**' (Auditory Neuropathy Spectrum Disorder), 'normal hearing'ⁱⁱ, 'other' and 'don't know'.

No cases were recorded as 'other' or 'don't know' while 15% of cases on the right and 12% on the left were listed as 'normal hearing'.

percent of right ears and 4% of left ears were recorded in the ANSD category.

Prevalence of ANSD among those with permanent hearing loss is likely to be 10%, according to a 2015 review by Rance³³.

An analysis of the types of hearing loss reported by ethnicity found significant differences between Māori and New Zealand European (Fishers exact test: $p=.0037$). More Māori had 'mixed' hearing

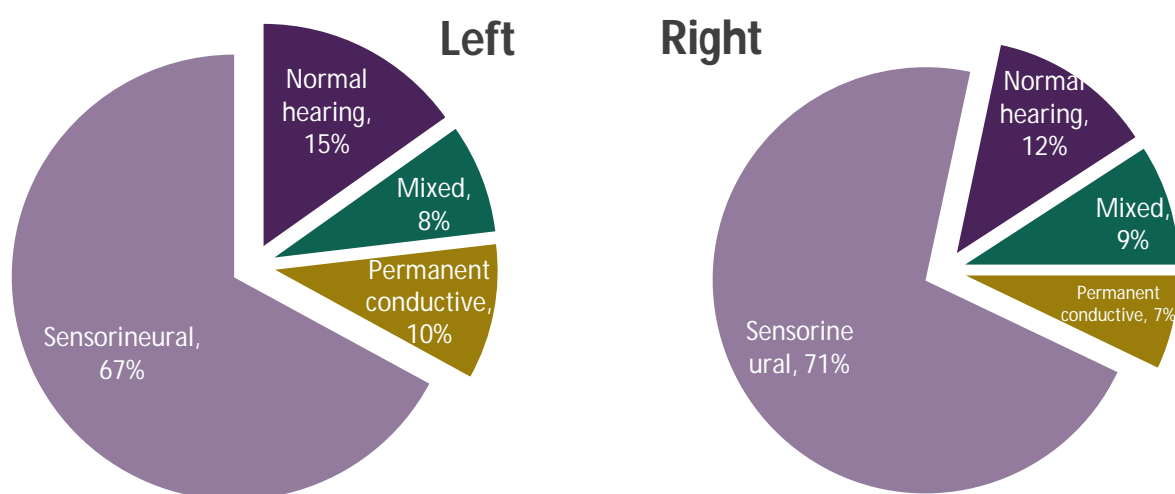


Figure 4: Type of hearing loss (2010-2015)

Please note that the ANSD group have sensorineural hearing losses; that is, this group is effectively a subgroup of the sensorineural category.

The most commonly reported type of hearing loss contained in notifications was sensorineural (11% in the right ear and 67% in the left), followed by normal hearing (15% in the left ear and 12% in the right), permanent conductive losses in the left ear (10%) and mixed losses in the right (9%). Two

losses than expected (11.9% for Maori vs 6.1% for New Zealand Europeans, $p=.0317$ - Z-test for proportions), and fewer Māori were recorded as having 'permanent conductive' hearing losses than expected (6.5% for Maori vs 12.1% for New Zealand European, $p=.0313$)ⁱⁱⁱ.

Given that Māori in our sample have more bilateral losses than their New Zealand European counterparts, it was unsurprising to see that Māori were less likely to have 'normal hearing' in one ear.

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

ⁱⁱ Those notifying cases could also select normal hearing for the hearing ear in children and young people with unilateral hearing loss.

ⁱⁱⁱ Data for those with missing hearing loss type data was excluded from this analysis.

Family History

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

The new question in the DND relating to family history is 'Does an immediate family member (only a mother, father or sibling) have a permanent hearing loss? (or had a permanent hearing loss if they have died).' The results for this question are shown in Figure 5.

Please keep in mind that data from 2014 contains information from approximately half the notifications for that period, as the question was changed in the middle of the year, hence we have included data from 2015 and 2016 in Figure 5.

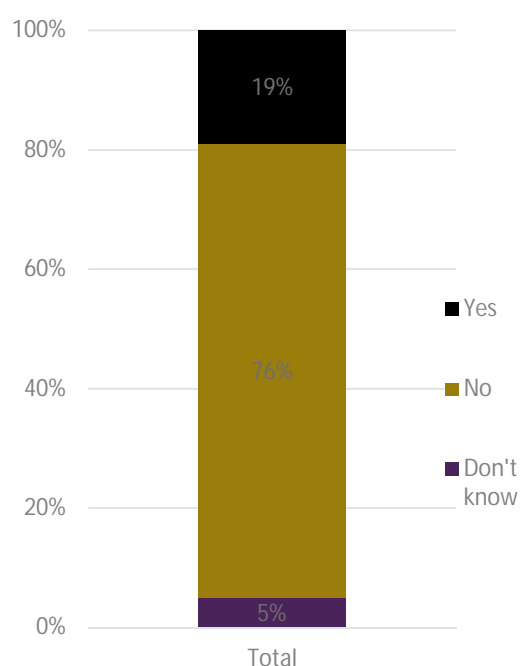


Figure 5: Immediate family member with hearing loss (2015 and 2016)

ⁱ The DND reports prior to 2005 note that a relatively high proportion of cases recorded 'family history' as the cause of the hearing loss (family history was reported as the cause of the hearing loss in 24-32% of cases between 2001 and 2005).

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

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

Ethnicity

Representation

The notification form records information about the ethnicity/ethnicities of tamariki diagnosed with hearing loss.

Options available are New Zealand European, Māori, Pacific Peoples, Asian and MELAA. The MELAA category relates to tamariki of Middle Eastern, Latin American or African ethnicity.

Of the 1344 notifications in the main dataset, all but 21 (2%) contain at least one ethnicity code. The majority (89%) contain one code, and a smaller proportion (8% and 1%) contain two or three codes respectively. Please keep in mind that the multi-code system means that some records (less than 10%) contain more than one code for ethnicity, and so they appear in more than one group.

A smaller proportion of notifications specified more than one ethnic group when compared with 2013 Census data. This suggests that coding for ethnicity is less complete for deafness notifications than for the Census data, and this may affect the validity of this comparison. It may also suggest a need for clearer instruction on how best to code for ethnicity.

The majority of notifications provided to the database since its re-launch in 2010 relate to tamariki of New Zealand European and/or Māoriⁱ ethnicity.

Multi-coded 2013 Census data is included for comparison in Figure 6. As individuals may identify with more than one ethnicity, the totals add to more than 100%. This figure shows the total

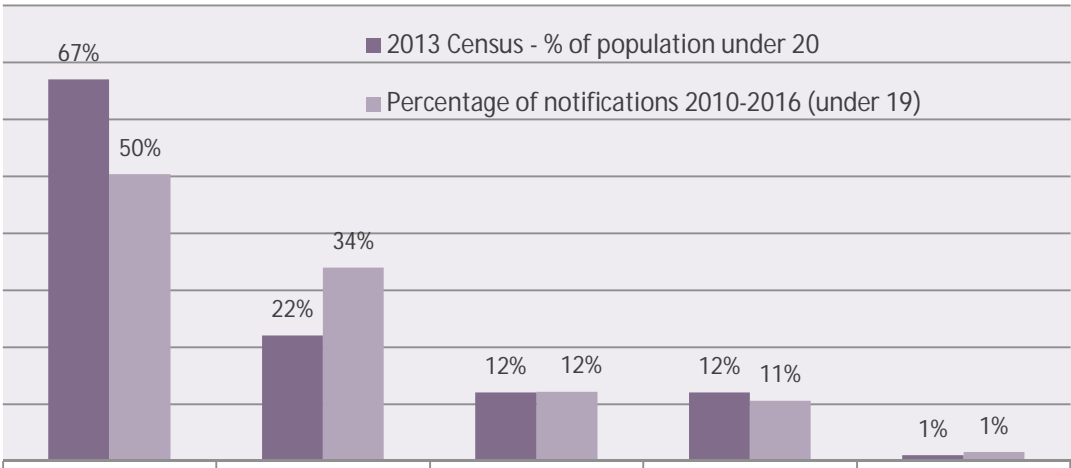


Figure 6: Notifications by ethnicity (2010-2010) compared with Census data (2013)

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

response count for ethnicity from the 2013 Census (for those under the age of 20) and compares this to the ethnicity breakdown for deafness notifications from 2010-2016, which includes those under the age of 19ⁱ.

Ethnicity and prevalence

Prevalence data

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

Nevertheless, Māori may still be underrepresented in DND statistics because of their greater chance of having a less severe (mild or moderate) hearing loss, as these are more likely to go unidentified. It may also be that disparities in access to, and within, the health system for Māori³⁴ may mean fewer cases are found or notified when compared with the New Zealand European population.

A number of other sources support the difference in prevalence of hearing loss between Māori and New Zealand Europeans which is also shown in DND data described in Figure 6: Notifications by ethnicity (2010-2010) compared with Census data (2013):

- Diagnoses from the newborn hearing screening programme show that Māori infants who are screened and for whom diagnostic information is available have higher rates of hearing loss³⁵.
- The Household Disability Surveys (1991-2006³⁶) suggest Māori may have higher rates of hearing disability (tamariki and adults) and higher rates of unmet need for technology and equipment when compared with non-

Māori³⁷. (For information about the limitations of this data please see the 2011 DND Report³⁸.)

- Findings from Digby *et al.* (2014) indicated young Māori have higher rates of permanent hearing loss than their New Zealand European peers, based on the previous DND dataset (1982-2005)³⁹.
- B4 School Check data:
 - » Referral rates from the B4 School Checkⁱⁱ (2011) analysed by Searchfield *et al.*, show higher rates of referral from hearing screening for Māori tamariki (9%) compared with non-Māori (5%)⁴⁰. It is important to note that high referral rates for Māori may relate to higher rates of ear disease, as these figures do not just relate to permanent hearing loss.
 - » The overall referral rate for Māori tamariki (2015/2016) completing their hearing screening was 7.4%, considerably higher than for New Zealand European tamariki, at 3.9%⁴¹.

For further information on ethnicity coding within the database, please refer to *Appendix B: Notifications and ethnicity*, on page 61.

Service access

Disparities documented in other parts of the health system demonstrate Māori have poorer access to, and through, the health system^{34,42}.

An article by McCallum *et al* (2015) in the *New Zealand Medical Journal*⁴³ examined both hospital admissions for under 15-year olds (2002-2008) and first ENT appointments (2007-2008) and found disparities in access to ventilation tubes for 0-4 year-olds, with the greatest inequalities being

i Individual year age data for ethnicity is not freely available from Statistics New Zealand.

ii For more information on the B4 School Check, please click [here](#) or view the glossary on page 75.

for Māori, Pacific and Asian tamariki living in deprived areas.

While the specific nature of the barriers to access were not described, research into whether such disparities exist for tamariki accessing other hearing services, such as those provided by audiologists, is needed.

Such investigations are particularly important as there is no service specification for audiology services nationally, meaning that services offered by district health boards (the DHBs see most tamariki with hearing loss) differ, as do waiting times.

Unilateral and bilateral hearing losses

Of 2010-2016 cases, including those with interpolated audiometric data, 68% are recorded as bilateral, while the remaining 32% are unilateral.

Figure 7, below, shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group during 2010-2016. These data include not only interpolated cases, but also those with one or more frequencies missing. As a result, more cases can be included in the comparison than presented in previous reports.

The significant difference between Māori and New Zealand European rates of bilateral loss (found on analysis of the now larger sample) supports the conclusions from the 2014 paper by Digby *et al* which found a larger proportion of bilateral hearing losses among young Māori when compared with their New Zealand European counterparts³⁹.

