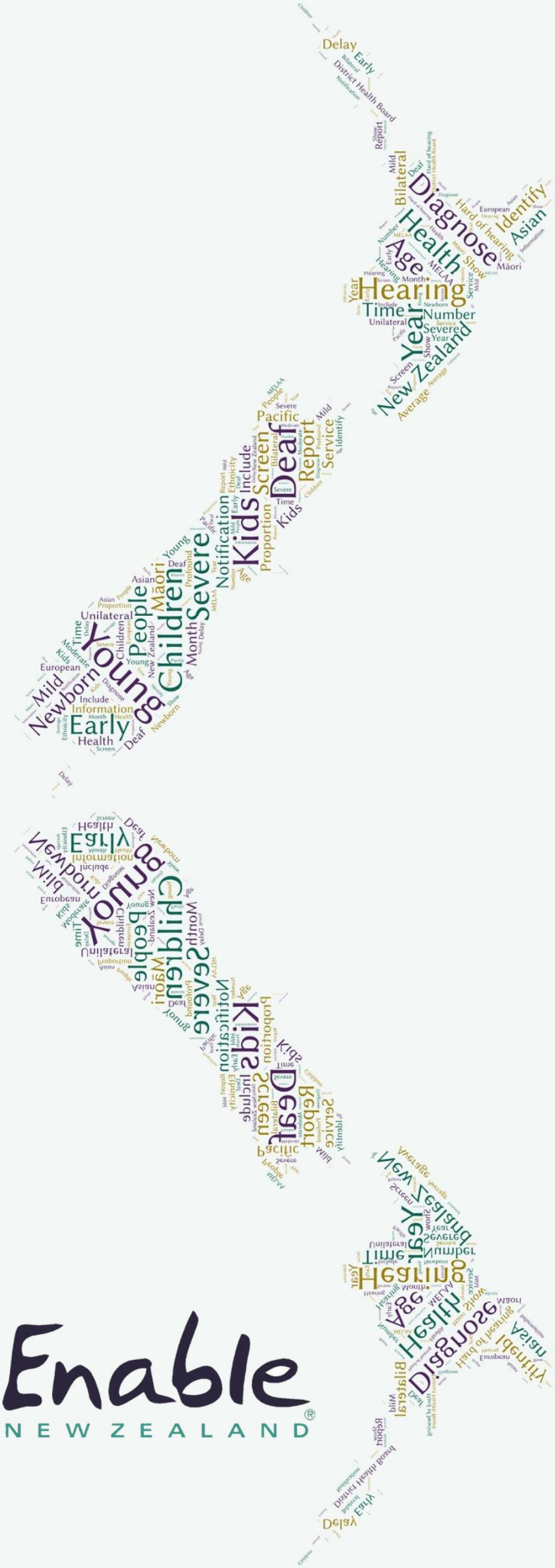


Enable NEW ZEALAND®

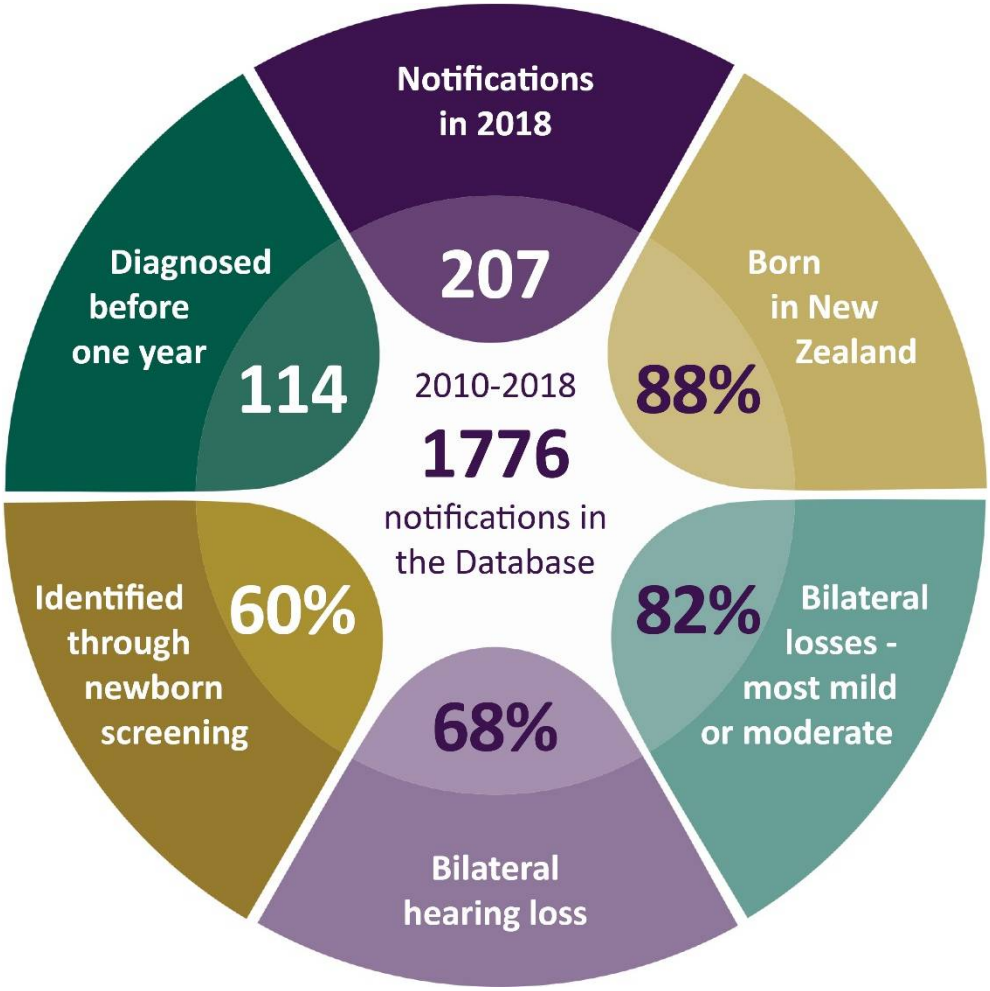
Deafness Notification Report 2018

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

Janet Digby, Levare Limited

December 2019

Summary



ISSN 2624-0025

Citation: Digby JE, Purdy SC, Kelly AS (2019) *Deafness Notification Report (2018) Hearing loss (not remediable by grommets) in New Zealanders under the age of 19*. Enable New Zealand. Auckland, New Zealand.

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

Contents

The Deafness Notification Database

- **The Deafness Notification Database (DND) was established in 1982 to collect information on newly diagnosed children and young people under the age of 19.**
- **The Database was relaunched in 2010 and now includes those born overseas and those with unilateral hearing losses.**
- **Our sincere thanks to the whānau(families)/kaitiaki (caregivers) and rangatahi (young people) who consented to share details of their child's/their own hearing for the Database.**
- **This information has helped us understand more about the nature of hearing losses being diagnosed in New Zealand and, in turn, is being used to inform those newly diagnosed and their families, assist researchers and help with resource allocation.**

Introduction

Nau mai, afio mai – welcome to the ninth in this series of annual reports describing notifications to the New Zealand Deafness Notification Database (DND). This report includes data for diagnoses made during the 2018 calendar year.

The 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.

Where the parents/mātua provide consent for this information to be shared, audiologists and some audiometrists from around the country send notifications electronically when they diagnose a child or young person with hearing loss.

Please refer to Appendix A: Making notifications to the Database on page 60 if you are an audiologist or audiometrist and wish to learn more about how to make notifications.

The analyses contained in this report generally pertain to 1776 children and young people notified with a hearing loss diagnosed between the start of 2010, when the DND was relaunched, and the end of 2018i.

The information contained in the Database has enabled us to shed light on the nature of hearing losses among children and young people, including helping us to understand that Māoriii are more likely to have less severe and bilateral hearing losses than their European counterparts.

"Ka mua, ka muri"

This Māori proverb translates to 'walk backwards into the future' and is about learning from the those who have gone before us.

Recent Database notifications have also shown that there is a growing number of children being

i There are additional notifications which have been included in the main dataset from years 2003-2009 and from early in 2018, which brings the total number in the dataset at the time of writing to 1914 children and young people who were initially diagnosed with a hearing loss between 2003 and 2018.

ii In this report the New Zealand Māori ethnic group is referred to as Māori, and New Zealand European ethnic group is referred to as European.

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 the result of nationwide newborn hearing screening.

Since 2010, the Database has included children and young people 18 years or younger, born in New Zealand or overseas, with:

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

Steps have been taken to allow data contained in this report to be compared with previous deafness notification data. However, in some cases questions have been amended to make these more specific and/or to reflect improved understanding in a specific area, such as family history. As a result, some longitudinal comparisons are not possible[ii].

For further information, please see the document's appendices and glossary:

- *History of the Database and changes to the inclusion criteria - see* Appendix B: History of the Database*, on page 61*
- *Terminology used in this report to describe hearing losses - see* Appendix E: Terminology used in this report *on page 64.*
- The completeness of notifications – see Appendix C: Completeness of notifications on page 63.
- Commonly used terms can be found in the Glossary which begins on page 65 of this report.

Acknowledgements

Our sincere thanks to the 207 whānau/kaitiaki and rangatahi who consented to share details of their child's/their own hearing for the Database.

As a result of their willingness to share basic information about these diagnoses, service providers can be better informed about current and future demand for services, factors most likely to result in delays in identification and other information that will help them better serve the needs of children, young people and their families/whānau and caregivers.

The time taken by audiologists and audiometrists to make notifications is appreciated.

This report has been funded by Enable New Zealand, through a contract with the Ministry of Health (MOH). The reports' authors would like to thank the MOH for funding the management, analysis and reporting of the Database from 2012.

The primary author 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. Tēnā korua.

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

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

- notifications have been reported for each calendar year throughout 1982-2005 and since the Database's relaunch, for 2010-2018;
- 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.

Mr Colin Brown is also acknowledged for his contribution to, and interest in, the DND over the years. This year, we extend our special thanks to Professor Russell Snell for his guidance on genetics and for introducing us to OMIM.

Contact details

The authors of the report hope that ongoing changes made to these reports will improve the value of the reports over time.

This year we have done a survey of readers to seek feedback on the future direction of the reports. A summary of the results is here.

This feedback has resulted in a number of changes to this year's report, including the addition of key points at the beginning of each section.

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 here.

Notifications

- **Notifications were made for 207 children and young people diagnosed during 2018, most of whom were born in New Zealand.**
- **Males are more likely than females to be diagnosed with a hearing loss and notified to the DND; they comprise 55% of notifications, similar to patterns found in similar jurisdictions overseas.**
- **The presence of one or more so-called additional disabilities (ADs) can have a significant impact on outcomes for children/young people with a hearing loss. One in five tamariki (children)/rangatahi (young people) notified to the Database had one or more confirmed 'additional disabilities' at the time their hearing loss was diagnosed, the most common types are syndromic, medical and neurodevelopmental in nature.**
- **A little over two thirds of notifications to the DND are for bilateral hearing loss, the remaining third for unilateral hearing loss.**
- **Research suggests that mild and unilateral hearing losses (UHL) are also associated with poor outcomes.**
- **Māori are more likely to have bilateral hearing losses than their European counterparts as well as more 'mixed' and less permanent conductive losses than their European counterparts.**
- **One in five tamariki/rangatahi whose information was notified to the DND have an immediate family member with a permanent hearing loss.**

General information

Two hundred and seven notifications pertaining to cases first diagnosed during the 2018 calendar year, and meeting the criteria for inclusion, were received by 15th March 2018, this year's cut off for new notifications[i, ii].

These notifications were received from a total of 61 audiologists, with notifications from 19 of the 20 district health boards (DHBs)[iii].

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

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

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 a 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 was the case in previous years.

iii 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 category.

iv 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 and extension of the deadline for notifications, these are submitted more evenly throughout the year, again with the number peaking between May and September, and then again before notifications close in March.

Because only a small number of cases met the criteria for the high frequency category in previous years[1], we have not described this group in this year's report, and we won't be seeking these notifications in future.

This report and the Database exclude children with Auditory Processing Disorders. For those interested, New Zealand Guidelines were published in 2019[2].

Number of notifications

Figure 1 shows the number of notifications which meet the criteria for the main dataset in each year[i,ii]. This shows the number of notifications that met all inclusion criteria at the time and were included in each of the Database's annual reports[iii].

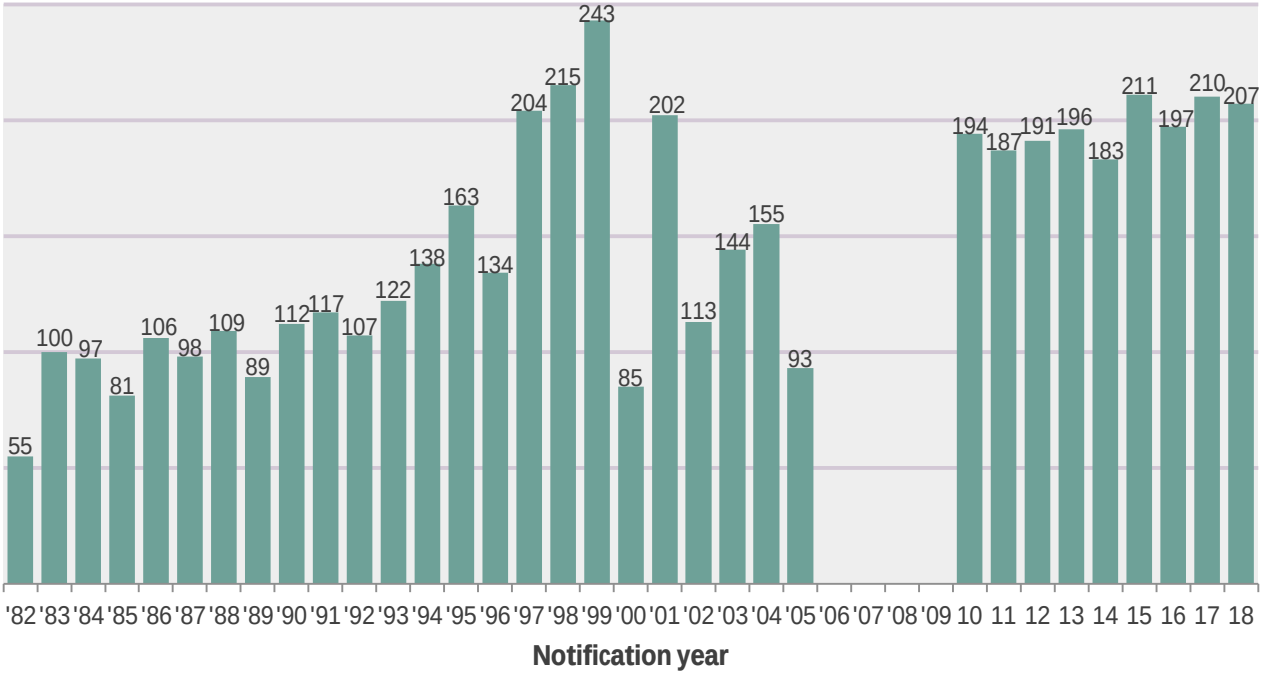


Figure 1: Notifications by year 1982-2005 and 2010-2018

i High frequency hearing losses, which were collected between July 2011 and July 2019, are not included in these figures.

ii The following types of notifications are not accepted into the dataset due to the inclusion criteria: 1) slight losses (those not meeting the 26 dB HL average across four frequencies - 0.5, 1.0, 2.0 and 4.0 kHz); 2) high frequency hearing losses which don't meet the 26 dB HL average noted above; 3) 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); 4) notifications with significant missing information (such as date of diagnosis, date of birth, location, audiometric data) where no further information was provided on request; and 5) notifications that didn't state that consent had been provided by the parent/caregiver, either through the UNHSEIP or through a consent specifically for the DND.

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. The figures now show the total number of notifications that met criteria for inclusion that were in place at that time.

This figure illustrates variability in the number of notifications provided to the original Database, particularly in the last six years of its operation[i, ii]..

Gender

Of the 1776 cases (2010-2018) contained in the main dataset, 45% of these are listed as female (n=803) and 55% male (n=973). This represents a ratio of 1: 1.21.

This gender difference was particularly noticeable in 2016 notifications with only 38% of notifications recorded as female, and 62% male[iii].

Overseas data

In overseas research, males are commonly found to have higher rates of hearing loss than females. These figures range between 51.5% and 58% for males (1:1.06 and 1:1.38) in various jurisdictions, as reported in the 2011 Comprehensive Handbook of Pediatric Audiology[3] and also in Feder et al.'s 2017 Canadian study on the prevalence of hearing loss among children and young people aged 3-19 years[4].

Australian Hearing's data on those under the age of 21 who have hearing aids or cochlear implants[iv]. show a similar pattern[5], with higher numbers of hearing loss among males (52.6%) than females (47.4%)[v].

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, for which fewer than half of cases were male (48.4%).

Birthplace

Tamariki born outside New Zealand have been formally included in the Database, and therefore its main analysis, since 2010.

Figure 2 shows the proportion of cases notified by birthplace for the 2010-2018 period. During that time, an average of 6% of children and young people notified have been born overseas, with the birthplace of an additional 6% being uncertain.

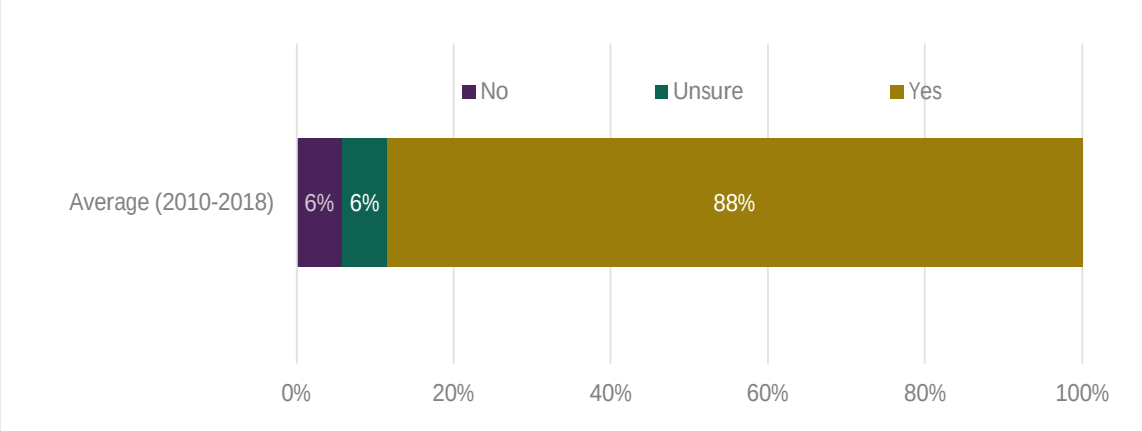


Figure 2: Proportion of cases born in New Zealand (2010-2018)

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.

ii Data from 2010 to 2018 have been revised slightly from previously reported figures, as further information about existing notifications is received, and as small numbers of retrospective notifications are provided to the Database. For example, in some cases an audiologist may not be able to notify a case in the year the diagnosis was made as they are unable to gain consent from the family/whānau by the deadline for notifications. 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.

iii From 2018, a third option has been available for selection in the notification form, in which the notifying professional can specify an additional gender option. This option has not yet been selected in any notifications.

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

v 0.1% of cases were of unknown gender.

The number of children for whom the audiologist was uncertain about the location of their birth has dropped from a high of 12% in 2010 to 2% in 2017 and 2018. This may be because audiologists are more likely to have information about the child's birthplace in cases where they are identified as a result of newborn hearing screening.

Of the 207 notifications to the Database in 2018, 4% were known to be born outside New Zealand, with birthplace listed as uncertain in a further 2% cases.

DHB representation

Table 1 contains the percentage of 2018 notifications from each DHB and compares these with the percentage of the population under the age of 20 from the 2013 Census[i]. The third column in that table shows the percentage of notifications received for 2010-2018 from each district health board – this can be compared with the percentage of the population under the age of 20[ii].

Tamariki and rangitahi in the Database are more likely to be of Māori ethnicity than their proportion in the general population would predict.

DHBs with more than 20% of their population identified as Māori are shown with shading in Table 1[6].

It is worth noting that, historically, many clinicians believe there is a preponderance of deafness in Auckland and Christchurch as 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 Waitematā[iii] – 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 that applied from the middle of the reporting year and were outlined in the 2015 report;
- Wairarapa, Whanganui, Lakes and West Coast – there are relatively small numbers of diagnoses of hearing loss each year.

In addition to these factors, and natural fluctuations in the number of hearing losses diagnosed among tamariki and rangitahi in each year, other factors influencing notification levels, are likely to include:

- the size of each DHB population within the age range for the Database;
- that distribution of young people by DHB has likely changed since the 2013 Census;
- the prevalence of hearing losses within DHB populations;
- 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 working within each DHB catchment area;
- the workload of these hearing professionals; and
- the level of commitment and capacity among staff to making notifications to the Database.

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

ii Please note, these percentages are rounded.

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

District Health Board	Percentage of notifications received in 2018 (under 19 years)	Percentage of population under the age of 20 (Statistics New Zealand, 2013 Census)	Percentage of notifications received 2010-2018 (under 19 years)
Auckland	12%	11%	5%
Bay of Plenty	6%	4%	12%
Canterbury	14%	11%	8%
Capital and Coast	7%	7%	11%
Counties Manukau	8%	13%	13%
Hawke's Bay	3%	3%	2%
Hutt	2%	3%	4%
Lakes	3%	2%	1%
Midcentral	2%	4%	2%
Nelson Marlborough	3%	3%	3%
Northland	7%	3%	5%
South Canterbury	2%	1%	2%
Southern	5%	7%	9%
Tairāwhiti	3%	1%	1%
Taranaki	3%	2%	2%
Waikato	9%	9%	10%
Wairarapa	2%	1%	0%
Waitematā	11%	13%	6%
West Coast	0%	1%	2%
Whanganui	0%	1%	0%

Table 1: Percentage of notifications (2018) compared with the estimated percentage of population under 20 years of age by district health board (2013 Census) and the proportion of notifications by DHB (2010-2018)

Children/young people – seven years later

A research project funded in 2017 by the Oticon Foundation of New Zealand aimed to improve understanding of what happens to children and young people following diagnosis, by requesting follow-up information from notifying clinics seven years following the initial diagnosis and notification of a hearing loss to the DND.

Of those clinics approached, all but two participated in the research, resulting in at least some data being received on 163 of the 194 children and young people notified in 2010 (84%)[i].

This research showed that only 56% of children/young people were still in the care of the notifying clinic at the time the follow-up data was provided. For those who were still in the care of the notifying clinic, 31% had not been seen by that clinic since 2016 or earlier.

[i] Data for 78% of notifications where the diagnosed child or young person was listed as Māori were received, compared with 81% of non-Māori.

For the 44% who were no longer in the care of the notifying clinic:

- Clinics had no information on who was currently providing service to 36% of the children or young people.
- 1% hadn't been seen since the clinic's original diagnosis.
- 7% had some information on where they had moved to or who was now looking after them.

We understand from speaking with audiologists on the Paediatric Technical Advisory Group (PTAG) that it is possible that DHBs who provided the original notifications may have been asked for information on the child or young people by their new provider (with communications moving between their medical records departments for example), without the original audiologist's knowledge and therefore without them knowing where the child or young person was now receiving care.

Some district health board audiology services have the ability to look for people outside their catchment (e.g. there is a database for those in the South Island which is searchable) while others do not.

This demonstrates the importance of clinic information systems and communications between clinics to ensure children are not lost to follow-up.

Additional disabilities

The presence of one or more so-called 'additional disabilities' can have a significant impact on outcomes for tamariki and rangatahi, and also on the level of support they may require, particularly from *Learning Support*, Ministry of Education (previously Special Education).

Children with such additional disabilities are sometimes referred to as being 'deaf plus'. We are yet to come across a term that is inclusive given the broad range of conditions and issues that are included in this section and which is ability focused. Suggestions for a better term are most welcome.

Full dataset

When considering the children and young people included in 2010-2018 notifications, the majority (79%) have no additional disability. Eleven percent have a confirmed additional disability and a further 9% are listed with a possible although as yet unconfirmed additional disability. Just over 1% of cases (n=21) contained no data on whether an additional disability was known to be present.

Additional disability	Number of cases	Percentage
Yes	199	11%
Unsure whether AD exists, no confirmed diagnosis	163	9%
No additional disability	1393	79%
No data	21	1%
Total	**1776**	**100%**

Table 2: Proportion of cases by additional disability status (2010-2018)

2018 data

Of 2018 notifications, 14% of children and young people were known to have disabilities *in addition* to their hearing loss at the time the notification was made. In a further 10% of cases there was uncertainty regarding whether the child or young person had an additional disability[i].

The proportion of children and young people diagnosed in 2018 with either a confirmed or possible additional disability (at the time of diagnosis) has increased to levels not seen since 2012.

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

Comparison with previous data

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 been confirmed.

Column four of Table 3 shows the total proportions of both confirmed and unconfirmed of cases with an additional disability. This figure is more consistent with those reported before the Database's re-launch in 2010.

Recently there has been criticism of immunisation rates, which have fallen and are now down 4.5% from their peak in 2016. These rates are particularly low for Māori children and those who live in poverty[7].

Previously, the authors of this report believed that the earlier identification of tamariki with hearing loss was the likely reason behind drop in the proportion of those with confirmed additional disabilities reported at the time of diagnosis of the hearing loss. More recent DND data (shown in Table 3) suggest the general downward trend from 2012-2016 may have reversed, but there are a number of possible contributing factors to changing data and it is not possible at this time to determine what might be having an effect.

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 (2010-2018)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	13%	10%	23%
2011	15%	5%	20%
2012	15%	11%	26%
2013	13%	11%	24%
2014	15%	8%	23%
2015	10%	10%	20%
2016	9%	10%	18%
2017	12%	8%	21%
2018	14%	10%	24%
Average 2010-2018	**11%**	**9%**	

Table 3: Proportion of cases with a known additional disability (2002-2018)

The rationale at the time was that tamariki may have not yet been diagnosed with these conditions, or they have conditions that have not yet developed at the time the notification to the Database was made.

For example, diagnoses of autism spectrum disorder are typically not made in the first year of life. Other possible reasons for what was previously a general downward trend in the proportion of tamariki reported with additional disabilities

included higher recent immunisation coverage[i, 8] and that tamariki with hearing loss in New Zealand are not all routinely assessed by a paediatrician.

Most common types of additional disabilities

A wide variety of reported conditions are contained in notifications, including those related to a specific syndrome, cerebral palsy, general or global developmental delays, intellectual disability and vision problems [9, ii]. Some children and young people have more than one additional disability listed on their notification form.

In an attempt to better describe the range of additional disabilities seen among children and young people whose data is contained in the Database, we have implemented a new codeframe to group these responses by type and applied this to all records, as seen in Table 4. This shows the type of disability, how we determined what is included in each category, the number of cases and the proportion of *all children/young people listed as having an additional disability* by category of disability.

Type of additional disability	Inclusions	Number of cases	Proportion of cases with AD
Syndromic	A diagnosed syndrome or syndromes. At this time the notification form doesn't seek information on the severity or specific implications of the syndrome(s) mentioned	55	28%
Medical	Medical conditions and issues, such as cardiac problems, bladder issues, renal issues and lung issues.	54	27%
Neurodevelopmental	Issues with the growth and/or development of the brain or central nervous system, such as ADHD, autism, developmental delays and intellectual disabilities	52	26%
Sensory	Issues relating to the sensory system, which don't relate to the child or young person's hearing. By far the most common of these among this cohort is vision problems (ranging from cataracts and blindness to amblyopia and refractive errors and structural changes within the eye), but there are also children and young people with other conditions such as sensory integration difficulties in this category	39	20%
Neurological	Issues relating to the brain, spine and the nerves that connect them, such as cerebral palsy, epilepsy, microcephaly, missing brain structures and issues with myelination	29	15%
Medical-developmental	Medical conditions and issues related to development such as hydrocephalus and cleft palate	15	8%

Table 4: Number of cases by type of additional disability (2010-2018)

Of the 1776 records in the Database for 2010-2018, the majority (78%) had no additional disability confirmed at the time the child or young person's hearing loss was diagnosed. The majority of those who were listed as having an additional disability had one or more disabilities in one category, while smaller numbers had one or more additional disabilities listed in two, three or even four categories.

Table 5 shows the number children/young people who are listed as having each additional disability code. For example, those listed with two additional

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 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.

Number of additional disability codes / category	Number of children/young people	Percentage
No additional disability	1393	78.4%
One additional disability code	162	9.1%
Two additional disability codes	30	1.7%
Two additional disability codes	6	0.3%
Three additional disability codes	1	0.1%
Unconfirmed AD	163	9.2%
No data	21	1.2%
Total	**1776**	**100%**

Table 5: Number of cases by number of additional disability code types (2010-2018)

disability codes includes some with a disability that is medical and one that is neurodevelopmental in nature. Others listed with one disability code may have two additional disabilities listed, both in the same category.

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 6. The first paper listed shows the huge variability in rates, presumably at least in part 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.

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, syndromes not entailing developmental delay, or medical disorders[10].

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.

More recently, Cupples *et al.* (2018) analysed language ability in 67 children who were enrolled in the LOCHI study at three and five years of age, using a number of standardised assessments. While across the entire cohort these children had stable outcomes, the authors note that children with autism, cerebral palsy and/or developmental delay showed a decline in standard scores during this time. They conclude that the type of additional disability can indicate expected language development where formal assessment of cognitive ability isn't possible[11].