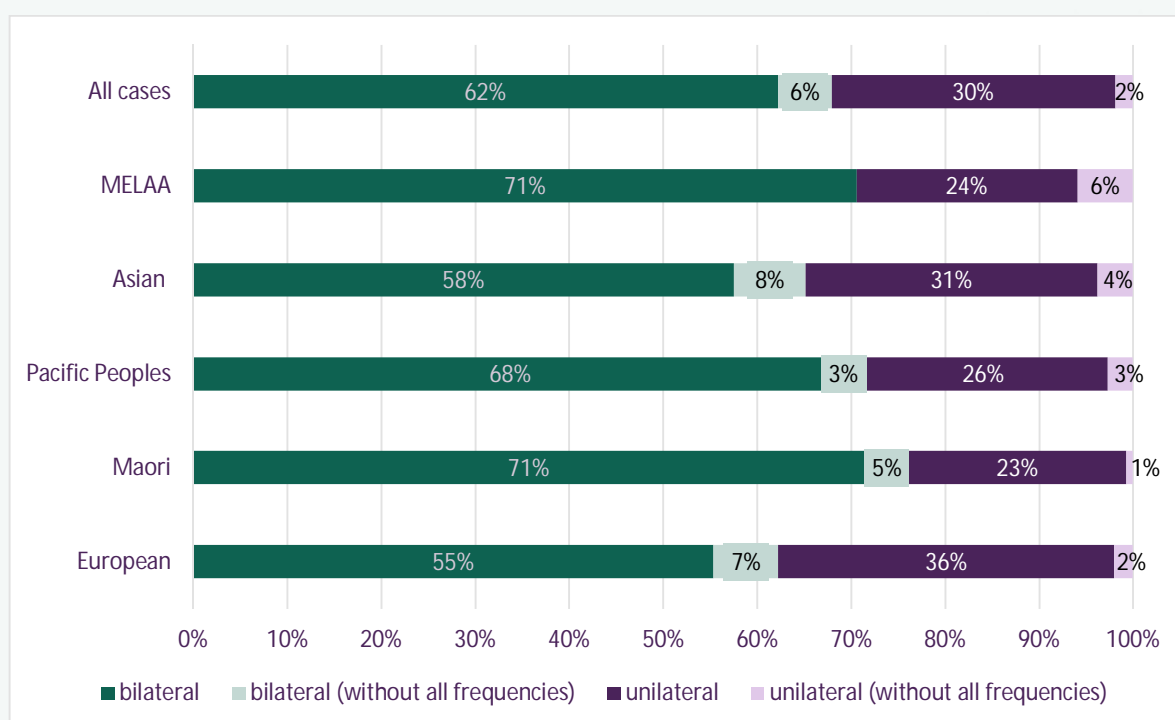


Figure 7: Proportion of unilateral and bilateral hearing losses by ethnicity (2010-2016) based on interpolated data and manual checks to determine bilateral/unilateral status

This difference can also be seen when comparing bilateral losses among Māori tamarikiⁱ notified between 2010 and 2016 (75%), with those who are New Zealand Europeanⁱⁱ (55%), and those with whose ethnicity is described using both Māori and European ethnicity (56%).

Please note that MELAA figures relate only to a very small number of cases (n=17).

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

Hearing loss present at birth

Out of all 2010-2016 cases, nearly 99% contained information indicating whether the audiologist believed the child's hearing loss was likely to have been present at birth.

Of those where a code was provided, the audiologist indicated they were 'unsure' in 48% of cases, with the hearing loss likely to have been present at birth in 38% and unlikely to have been present at birth in 14% of cases.

Further analysis shows that the proportion of New Zealand Europeans *without* 'hearing loss thought to be present at birth' was significantly higher

than for Māori (Z Test: 95% CI (.054, .132), $p < .0001$). Because of the number of 'unsure' answers for this question, one cannot assert that Māori therefore have more hearing losses present at birth.

The percentage of New Zealand European tamariki where the audiologist was unsure whether the hearing loss was present at birth, or where these data were missing, was 7.2% lower than that for patients of Maori ethnicity (Z Test: 95% CI (-13.3, -1.1), $p = .0202$).

Ethnicity	Hearing Loss Thought to be Present at Birth			n
	No	Yes	Unsure/Missing	
Māori	34 (8.4%)	156 (38.6%)	214 (53.0%)	404
New Zealand European	118 (17.7%)	244 (36.5%)	306 (45.8%)	668
Both	9 (10.6%)	39 (45.9%)	37 (43.5%)	85

Table 5: Frequency (percentage in parentheses) for hearing loss thought to be present at birth by ethnicity.

ⁱ Ethnicity is self-selected and is a reflection of the ethnicity the parents/children identify with as opposed to being a measure of racial heritage.

ⁱⁱ New Zealand European refers to an ethnicity of which members are predominantly of European descent; that they or their forebears originated in Europe.

Deprivation

Introduction

This year, for the first time, we have included deprivation data which has been provided by the Ministry of Health in our analyses.

The deprivation data is based on data from The New Zealand Index of Deprivation devised and calculated by the University of Otago (Wellington). ‘NZDep2013’ is the latest in the series which began in 1991. It draws on New Zealand Census data relating to income, home ownership, employment, qualifications, family structure, housing, access to transport and communications, allocating a deprivation score to every area in New Zealand.

The variables used to determine the deprivation score (NZDep2013) for a specific meshblock (small area) are contained in Table 6.

These areas (meshblocks) are small, containing a median of 81 people, and the scores allocated to

each are between 1 and 10, with scores of 1 being allocated to the 10% of areas which are the least deprived, and scores of 10 allocated to the 10% of areas which are the most deprived⁴⁴.

The deprivation scores allocated to the primary addresses associated with each National Health Identifier are used in this analysis. Please note that NZDep2013 relates to the *addresses at which tamariki were living according to their NHI*ⁱ – it doesn’t relate to the individual’s specific level of deprivation.

Of the 1344 tamariki in the main dataset, all but 25 (nearly 2%) had deprivation data available. Data were unavailable for tamariki whose: NHI was not valid (n=20), who currently live outside New Zealand (n=2), whose NHI was not provided until after the search was completed (n=1) and those who had no NHI listed (n=2).

Area	Variable in order of decreasing weight in the index
Communication	People aged <65 with no access to the Internet at home
Income	People aged 18–64 receiving a means tested benefit
Income	People living in equivalised households with income below an income threshold
Employment	People aged 18–64 unemployed
Qualifications	People aged 18–64 without any qualifications
Owned home	People not living in own home
Support	People aged <65 living in a single parent family
Living space	People living in equivalised households below a bedroom occupancy threshold
Transport	People with no access to a car

Table 6: Deprivation variables used for NZDep2013

ⁱ As at the date of extraction, in April 2017.

Tamariki in the database

In general, tamariki in New Zealand with one or more disabilities are more likely to live in higher levels of deprivation than those without.

The New Zealand Child Poverty Monitor noted that New Zealand children under the age of 17 are more than twice as likely to be living in income poverty than adults over the age of 65 years⁴⁵.

Child Poverty Action Group (2015) stated⁴⁶ disabled children are at increased risk of living in low-income households. Overall, 11% of children under the age of 15 have a disability. Māori are more likely to have a disability, while Pacific children have lower reported rates of disability⁴⁷.

Notifications

Tamariki in our main dataset (with a hearing loss averaging 26dBHL over four frequencies in one or

both ears) are much more likely to live in high deprivation areas than lower deprivation areas.

Tamariki who live in the most deprived areas are much more likely to be of Māori and/or Pacific ethnicities, and much less likely to be New Zealand European than those in the least deprived areas.

Further analyses relating to deprivation status can be found later in this report.

The developers of the NZ Deprivation Index have kindly provided data on the national deprivation (NZDep2013) distribution of tamariki in relevant age groups, so we could compare this with the distribution for children and young people whose diagnosis was notified to the database⁴⁸. Figure 9 shows these comparisons, for children 0-5 years of age, and those 6-17 years of ageⁱ.

ⁱ The ages of children/young people notified to the DND have been determined by establishing the age of each as at April 2017, when the deprivation code search was completed. This

is not the date at which NZDep2013 meshblock scores were allocated.

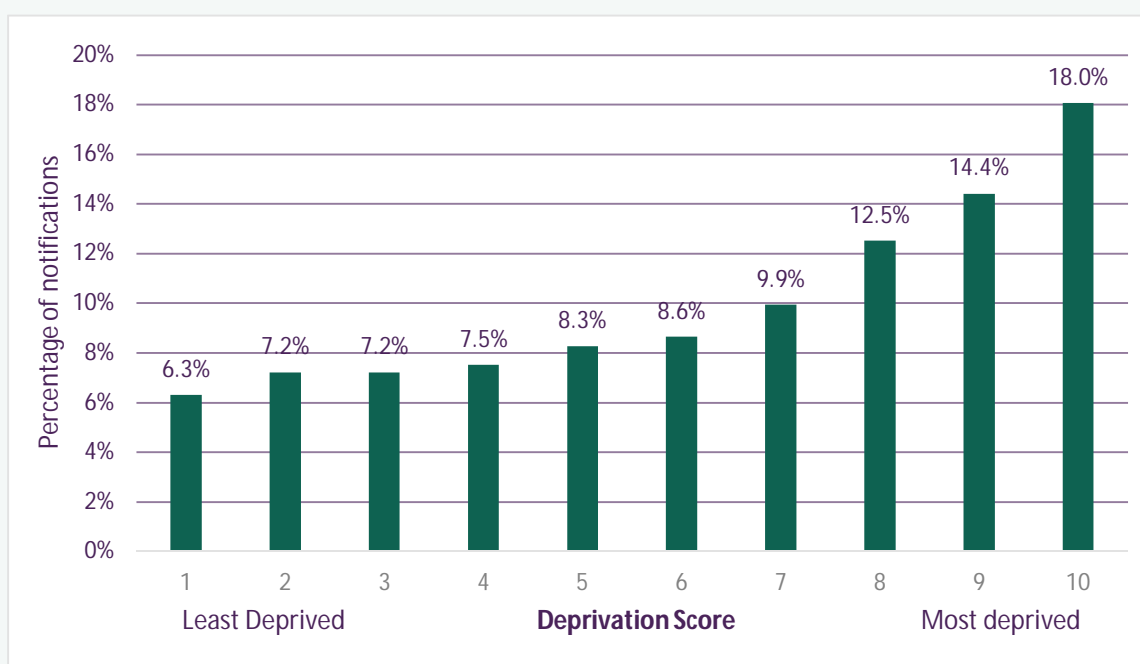


Figure 8: Deprivation scores of children and young people in the DND (2010-2016)

Both DND distributions are skewed towards the higher deprivation scores than the national distribution for tamariki of the same ageⁱ. This is particularly the case for tamariki notified to the database and aged 6-17, which contains a preponderance of those living in the four most deprived area groupings when compared to the national figures.

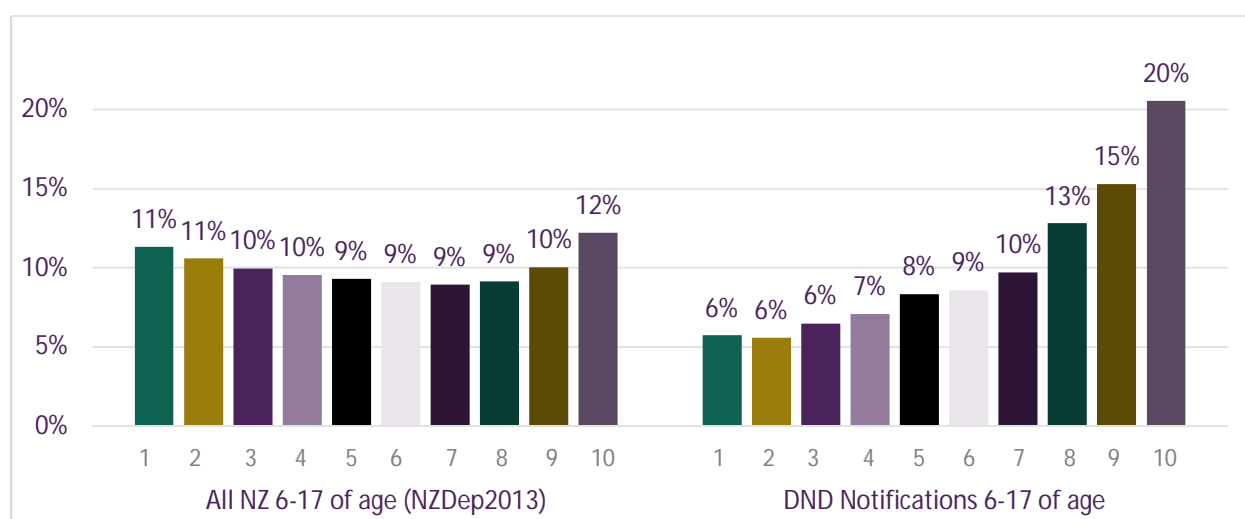
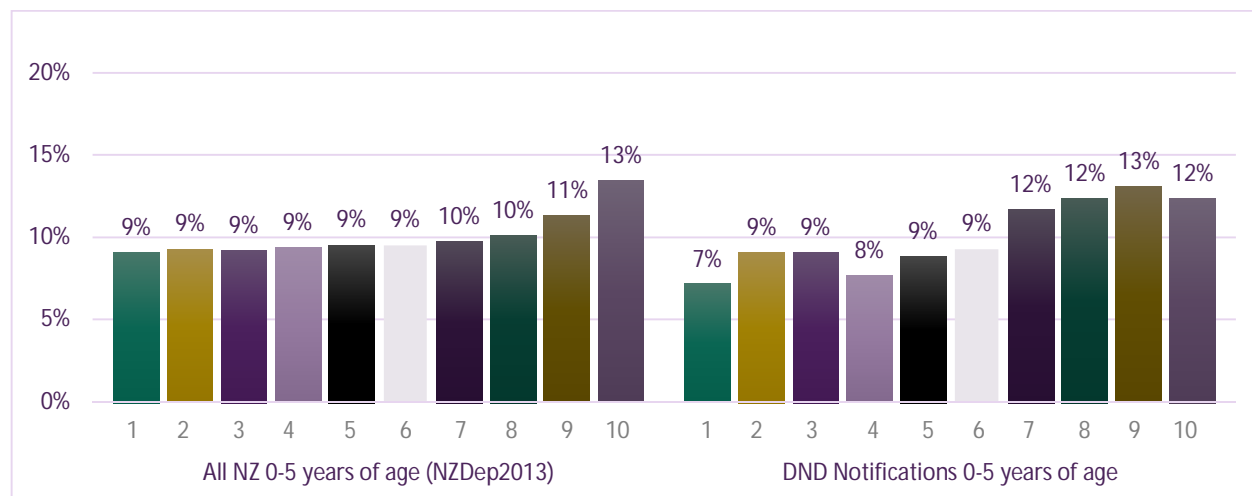