Source	Date	Location	Details	Rates
LOCHI[12]	2013	Australia	Study examining 260 children in Australia born with hearing impairment	18% of children in their sample have one additional disability, 10% with two and 9% with three or more
Ear Foundation for National Deaf Children's Society[13]	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: • visual impairment (4-57% depending on the definition) • neurodevelopmental disorders (2-14%) • speech language disorders (61-88%)
The Consortium for Research into Deaf Education[14]	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
Fortnum *et al.*[15]	2002	UK	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis[16]	1997	UK	Trent region study of permanent congenital hearing impairment	38.7% of children found to have one or more additional clinical or developmental problems, although this study used a wide definition of additional needs.
Holden-Pitt and Diaz[17]	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

Table 6: Additional disabilities, selected overseas rates for comparison

Bilateral and unilateral loss

Background

Unilateral hearing prevents the auditory system from processing and integrating input from both ears, which is important for improved understanding of speech in noisy situations and for sound localisation[18, 19].

A series of studies in the United States in the early 1980s caused the significance of unilateral hearing losses (UHL) to be re-evaluated by professionals, who had often minimised the implications of unilateral hearing loss in children[20, 21, 22].

There is evidence that children with unilateral hearing losses have reduced educational performance, language delays and higher rates of behavioural issues, which are reported as significant in about a third of all cases[23, 24, 25, 26, 27].

Some research suggests that children with mild hearing loss may have worse outcomes than those with hearing losses of greater severity, likely due to the fact that children with these hearing losses often have them identified later and receive fewer support services[28].

To reflect the acknowledged importance of unilateral loss, cases where these average more than 26 dB HL in the child/young person's hearing-impaired ear[i] have been included in the DND since its re-launch in 2010[ii,iii].

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 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.

Bagatto *et al*. (2019)[29] completed a review paper that draws on the views of an international panel of experts along with a parent advocate and a review of the literature. This review defines unilateral hearing loss as any degree of permanent hearing loss in one ear (using pure tone averages over 0.5, 1.0 and 2.0kHz) that is greater than 15dB, regardless of aetiology, with normal hearing in the opposite ear. This paper notes that the majority of cases of UHL are due to cochlear malformations and Mondini dysplasia, and that environmental causes are also commonly implicated. As a result, aetiologic assessment following diagnosis, including complete ontologic evaluation including imaging, is recommended.

Prevalence

Prevalence of unilateral hearing loss (UHL) is difficult to understand, not least because the definition for UHL differs between studies, and samples often don't include the complete group being described[30].

Newborn hearing screening programme data from overseas suggest around one in 1000 babies are born with a UHL, about a third of the total babies identified with a hearing loss[31].

As described by Vila and Lieu in 2014, one in ten or more of the children diagnosed with UHL will see this hearing loss progress to affect their other ear[32, 33, 34].

Prevalence rates rise with age to between 3.0 and 6.3% among children 6-19 years of age, according to Ross *et al.*[35]

Recommendations

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.'[36] 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 has a supplement (2013) that states that all children with unilateral or bilateral hearing loss should be referred to early intervention services for evaluation and consideration of enrolment. It stated that most infants and children with bilateral hearing loss and many with unilateral hearing loss benefit from some form of personal amplification device[37].

The American Academy of Audiology recommended in 2013 that children with unilateral hearing loss should be provided with hearing aids on a case by case basis[38].

In New Zealand, Project HIEDI recommended in 2010 that families of children with unilateral hearing loss be offered advisory services (from an Advisor on Deaf Children) and that such children be regularly assessed to quickly determine if they are beginning to fall behind and to determine what support is appropriate[39].

Management and research

While there is limited high-quality evidence on how to best manage unilateral hearing loss in young children, consensus-based principles of technology management for children with UHL are described in Bagatto *et al.*'s recent review[29].

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.

Proportions of unilateral and bilateral hearing losses in the Database

Bilateral and unilateral hearing losses[i]:

- two thirds of cases for 2010-2018 had full audiometric data enabling them to be coded as bilateral (67%) and unilateral (33%); and
- when cases with missing frequencies were included, and interpolation used, the proportion of 2010-2018 cases that were bilateral/unilateral was 68:32 (see Figure 3, below).

Other influences

While immunisation coverage (including for conditions such as mumps) in New Zealand rose significantly from 45% in 2007 to 92% in 2012[40] there was 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.

More recently, concerns about falling immunisation rates have been raised, with particular concern expressed about those for Māori and those living in poverty[7].

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 loss[41].

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

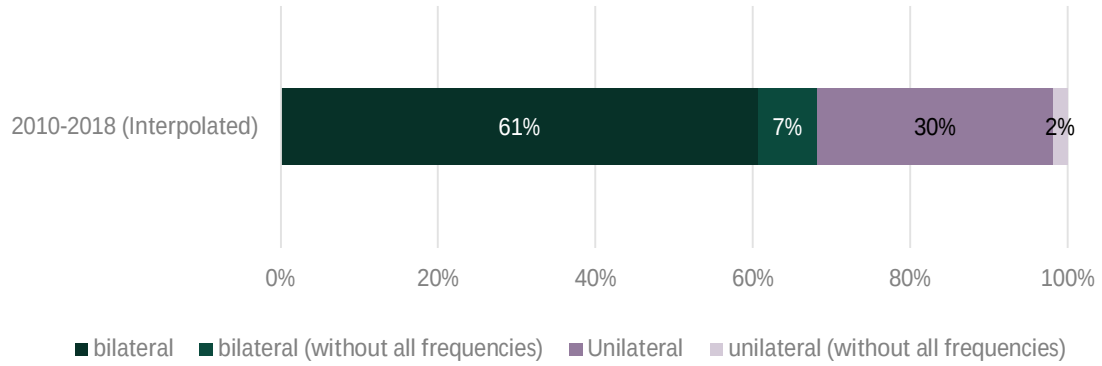


Figure 3: Proportion of bilateral and unilateral cases (2010-2018)

Progression to bilateral from unilateral

The NZ study mentioned in *Children/young people – seven years later* on page 12, followed up 163 of the 189 children and young people notified to the DND in 2010 seven/eight years later.

Of those with recent data, 32% of those children or young people with a unilateral hearing loss had progressed to a bilateral hearing loss.

Accurate diagnosis of young children is inherently difficult. One explanation for this is that the testing became easier over time as testing young children becomes easier as they develop.

There can also be a difference between ABR and behavioural results that is greater at low frequencies. Another possibility suggested by PTAG is that the quality of paediatric audiology may have improved with the introduction by NZAS of compulsory paediatric certification.

i From 2015, these reports describe the proportion of bilateral and unilateral hearing losses based on cases with and without all data-points and also on interpolated figures using manual checks for those records that cannot have data interpolated. This change means we can now report on the number of ears affected by hearing loss in more cases.

Single sided deafness

Background

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

While there are few studies on children and young people with a diagnosis of this type, a recent review focused on adult research (2016) concluded that no recommendations for the management of unilaterally deaf adults could be made based on the current evidence[42].

Different case definitions of (SSD) are used internationally; for example, some definitions include only those with severe or greater hearing loss in the worse ear and others only those with profound loss[43, 44]. 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, those with SSD may be more likely to receive cochlear implants compared with those with less severe degrees of hearing loss, who may receive a bone conduction hearing aid.

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

This category is effectively a subgroup of the unilateral hearing category discussed elsewhere in this report.

Notification Year	Proportion of cases with single sided deafness
2010	6%
2011	4%
2012	8%
2013	10%
2014	8%
2015	6%
2016	5%
2017	6%
2018	3%
Average 2010-2018	**6%**

Table 7 Single Sided Deafness Cases by Year (2010-2018)

DND data

The proportion of 2010-2018 unilateral hearing loss cases[ii] which met the DND's criteria for SSD is 22%.

The data contained in Table 7 show the proportion of total notifications each year that met the DND's definition for SSD[iii].

i These average thresholds have been chosen considering the ASHA (American Speech-Language-Hearing Association) codeframe for severity, and because 26 dB HL is the lower limit for average notifications to be accepted into the database and as a 70 dB HL 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.

iii 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.

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 ear[i]. Options provided are: 'sensorineural', 'mixed', 'permanent conductive', 'normal hearing[ii]', 'other' and 'don't know'.

'ANSD' (Auditory Neuropathy Spectrum Disorder) is offered as an option within sensorineural hearing loss and is not split out in the graph below.

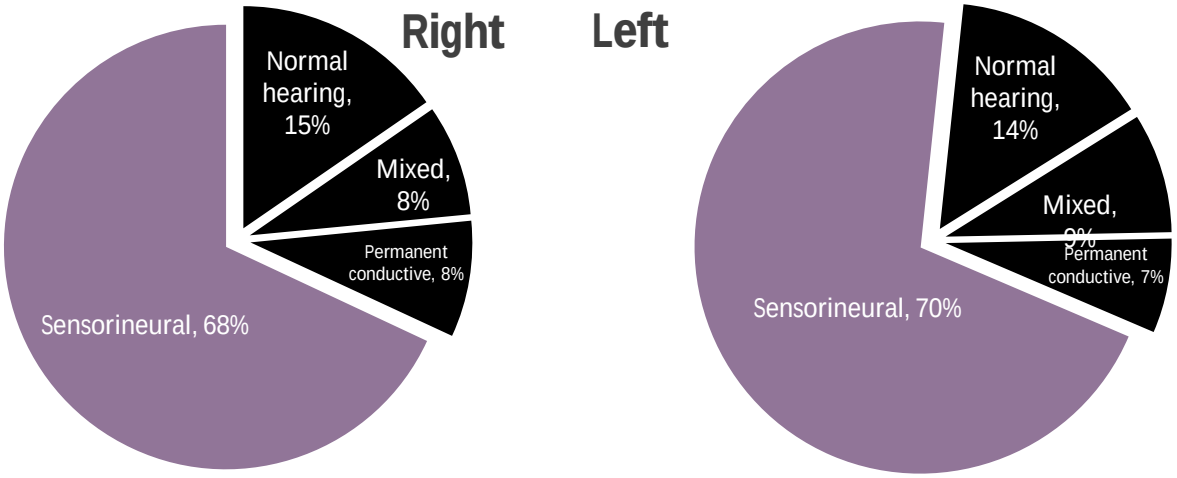


Figure 4: Type of hearing loss (2010-2018)

The most commonly reported type of hearing loss contained in notifications was sensorineural (70% in the left ear and 68% in the right), followed by normal hearing (14% in the left ear and 15% in the left). See Figure 4 for full detail.

Three percent of right ears and four percent of left ears were recorded in the ANSD category.

Prevalence of ANSD among those children with permanent hearing loss is likely to be approximately 10%, according to a 2015 review by Rance[45]. Among those from the Avon newborn hearing screening programme in England[46] 15.7% were identified to have abnormal air and bone conduction thresholds and were found to have ANSD.

These figures seem to suggest that New Zealand may have lower rates of ANSD than other similar jurisdictions. This could be suggestive of differences in our New Zealand population, also suggested by our lower proportion of severe and profound hearing losses.

An analysis of the types of hearing loss among 2010-2016 notifications included in the previous report[47] found significant differences in the type of hearing loss between Māori and Europeans (Fishers exact test: p=.0037). More Māori had 'mixed' hearing losses than expected (11.9% for Māori vs 6.1% for Europeans, p=.0317, Z-test for proportions), and fewer Māori were recorded as having 'permanent conductive' hearing losses than expected (6.5% for Māori versus 12.1% for European, p=.0313)[iii].

Given that Māori in our sample have more bilateral losses than their 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

Background

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

The 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?[i] (or had a permanent hearing loss if they have died).' The results for this question are shown in Figure 5.

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 2015-2018 in Figure 5.

Full dataset

Figure 5 shows data from 2015-2018 notifications. The proportion of notifications pertaining to children and young people who are listed as having no immediate family member(s) with a permanent hearing loss ranged from 73 to 78% during that time, with between 17 and 22% listed as having one.

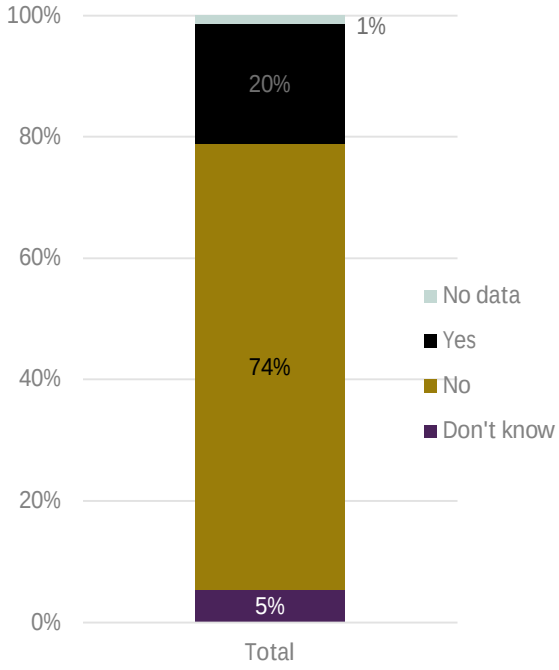


Figure 5: Immediate family member with hearing loss (2015-2018)

i The DND reports prior to 2005 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

- **Almost all records in the Database contain ethnicity information about tamariki and rangatahi diagnosed.**
- **Disparities across the health system have been well-documented for Māori in terms of access to and through the health system.**
- **The largest number of notifications are listed as European although there are fewer than would be expected based on the size of their population under 20 years of age, and the number of notifications from those of Māori ethnicity are higher than expected based on their population.**
- **Other sources further confirm higher rates of permanent hearing loss among Māori compared with their European counterparts.**
- **Children and young people of European ethnicity are more likely to be without hearing loss at birth when compared to non-European.**

Representation

Background

The DND notification form records information about the ethnicity/ethnicities of tamariki/ rangatahi diagnosed with hearing loss. Options available on the form are: European, Māori, Pacific Peoples, Asian and MELAA[i].

Please keep in mind that the multi-code system used for the DND means that some records contain more than one code for ethnicity, and so they appear in more than one group. The authors of this report believe this system of coding is a more complete reflection of ethnicity than those that either force participants to provide one code or use a prioritisation framework to re-code down to one code per participant.

Full dataset

Of the 1776 notifications in the main dataset (covering 2010-2018 notifications) all but 23 (1%) contain at least one ethnicity code. The number of notifications containing no ethnicity codes has reduced from a high of 5% in 2010 to less than 0.5% in 2017 and 2018. The majority of notifications (89%) contain one code, and a smaller proportion (9% and 1%) contain two or three codes respectively.

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 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 2010-2018, which includes those under the age of 19[ii].

The majority of notifications provided to the Database since its re-launch in 2010 relate to tamariki of European and/or Māori[iii] ethnicity.

i The MELAA category relates to tamariki of Middle Eastern, Latin American or African ethnicity. An 'other' category is also listed for situations where the notifying audiologist is unsure which category a specific ethnicity falls into. These are recoded before analysis is completed.

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

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

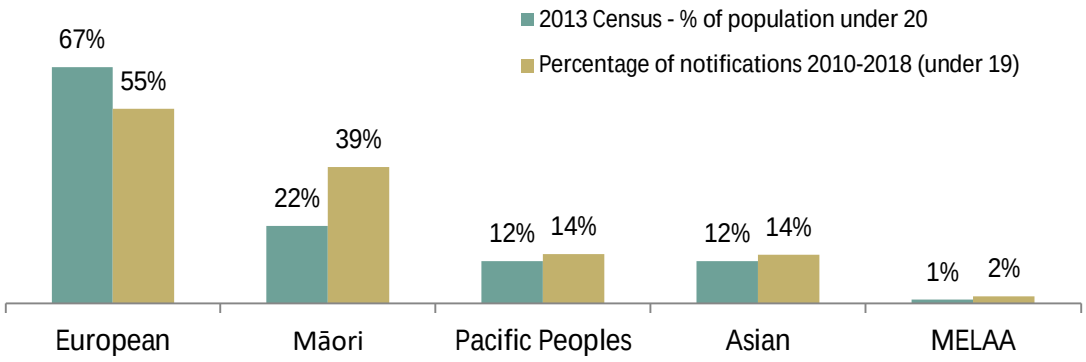


Figure 6: Notifications by ethnicity (2010-2018) compared with Census data (2013)

Ethnicity, prevalence and other characteristics

Prevalence data

Compared to the general population, the proportion of notifications from those of 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.

A number of sources reinforce the higher prevalence of hearing loss between Māori and Europeans which is also shown in DND data described in Figure 6:

- 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 loss[48].
- The Household Disability Surveys:
  - » 1991-2006 Surveys[49] suggest Māori had higher rates of hearing disability (tamariki and adults) and higher rates of unmet need for technology and equipment when compared with non-Māori[50]. (For information about the limitations of this data please see the 2011 DND Report[51].)
  - » The 2013 Survey continues to suggest Māori had higher unmet need for technology and equipment when compared with non-Māori[52] but also that they now have *lower* rates of hearing disability compared with their European counterparts across all age groups[53], although this seems to relate to the lower age profile for Māori (younger people have fewer disabilities).
- Findings from Digby *et al*. (2014) indicated young Māori have higher rates of permanent hearing loss than their European peers, based on the previous DND dataset, which included notifications from 1982-2005[54].
- B4 School Check data:
  - » Referral rates from the B4 School Checki analysed by Searchfield et al. (2011), show higher rates of referral from hearing screening for Māori tamariki (9%) compared with non-Māori (5%)[55]. 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 who completed their hearing screening was 7.9% in 2016/17, considerably higher than for European, at 3.5%[56]. Post screening diagnostic results are not available. Rates were similarly high for Māori when compared with European since 2010/11.

Despite all these sources pointing to higher rates of hearing loss among young Māori, this group may still be underrepresented in DND statistics because of their greater chance of having a less severe hearing loss, and it is probable that less

i For more information on the B4 School Check, please click here or view the glossary on page 75.

severe (especially mild) hearing losses are less likely to be identified.

This is particularly likely for older Māori children and young people who were not screened as newborns and for those children and young people who develop a hearing loss after birth. Programmes, including New Zealand's UNHSEIP[i] often do not identify mild hearing losses[57]. The B4 School Check targets mild and greater hearing losses – this screening is focused on children before they reach school age[58].

It also seems likely that disparities in access to, and within, the health system for Māori[59] may mean fewer cases are found or notified when compared with those in the European population. (See the section on Service access, below.)

Those listed with Asian ethnicity are also over-represented in the data, with 18% of notifications coming from this group, while only 12% of the population were categorised as 'Asian'. This may be related to recent growth in the population since the 2013 Census; the 2018 Census data will be used as our comparison as soon as this is available.

For further information on ethnicity coding in 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[59, 60].

An article by McCallum *et al.* (2015) in the *New Zealand Medical Journal*[61] 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 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.

Screening coverage rates for programmes, such as the UNHSEIP, show those listed as Māori are less likely to have their screening completed than their European counterparts[48].

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

Unilateral and bilateral hearing losses

Of 2010-2018 cases, including those with interpolated audiometric data, 68% are recorded as bilateral, while the remaining 32% are unilateral.

Figure 7 shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group during 2010-2018. 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 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 European counterparts[54].

This difference can also be seen when comparing bilateral losses among Māori tamariki[ii] notified

i "The UNHSEIP is not designed to identify babies with mild hearing losses." Ministry of Health's 2016 Universal Newborn Hearing Screening and Early Intervention Programme: National policy and quality standards: Diagnostic and amplification protocols.

ii 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.

between 2010 and 2018 (78%), with those who are European[i] (63%), and those described as both Māori and European (67%).

Please note that MELAA figures relate only to a very small number of cases (n=30).

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

Hearing loss present at birth

Of all 2010-2018 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 for 'likely present at birth' was provided, the audiologist indicated they were 'unsure' in 43.7% of cases, with the hearing loss likely to have been present at birth in 42.9% and unlikely to have been present at birth in 13.5% of cases.

Analysis of 2010-2016 cases in the 2016 report found that the proportion of Europeans *without* 'hearing loss thought to be present at birth' was significantly higher than for Māori (Z Test: 95% CI (0.054, .132), p<.0001). Because of the number of 'unsure' answers for this question, one cannot assert that Māori have more hearing losses present at birth. Further research is needed to determine whether progressive hearing loss is more common among non-Māori.

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

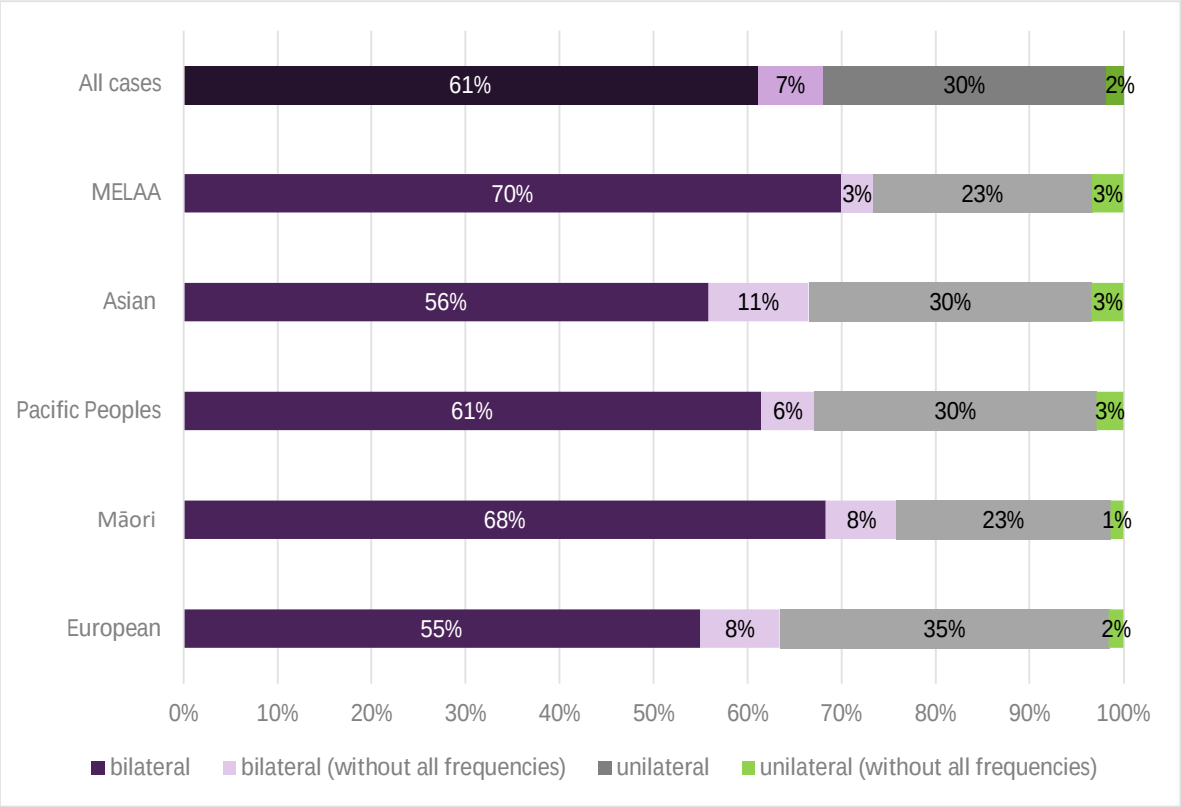


Figure 7: Proportion of unilateral and bilateral hearing losses by ethnicity (2010-2018) based on interpolated data and manual checks to determine bilateral/unilateral status

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

Deprivation

- **Deprivation scores draw on Census data, which indicate the level of deprivation for each small area in New Zealand.**
- **In general, New Zealand deprivation data show that children in the general population under the age of 17 are more than twice as likely to be living in income poverty than those over the age of 65 years. Those with one or more disabilities are also more likely to live in areas with higher deprivation than those without.**
- **DND data indicates that those notified to the Database are more likely to live in areas of high deprivation, more-so than the national distribution for children and young people of the same age.**
- **Children and young people notified to the Database who are of European ethnicity are much more commonly living in the least deprived areas than those of Māori and/or Pacific ethnicity.**

Introduction

Deprivation data provided by the Ministry of Health has been included in our analyses since the 2016 report.

Deprivation data used 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 8.

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 deprived[62].

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 1776 tamariki in the main dataset, all but 29 (≈ 98%) had deprivation data available. Data were unavailable for tamariki whose: NHI was not valid (n=24), who 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).

Tamariki and deprivation

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 years[63].

In general, tamariki in New Zealand with one or more disabilities are more likely to live in areas with higher levels of deprivation than those

i As at the date of extraction, in July 2018.

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 8: Deprivation variables used for NZDep2013

without. Child Poverty Action Group (2015) stated[64] 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 disability[65].

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 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 kindly shared 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 Database[66].

The 2016 report shows these comparisons, for children 0-5 years of age, and those 6-17 years of age[i]. Both DND distributions skew more towards the higher deprivation scores than the national distribution for tamariki of the same age[ii]. This was particularly the case for tamariki notified to the Database during 2010-2016 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 8 shows the distribution of cases by deprivation status with splits by ethnicity[iii]. 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 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.

ii 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.

iii 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.

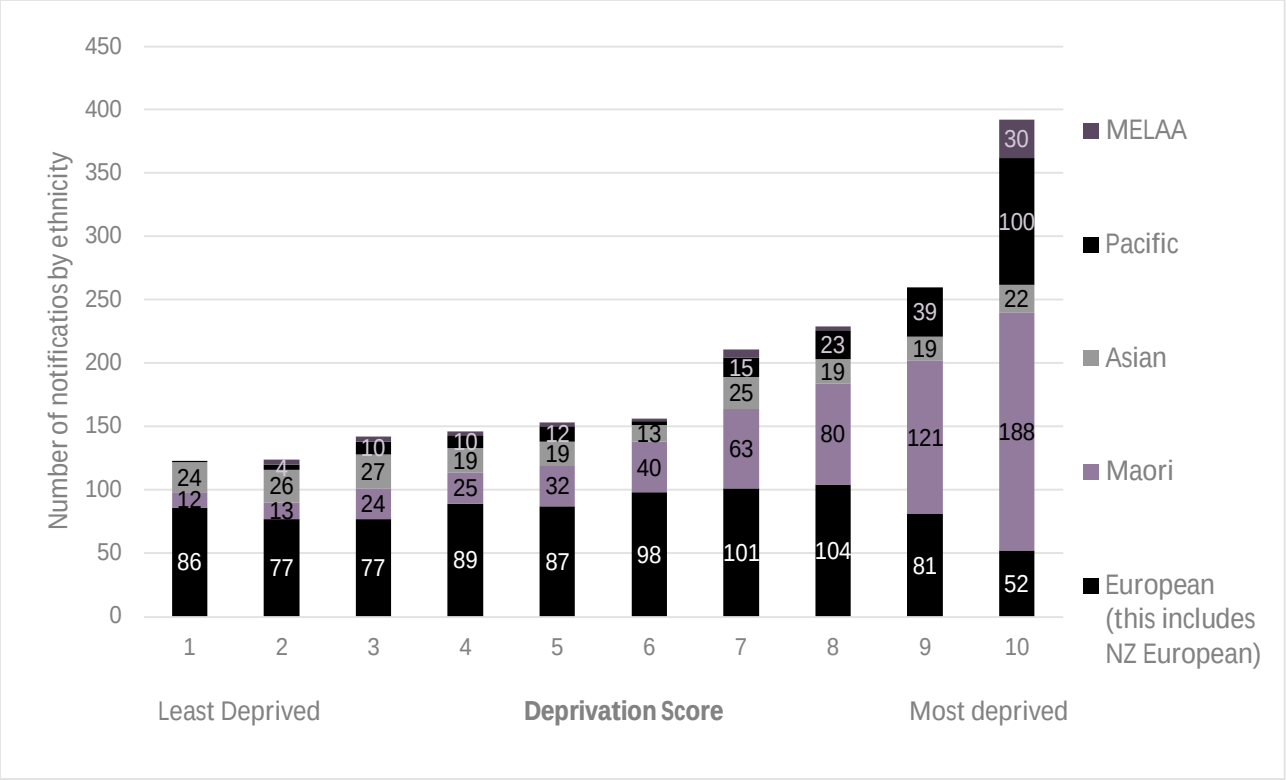


Figure 8: Deprivation scores of tamariki and rangatahi in the DND by ethnicity (2010-2018)[i]

A logistic regression was conducted for 2010-2016 notifications to see whether a linear or non-linear relationship exists between a tamariki having other known disabilities and level of deprivation. No association was found (p=0.7801).

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

- **The aetiology of hearing loss is either genetic or non-genetic in nature. The proportion of hearing losses with a confirmed genetic cause is increasing.**
- **Almost 99% of the records in the Database contain information about whether the cause of the hearing loss is unknown or known, and 89% of these have hearing loss with an unknown cause.**
- **The proportion of hearing losses where the cause was known has been falling since the relaunch of the Database in 2010 and particularly from 2014, likely in part due to the reducing age of identification resulting from nationwide implementation of newborn hearing screening, which began in 2007.**
- **Just over 3% of the children and young people in the Database are reported to have 27 specific syndromes, the most common being Down Syndrome.**

Causes of deafness

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

The American College of Medical Genetics and Genomics estimated in 2014 that 30% of genetic deafness is syndromic[67]. In non-syndromic deafness with a genetic cause, the most common genetic mutations found are in the GJB2 and Pendrin genes. The Otoferlin gene has been implicated in cases of ANSD[68].