Figure 9: Comparison of deprivation distribution for New Zealand children (NZDep2013) and those notified to the DND by age group (2010-2016)

Figure 10 shows these the distribution of cases by deprivation status and splits these by ethnicityⁱⁱ. NZ European tamariki are more likely to be living in the less deprived areas of the country, while Māori and Pacific tamariki are more likely to be living in more deprived areas.

ⁱ Comparisons were made for 0-5 and 6-17-year age groups. These both showed fewer children in the lower deprivation scores and more in the higher deprivation areas than in the general New Zealand population for each age group.

ⁱⁱ The ages of children/young people notified to the DND have been determined by establishing the age of each as at April 2017, when the deprivation code search was completed. This is not the date at which NZDep2013 meshblock scores were allocated.

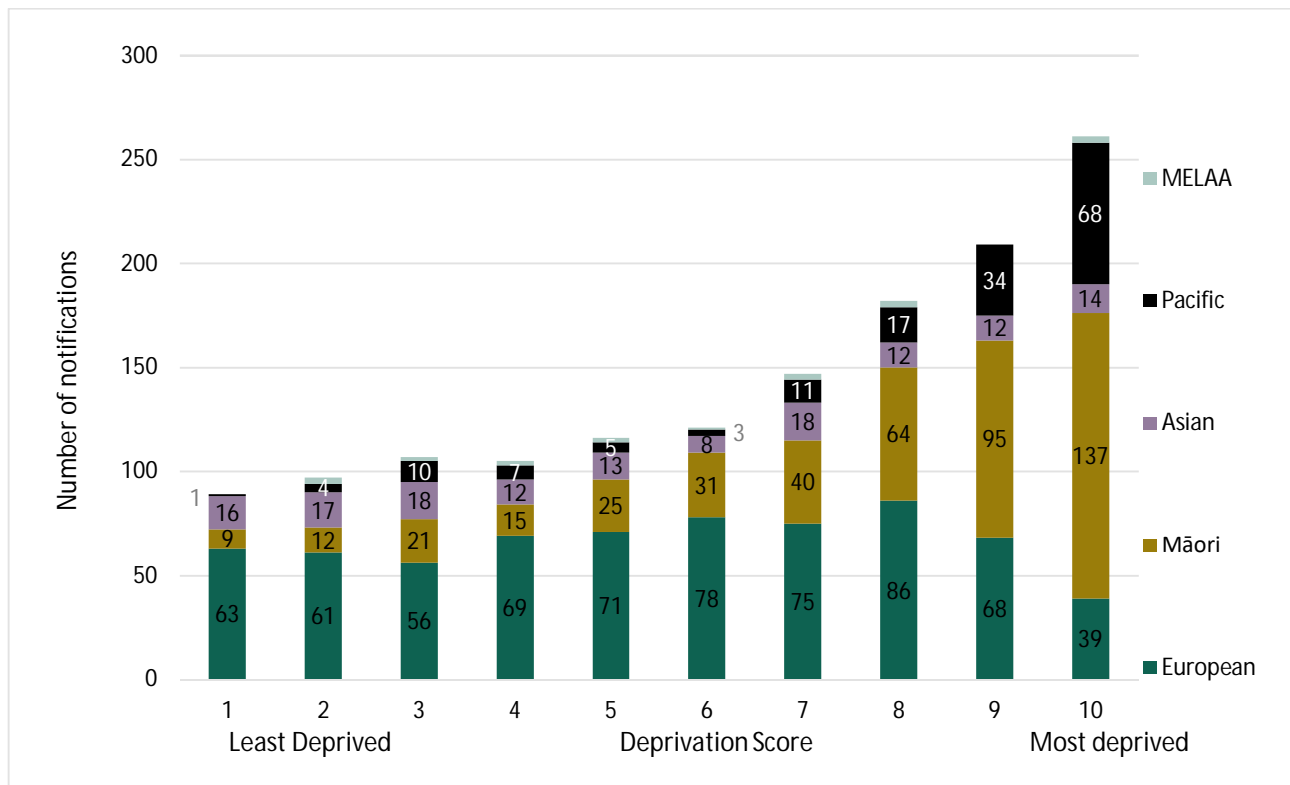


Figure 10: Deprivation scores of tamariki and young people in the DND by ethnicity (2010-2016)ⁱ

ⁱ MELAA labels have been removed as these are difficult to show clearly on this graphic due to the small numbers in each deprivation grouping. From left to right, these are: 0,3,2,2,2, 1,3,3,0,3.

Aetiology

Causes of deafness

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

The American College of Medical Genetics and Genomics estimated in 2014 that 30% of genetic deafness is syndromic⁴⁹. In non-syndromic deafness with a genetic cause, the most common genetic mutations found are in the GJB2 and Pendrin genes. The Otoferlin gene has been implicated in cases of ANSD⁵⁰.

The proportion of hearing losses with a confirmed genetic cause is increasing over time^{51,52}, as more hearing losses are better understood in terms of their aetiology, and as genetic testing becomes cheaper and more widely available.

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

In tamariki, mumps is thought to be the commonest cause of unilateral acquired sensorineural deafness and is usually sudden in onset and profound in severity⁵³.

Cytomegalovirus (CMV) is a significant cause of deafness among tamariki and young people in overseas studies, causing 10-20% of cases in those under the age of 5⁵⁴. In New Zealand, CMV seroprevalence was assessed from 9343 first-time New Zealand blood donors. The highest

prevalence was found among Pacific Islanders (93.2%) and the lowest in Caucasians (54.8%)^{55,56}.

Internationally, as reported by Davis and Davis¹, it is common for a high proportion of cases (between 15% and 57%) of hearing loss to be of unknown aetiology. **Aetiology** is reported as more likely to be investigated in cases of bilateral hearing loss, and where the hearing loss is more severe in nature, compared with unilateral cases or those which are less severe⁵⁷.

It is worth noting that identification of one aetiology does not exclude the presence of an underlying genetic predisposition. For example, the A1555G mitochondrial mutations may predispose a patient to hearing loss, and this hearing loss is expressed when certain antibiotics are used⁵⁸.

New Zealand data

Almost 99% of the 1344 records in the main dataset contain information about the aetiology of the child or young person's hearing loss, that is, whether the hearing loss is of known or unknown cause. Of those that do contain aetiological information, 88% are of unknown cause, with the remaining 12% being listed as having a known cause.

As seen in Figure 11, the proportion of hearing losses where the cause was thought to be known has decreased significantly in the years 2010 - 2016, when compared with figures from before 2006. At least some of this difference is thought to be the result of changes in the cause information

requested, as the notification form has been made more specific, asking for confirmed, and not suspected cause.

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

As mentioned earlier in this report, immunisation coverage in New Zealand has risen significantly since vaccination for tamariki became a

Performance Programme indicator in 2006. This is likely to be reducing rates of hearing loss from diseases such as meningitis (which presents with additional disabilities, although the numbers are likely to be smallⁱ), measles, mumps and rubella.

The importance of cytomegalovirus (CMV) in causing deafness among tamariki in New Zealand is not yet understood. There may be differences in rates of exposure to CMV among different ethnic groups in New Zealand, with serologic information from three-year-olds indicating higher levels of exposure among Māori and Pacific Peoples when compared with their New Zealand European counterparts⁵⁹.

Mumps, measles and meningitis were previously often considered by audiologists as possible causes of hearing loss; however, this has become less common as a result of increased immunisation coverage.

ⁱ It is difficult to compare the number of cases of meningitis over time as this information was not collected prior to the re-launch of the database in 2010 and as there is no specific

question related to meningitis in the current notification form.

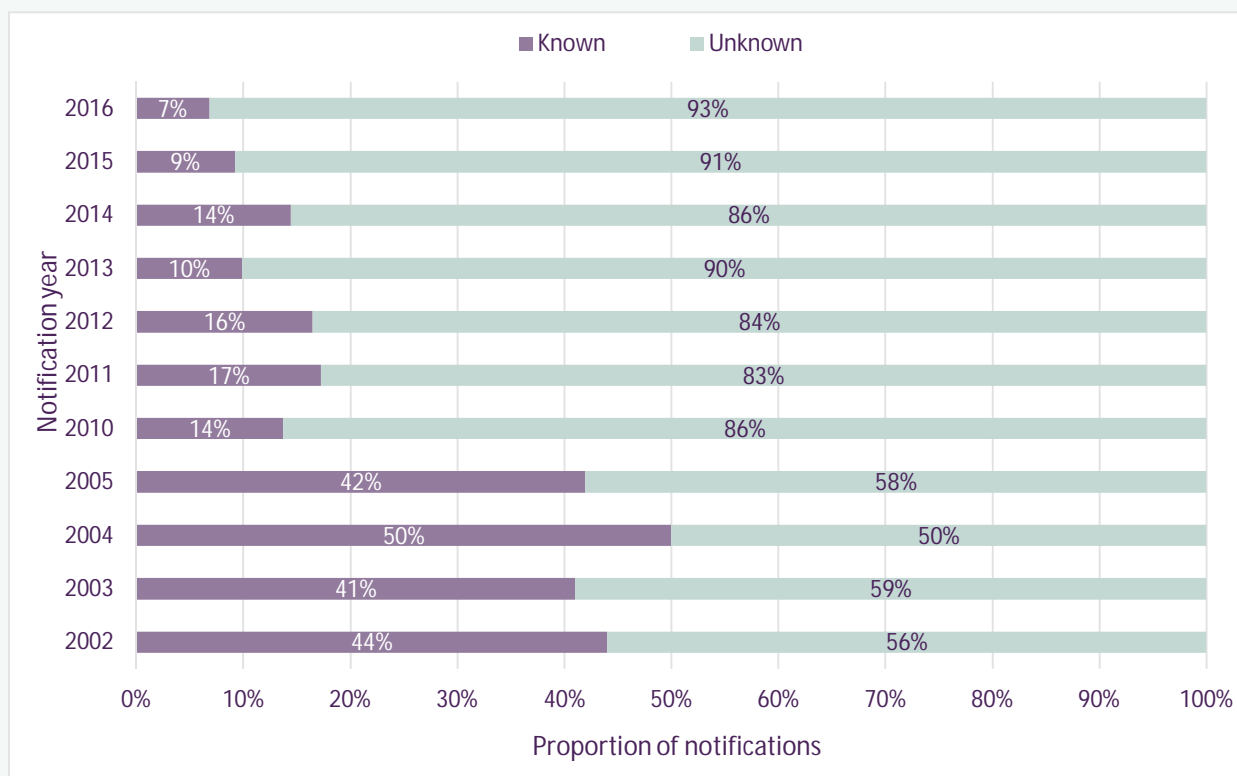


Figure 11: Proportion of hearing losses of known and unknown cause notified to the DND by year diagnosed (2002-2005 and 2010-2016)

It is worth noting that the current concern regarding mumps incidence in New Zealand, which is thought to relate to immunisation dose timing and coverage rates, may be having an impact on incidence and should again be a clinical consideration⁶⁰.