The proportion of hearing losses with a confirmed genetic cause is increasing over time[69, 70], as more hearing losses are better understood in terms of their aetiology, and as genetic testing becomes cheaper and more widely available. Hereditary hearing loss is clinically and genetically varied, and even with the large number of genes that have been associated with hearing loss, many cases still remain unexplained[71].

'Genetic defects[i]' were estimated by Morton and Nance in 2006 to result in 68% of the cases of hearing loss present at birth and 54% at 4 years[72].

Non-genetic aetiologies resulting in an early onset of hearing loss include prematurity and infections during pregnancy, such as cytomegalovirus. The influence of non-genetic aetiologies is known to increase with age at onset, as infections (including rubella), medication, exposure to trauma, diseases such as meningitis and mumps, and noise-exposure become factors[72].

In tamariki, mumps is thought to be the commonest cause of unilateral acquired sensorineural deafness and is usually sudden in onset and profound in severity[73].

Cytomegalovirus (CMV) is a significant cause of deafness among children and young people in overseas studies, causing 10-20% of cases in those under the age of five[74].

i The term 'genetic defects' is used in the paper and has a specific meaning in the literature.

Internationally, as reported by Davis and Davis[3], it is common for a high proportion of cases (between 15% and 57%) of hearing loss to be of unknown aetiology. Aetiology is reported as more likely to be investigated in cases of bilateral hearing loss, and where the hearing loss is more severe in nature, compared with unilateral cases or those which are less severe[75].

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 used[76].

New Zealand data

Almost 99% of the 1776 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, 89% are of unknown cause, with the remaining cases listed as having a known cause.

As seen in Figure 9, the proportion of hearing losses where the cause was thought to be known has decreased significantly since 2012, 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 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.

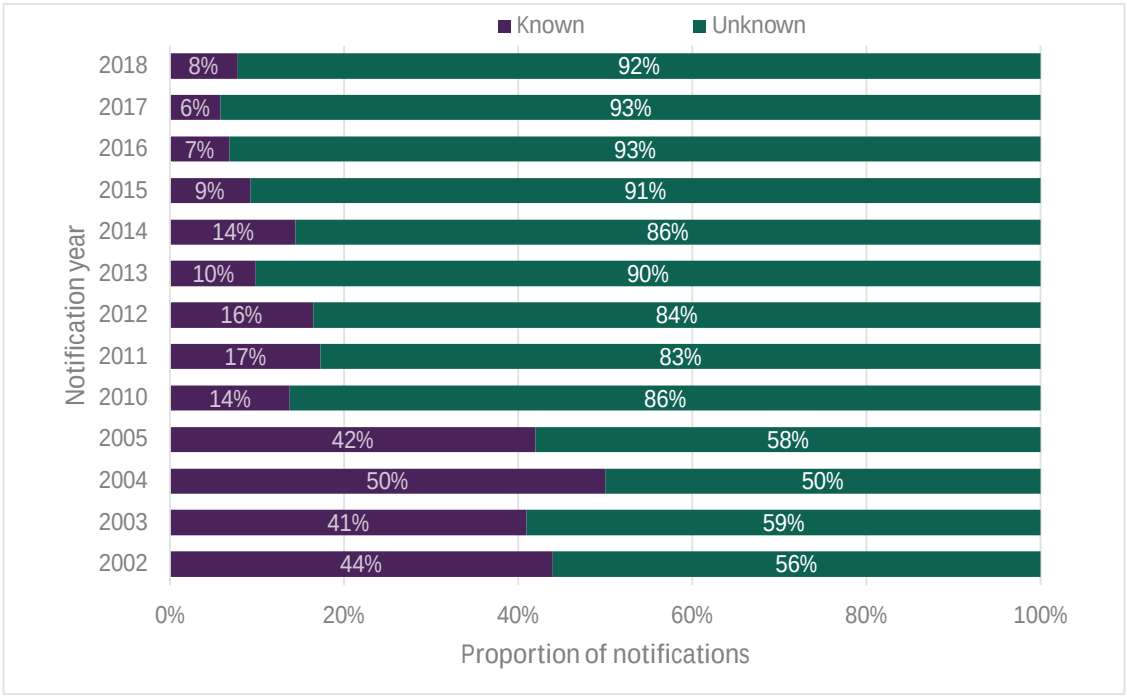


Figure 9: Proportion of hearing losses of known and unknown cause notified to the DND by year diagnosed (2002-2005 and 2010-2018)

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 generally increased immunisation coverage. The impact of the current measles epidemic[77] is not yet known. 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 consideration[78].

The importance of cytomegalovirus (CMV) in causing deafness among tamariki in New Zealand is not yet understood. CMV seroprevalence was assessed from 9343 first-time New Zealand blood donors in 2009. The highest prevalence was found among Pacific Islanders (93.2%) and the lowest in Caucasians (54.8%)[79, 80].

In New Zealand during the 2010-2018 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.

Aetiology types

In an attempt to better describe children and young people with a known aetiology, we have categorised these and in the section *Most common types of additional* disabilities on page 15.

Children and young people with syndromes

Among the 1757 children and young people in the 2010-2018 dataset, twenty-seven specific syndromes had been confirmed, affecting 55 children and young people. This number represents just over 3% of the total.

The most common syndromes identified were Down Syndrome (also referred to as Trisomy 21), which was identified at the time of the notification for 14 children and young people, Goldenhar Syndrome and Pierre Robin Syndrome/Sequence which were present in five children/young people each.

For information on syndromes we recommend the OMIM Catalog of Human Genes and Genetic Disorders. It provides comprehensive and well referenced online information on a large variety of genes and genetic disorders, and is freely accessible to the public and clinicians. The links to the most common syndromes listed above take the reader to their respective pages in this catalogue. It may be helpful for audiologists to better understand syndromes of those in their care so they can determine an appropriate plan for clinical management.

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 scans[81].

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 services[82].

Identification of hearing losses

- **Behavioural methods for identifying hearing loss among young children are not reliable so before implementation of objective newborn hearing screening across New Zealand, average ages at the time of identification were very high.**
- **Historically, for those born in New Zealand, it was parents who were the most likely to suspect their child's hearing loss.**
- **Since 2013, newborn hearing screeners have been the most likely group to first suspect hearing losses among New Zealand children and young people, with 60% of all diagnoses resulting from a screening referral.**
- **85% of the 110 cases notified in 2018 were notified by the internationally recommended age of three months.**
- **There are two peaks for identification of hearing losses among New Zealand tamariki and rangitahi – from newborn hearing screening and from the B4 School Check – between the age of four and five.**
- **An estimated 94% and 99% of the eligible population are checked by UNHSEIP and B4SC.**
- **Since implementation of newborn hearing screening, the proportion of children and young people whose hearing losses have been identified before the age of one has more than quadrupled to 114.**
- **Those born overseas, with mild, acquired and/or unilateral hearing losses and those who are of Pacific ethnicity are at greater risk of having their hearing loss identified later.**

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 2018.

Table 9 shows the top three groups that first suspected the hearing loss among notified cases *during selected years* since 2010.

Parents/caregivers have gone from being most likely to first suspect a child or young person's hearing loss – in more than a third of cases (37% in 2010) – to being first in only 9% of cases.

The proportion of 2018 cases first suspected by parents or caregivers is significantly below historic levels reported in the original Database, in which

	2010	2014	2016	2018
Most likely to suspect	Parent or caregiver (37%)	Newborn hearing screener (39%)	Newborn hearing screener (56%)	Newborn hearing screener (63%)
Second most likely to suspect	VHT (17%)	Parent or caregiver (22%)	VHT (16%)	VHT (10%)
Third most likely to suspect	Medical professional (10%)	VHT (13%)	Parent or caregiver (6%)	Parent or caregiver (9%)

Table 9: Groups most likely to first suspect hearing loss (Selected years, born in NZ)

between 34% and 52% of cases were first suspected by parents during the 2000-05 period.

Newborn hearing screeners were not in the top three groups to suspect a hearing loss in 2010 or 2011[i] and yet they are now first to suspect more than half of all cases of hearing loss notified to the Database.

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[83, 84].

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 from developing or to cause significant delays in speech development.

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

Age at diagnosis

Figure 10, below, shows the number of cases identified grouped by the age of the child[ii]. 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 during the first year after birth is more than four times as high in 2018 (n=114) as it was in 2010 (n=24), when the Database was re-launched.

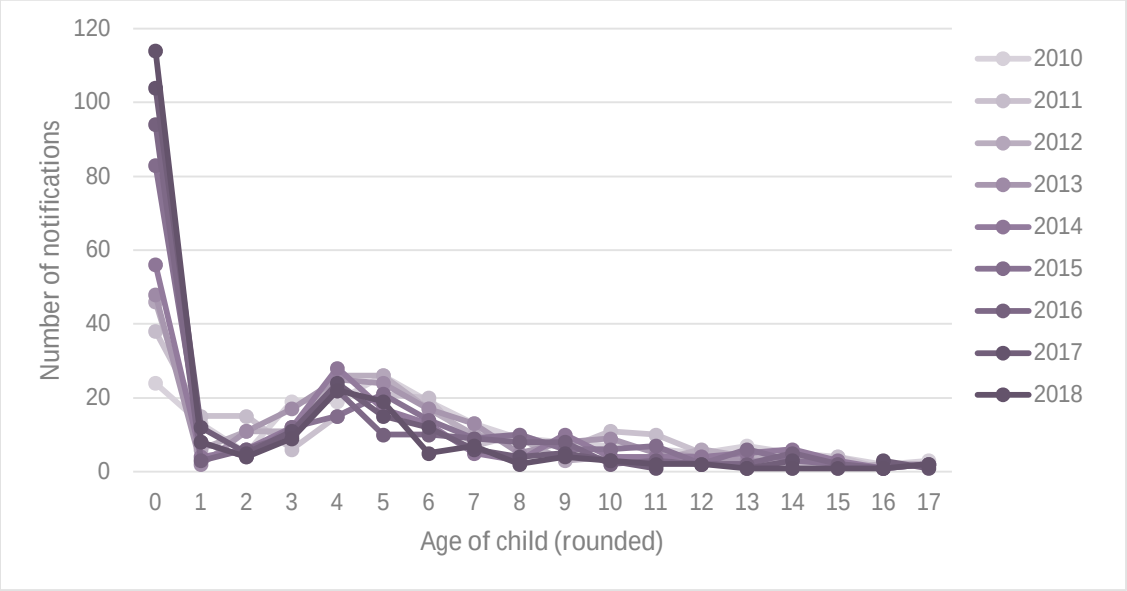


Figure 10: Number of children diagnosed by age (2010-2016)

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

ii 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.

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[i, 85]. 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 43 for more details about the B4 School Check). This suggests that some children who were previously being identified by hearing screening around 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 losses that are late onset (such as progressive and acquired hearing losses).

It is important to remember this average age includes all children diagnosed in the notification period, for whom specific confirmation age data was available[ii], 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 10. 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 Table 10 have been slightly revised compared to those reported previously, to account for some notifications that were later removed from the Database as more information became available and others that have been added retrospectively. These changes are small.

	2010	2011	2012	2013	2014	2015	2016	2017	2018
Average all cases	65	57	61	60	60	53	44	37	34
Average born in New Zealand	62	53	56	54	53	47	37	32	31

Table 10: Average ages of diagnosis for all cases in months (2010-2018)

i 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 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.

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

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 11 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 11: Early and late average ages of identification (2010-2018)

Age at diagnosis by severity of hearing loss

Table 12 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 (2010-2018)	Total number of cases
mild	62	543
moderate	40	293
moderately severe	27	77
severe	27	36
profound	12	65

Table 12: Average age at diagnosis, in months, for *bilateral* hearing losses by degree (ASHA codeframe) using interpolated data with manual checks (2010-2018)[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 from the audiologist and interpolated data-points.

The average age at which bilateral hearing losses (including those for children born outside New Zealand) is confirmed has dropped from an average of 63 months in 2010 to 33 months in 2018. Please note that these changes may relate to cohort differences as well as overall improvements in the age of identification resulting from the implementation of newborn hearing screening around the country.

Children under the age of four are more likely to be missing some severity data[ii], meaning they could not be classified for Table 12. 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 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 European children, although

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

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

this difference was not reported in every one of these reports[i].

Table 13 shows the average identification ages (2010-2018) for each ethnic group, for all children and young people notified, where ethnicity information was provided.

Ethnic Groups	Average months at diagnosis (2010-2018)
European	50
Māori	51
Pacific Peoples	64
Asian	38
MELAA	65

Table 13: Average months at diagnosis by ethnicity (2010-2018)

When viewing data on ethnicity, please keep in mind that Table 13 is based on multi-code data, hence a small number of cases are in two or more ethnicity groups at one time.

Children and young people listed with Pacific Peoples and/or MELAA ethnicity consistently have the highest average age at diagnosis when compared with the other groups in the sample.

The average age at detection across all years is of particular concern for Pacific tamariki, at 64 months, although recent years have seen a drop from a high of 84 months in 2012 to 33 months in 2018, which suggest improvements for this group, although may be related to differences in the cohorts over time.

Māori children and young people have been identified at an average of 52 months over the full period, the same as their European counterparts. The average age has dropped from a high of 65 months in 2013 to 30 months in 2018.

MELAA also have a high average at 65 months. While these data are included below it is worth keeping in mind that this group is historically very small, so large variations exist in the averages over time although generally these averages are high.