In New Zealand during the 2010-2016 period, bilateral hearing losses and those which were recorded as severe or profound in severity were

more likely to have a known aetiology than those categorised as mild and/or unilateral in nature.

Figures in this year's report are slightly different from those reported in last year's report, as a review of 'known' and 'unknown' codes provided by audiologists was undertaken in the context of the specific details provided. This meant a small number of codes changed from 'known' to 'unknown'ⁱ.

Testing for aetiology in New Zealand

During the last few years there has been a drive among the New Zealand based ENT specialist community to increase the proportion of hearing losses that undergo aetiological investigations, such as genetic testing, MRI and CT scans⁶¹.

Although practice varies, ENT specialists generally refer young people/families of children with

hearing loss for genetic testing where there is no clear explanation for the cause of the hearing loss.

Over time, more genes and mutations are being added to those for which testing is available in New Zealand. ENT specialists request the tests and counsel patients about the results. If multiple or unusual mutations exist, ENT specialists refer to genetic services⁶².

ⁱ For example, one case which was categorised as having a known cause by the notifying audiologist was changed in this way after reviewing the details which noted "Genetic cause

(strong family history of hearing loss on mother's side) results of genetic testing pending."

Identification of hearing losses

Who first suspected the hearing loss?

Information on who first suspected the child or young person’s hearing loss was recorded for all tamariki born in New Zealand and diagnosed in 2016.

Table 7 shows the top three groups that first suspecting the hearing loss among notified cases during selected years since 2010.

Parents/caregivers have gone from first suspecting hearing loss in more than a third of cases (37% in 2010) to being first in 23% of cases. Newborn hearing screeners were not in the top three groups to suspect a hearing loss in 2010 or 2011ⁱ and yet they are now first to suspect more than half of all cases of hearing loss notified to the database. For the second time, Vision Hearing Technicians (VHTs) are not listed in the top three groups most likely to first suspect a hearing loss. This is likely to be because many hearing losses previously identified first by the VHTs are now being identified as a direct result of newborn hearing screening.

Strong evidence exists that behavioural methods previously relied upon 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^{63,64}.

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

In addition, the challenges parents face in trying to identify their child’s hearing loss are considerable, particularly when their hearing loss is not so severe as to prevent speech 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 tamariki, over the last three years, towards those using objective methods, particularly those using these measures in newborn hearing screening.

The proportion of 2016 cases first suspected by parents or caregivers is again lower than at any time since the database re-launch in 2010. It is also below historic levels reported in the original database, which showed between 34% and 52% of cases were first suspected by parents during the 2000-05 period.

	2012	2014	2016
Most likely to first suspect	Parent or caregiver (23%)	Newborn hearing screener (40%)	Newborn hearing screener (57%)
Second most likely to suspect	VHT (17%)	Parent or caregiver (22%)	VHT (15%)
Third most likely to suspect	Newborn hearing screener (17%)	VHT (12%)	Medical professional (other than GP) (8%)

Table 7: Groups most likely to suspect hearing loss (Selected years, born in NZ)

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.

Age at diagnosis

Figure 12, below, shows the number of cases identified by the age of the childⁱ. There is a notable peak in the number of notifications during the first year of life – this is undoubtedly the effect of the universal newborn hearing screening programme. The peak for diagnosis in the first year of life is now almost four times as high as it was in 2010, when the database was re-launched. This is a positive trend, as it indicates more and more tamariki are being diagnosed early. A further, smaller peak can be seen for four, five and six-year-olds; this is likely to correspond to the B4 School Check^{ii,65}. The number of tamariki being identified at this time has fallen by almost half since 2010, although screening coverage for the hearing portion of the B4 School Check has been rising during this time. See page 41 for more

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

ii The B4 School Check aims to screen all tamariki before they reach school, and to identify and provide intervention to those tamariki identified with targeted conditions. Part of this Check involves screening tamariki for hearing loss. This screening should be completed on all tamariki not already

details about the B4 School Check. This suggests that some children who were previously being identified by hearing screening close to school age are now being identified through newborn hearing screening.

Overall age at identification

Caution: There are several issues with reporting the average age at identification (diagnosis) for all groups of tamariki. While this may have some relevance to measures used before 2006, as it describes the average age at which providers will, on average, begin working with tamariki to provide interventions of some type, the average relates to all newly diagnosed tamariki, as it is not possible to separate out tamariki with hearing

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.

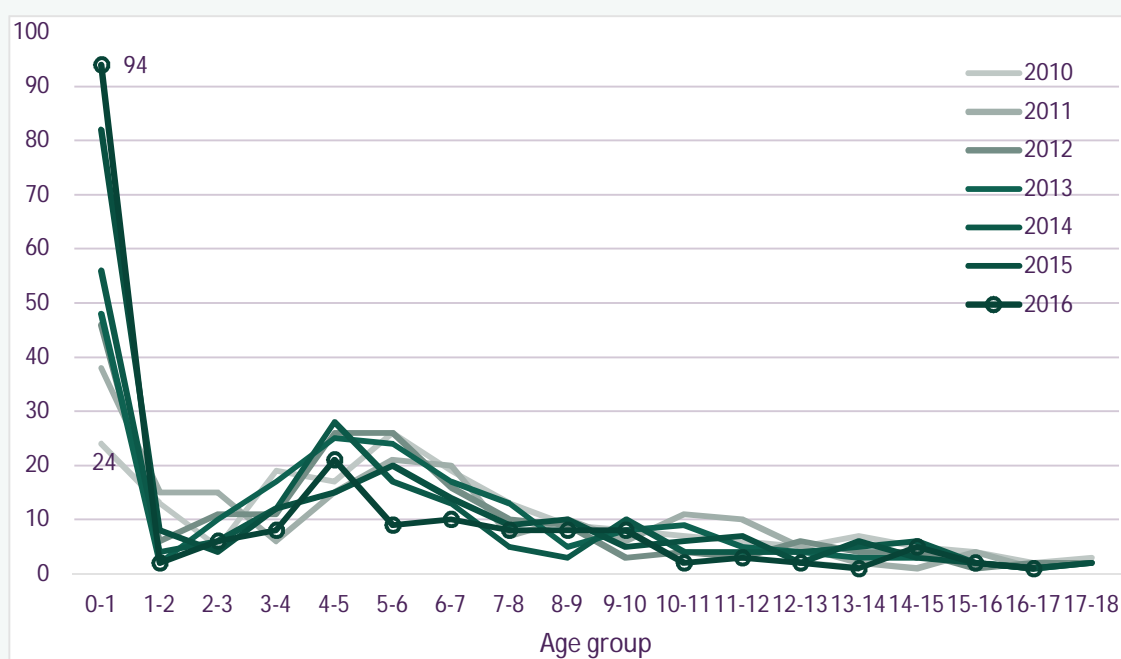


Figure 12: Number of children diagnosed by age (2010-2016)

losses that are late onset (such as progressive and acquired hearing losses).

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

Keeping this in mind, the average ages at diagnosis for children diagnosed and notified to the database are described in Table 8. This table shows that, although there has been a fall in the overall average age of confirmation, the reduction is quite

slow and seems to have been influenced by the increase in the number of notifications around five years of age for 2012 and 2013 as well as the increases at ten years of age for 2013 and at 10-11 years for 2011. Those born in New Zealand have a more marked drop in the average age than the full sample, which includes those born overseas and a small number where the place of birth was not provided on the notification form.

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

	2010	2011	2012	2013	2014	2015	2016
Average all cases	65	57	61	60	60	53	43
Average born in New Zealand	62	53	56	54	53	47	36

Table 8: Average ages of diagnosis for all cases in months (2010-2016)

For the purposes of comparison with previous data, the average age at diagnosis is presented, but those groups who are more and less likely to be identified later can be found in Table 9 below.

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

Table 9: Early and late average ages of identification (2010-2016)

ⁱ 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.

Age at diagnosis by severity of hearing loss

Table 10 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.

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2010-2015)	Total number of cases
mild	69	413
moderate	45	225
moderately severe	24	56
severe	22	29
profound	15	50

Table 10: Average age at diagnosis, in months, for *bilateral* hearing losses by degree (ASHA codeframe) using interpolated data with manual checks (2011-2016)ⁱ

Please note that, as a number of records in the database contain incomplete severity information, we have included those determined to be bilateral using both data by the audiologist and interpolated data-points. Also, please note that Table 10 only includes cases of bilateral hearing loss.

Children under the age of four are more likely to be missing some severity dataⁱⁱ, meaning they could not be classified for Table 11. This may be the reason why reductions in average age of diagnosis are not as clear in these data.

The greatest variability in the age at diagnosis is for mild and moderate hearing losses – understandable given that these losses can be difficult to identify. The database does not include

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.

information about the proportion of losses which are thought to be progressive in nature.

Age at diagnosis and ethnicity

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 reportⁱⁱⁱ.

Table 11 shows the average identification ages for each ethnic group by year, for all children and young people notified, where ethnicity information was provided.

Ethnic Groups	Average months at diagnosis (2010-2016)
NZ European	54
Maori	57
Pacific Peoples	72
Asian	45
MELAA	81

Table 11: Average months at diagnosis by ethnicity (2010-2016)

When viewing data on ethnicity, please keep in mind that Table 11 is based on multi-code data, hence a small number of cases are in two or more ethnicity groups at one time.

MELAA data are included below but this group is historically very small, hence variation in the averages over time are greater although generally these averages are high.

While Māori are more likely to have bilateral hearing losses (which are on average identified earlier than unilateral losses), they are also more likely to have mild and moderate hearing losses than their New Zealand European peers, and mild and moderate losses are on average identified later than severe and

ⁱⁱ A number of factors may influence this pattern, including that babies can wake during testing and that younger tamariki can be difficult to test.

ⁱⁱⁱ For example, the 1997 DND report noted a similar age of identification between Māori and non-Māori while the 2002 – 2004 reports noted a difference, with New Zealand European tamariki being identified, on average, earlier than Māori and Pacific tamariki.

profound losses³⁹. These opposing effects make it difficult to understand how effectively the system is working to detect hearing losses early among Māori children and young people. It is worth noting that the proportion of cases reported as Māori in the database has grown since 2010 – this could be an indication of some improvement in accurate coding of ethnicity in some areas, although we have no evidence to support this suggestion.

Another way to examine age at diagnosis is by splitting cases into Māori, New Zealand European or both.

We have done this using the more accurate measure of days old at diagnosis. This information wasn't required on the notification form until part way through 2011, so isn't available for 204 of the 1344.

Examining the variance in this way, using ANOVA, the age at confirmation is significantly older for those listed as Māori compared to those:

- listed in *both* the Māori and New Zealand European categoriesⁱ (95% CI: (82, 870)) - by 477 days, and those
- listed in the New Zealand European category only (95% CI (82, 870)) - by 230 daysⁱⁱ.

Pacific Peoples and MELAA consistently have the highest average age at diagnosis when compared with the other groups in the sample. We hope future analyses will shed light on the types of

hearing losses which are common among these groups, so we can better understand the reasons for their later average diagnoses.

Newborn hearing screening

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

Screening status

Table 12 shows the screening status of New Zealand-born children notified to the database (and therefore diagnosed) in the period 2010 to 2016.

As expected, the proportion of children being diagnosed as a direct result of referral from the UNHSEIP is increasing, and the proportion of notifications not offered screening is falling. Please note that this table shows children diagnosed at varying ages, so almost one in seven (13%) of the cases notified for 2016 were not screened as newborns as no [UNHSEIP](#) service was available in their area at the time of their birth.

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.

ⁱ There was no significant difference between New Zealand European only and those who are both New Zealand European and Māori (p value 0.2044).

ⁱⁱ We have repeated this analysis using the alternative code for months old at diagnosis, which contains complete data but is less accurate as it is not based on date of diagnosis. This ANOVA showed no significant difference in age at diagnosis between Māori and New Zealand European (p=.071).

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

Referrals from the UNHSEIP

Overseas, a number of comparable newborn hearing screening programmes (such as those in the UK and Australia) seem to be converging at a birth prevalence of approximately 1.0 to 1.1 per thousand babies for bilateral hearing losses, and approximately an additional 0.5 per thousand unilateral hearing losses^{66,67}. Using these overseas rates and including unilateral hearing losses, we might expect approximately 100

diagnoses directly from the newborn screening programme each year, based on a figure of 62,000 births per yearⁱ.

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

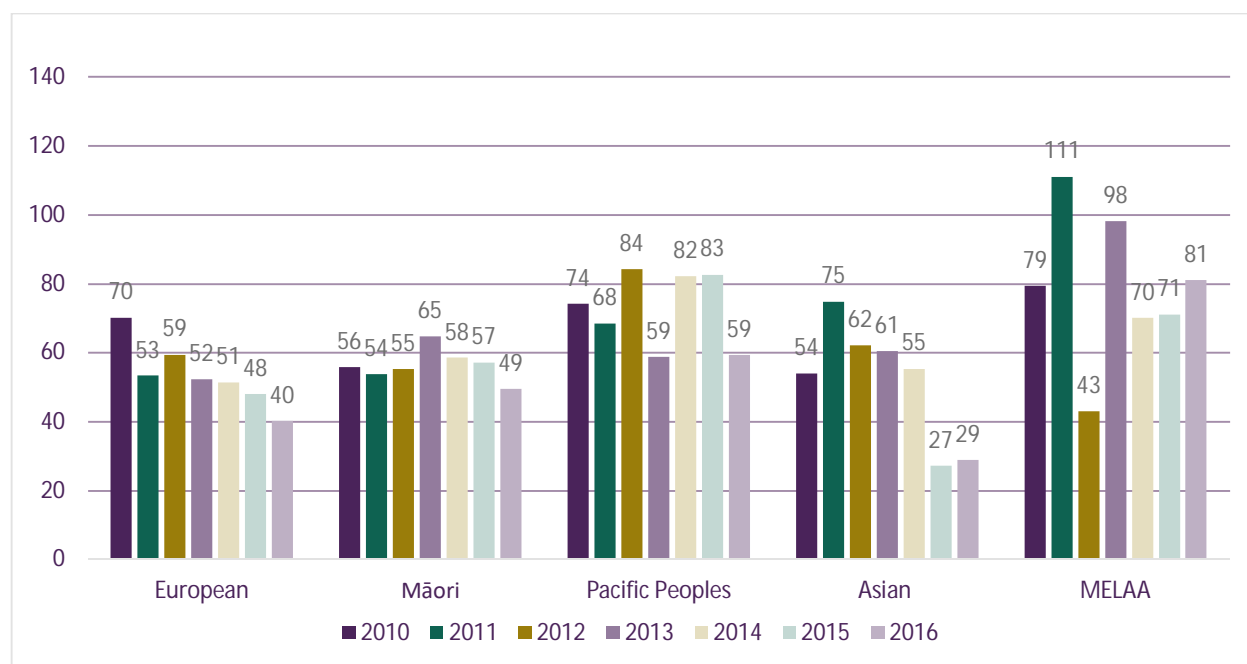


Figure 13: Average age of diagnosis by ethnicity in months (2010-2016)

Now we have local data from the Deafness Notification Database suggesting that young Māori have higher rates of hearing loss than is found in the New Zealand European population. In addition, our national screening programme for newborns (UNHSEIP) demonstrates our rates of

hearing loss at birth are somewhat higher than those reported in similar jurisdictions overseas, at 1.2 cases of bilateral hearing loss per thousand babies screened, plus an additional 1.1 per thousand cases for unilateral hearing loss per thousand babies screened³⁵.

ⁱ This is an approximation of the number of births reported in 2010.

Was universal newborn hearing screening (using aABR or aOAE) offered to this family after this child or young person's birth?		2010	2011	2012	2013	2014	2015	2016
No	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	4%	2%	4%	1%	4%
	No, this service was not available at the time (at the time of diagnosis)	68%	53%	54%	49%	38%	27%	13%
Unsure	Unsure whether screening was offered to this family	7%	4%	6%	7%	5%	5%	2%
Yes	Yes, a screening programme was in place, but the child was directly referred to audiology due to atresia	0%	0%	1%	0%	1%	1%	5%
	Yes, screening was offered but this child was not screened	1%	1%	1%	1%	1%	3%	2%
	Yes, the child was screened and referred but follow-up did not occur at the time, and so this is a delayed diagnosis	1%	4%	2%	5%	1%	2%	5%
	Yes, this child was screened and passed	1%	7%	6%	5%	13%	16%	16%
	Yes, this diagnosis is a result of a referral from screening	18%	27%	28%	32%	39%	46%	53%

Table 12: Screening status of children born in New Zealand and diagnosed 2010-2016ⁱ

A total of 88 of the 2016 notifications (53%) related to children born in New Zealand who were diagnosed as a direct result of newborn hearing screening. This percentage has more than tripled since the database was re-launched in 2010.

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

	2010	2011	2012	2013	2014	2015	2016
Number of diagnoses resulting from universal newborn hearing screening	34	44	48	53	59	82	88
Diagnoses as a proportion of total notifications	18%	27%	28%	32%	39%	46%	53%

Table 13: Diagnoses resulting from newborn hearing screening in New Zealandⁱⁱ, 2010-2016

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

ⁱⁱ Please note that the table shown in the 2011 report contained data for all cases, whereas this table contains data only for tamariki born in New Zealand.

The most recent NSU monitoring report³⁵, includes screening data from 1 January to 31 December 2015, and audiology data to 9 August 2016 for tamariki screened in 2015, reports thatⁱ:

- 91.5% of babies born during 2015 completed screening within the period. Of those babies who completed screening, approximately 83.1% did this by the target of one month (corrected age) – 1-month screening completion rates for DHBs ranged from 50% to 98%;
- 2.4% of babies screened during the period were referred to audiology, and an additional 2.2% of tamariki who passed their screening were referred for surveillance owing to the presence of one or more risk factors for hearing loss;
- 66.5% of the 1,318 babies referred to audiology had audiology assessment data reported to NSU by the date of data extraction for the report (August 2016). 56.4% of babies referred had their assessment completed by the target time of three months of age, well below the target of 90%;
- An additional 7.9% (n=104) were categorised as either DNA, lost contact or declined, 79 of these tamariki were Māori;
- Māori and Pacific babies were less likely to be screened, more likely to be referred and less likely to have audiological assessments completed compared with Asian or Other ethnic groups.
- Rates of permanent congenital hearing loss (both bilateral and unilateral) were higher for Asian babies (2.8 per 1,000 babies) Māori babies (2.7), Pacific babies (2.6) than their Other category counterparts (1.8);

- Given that Māori and Pacific babies are less likely to be screened, more likely to have a hearing loss and less likely to complete audiological assessments than the Other group, the true prevalence of hearing loss in these groups are likely to be higher; and
- 121 babies in total were identified with some type of hearing loss during the period.
- 97.7% of referrals to early intervention services began receiving services by six months of age, higher than the 90% target. [At this time, we have no data to help us understand ongoing access rates.]

Key screening goals – age at diagnosis

New Zealand's UNHSEIP was implemented to reduce the age of intervention for children born with hearing loss, as this approach had been successful overseas in improving outcomes. Such 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 tamariki 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 tamariki 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 DND reports provide useful data to show how the overall age at identification changes over time.

ⁱ Please note that the most recent report contains data from 2015, and so doesn't align with the DND reporting period (calendar year 2016).

There has been a pleasing reduction in the average age at diagnosis of cases referred from newborn hearing screening in New Zealand (therefore born in New Zealand), from ten months in 2010, and dropping by a month each year to four months in 2016.

Of the 88 cases notified in 2016 that were identified as a direct result of newborn hearing screening in New Zealand, 69 (78%) were diagnosed by the internationally recommended age of three months. This is a pleasing improvement on previous levels.

	2010	2011	2012	2013	2014	2015	2016
Number of diagnoses resulting from universal newborn hearing screening	34	44	48	53	59	82	88
Average months at diagnosis	14	8	6	7	5	6	4

Table 14: Newborn hearing screening diagnoses and age at diagnosis (2010-2016)

Identification of false negatives

The DND likely provides the only method for identifying potential **false negatives** from the newborn hearing screening programme⁶⁸.

In 2012, there was a Ministry of Health initiated recall of 3,422 babies, 2,064 of whom had potentially been incorrectly screened; 901 of these tamariki had been rescreened by 28 November, 2012⁶⁹. In 2016, no cases notified to the database were explicitly identified as having wrongly passed their New Zealand based newborn screening^j, meaning we have no confirmed false negative cases for this year. This isn't to say that one or more babies diagnosed in 2016 weren't incorrectly passed at their newborn hearing screening, just that none were recorded as such in the notifications.

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

Twenty-seven of the tamariki identified with hearing loss during 2016 had been screened previously and passed this screening. This figure, and the fact that it is rising, is not necessarily a concern, as many tamariki develop hearing losses after their initial diagnosis, and over time more tamariki are being screened.

Of these 27 cases, there are two groups that may be useful to remove to help us identify potential false negatives. The first of these have known acquired hearing loss, while the second is those with hearing losses where there is some uncertainty – they were either suspected to have been present at birth, or the diagnosing professional was unsure whether the hearing loss was likely to have been present at birthⁱⁱ. As the second of these groups is based on a relatively subjective assessment by the clinician, these cases may or may not provide cause for concern. It is possible New Zealand has a greater prevalence of progressive hearing losses because of our high rate of CMV⁷⁰.

ⁱ One child was reported to have been screened in Australia and was referred. This child then passed the rescreening, and then later was discovered to have a hearing loss in New Zealand. Not enough detail was provided to ascertain whether this case was a likely false negative.

ⁱⁱ 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.

	2010	2011	2012	2013	2014	2015	2016
Total cases identified by year who were screened previously (i.e. are not currently referrals from the UNHSEIP) and who passed this screening	2	11	10	9	19	28	27
Number of cases from regional screening programmes, or from the UNHSEIP, which passed screening, which were not thought to be acquired loss, and where the notifying professional answered 'yes' or 'unsure' to the question about whether the loss was thought to have been present at birth and who were born in NZ	2	5	4	6	10	19	17

Table 15: Potential false negatives and cases previously referred from hearing screening, 2010-2016, born in New Zealand only

Of the seventeen 2016 cases identified as potential false negatives in Table 15, the age of identification for these tamariki ranged from two years to eight years two months of age.

The audiologists had difficulties getting confirmed diagnoses in four of these cases. Further details were provided about two:

- "Passed BC NBHS but retained for follow up due to suspicion of mild loss."
- "Child passed newborn hearing screening test. No hearing loss was suspected until she did

not pass a test that was done at school, so she was referred for a full audiological assessment."

Of the 17 cases listed, one of these was coded 'yes' to the question about whether the loss was thought to have been present at birth (no further information was included in that specific record about what might have occurred.) This is the category that is most likely to include false negatives, although false negatives could exist anywhere in the full group of 17.

B4 School Check

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

required) are administered by Vision Hearing Technicians around the country.

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

Outcome	Description	2010/11	2011/12	2012/13	2013/14	2014/15	2015/16
Pass Bilaterally	The child was screened and passed.	58%	67%	71%	74%	79%	80%
Referred	The child was screened and referred to a relevant service.	5%	5%	5%	5%	5%	5%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	7%	8%	7%	6%	6%	6%
Under care	The child is already under the care of a relevant service.	1%	3%	3%	3%	3%	4%
Decline	The hearing check was declined by the caregiver.	4%	5%	4%	3%	1%	1%
Not Checked	The child did not receive a hearing check.	24%	16%	11%	12%	6%	5%
Population	Derived from the PHO enrolled population.	63,585	65,692	64,911	65,335	63,730	62,785

Table 16 B4 School Check Hearing Screening data (those tamariki screened in 2010-2016)^{i,41}

The overall referral rate for tamariki completing this screen is 5.3% (2015/2016). Māori and Pacific tamariki have higher rates at 7.4% and 8.9% respectively, and 4.5% of Asian tamariki were referred. The lowest referral rate was for New Zealand European tamariki, at 3.9%.

The coverage rates in 2015/16 for each ethnic group also vary, with only 1.5% of New Zealand Europeans not checked, compared with 8.3% of Asians, 7.0% of Pacific Peoples, and 7.3% of Māori.

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

Delays in diagnosis

Information about delays

Those notifying cases were asked to provide information about the length of delay in identifying hearing loss and reasons for the delay, where one existed. Not all 2016 cases for which there was a delay had one or more reasons for the delay listed.

The average delay in 2016, between first suspicion of the hearing loss and confirmation of the child or young person's hearing loss, *including* those born overseas, and mild, acquired or unilateral hearing lossesⁱ was nine months.

Almost half the delays recorded in 2016 were one month or less in duration.

Year	Delay in months
2016	9
2015	11
2014	13
2013	12
2012	10
2011	16
2010	23

Table 17: Delay in months by year, 2010-2016

While 9 months is a significant average delay between first suspicion of a hearing loss and confirmation of this loss, average delays in the last five years are improvements on 2010 and 2011 figures. This is likely to be owing to the introduction of, and improvements in, newborn hearing screening around the country.