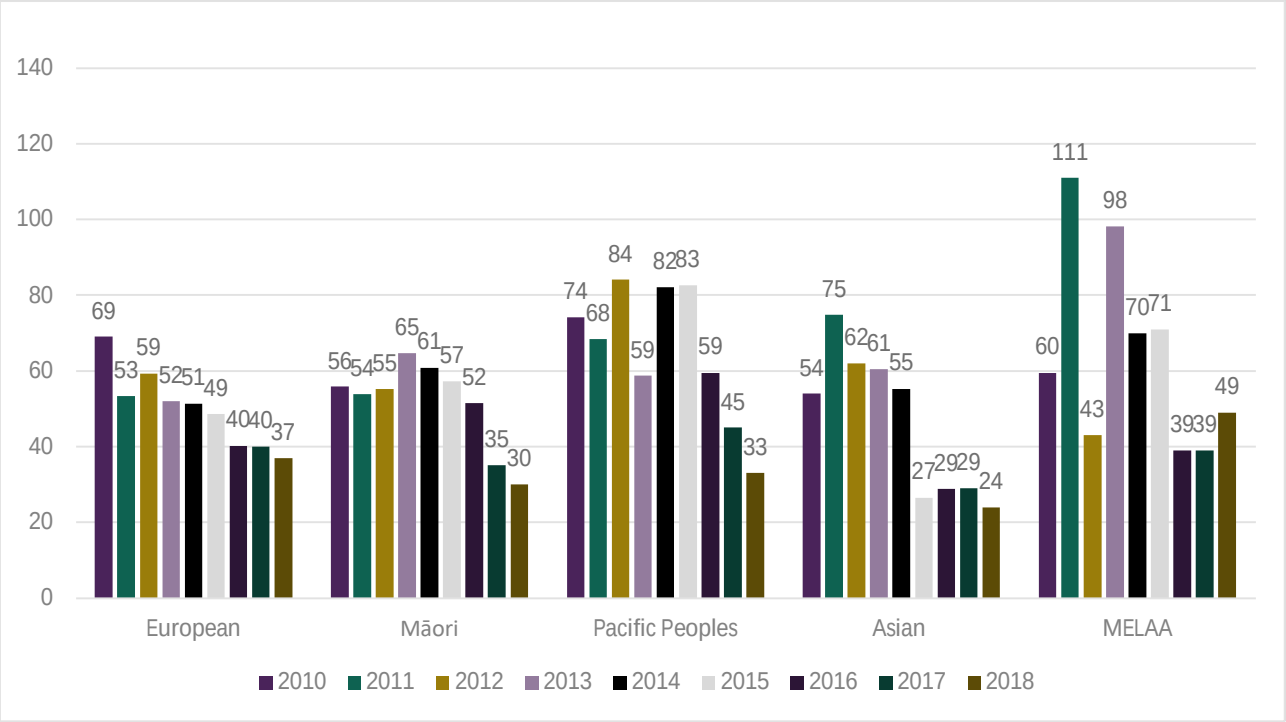


Figure 11: Average age of diagnosis by ethnicity in months (2010-2018)

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

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 European peers, and mild and moderate losses are on average identified later than severe and profound losses[54]. 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.

We hope future analyses will shed light on the types of hearing losses that are common among these groups, so we can better understand the reasons for their later average diagnoses.

Another way to examine average ages at diagnosis is to split cases into 'Māori', 'European' or 'both'[i]. In 2016 the variance was examined 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 European categories[ii] (95% CI: (82, 870)) - by 477 days, and those
- listed in the European category only (95% CI (82, 870)) - by 230 days[iii].

Newborn hearing screening

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

Screening status

Table 14 shows the screening status of New Zealand-born children notified to the Database (and therefore diagnosed) in the period 2010 to 2018.

As expected, the proportion of children being diagnosed as a direct result of referral from the UNHSEIP is increasing, and the proportion of children notified who were not offered screening is falling.

Please note that this table shows children diagnosed at varying ages, so not all were screened as newborns because 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 UNHSEIP is still incomplete, the true extent of loss to follow-up in the UNHSEIP cannot be ascertained.

The most recent NSU UNHSEIP Summary Report[48], includes data for babies screened from **1 January to 31 December 2017** (a year earlier than for the DND data contained in this report), and, reports that[v]:

i 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 1561.

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

iii 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 European (p=.071).

iv 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.

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

Was universal newborn hearing screening (using aABR or aOAE) offered to this family after this child or young person's birth?		2010	2012	2014	2016	2018
No	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	4%	4%	1%
	No, this service was not available at the time (at the time of diagnosis)	68%	54%	38%	13%	6%
Unsure	Unsure whether screening was offered to this family	7%	6%	5%	2%	6%
Yes	Yes, a screening programme was in place, but the child was directly referred to audiology due to atresia	0%	1%	1%	5%	3%
	Yes, screening was offered but this child was not screened	1%	1%	1%	2%	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%	2%	1%	5%	3%
	Yes, this child was screened and passed	1%	6%	13%	16%	19%
	Yes, this diagnosis is a result of a referral from screening	18%	28%	39%	53%	60%

Table 14: Screening status of children born in New Zealand and diagnosed 2010-2018, selected years[i]

- 94% of babies born during 2017 completed screening within the period. Of those babies who completed screening, approximately 89% did this by the target of one month (corrected age).
- 1% of babies who completed screening during the period were referred to audiology, this is a drop from previous periods and may indicate improvements in screening practice as these figures are more in line with those found in established programmes overseas.
- 70% of the babies referred to audiology had audiology assessment data reported to NSU by the date of data extraction for the report. 64% of babies referred had their assessment completed by the target time of three months of age, well below the target of 90%.
- Māori babies were less likely to be screened (when compared with non-Māori and non-Pacific babies) and when they were screened they were less likely to be screened by one month of age.
- Māori and Pacific babies were more likely than other ethnicities to be referred to audiology.
- Māori and Pacific babies were less likely to complete diagnostic audiology and to complete diagnostic audiology by three months when compared with non-Māori and non-Pacific babies.
- Rates of permanent congenital hearing loss were 2.0 per thousand babies overall, (both bilateral 1.2 per thousand and unilateral 0.8 per thousand) meaning there were 112 babies diagnosed as a result of screening within 2017. By ethnicity these were: Māori babies (2.8 per thousand), Pacific babies (2.2), Other babies (1.7) and Asian babies (1.5 per 1,000 babies).

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.

- 90% of referrals for early education intervention began receiving services by one month of age, meeting the 90% target. [At this time, we have no data to help us understand ongoing access rates.]
- 98% if the children referred to the Ministry of Education through the UNHSEIP under six months of age began receiving early intervention support before six months of age.
- 47% of the children who were identified through the UNHSEIP demonstrated language abilities at or above their current age level when compared with peers of the same age, when assessed at four and a half years. 63% of the children assessed presented with language abilities that did not indicate a significant language delay when compared with same age peers.

Referrals from the UNHSEIP

The Universal Newborn Hearing Screening and Early Intervention Programme has provided much needed local data helping us understand birth prevalence of the types of hearing losses that are the target of this screening.

This 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 0.8 per thousand cases for unilateral hearing loss per thousand babies screened[48]. This is higher than many of the reported rates from overseas screening programmes[i].

These higher prevalence rates are consistent with the higher rates of hearing loss seen among young Māori whose information is notified to the DND, in comparison to their European counterparts.

A total of 110 of the 2018 notifications (60%) were for children born in New Zealand who were diagnosed as a direct result of newborn hearing screening. This percentage has risen from 18% in 2010 but has remained static during 2017 and 2018.

It is worth remembering that the number of cases of hearing loss that 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 - is unknown.

	2010	2012	2014	2016	2018
Number of diagnoses resulting from universal newborn hearing screening	34	48	59	88	110
Diagnoses as a proportion of total notifications	18%	28%	39%	52%	60%

Table 15: Diagnoses (born in NZ) resulting from newborn hearing screening in New Zealand[ii], 2010-2018, selected years

i 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. Using these overseas rates and including unilateral hearing losses, we might expect approximately 95 diagnoses directly from the newborn screening programme each year, based on an average figure of 59,803 births per year in the period 2010-2017.

Because overall population prevalence in New Zealand is not known for the types of permanent hearing loss included in the database, 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.

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.

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.

There has been a pleasing overall reduction in the average age at diagnosis of cases referred from newborn hearing screening in New Zealand (therefore born in New Zealand), from fourteen months in 2010, to four months in 2018.

Of the 110 cases notified in 2018 that were identified as a direct result of newborn hearing screening in New Zealand, 85% were diagnosed by the internationally recommended age of three months.

Table 16 shows the changes in the average age at diagnosis since 2010.

	2010	2011	2012	2013	2014	2015	2016	2017	2018
Average months at diagnosis	14	8	6	7	5	6	4	5	4

Table 16: Age at diagnosis for children referred from and diagnosed as a result of the newborn hearing screening programme (2010-2018)

Identification of false negatives

The DND likely provides the only method for identifying potential false negatives from the newborn hearing screening programme86,i.

In 2018, no cases notified to the Database were explicitly identified as having wrongly passed their New Zealand based newborn screeningii, meaning we have no confirmed false negative cases for this year. This isn't to say that one or more babies diagnosed in 2017 were not incorrectly passed at their newborn hearing screening, just that none were recorded as such in the notifications.

There was one case that the notifying audiologist noted on the form was a likely false negative:

> *"This baby referred from newborn hearing screening in her left ear and the results indicate a sensorineural hearing loss in this ear. She actually passed screening in her right ear, but there are strong indications of a sensorineural hearing loss also being present in this ear, despite the screening [result]. Unfortunately, I was only able to get limited ABR results for this baby in her right ear due to her older age and limited*

i 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, 2012.

ii 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.

sleep. The family did not attend their first appointment for ABR and two appointments were required to gather the results that were obtained. These factors delayed the diagnosis. It was decided to defer further ABR assessment at this time due to the distance the family need to travel to these appointments and baby's older age for ABR under natural sleep. Our DHB will continue to monitor her hearing."

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.

Thirty-four of the tamariki who were born in New Zealand and identified with hearing loss during 2018 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 as over-time more tamariki are being screened.

Of those 34 cases, it is possible to remove two groups to help us narrow the focus on the most likely potential false negatives; this has been done in Table 17. The first of these have known acquired hearing loss, while the second is those with hearing losses where there is some uncertainty – they were either suspected to have been present at birth, or the diagnosing professional was unsure whether the hearing loss was likely to have been present at birth[i]. As the second of these groups is based on a relatively subjective assessment by the clinician, these cases may or may not provide cause for concern. (It is possible New Zealand has a greater prevalence of progressive hearing losses because of our high rate of CMV[79].)

Of the nineteen 2018 cases identified as *potential* false negatives in Table 17, the age of identification for these tamariki ranged from two to ten years of age.

The families experienced a wait to see a hearing professional in two of these cases, while in four cases the audiologist had difficulties getting a confirmed diagnosis. Parents/the young person/carers or educators suspected something other than hearing loss in two cases. In one case the child referred unilaterally from their newborn hearing screen and the ABR was misinterpreted as a temporary conductive hearing loss on that ear. After referring on the hearing screen at the B4SC the child was referred back to audiology and at that time the permanent hearing loss was diagnosed.

	2010	2011	2012	2013	2014	2015	2016	2017	2018
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	10	19	28	28	32	34
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	18	18	19

Table 17: Potential false negatives and cases previously referred from hearing screening, 2010-2018, born in New Zealand only

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

Of the 19 cases listed, 14 were coded 'unsure' to the question about whether the loss was thought to have been present at birth. The remaining five were listed as 'yes' to that question.

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 alternating cohorts from 2010-2017 are shown below. See previous reports in this series for data from other years.

A recent paper by Gibb *et al*. (2019) from the *British Medical Journal* found Māori and Pacific children were less likely to complete the checks than non-Māori and non-Pacific children along with other disadvantaged groups, such as those living in socio-economic deprivation, the young, those tamariki with younger mothers, and those with worse health status. The authors note that the 'patterns of non-participation suggest a reinforcing of existing disparities, whereby the children most in need are not getting the services they potentially require', and the authors suggest increased efforts to ensure all children are screened[87].

Outcome	Description	2011/12	2013/14	2015/16	2017/18
Pass Bilaterally	The child was screened and passed.	65%	72%	80%	84%
Referred	The child was screened and referred to a relevant service.	5%	5%	5%	5%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	8%	6%	5%	6%
Under care	The child is already under the care of a relevant service.	3%	3%	4%	4%
Decline	The hearing check was declined by the caregiver.	5%	3%	1%	1%
Not Checked	The child did not receive a hearing check.	16%	12%	5%	1%
Population	**Derived from the PHO enrolled population.**	**65,692**	**65,335**	**62,581**	**61,005**

Table 18 B4 School Check Hearing Screening data (those tamariki screened in alternating years from 2010-2018)[i, ii, 56]

Please note that the data used for this paper were from 2014/15 and the numbers not checked have reduced since that time from 11.6% to 1% in the 2017-18 year. All ethnic groups have seen these improvements. However, it is also worth noting that the denominators for the B4 School

i The Ministry of Health notes that the population used is the PHO enrolled population. We use this rather than SNZ due to the better inter census accuracy, and as Statistics New Zealand population projections only include 5-year age groups.

ii Note that column figures don't sum to 100% due to rounding.

Check comprise children who are enrolled with a PHO. Some children who are not enrolled are also screened but the data make it difficult to understand the overall coverage rate for the hearing screening within this Check. In this period, Pacific children have the highest proportion not screened at 3.8%, followed by European at 3.7%.

The overall referral rate for tamariki completing this screen is 5% (2017/2018). As with previous years, Māori and Pacific tamariki have higher referral rates (7.3% and 9.7%), and Asian and MELAA tamariki lower rates than the average (4.3% and 4.6%). The lowest referral rate was for European tamariki, at 3.8%.

Delays in diagnosis

- **The average delay between first suspicion of a child or young person's hearing loss and its confirmation is now six months, down from 26 months in 2010, undoubtedly the result of nationwide implementation of the newborn hearing screening programme.**
- **An earlier analysis of 2010-2016 data showed the average delay for Māori was 7.5 months higher than for European and that Māori had 1.32 times more reasons listed for their delay.**
- **Every one-unit average increase in the child/young person's deprivation score (based on the area in which they lived) was associated with an additional month of delay in diagnosis.**
- **Just over half the children and young people diagnosed in 2018 had no delay or a delay of one month or less listed in their record.**
- **Audiologists having difficulty getting a confirmed diagnosis was the most commonly mentioned reason for the delay in diagnosis. This can be the result of conductive overlay or the child being unwell. Parents not attending appointments (for any reason), was the second most common, followed by the waiting time to see a hearing professional.**

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 cases for which there was a delayed diagnosis had one or more reasons for the delay listed.

The average delay in 2018, 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 losses[i] was six months, down from nine months in 2017.

Just over half (52%) of 2018 notifications had no delay or a delay of one month or less. When all records for 2010-2018 are considered, 63% of notifications have a diagnostic delay of one month or more listed based on the age at suspicion and date of diagnosis[ii].

Year	Delay in months
2010	26
2011	16
2012	10
2013	12
2014	13
2015	11
2016	8
2017	9
2018	6

Table 19: Delay in months by year, 2010-2018[iii]

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.

ii It isn't easy to determine whether a delay exists for a specific case. For example, if a baby is referred to audiology and is unable to see an audiologist for two months this may be considered a delay, while for a 16-year-old some audiologists may not consider a two month wait to constitute a delay. In addition, some audiologists may mark a delay as existing and provide reasons where the delay is a week or two, while another may have a significant delay but not provide any reasons for this delay.

iii Please note that some figures have changed slightly to those reported previously due to inclusion of retrospective notifications in the main dataset.

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

Please keep in mind that these delay figures are not always directly comparable with previous years owing to the changing composition of 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.

A previous examination of 2010-2016 notification data showed that:

- that the average delay for Māori was 7.5 months higher than those for European tamariki.
- that the average delay for Māori tamariki was 15.66 months higher than those tamariki who identified as both Māori and European
- Māori tamariki were 1.60 times more likely to have one or more reasons for the delay listed in their notification form when compared with their European counterparts. 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[i].