ⁱ Some previous reports (prior to 2006) included only children with moderate or greater losses, which were not thought to be acquired in nature, for children born in New Zealand.

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

Examining the differences in delay by ethnicity showed that the average delay for Māori tamariki was:

- 7.5 months higher than those for New Zealand European tamariki (One-way ANOVA: 95% CI (0.65, 14.40)).
- 15.66 months higher than those tamariki who identified as both Māori and New Zealand European (One-way ANOVA: 95% CI (2.71, 28.60)).

Māori tamariki were 1.60 times more likely to have one or more reasons listed in their notification form when compared with their New Zealand European counterparts (Logistic regression: 95% CI (1.24, 2.05)). In addition, Māori had a higher average number of reasons listed for a delay by a factor of 1.32.

There was also a significant association between the average length of delay and deprivation, with each one-unit increase in deprivation being associated with a one month increase in the average delay in diagnosis ($p=.0012$).

Delay causes

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

In 2016, 41% of all cases had one or more reasons for delay listed, with 29% having one reason, and 12% having two or more reasons for the delay listed. The number of cases with no reasons listed for the delay has risen during the last four years.

Again, this year the analysis in Table 18 examines the reasons for delay where one or more reasons are listed *and* where the delay was reported to be greater than six months, measured from the time the hearing loss was first suspected until the time when the hearing loss was diagnosed. For cases diagnosed in 2010-2016, Table 18 shows the most commonly cited reasons for delays in diagnosis.

Table 18 also contains some possible approaches to reducing the various types of delay.

Rank (most frequently mentioned)	Reasons for delay	The authors have identified some potential ways to reduce the length of delay
1st	Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)	<ul style="list-style-type: none"> efficient clinical practice to complete assessments over fewer appointments (Following 2016's Diagnostic and amplification protocols⁷¹ which can be found on the National Screening Unit website and which used to be referred to as Appendix F) prompt referral from newborn hearing screening
2nd	Parents did not attend appointments (for any reason)	<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 e.g. work to reduce 'do not attend rates' audiology services closer to home for families (e.g. community based clinics or outreach) reduced waiting times
3rd	Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)	<ul style="list-style-type: none"> better funding for audiology resources/DHBs to prioritise newborn hearing screening referrals and other paediatric cases better communication with parents, more attempts to contact families before discharging from service e.g. work to reduce 'do not attend rates'
4th	Parents suspected something other than hearing loss (e.g. speech delay, developmental delay)	<ul style="list-style-type: none"> better education for parents so they can identify signs of a possible hearing loss (including before baby is born through newborn hearing screening materials and using these as an opportunity for discussion) clear guidance on pathways for assessment for parents
5th	Difficulty getting a referral to audiology (e.g. GP or other health professional dismissed parent concern and no referral was made)	<ul style="list-style-type: none"> education for non-hearing professionals such as doctors, well-child nurses, preschool teachers, childcare workers, speech language therapists (SLTs) and specialists
6th	Follow-up lost in the system and did not occur as scheduled (between professionals or review or follow up appointment not made) OR Referral not made between professionals	<ul style="list-style-type: none"> better systems and processes for scheduling and seeing follow-up occurs
7th	Child was born or lived overseas, and hearing loss not diagnosed there	<ul style="list-style-type: none"> audiologists and others 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 18: Top reasons for delay for those with diagnosis delays of more than six months and possible remedies (2010-2016)

Comments received regarding 2016 diagnoses sometimes contained information about the reason for delays, and some of these are reproduced below. These comments demonstrate the complexity of reasons for delayed diagnosis including both challenging situations for families and audiologists working in less than ideal conditions:

“Unable to obtain hearing thresholds at 1kHz as baby was a restless sleeper, several attempts at ABR required to get the notified results.”

“Baby screened in [one DHB] but family DNA-d diagnostic apts. Baby transferred to family in [another DHB] in view of social concerns, paed[s] [there] sent referral directly to Audiology in view of confirmed CMV”

“Child did not receive NBHS [newborn hearing screen]. Failed B4SC Hearing Screening but referral for formal audiology did not occur... Referred to us by ORL due to ongoing parental concerns re hearing and speech (no middle ear fluid found on grommet reinsertion).”

“A number of factors contributed to this delayed diagnosis - non-attendance, baby not settling, interference from construction work at hospital, challenges on VRA due to excessive false positives from patient. Eventual diagnosis was a combination of VRA testing and ABR under sedation.”

Delays attributed to newborn hearing screening

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

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

More information about the causes of delays in all groups can be found in the section on *Delay causes*, beginning on page 44.

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 three months, by which time ABR becomes more difficult due to babies being less likely to sleep without sedation or anaesthesia. Without early ABR testing for these tamariki it can be more difficult to obtain a diagnosis for this group until they can be tested using Visual Reinforcement Audiometry (VRA) at six months to two years of age. (Some tamariki may not be testable using VRA until after six months due to other developmental difficulties.)

Short Case Study: Reducing ‘Do Not Attend’ rates in Taranaki

April 2017’s [Screening Matters](#) contained an article on the Come Here project, which aimed to improve health outcomes for Māori tamariki by reducing do not attend (DNA) rates at DHB audiology clinics. This programme, led by Mary Bird (Service Improvement Advisor and UNHSEIP Coordinator) and her team, has seen reductions in the DNA rate for Māori, although keep in mind that these rates fluctuate as the numbers are small.

Average annual DNA rates are now 17% and are continuing to stay low even after the project has

ⁱ These six cases contained seven reasons for the delay.

moved into maintenance mode. This compares very favourably to the 20-31% DNA rates before the project begun. Recent months have seen 2-3% DNA rates. Given that families might have to travel up to 1.5 hours to get to Taranaki Base Hospital from places such as Waverley, those figures are particularly impressive.

The Māori health team is involved, flyers have been developed and include a version in te reo Māori on the reverse, and these were distributed to Tui Ora, Nga Ruahine and Ngati Ruanui for the Whanau Ora nurses. Awareness of the 1:3:6 screening benchmarks has improved among

audiology staff and the Māori Health service team members, and the audiology service is now more aware of cultural requirements. There is also an information loop which means that the DNA rate is monitored and reported monthly by the Audiology Administrator. The Administrator role has been found to be critical to the success of the new pathway.

This work is relevant to delays for identification of newborn hearing screening, as parents not attending appointments (for any reason) is the second most commonly cited reason for delays in identification of hearing loss.

Severity

Audiometric data

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

Notifying clinicians are encouraged to provide as much audiometric data as possible for each case they are notifying to the database.

Those notifying cases were approached where significant information was missing, and were able to fill in some gaps. Of the cases that still contained missing data, data are more commonly reported for 0.5 kHz and 2.0 kHz and less likely to

be reported for 4.0 kHz and 1.0 kHz frequencies. This demonstrates that frequencies which are typically tested at the end of the protocol for testing young tamariki are less likely to be complete (i.e. 4.0 kHz and 1.0 kHz).

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

As shown in Table 13, below, the proportion of cases for which the thresholds were determined through ABR is rising, from 21% in 2010 to 50% in 2016. This is thought to indicate that fewer tamariki 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 losses are diagnosed at younger ages, on average.

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

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

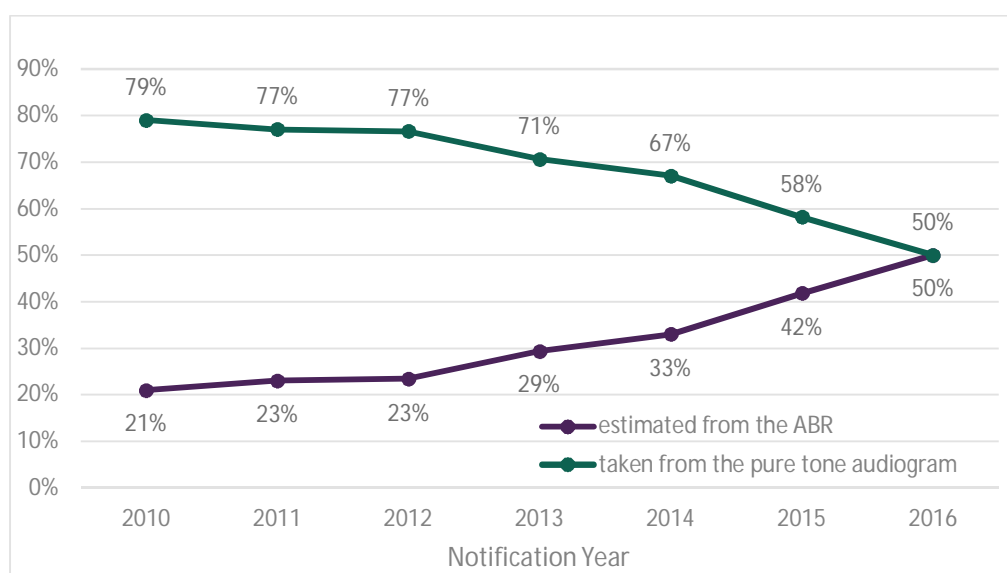


Figure 14: Proportion of cases containing thresholds from ABR (as opposed to being taken from the PTA), by notification year 2010-2016

Classifications

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

Degree of loss	Clark 1981 (ASHA) ⁷²
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 19: Clark's 1981 ASHA severity codeframe

Calculating severity for notifications

While the New Zealand DND collected some audiometric data for a number of years until the end of 2005, this information was insufficient to allow comparisons to be made easily with data from other jurisdictions. From 2010, the re-

launched database has requested full audiometric data from those notifying cases, in the hope that more meaningful comparisons can now be made with overseas data.

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

Interpolation

Table 20 shows the severity of hearing losses diagnosed for the first time in 2014, which is calculated in two ways. The first of these is using data containing all eight data-points, while the second includes interpolation.

While only cases where all eight-audiometric data-points are present can be included in most severity calculations, interpolation of data has

been used in some cases to provide a more complete picture of the severity of hearing losses notified. Interpolation is only used where three of the four data-points are provided for one ear, and where both data points surrounding the interpolated point are provided. This technique is becoming increasingly useful as more tamariki are being diagnosed earlier, meaning they cannot have their hearing assessed behaviourally.

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

Key findings include:

- that the proportion of less severe hearing

losses is higher among bilateral cases when compared with those pertaining to only one ear;

- the number of bilateral hearing losses for which severity can be calculated rises when interpolation is used;
- that the proportion of mild bilateral losses drops when these cases are removed, increasing the proportion of moderate and greater hearing losses; and
- the proportion of moderate and moderately severe losses rises for unilateral cases.

The table below compares the proportion of bilateral/unilateral cases, comparing those that have not been interpolated or had manual checks to those that have. Please note that this includes all notifications from 2010-2016 while last year's report only contained 2014 data.

Degree of loss using ASHA severity codeframe	Bilateral 2010-2016	Bilateral 2010-2016 (interpolated and manual checks)	Unilateral 2010-2016	Unilateral 2010-2016 (interpolated and manual checks)
Mild	61%	54%	49%	46%
Moderate	27%	29%	18%	18%
Moderately severe	5%	8%	11%	12%
Severe	3%	4%	5%	7%
Profound	5%	6%	18%	18%
Sample size	n=596	n=796	n=352	n=380

Table 20: Comparison of severity classifications by methodology, 2010-2016

Severity profile differences between bilateral and unilateral hearing losses

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

This year, a similar graph is included, but this time we have included the severity profiles for bilateral and unilateral hearing losses for cases in which missing audiometric data could be interpolated (meaning more cases can be classified by their

severity) and where a manual determination of whether the loss was bilateral or unilateral could be made based on available data. The authors of this report believe this shows a more accurate picture, and this method of analysis will be used in future.

Figure 15 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).