Delay causes

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

In 2018, 38% of all cases had one or more reasons for delay listed. Seventy-two percent of those had one reason listed for the delay, and 28% 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 – this is not surprising given the reducing average age at identification and rising number of cases with no delay.

The analysis in Table 20 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. This includes all cases diagnosed in 2010-2018 and shows the most commonly cited reasons for delays in diagnosis, as well as some possible approaches to reducing the various types of delay.

When the 2018 cases are examined separately the order of the most common causes of delay remain generally the same, although a different item moves into fourth place "Child or Young person had other medical issue(s) which took precedence.

Comments received regarding 2018 diagnoses sometimes contained information about the reason for delays, and some of these are included below.

These comments demonstrate the complexity of reasons for delayed diagnosis, including issues with systems and equipment and underlying middle ear issues making diagnosis difficult and difficulties getting families in/back to clinic for appointments:

> *"Had first hearing test in 2012 but discharged as soundfield testing was normal with absent DPOAEs. GA-ABR was offered but the parents did not want that at the time. Re-referred by VHT in 2017." [73 month delay]*

i See the 2016 report for further detail.

Rank (most 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)	• efficient clinical practice to complete assessments over fewer appointments (Following 2016's Diagnostic and amplification protocols,[88] which can be found on the National Screening Unit (NSU) website and which used to be referred to as Appendix F) • prompt referral from newborn hearing screening
2nd	Parents did not attend appointments/delayed or rescheduled these (for any reason including service failed to engage family)	• better communication with parents, flexible appointments for families, 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 • increasing the responsibility of health professionals for differential quality of care between Māori and non-Māori, reducing a culture of blaming Māori for the state of their health[89] and acknowledging Pākehā privilege within health services[90] • assistance with travel costs
3rd	Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)	• better funding for audiology resources/DHBs to prioritise newborn hearing screening referrals and other paediatric cases • better communication with parents, more attempts to contact families before discharging from service, e.g. work to reduce 'do not attend rates'
4th	Parents or educators suspected something other than hearing loss (e.g. speech delay, developmental delay)	• 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	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	• better systems and processes for scheduling and seeing follow-up occurs

Table 20: Top reasons for delay for those with diagnosis delays of more than six months and possible remedies (2010-2018)

"Family attended initial ABR testing, but incomplete results were obtained as baby did not sleep. Family did not attend follow-up appointment and could not be contacted by schedulers. Lost to follow-up." [67 month delay]

"[The] child was a unilateral (right) refer from NBHS [newborn hearing screening]. Passed left ear, and ABR was misinterpreted as a temporary conductive hearing loss on the right. Chloe was referred back into our system after failing B4 School hearing testing on the right, with a recent confirmation of profound SNHL on that side." [50 month delay]

"Delay due primarily due to overlying conductive component and ABR equipment not having been calibrated to test bc at 4000Hz at the time. Would have known earlier if able to test this then." [36 month delay]

"Family did not attend several appointments (5x DNA in Audiology, several more DNAs in ENT and Paeds). Once family started attending behavioural testing was complicated by persistent middle ear dysfunction and variable reliability. Therefore, ABR under GA was arranged to coincide with ventilation tube insertion." [46 month delay]

"Medical professionals suspected a different cause for hearing loss (otitis media rather than cholesteatoma)." [17 month delay]

"Bilat refer on NBHS. DNA'd diagnostic audiology. Bilat refer on B4school check.

DNA'd diagnostic audiology x 2. Attended third appointment." [17 month delay]

"Parents cancelled / declined 2nd ABR attempt but agreed for VRA." [7 month delay]

Delays attributed to newborn hearing screening

Of the sixteen tamariki whose 2018 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 ten cases:

- audiologist having difficulties getting a confirmed diagnosis (n=5);
- waiting time to see hearing professional (e.g. DHB waiting list to see audiologist, for GA ABR, no audiology staff at the DHB, limited staff resource, referred to another DHB for service) (n=1);
- child or young person had other medical issue(s) which took precedence (e.g. feeding issues, medically fragile) (n=2);
- parents did not attend appointments/delayed or rescheduled these (for any reason including distance, ill family member, cost, declined offer(s) of appointments) (n=5);
- Follow-up lost/referral in the system and did not occur or was delayed (e.g. annual review or follow up appointment not made or referral not received or sent) (n=1)

More information about the causes of delays in all groups can be found in the section on Delay causes, beginning on page 46.

One important consideration for newborn hearing screening referrals is the importance of prompt referral from the UNHSEIP to audiology, and the high priority given to 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 because babies are 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[i].

[i] Some tamariki may not be testable using VRA until after six months due to other developmental difficulties.

Severity

- **Severity data is now much more likely to be estimated from the ABR as children are being diagnosed at younger average ages.**
- **Many different frameworks are used to categorise severity of hearing loss around the world. Here in New Zealand the Clark (ASHA) framework is most common.**
- **The proportion of notifications classified as less severe is higher among bilateral cases.**
- **A number of factors are likely to contribute to New Zealand's lower proportion of more severe hearing losses when compared with other similar jurisdictions, including higher numbers of milder degrees of hearing loss among Māori.**

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 (NSU) 2009 Universal Newborn Hearing Screening and Early Intervention Programme National Policy and Quality Standards[i].

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

Those professionals who notified 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 that 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 form[ii].

As shown in Figure 12, below, the proportion of cases for which the thresholds were determined through ABR is rising, from 21% in 2010 to 59% in 2018. This strongly suggests that over time 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 and follow up rates continue to increase, meaning 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.

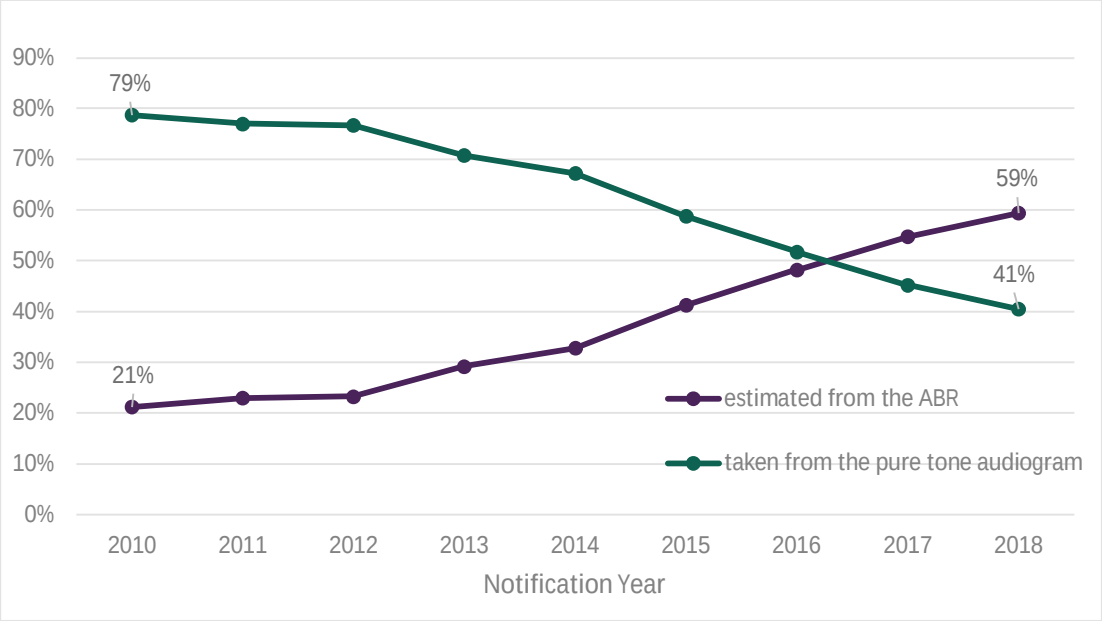


Figure 12: Proportion of cases containing thresholds from ABR (as opposed to being taken from the PTA), by notification year 2010-2018

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 F: Severity codeframes, on page 64.

Degree of loss	Clark 1981 (ASHA)[91]
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 21: 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[i]. 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.

Interpolation

Table 22 shows the severity of hearing losses notified between 2010 and 2018, calculated in two ways. The first of these is using data containing all eight data-points, while the second includes interpolation[ii].

While only cases where all eight-audiometric data-points are present can be included in most

i 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.

ii Please note that while the label in last year's report indicated that the data in this table covered 2010-2017, it actually included only 2016 data.

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:

- the proportion of less severe hearing losses is higher among bilateral cases;
- the number of bilateral hearing losses for which severity can be calculated rises when interpolation is used;
- the proportion of mild bilateral losses drops when interpolated 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.

Table 22 compares the proportion of bilateral/unilateral cases, comparing those that have not been interpolated or had manual checks with those that have. Please note that this includes all notifications from 2010-2018 while last year's report only contained 2014 data.

Degree of loss using ASHA severity codeframe	Bilateral 2010-2018	Bilateral 2010-2018 (interpolated and manual checks)	Unilateral 2010-2018	Unilateral 2010-2018 (interpolated and manual checks)
Mild	61%	54%	52%	45%
Moderate	26%	29%	7%	16%
Moderately severe	5%	8%	9%	9%
Severe	3%	3%	6%	6%
Profound	4%	6%	17%	23%
Sample size	n=714	n=1037	n=433	n=509

Table 22: Comparison of severity classifications by methodology, 2010-2018

Severity profile differences between bilateral and unilateral hearing losses

Most previous reports have 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.

For 2017 and this year's report, 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 believe this provides a more accurate picture, and this method of analysis will be used in future.

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

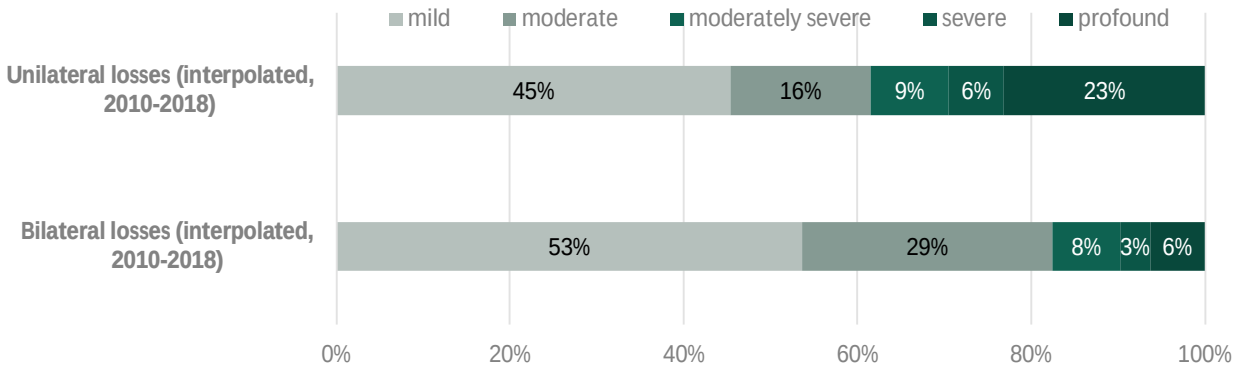


Figure 13: Unilateral and bilateral hearing losses by degree (2010-2018)[i]

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 losses[ii];
- 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 notifications using the DND severity codeframe (1996-2005), a longitudinal comparison of the proportion of tamariki and rangatahi 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 2018 figures shown here match those exclusions from the earlier Database.

Table 23 shows the average proportion of hearing loss notifications in each category between 2010 and 2018 and compares this with data from 2001 to 2004[iii].

Proportion of cases notified by degree of hearing loss	Average 2001-2004	Average 2010-2018
Mild	48%	54%
Moderate	35%	34%
Severe	10%	7%
Profound	6%	6%

Table 23: 2001-2004 DND data compared with interpolated 2010-2018 notification data, selected cases only (1996-2005 DND severity codeframe)

i Please note that in the 2017 report this graph was mislabelled in the plot area as 2010-2017 data, when it was in fact 2017 data only as described in the graph caption.

ii 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.

iii 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 or not 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 23, which includes more cases by using interpolated and manually checked thresholds.

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 smaller.
- Some cases with audiometric data points in the severe and profound range did not contain complete audiometric data and these have not been included in this table, meaning severe losses (and other degrees too) may be under-represented[i].
- Often children diagnosed with hearing loss have a sloping hearing loss and the better thresholds reduce the average degree of hearing loss.
- As noted previously, vaccination programmes have reduced rates of meningitis in New Zealand and this reduction is expected to have led to a reduction in rates of (more severe) hearing loss[92]. However, the reduction in the number of more severe cases due to meningitis is likely to be small.

A number of viral infections can cause hearing loss, which can be congenital or acquired, unilateral or bilateral and is typically sensorineural[93], although mumps, for example, almost always causes single-sided deafness.

Ethnicity and severity profiles

Historically, DND reports have shown that the greatest number of notifications pertain to European and Māori and children, and that milder degrees of hearing loss are more commonly reported among Māori[51, 94].

These findings have been confirmed by analysis of: 1982-2005 data[54, ii] and 2010-2016 data[iii].

A previous analysis of cases that were coded only as Māori or European was also completed, showing the proportion of cases of moderately severe or greater severity was 8% among Māori, compared with 14% among European. It was 7% among those listed as both Māori and European.

Together, these examinations suggest that young Māori have fewer severe and profound hearing losses than their European counterparts.

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.

ii Young Māori in the database are more likely to have mild or moderate hearing losses when compared with their European peers

iii A 2016 analysis showed the proportion of cases in each of the severity categories, split by ethnicity grouping, and found Māori had a higher proportion of mild and moderate cases than their European peers.

Comparisons with international data

Several analyses have been conducted for previous DND reports to compare the notifications to the DND and their 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, at least in part, due to the fact that Māori have a different severity profile to other groups.

In addition, recent research suggests those children with milder degrees of hearing loss who were previously unaided, demonstrated deficits in phonological memory and morphosyntactic skills, suggesting issues with leaving mild hearing loss untreated[95], although research focusing on mild hearing losses is limited.

As a result of this apparent difference, clinicians might keep in mind that those children and young people with milder degrees of hearing loss are at increased risk of not wearing hearing aids prescribed to them[96, 97], and that those families with children who have cochlear implants are managing and promoting device use more than those with hearing aids[98].

Intervention and support

- **The Ministry of Education provide services to students who are deaf and hard of hearing through groups such as Advisors on Deaf Children and other specialist educators. They provide services to approximately 1,700 children under the age of eight, including 690 babies and young children identified as a result of UNHSEIP.**
- **Deaf education centre's based in Auckland (Kelston) and Christchurch (van Asch) provided services in the 2018 year to 2,475 deaf and hard of hearing students nationwide.**
- **Children and young people diagnosed with a hearing loss since 2010 and notified to the Database are most likely to be fitted with two hearing aids.**
- **Children and young people around the country received 97 publicly funded cochlear implants (a total for northern and southern regions) during the 2018 calendar year, and 1,299 children and young people received hearing aids through MOH funding.**

Ministry of Education

Although the Ministry of Education has not yet been able to provide data related to the UNHSEIP, or for language outcomes of identified children and young people these are being developed and they hope to share these for future reports.

For the first time, the Ministry have been able to share with us information about the numbers of children and young people who are deaf or hard of hearing who are receiving services from Ministry of Education staff. We are delighted to share these figures with you (see below).

In 2018 the Ministry of Education, Learning Support provided support to approximately 1,700 children who are deaf and hard of hearing, birth to eight years of age (Year 3 at school) through the Adviser on Deaf Children Service.

This included support to children in the following areas:

- Support for 690 babies, infants and children under the age of five identified as deaf and hard of hearing through the Universal Newborn Hearing Screening programme (UNHSEIP) and their families and whānau.
- Support for 240 babies, infants and children under the age of five and their families identified as deaf and hard of hearing not through the Universal Newborn Hearing Screening programme (UNHSEIP) and their families and whānau.
- Support for 770 school aged children (Year 1 to Year 3 at school) identified as deaf and hard of hearing with moderate communication and learning needs.

The Ministry also funds support for children and young people who are deaf and hard of hearing birth to Year 13 at school through:

- First Signs support (Deaf Aotearoa), birth to five years of age,
- Cochlear Implant Habilitation programmes, habilitation support, and

The Deaf Education Centres (Kelston Deaf Education Centre and van Asch Deaf Education Centre), regional and core services.

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 DECs 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 have rewritten this section in the hope we can make the categories of service easier to understand, particularly for those not familiar with the terminology. Terms such as ORS (Ongoing Resource Scheme) are defined in the glossary on page 65.

As at 1 December 2018, the Deaf Education Centres provided services to 2475 tamariki and rangatahi. These services can be broken down into the categories[99] set out in Table 24.

Hearing aids

All but one of the 207 cases notified to the Database in 2018 contained information about whether hearing aids were to be fitted[i].

Audiologists were asked "How many hearing aids are to be fitted?". The resulting data represent the audiologist's stated plan at the time of notification. We have no data on what hearing aids, if any, were actually provided. There are several reasons why the plan may not be followed in individual cases (e.g. parental preference, worsening hear loss, diagnosis of additional needs).

As has been the case with data since 2010, children and young people whose cases were diagnosed in 2018, are most likely to be fitted with two hearing aids (50%). This reflects the preponderance of bilateral losses notified to the Database.

Figure 14 shows the proportion where the plan is to prescribe one or two hearing aids 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, likely for the same reason.

When data for all children and young people notified from 2010 to 2018 are considered, the audiologist's intention was to:

- fit 83% of bilateral losses with one or two hearing aids, while 5% were not expected to receive any aids;
- fit 40% of unilateral hearing losses with one hearing aid, while 18% were not expected to receive any aids.

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 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.

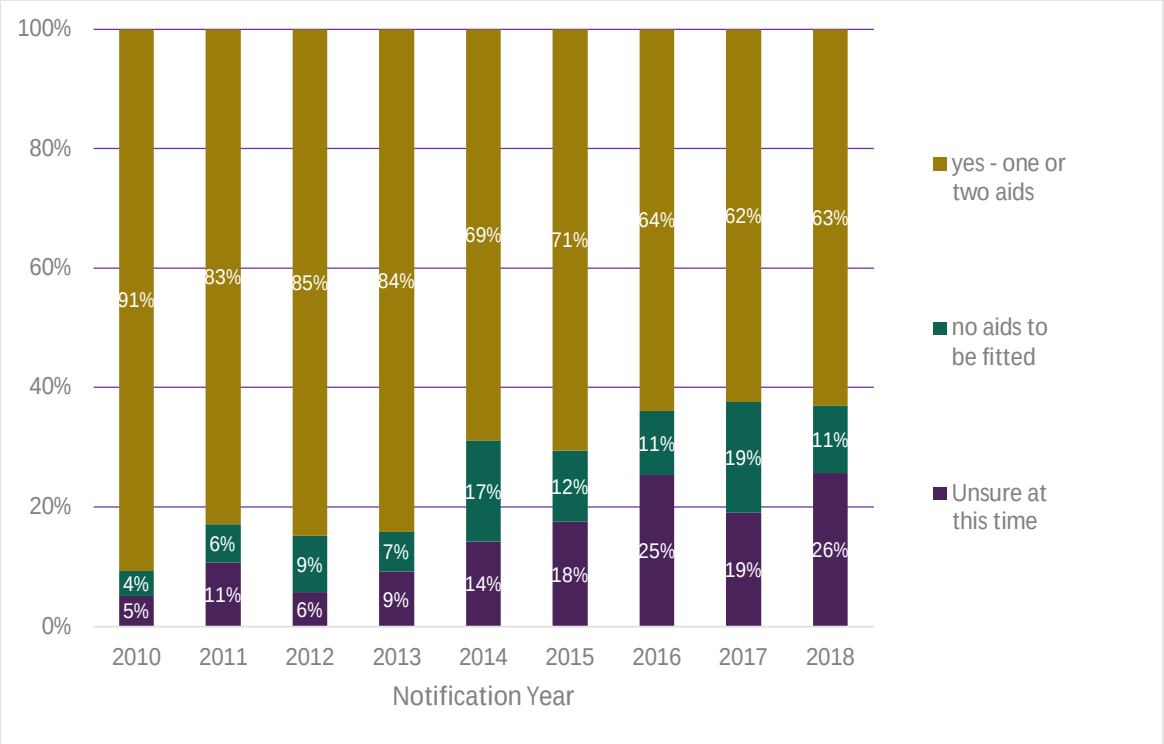


Figure 14: Hearing aids to be fitted by notifications (2010-2018)

Intention to fit, ethnicity and deprivation

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

Chi squared analyses completed and described in the 2016 report – which held severity constant – showed:

- there are significantly more 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 Europeans and fewer Māori with missing data than expected – this could be due to the fact that 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 was 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 in Table 24[i]. 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 2018 calendar year[ii, iii].

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.

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
Māori	72	42	291	50	455
European	59	62	312	90	523
Pacific	34	20	107	25	186
Other	20	15	86	14	135
Total	185	139	796	179	1299

Table 24: MOH Funding of Children's Hearing Aids, Calendar Year ending 31 December 2018, EnableNZ[100]

These and 2017 figures are different from those reported by the previous provider and this is thought to be due to significant differences in what is counted in these figures.

The current provider does not include repair or replacement requests, bone-anchored hearing aids, RM systems, or funding for parts, moulds or accessories in these data.

A total of 1299 unique service users (tamariki) received hearing aid(s) during this period.

A recent study in the US examined language outcomes for 290 children between two and seven years of age with mild to severe hearing loss. Those fitted after 18 months of age improved in their language abilities as a function of the amount of hearing aid use[101]. Risks of oral language development delays were found to be moderated by early and consistent access to well-fit hearing aids which provided optimised audibility.

Prescribing and usage

The NZ study mentioned in *Children/young people – seven years later* on page 12, followed up 163 of the 189 children and young people notified to the DND in 2010 seven/eight years later.

For those children and young people for whom the audiologist was intending to fit two hearing aids, and for whom 2018 follow-up data was provided (n=110), 18% received no hearing aids while 4% received one. The remaining 78% of children and young people received two devices as the audiologist intended.

For the 163 children and young people for whom follow-up data were provided:

- 40% had been wearing their device or devices consistently since they were fitted. Most of these young people (52%) were in the 12-16 year category, with 30% eight to 12 years old;
- 9% had no devices and so had no device usage information;
- 9% of cases did have devices but the clinic listed no device usage information;
- clinics 'did not know' about device usage in 13% of cases; and
- 29% had either used their device 'inconsistently', 'seldom' or 'never'. The age spread for this group shows most (40%) in the 12-16 year category, with 27% being eight to 12 years old;
- Of these children and young people, the most common reasons for poor or inconsistent usage were that:
  - » the child felt conscious/didn't like them/refused to wear the device(s) – 25%;
  - » no reasons were provided (15%);
  - » the initial diagnosis seems to have been inaccurate or the loss was temporary and so the device(s) are now no longer needed 13%;
  - » and the family felt the device provided no or limited benefit 8%;
  - » the children who were not wearing their device(s) for these reasons were mostly under the age of 16, with the largest group being between 12 and 16 years.
  - » 46% of children who were recorded as Māori had inconsistent, seldom or no device use, compared with 23% of Pākeha.

A recent study in the US examined oral language outcomes for 290 children between two and seven years of age with mild to severe hearing loss. Those fitted with hearing aids after 18 months of age improved in their language abilities as a function of the amount of hearing aid use[101].

Please note that while we have information from the UNHSEIP on the proportion of children who are screened by one month and who have diagnosis by three months, we do not have information on the proportion who receive hearing aids by six months of age.

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.

Most children receiving cochlear implants have severe or profound hearing losses, or progressive hearing losses that 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.

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 Taupō, and the Southern Hearing Charitable Trust covers the area south of this line.

During the 2018 calendar year there were 49 publicly funded cochlear implants provided in the Northern Region and 60 in the Southern Region, to children and young people under the age of 19. These implants are provided based on Ministry of Health candidacy criteria for children and young people who are assessed by the cochlear implant teams[i].

Children receiving cochlear implants	Southern Cochlear Implant Programme[102]		Northern Cochlear Implant Programme[103]	
	Ears	Children	Ears	Children
ACC cases	1	1	2	2
Public Funding - (1 Jan to 31 December)	54	28	43	23
Private procedures	3	3	3	6
Re-implants – recalled devices, failed integrity tests, or soft failures	0	0	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)	0	0	0	0
	60	33	49	32

Table 25: Publicly funded cochlear implants in New Zealand during (2018)[ii]

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: 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 are encouraged to make future notifications to the Database by following this link. Audiometrists are encouraged to make notifications for cases of hearing loss where they were the first to diagnose among those who are over the age of sixteen-years.

Notes for those completing notifications:

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 wherever possible, before the end of the notification period in mid-March of the following year.

   This ensures these reports contain accurate information about those children and young people notified during each 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 UNHSEIP 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 to make this possible.

   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.

Questions: If you are in doubt about whether or not a case meets the criteria for inclusion, please notify the case. For answers to any questions at all, please email Janet Digby.

Appendix B: 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 rangatahi from 1982 to 2005. This system included data on the number and ages of tamariki diagnosed with permanent hearing loss and annual reports describing collected notifications were released.

The data presented in reports before 2006 contained notifications provided to the Database within a specific year; that is, they pertained to cases notified to the Database in a calendar year, rather than those who are identified in that yearError! Bookmark not defined.. 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.

That 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 (UNHSEIP). Information from the DND was known to 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 with support from Dr Andrea Kelly and Professor Suzanne Purdy 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, Janet Digby and Andrea Kelly and Professor Suzanne Purdy.

Inclusion criteria

The original criteria for inclusion in the DND were based on a 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)[104]."*

There was a strong view among audiologists consulted that the previous definition (above), which was used before 2006, was 'medically-focused' and didn't adequately acknowledge or include hearing losses, particularly mild and unilateral losses, where the family might not want hearing aids fitted or where hearing aids may not be appropriate.

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

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

In addition, this database now includes 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],
- regardless of their place of birth.

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 temporary hearing losses associated with otitis media.

Notifying cases

Notifications to the relaunched Database 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 and hard of hearing 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 be gratefully received by Janet Digby.

i This group comprised: 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 they are at increased risk for poorer educational and speech/language outcomes compared to children and young people with normal hearing in both ears.

Appendix C: 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 believe it is likely that the Database has been receiving 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.

Appendix D: 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 grouping[105]. 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 generally greater than the number of respondents.

Using this method provides a more detailed and accurate 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 group 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 is easily available to them in their files or databases.

Appendix E: Terminology used in this report

There are several terms used by young people with a hearing loss and their families/whānau. Those whose information is included in this report range from those whose hearing losses are unilateral and mild in severity, through to those whose hearing losses are bilateral and profound. The terms commonly used differ both 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 have 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' in these reports, generally moving away from the term 'hearing impaired' which has been used previously.

Appendix F: Severity codeframes

Differences between classification systems make it difficult for meaningful direct longitudinal and geographical comparisons of the proportion of tamariki in each severity category[i]. Unfortunately, there is no clear standard internationally for classifying hearing loss, or a consistent definition for where a hearing loss begins for the purposes of epidemiological comparison.

Table 24 shows some of the differences between local and overseas severity classifications (these systems use an average of the pure-tone thresholds at 0.5 kHz, 1.0 kHz, 2.0 kHz and 4.0 kHz)[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)[106]	World Health Organisation[107]	CDC[108]	Proposed code from Davis and Davis[3]
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 26: 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 26.

Glossary

Advisors on Deaf Children (AODCs): The Ministry of Education employs Advisers on Deaf Children to help families understand their child's hearing loss and to guide parents as they consider the technology and communication options available. Advisors also provide assessments and information about a child's development and behaviour to other professionals working with the family. They work closely with teachers from the two Deaf Education Centres[109]. 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 this refers to cause(s) of a child or young person's hearing loss.

Audiometric data: Audiometric data relates to a person's hearing acuity given variations in sound intensity and pitch (frequency). The Database collects information on the child's hearing thresholds at 0.5, 1.0, 2.0 and 4.0 kHz wherever possible.

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 causes 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.

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 one or more targeted conditions, including hearing loss. This screening takes place when the child is aged four, or five if they are not checked earlier.

Bilateral hearing loss: Hearing loss affecting both ears.

BLENNZ: Blind and Low Vision Education Network New Zealand is a school that comprises a national network of educational services for children and young people who are blind, deafblind or have low vision in New Zealand.

Confirmation of hearing loss: For the purposes of this report, 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.

District health board (DHB): 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 and which holds the contract for the management and reporting associated with the New Zealand Deafness Notification Database.

False negatives: False negative is a term used to describe anyone screened who is 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)

Kaitiaki: Trustee, minder, guard, custodian, guardian, caregiver, keeper, steward. (Māori Dictionary) In the context of this report, this refers to the caregiver of a child or young person whose information has been provided to the DND.

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 students.

Notifications: Notifications contain data about an individual child or young person, demographic information, and information on the hearing loss and its diagnosis. Information is provided to the DND 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 learning support, to help them join in and learn alongside other students at school. This funding provides Specialist Services staffing for students (who are ORS funded) including school counsellors. This scheme was previously 'reviewable'.

Single Sided Deafness (SSD): The DND defines this group as children and young people who meet the criteria for the DND and who have a hearing loss of more than 70 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the worse ear, and a hearing loss of less than 26 dB HL over four frequencies (over 0.5, 1.0, 2.0 and 4.0 kHz) in the better ear.

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.

Resource Teachers: Deaf (RTDs)[i]**:** 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 Year 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 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 (NSU) 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 directed 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, along with other Well Child providers, 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 VHTs 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.)

i This information is adapted from a very helpful description found on the KDEC website.

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