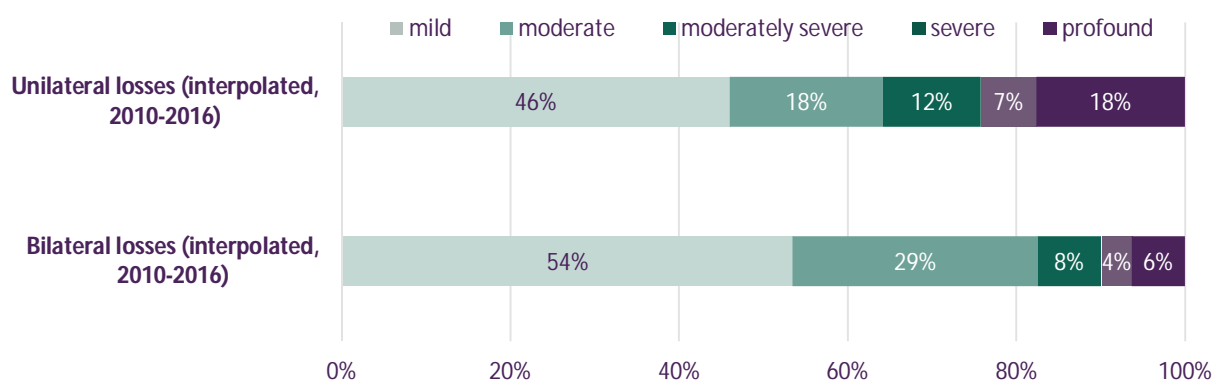


Figure 15: Unilateral and Bilateral hearing losses by degree (2010 - 2016)

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 in 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 lossesⁱ.

- 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 tamariki in each group is possible using data reported between 2001 and 2005 and more recent data.

The database at that time excluded cases of unilateral hearing losses, tamariki born overseas and those with acquired hearing losses. The 2010 to 2016 figures shown here match those exclusions from the earlier database.

Table 21 shows the average proportion of hearing loss notifications in each category between 2010

and 2016 and compares this with data from 2001 to 2004ⁱⁱ.

Proportion of cases notified by degree of hearing loss	Average 2001-2004	Average 2010-2016
Mild	48%	54%
Moderate	35%	33%
Severe	10%	7%
Profound	6%	6%

Table 21: 2001-2004 DND data compared with interpolated 2010-2016 notification data, selected cases only, 1996-2005 DND severity codeframe

ⁱ It is worth noting that as the average age for identifying hearing loss reduces as a result of newborn hearing screening, the severity distribution at the time of diagnosis for hearing losses should be shifting towards the lower severity categories.

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

In a previous reporting period we noted that the severity profile of cases had changed – we noted that we would be watching future data to see whether the profile returned to a pattern that more closely matched that seen before 2005. A return to historical patterns with fewer mild losses is not evident, either when cases containing full audiometric thresholds are considered, or when comparing data in Table 21, which includes more cases by using interpolated and manually checked thresholds.

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

- information about individual tamariki and young people are included in the dataset at the time of first diagnoses. A greater proportion of hearing losses are now being

identified earlier, thanks to the introduction of newborn hearing screening. As a result, progressive hearing losses have not yet had the time to worsen, meaning the proportion of more severe losses may be less;

- some cases with audiometric data points in the severe and profound range did not contain complete audiometric data and these have not been included in this table, meaning severe losses (and other degrees too) may be under-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 loss⁷³. However, the reduction in the number of more severe cases due to meningitis is likely to be small.

Ethnicity and severity profiles

Historically, DND reports have shown that the greatest number of notifications pertain to New Zealand European and Māori and children, and that milder degrees of hearing loss are more commonly reported among Māori^{38, 74}. A recent analysis of 1982-2005 data³⁹ confirmed that young Māori in the database are more likely to have mild or moderate hearing losses when compared with their New Zealand European peers.

This pattern is repeated with recent Deafness Notification data. Last year's analysis showed the proportion of cases in each of the severity categories, split by ethnicity grouping – which

showed Māori had a higher proportion of mild and moderate cases than their New Zealand European peers.

An analysis was completed using 2010 to 2014 interpolated data, and this showed the same pattern of results. In addition, an analysis of those cases that were coded only as Māori or New Zealand European was also completed, showing the proportion of cases of moderately severe or greater severity was 8% among Māori, compared with 14% among New Zealand European. It was 7% among those listed as both Māori and New Zealand European.

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

Several analyses have been conducted for previous DND reports to compare the DND's severity distribution with those from other countries and jurisdictions.

Despite differences in cohort and severity codeframes used, these analyses show a consistent pattern, with DND data showing a relatively higher number of cases with mild and/or moderate hearing loss, and a smaller number of cases with severe/profound hearing loss.

Details of these comparisons can be found in the reports noted:

- UK, Finland and the USA data with NZ data 2010-2012 (2012 report)
- Colorado data with NZ data 2010-2013 (2013 report)
- Australian data with NZ data from 2010 to 2015 (2014 report)
- Colorado data with NZ data 2010-2015 (2015 report)

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

Intervention and support

Ministry of Education

The Ministry of Education are to contribute data to future UNHSEIP Monitoring Reports. This means data for this year will not be included in this report, but will be included in the next UNHSEIP Monitoring Report and summarised in the next DND Report (2017).

Deaf Education Centres

Kelston Deaf Education Centre (KDEC) and van Asch Deaf Education Centre (vADEC) provide services to Deaf and hard of hearing students. The two DEC's have had a combined Board of Trustees since 2012; the strategic focus of this Board is on working together with families/whānau and the Deaf community to provide equitable and coordinated deaf education, so that deaf and hard of hearing students:

- contribute meaningfully to their communities;

- are socially well integrated; and
- are able to determine their future and fulfil their dreams.

The combined Board has prioritised the development of an accurate national picture of the deaf student population.

There are terms used in the education sector that may not be familiar to readers of these reports, who are largely health-based. The categories, funding streams and eligibility are all relatively complicated, and there have been changes to the terms used in recent years. As a result, the authors of this report have rewritten this section of the report in the hope we can make the categories of service easier to understand, particularly for those not familiar with education terminology. Terms such as ORS can be found in the glossary on page 63.

As at 1 December 2016, the Deaf Education Centres provided services to 2254 tamariki. These services can be broken down into the following categories⁷⁵:

Type of support	Description	KDEC	vADEC	Total
1. School Provision	<p>This category includes school aged children and young people who are supported by KDEC and vADEC staff (Teachers of the Deaf) in regular schools.</p> <p>Most of these children are ORS verified as 'high' or 'very high' needs.</p> <p>This category used to be referred to as 'Deaf Units'.</p>	n=98	n=30	n=128
2. Resource Teachers Deaf	<p>This category includes children over the age of 3 years although most children receiving this support are over the age of 4.5 years.</p> <p>Children in this category are not always ORS verified as they can't be verified until they begin school.</p> <p>Funding for this service comes from ORS funding (0.1 and 0.2 FTEs) and the DEC's have some allocation of RTDs under the moderate needs contract.</p>	n=379	n=310	n=689
3. Specialist support: DEC funded, and teacher supplied by student's school	<p>These ORS verified children (school aged children in mainstream schools and children in special schools) have funding which goes from the Ministry of Education to their schools, including KDEC or BLENZ.</p> <p>For example, this funding can be used for teacher aids and other specialist support (occupational support, physical therapists, speech language therapist, Kaitakawaenga etc.) where staff are employed by the MOE.</p>	n=83	n=72	n=155
4. ASSIST	<p>ASSIST stands for Assessment and Involvement of a Specialist Teacher. ASSIST professionals provide assessment services for children to determine their needs.</p> <p>Implementation of changes following the Wilson Report meant that children over the age of eight receive services from the Deaf Education Centres rather than from AODCs.</p> <p>Children can be just on ASSIST, they can be on ASSIST and receive DEC funding in their school, or they can be on ASSIST and receive support from an RTD.</p> <p>This category of service is relatively new, and so is still evolving.</p>	n=1037	n=364	n=1401
Total		n=1523	n=731	n=2254

Table 22 Children receiving support through the DEC's (2016 year)

Children and young people receiving services from vADEC/KDEC (n=853) are mostly not ORS verified (58%), followed by those who are verified high needs (29%) and very high needs (13%). (These are children who are ORS verified and receiving RTD support, or specialist support in categories 2 and 3 in the table above.)

The ethnic breakdown for children receiving Deaf Education Centre (DEC) services is shown in Figure

16. Please note that while the DEC's allow parents to nominate up to four ethnicities and this information is recorded in their database, for the purposes of the table below, the analysis shows preferred ethnicity chosen by the family/whānau. This means these figures are not directly comparable with DND data which allows children/young people to be listed in more than one category.

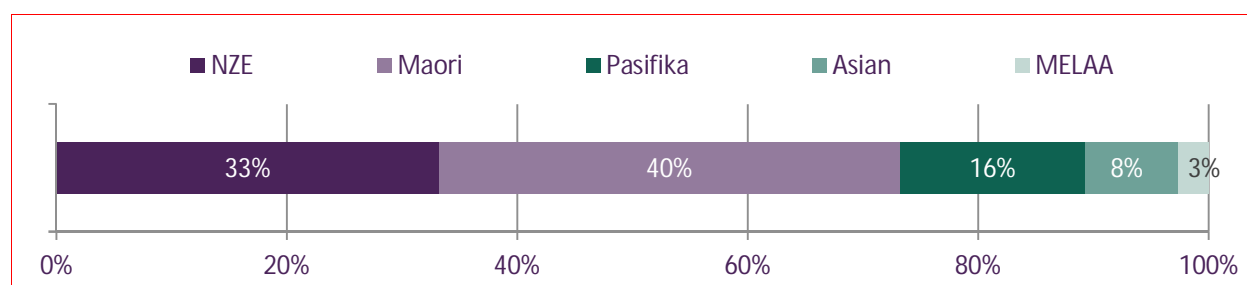


Figure 16: Ethnicities of all children receiving services via the Deaf Education Centres (2016)

Hearing aids

All cases notified to the database in 2016 contained information about whether hearing aids were to be fittedⁱ. Audiologists were asked "How many hearing aids are to be fitted?" These data represent the audiologist's stated plan at the time of notification. We have no data on what hearing aids are actually fitted. There are several reasons why the plan may not be followed in individual cases (e.g. parental preference, diagnosis of additional needs).

As has been the case with data since 2010, most children and young people whose cases were notified to the database and whose loss was first diagnosed in 2016 are to be fitted with two hearing aids (52%). This reflects the preponderance of bilateral losses notified to the database. Figure 17 shows this proportion is dropping, likely because the average age of diagnosis is falling. The proportion of cases in

which the professional notifying the case is unsure whether hearing aids will be provided is rising, for the same reason.

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

When data for all children notified from 2010 to 2016 is considered:

- bilateral losses - the majority of tamariki with bilateral hearing losses (84%) would be fitted with one or two hearing aids. Of these, 94% would receive two aids, and 6% one aid. The audiologist was unsure whether the child would receive aids in 11% of cases; and

ⁱ It is worth noting that some children with unilateral hearing losses were reported to be receiving more than one hearing aid. In these cases, we can confirm that is because, although the average threshold for the better ear does not meet the 26 dB HL average required for inclusion in the database, one

or more hearing thresholds are seen as sufficiently poor to warrant amplification in the better ear. This is indicative of one of the limitations related to classification systems that average hearing thresholds across four frequencies and categorise children into broad severity groups.

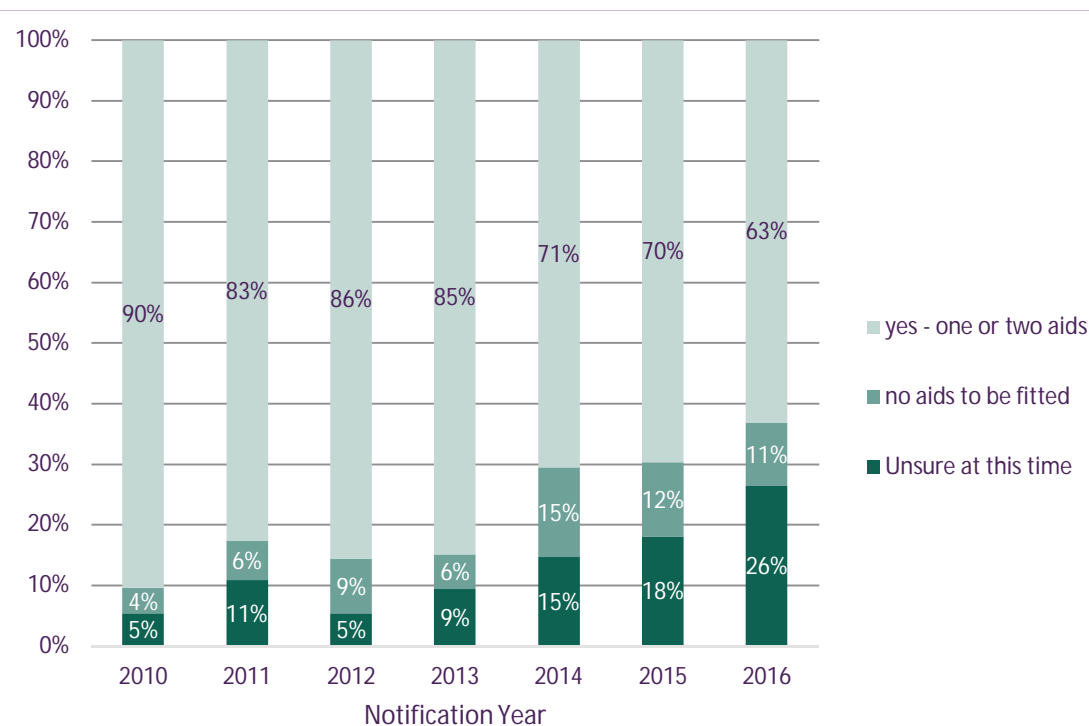


Figure 17: Hearing aids to be fitted by notifications (2010-2016)

- unilateral losses - 42% would be fitted with one hearing aid, and 24% would receive two aids (keep in mind that some have hearing loss in the other ear even if it didn't meet the 26dB HL pure tone average required to be considered a loss in the database) and 18% would receive no aids. The audiologist was unsure whether the child would receive hearing aid(s) in 17% of these cases.

Intention to fit, ethnicity and deprivation

Our data on the number of hearing aids to be prescribed are aligned with our previous assertions that Māori were more likely to have bilateral hearing losses than their New Zealand European counterparts.

Chi squared analyses which held severity constant showed:

- there are significantly more New Zealand European children with zero or one hearing aids being fitted than expected, and fewer with two hearing aids being fitted than expected;

- there are significantly fewer Māori with zero or one hearing aids being fitted than expected and more Māori with two hearing aids being fitted than expected.

Also, there are more New Zealand Europeans and fewer Māori with missing data than expected – this could be due to the fact that New Zealand Europeans are more likely to have unilateral losses, meaning the audiologist is less clear about the benefit of aids compared with the predominantly bilateral losses among Māori.

An analysis was also conducted to establish whether there is a relationship between the level of deprivation and whether hearing aids were to be prescribed. This analysis found no significant differences (ANOVA: $p=.8935$).

Funding for hearing aids

To provide some context for these figures, data provided by the Ministry of Health's providers for Hearing Aid Services during the period covered by

this report, are shown belowⁱ. Enable New Zealand began providing Hearing Aid Services on the 1st of July 2017, taking on this role from *accessible*. These data show MOH funded hearing aids for tamariki under the age of 19, and those in fulltime education and under the age of 21 during the 2016 calendar year^{ii,iii}.

A total of 2127 service users (tamariki) received hearing aid(s) during this period, down on the previous figures of 2494 in 2015, 2575 in 2014 and the 2659 in 2013^{iv}. Of those children receiving aids as below, 60% received aids in both ears, while the remaining 40% were received aids in only one ear.

These data make for an interesting comparison with the 2010-2016 DND data. Figure 17 shows that 52-70% of tamariki were to receive two aids between 2010 and 2016, and 12-19% one hearing aid. The difference between these figures and those from *accessible* and Enable NZ are likely to be at least in part owing to the fact that the DND data contains information about the number of aids to be provided *at the time of diagnosis*. These figures do not capture which of these cases then go on to receive more than the intended number of aids as they get older (as recorded in the notification form), either because their losses progress, or because the amplification strategy changes from the initial plan.

accessible: 01/01/2016-30/06/2016⁷⁶

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
NZ Māori	71	52	301	35	459
NZ European	92	62	341	62	557
Pacific	17	24	111	11	163
Other	56	25	140	25	246
Total	236	163	893	133	1425

Enable NZ: 01/07/2016-31/12/2016⁷⁷

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
NZ Māori	36	34	175	25	270
NZ European	55	26	147	36	264
Pacific	16	7	35	3	61
Other	12	11	66	18	107
Total	119	78	423	82	702

**Table 23: MOH Funding of Children's Hearing Aids
Calendar Year ending 31 December 2016, *accessible* and EnableNZ**

i Please note that "Hearing loss is defined as a permanent sensorineural or conductive hearing loss described by Clark 1981 Scale of Hearing Impairment, as used by the American Speech-Language-Hearing Association (ASHA) and the New Zealand Audiological Society Best Practice Guidelines July 2016." According to the Ministry of Health's Hearing Aid Services Manual, September 2017.

ii Domes and tubes, ear molds, remotes, FM (remote microphone hearing aid) systems, dry kits, and insurance excesses are excluded from these data.

iii Please note, these data pertain to all tamariki receiving hearing aids and not just to those receiving hearing aids for the first time.

iv Data provided in 2014 and reported in the report for 2013 was incorrect as these covered only a six-month period.

Cochlear implants

Although the DND notification form does not request specific information about [cochlear implants](#), it is useful to provide some information about the number of cochlear implants provided to children and young people in New Zealand, and some background on the funding for these implants.

Funding from the Ministry of Health is administered by two cochlear implant trusts. The Northern Cochlear Implant Trust covers the area northwards from an almost horizontal line extending roughly through Taupo, and the Southern Hearing Charitable Trust covers the area south of this line.

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

During the 2016 calendar year there were 55 publicly funded cochlear implants provided in the Northern Region and 50 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 teamsⁱ.

Children receiving cochlear implants during the 2016 calendar year	Southern Cochlear Implant Programme ⁷⁸		Northern Cochlear Implant Programme ⁷⁹	
	Ears	Children	Ears	Children
ACC cases	0	0	1	1
Public Funding - (1 Jan to 31 December)	39	22	46	29
Private procedures	1	1	6	6
Re-implants – recalled devices, failed integrity tests, or soft failures	8	8	1	1
Sequential or retrospective second cochlear implants (second ear for those under 6 already with one publicly funded ear - 1 January to 30 June)	3	3	1	1
	50	33	55	38

Table 24: Publicly funded Cochlear implants in New Zealand (2016)ⁱⁱ

While the DND may be missing some notifications for tamariki in the severe and profound categories, there are a number of other reasons DND figures are low compared with the number of tamariki implanted during the same period. One reason is that some tamariki who were notified to

the database with less severe hearing losses have a progressive loss which goes on to become more severe, something that is not tracked by the database.

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

ⁱⁱ In some years the number of cochlear implants provided exceeds the number of profound or severe cases notified to the database. While the DND may be missing some

notifications for children in the severe and profound categories, there are a number of other reasons why this figure is low compared with the number of children implanted during the same period. One is that some children who are notified to the database as having less severe hearing losses develop more significant losses over time, something which is not tracked by the database.

Appendices

Appendix A: History of the database

History of the DND

The original Deafness Notification Database was New Zealand's annual reporting system for new cases of hearing loss among tamariki and young people from 1982 to 2005. This system included data on the number and age of tamariki 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 tamariki notified to the database in a calendar year, rather than those who are identified in that year⁷⁴. During most of that time the database was managed by the National Audiology Centre on behalf of the Ministry of Health, and later by the Auckland District Health Board.

The database provided the only source of information from which the prevalence of permanent hearing loss among tamariki could be estimated, and from which the characteristics of new cases of hearing loss among tamariki 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 tamariki 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 could provide an important measure of changes in the age of identification and as the only way to identify potential false negatives within the screening programme.

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

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

Inclusion criteria

The original criteria for inclusion in the DND were based on the Northern and Downs definition below, and were applied to data until the end of 2005:

"Children under 18 years with congenital hearing losses or any hearing loss not remediable by medical or surgical means, and who require hearing aids and/or surgical intervention. They must have an average bilateral hearing loss (over four audiometric frequencies 500-4000Hz), greater than 26 dB HL in the better ear (Northern and Downs classification, 1984)⁸⁰."

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.

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 tamariki:

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

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

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

Notifying cases

Although the Database was restarted by the New Zealand Audiological Society, efforts have been, and continue to be made, to publicise the data-

base to non-members of the Society in an attempt to collect as many notifications as possible.

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

Future renaming of the database

During 2012, feedback on the name of the database was sought from parents of deaf tamariki, 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

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

ii While cases of unilateral hearing loss were technically excluded from the database until 2005, there were still large numbers of notifications sent to the administrators of the database, although these were not included in the main

analysis. Professionals consulted in the development of the re-launched database unanimously believed this group should be included in the database, at least in part as there is strong evidence that this group has increased risk for poorer educational and speech/language outcomes compared to children with normal hearing in both ears.

be gratefully received by Janet Digby, email:
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Appendix B: Notifications and ethnicity

The method used in this report to classify ethnicity is the total response method, where every person identifying with a specific ethnicity is included in that specific grouping⁸¹. 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 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 each ethnicity when compared with older survey methods, which required respondents to select only one ethnicity, the one with which they mostly identified, or where ethnicities are prioritised to include only one ethnic grouping per child. Using the total response method also aligns the database with The New Zealand Census, which began explicitly instructing

respondents that they could select more than one category for their ethnicity in 1996.

The proportion of notifications in each ethnic group was calculated differently in DND reports before 2006, with respondents being coded initially as belonging to one 'race' and later as one 'ethnic group'. Categories used have also changed. As a result, direct comparison with ethnicity data from before the re-launch in 2010 is not possible.

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

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

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

However, we can use a number of methods to provide some indication of the number of new

diagnoses of hearing loss annually among tamariki. 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 D: Severity codeframes

Differences between classification systems make it difficult for meaningful direct longitudinal and geographical comparisons of the proportion of tamariki in each 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 24 shows some of the differences between local and overseas severity classifications (these systems use an average of the pure-tone thresholds at 0.5 kHz, 1.0 kHz, 2.0 kHz and 4.0 kHz)ⁱⁱ. Audiologists in New Zealand are commonly using Clark's 1981 (ASHA) classifications in their clinical practice, as per the New Zealand Audiological Society practice guidelines.

Category	1996-2005 NZ DND	1982-1996 NZ DND	Clark 1981 (ASHA)	Jerger and Jerger (ASHA) ⁸²	World Health Organisation ⁸³	CDC ⁸⁴	Proposed code from Davis and Davis ¹
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 25: Comparison of audiometric severity classification systems

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

ⁱⁱ Australian Hearing uses the following codeframe (0-40dBHL, 41-60 dBHL, 61-90dBHL, 91dBHL+), but don't name the categories so these are not included in Table 25.

Glossary

Advisors on Deaf Children (AODCs): The Ministry of Education employs advisers on deaf tamariki 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 Centres⁸⁵. Implementation of changes proposed in the Wilson Report (2011) were completed in 2015, meaning AODCs are now working with an Early Years focus.

Aetiology: The cause or set of causes; in this report aetiology 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). The database collects information at 0.5, 1.0, 2.0 and 4.0 kHz wherever possible, and at higher frequencies for tamariki whose hearing loss meets the criteria for inclusion as a 'high frequency hearing loss'.

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

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

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

Bilateral hearing loss: Hearing loss affecting both ears.

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

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

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

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

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

False negatives: False negative is a term used to describe screened tamariki 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 tamariki 18 years or younger, born in New Zealand or overseas, with:

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

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

Learning Support: This is the new name for what was previously termed 'Special Education' services provided by the Ministry of Education. The name change was in [response to feedback](#) that terms like special education and special needs create barriers for tamariki.

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 loss, 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 tamariki at school. This funding provides Specialist Services staffing for students (who are ORS funded) including school counsellors. This scheme was previously reviewable (ORRS).

Special Education: Now referred to as Learning Support.

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.

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

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

An RTD is a trained specialist teacher who can:

- provide specialist 1:1 teaching;
- assist classroom teachers with curriculum adaptation and delivery;
- provide specialist advice, guidance and assistance for classroom environment and management;
- assist classroom teachers with the assessment of learning outcomes involving language and literacy achievement;
- liaise with all staff, support agencies and caregivers;
- monitor and support the use of audiological equipment;
- provide improved access to the curriculum for deaf and hard of hearing students.

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

Tamariki: Children and young people - normally used only in the plural. (Source: [Māori Dictionary](#).)

Unilateral hearing loss: Hearing loss affecting one ear. With regard to the DND, there may be minimal hearing

database. A limited analysis of data from high frequency hearing losses notified in 2013 can be found in, on page 47.

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

loss in the other ear but it qualifies as unilateral where the hearing loss in the other ear does not meet the 26 dB HL four frequency average criterion.

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

van Asch Deaf Education Centre (vADEC): van Asch Deaf Education Centre provides educational

programmes and services to Deaf and hard of hearing students, from roughly Taupo southwards.

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

Whānau: extended family, family group, a familiar term of address to a number of people - the primary economic unit of traditional Māori society. In the modern context the term is sometimes used to include friends who may not have any kinship ties to other members. (Source: [Māori Dictionary](#).)

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