Deafness Notification Report 2019

Rīpoata Whakamōhiotanga Turi

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

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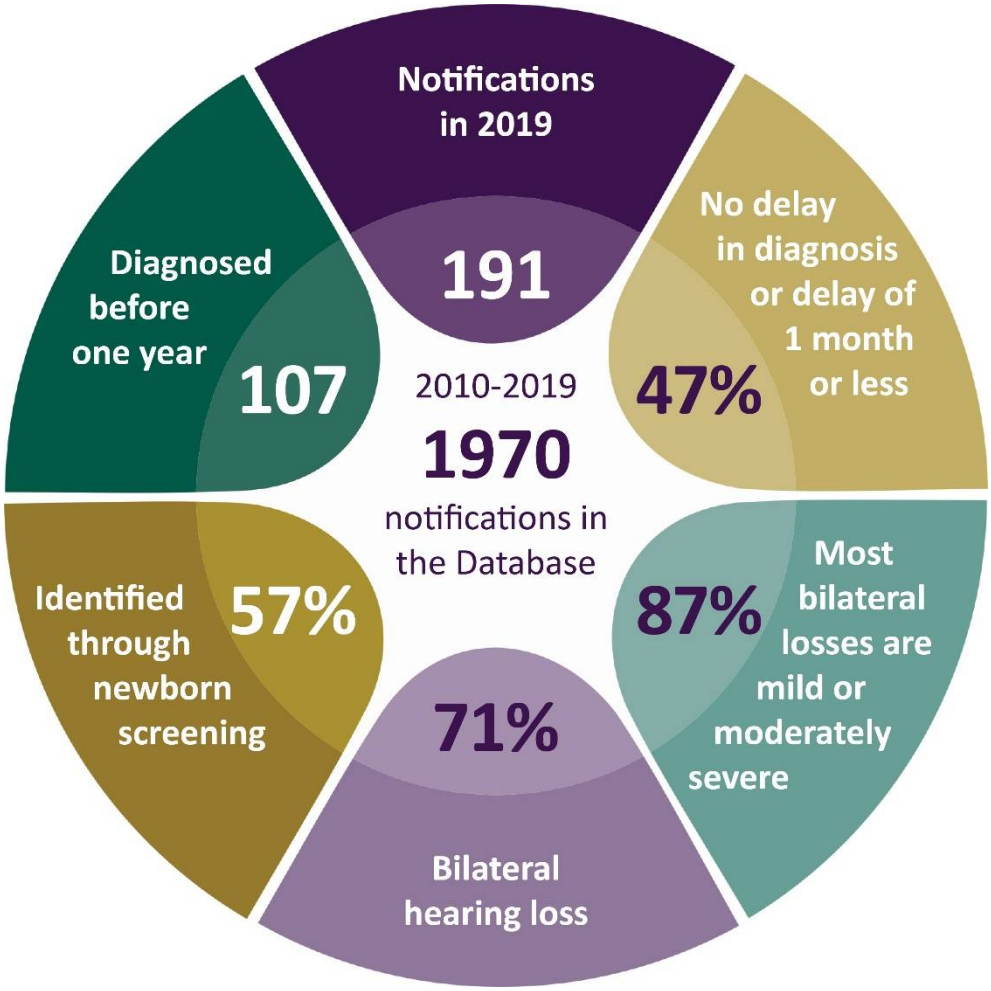
November 2020

Enable NEW ZEALAND

Summary

Whakarāpopoto



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

Contents

Ngā Ihirangi

The Deafness Notification Database

Te Pātengi Raraunga Whakamōhiotanga Turi

- **The Deafness Notification Database (DND) was established in 1982 to collect information on children and young people under the age of 19 who have been diagnosed with permanent hearing loss.**
- **After a hiatus from 2006, the Database was relaunched in 2010, and since that time has included those children and young people 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.**
- **These data have helped us understand more about those diagnosed with hearing loss in New Zealand and the nature of hearing losses being diagnosed; this, in turn, is being used to inform those who are newly diagnosed and their families, help researchers and assist with resource allocation.**

Introduction

Nau mai, haere mai ki te putanga tuangahuru o tēnei raupapatanga o ngā rīpoata ā-tau, e whakaahua ana i ngā whakaaturanga ki te Raraunga Turi o Aotearoa. Kei roto i tēnei rīpoata ngā raraunga mō ngā tamariki me ngā rangatahi i kohuratia i te tau 2019.

Welcome to the tenth in this series of annual reports describing notifications to the New Zealand Deafness Notification Database (DND). This report includes data for children and young people diagnosed during the 2019 calendar year.

The DND was established in 1982 and contains information on newly diagnosed permanent hearing loss among children and young people under the age of 19.

Where the parents (mātua) or caregivers (kaitiaki) provide consent for this information to be shared, audiologists and audiometrists from around the country send notifications electronically following diagnosis of a child or young person with hearing loss.

"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.

Please refer to Appendix A: Making notifications to the Database on page 65 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 1970 children and young people notified with a hearing loss diagnosed between the start of 2010, when the DND was relaunched, and the end of 2019[i].

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 Māori[ii] are more likely to have less severe and bilateral hearing losses than their European counterparts.

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 the European ethnic group refers to those of European ethnicity and includes New Zealand Europeans.

Recent notifications have also shown that there is a growing number of children being identified under the age of one year. This is pleasing as the earlier a child's hearing loss is identified the earlier intervention can be provided and/or monitoring can begin. This shift is undoubtedly 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).

This report and the DND generally exclude children with Auditory Processing Disorders. For those interested, comprehensive *New Zealand Guidelines* were published in 2019[1].

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, longitudinal comparisons are not always possible[ii].

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

- *History of the Database and changes to the inclusion criteria* - see Appendix B: History of the Database, *on page* 66.
- *Terminology used in this report to describe hearing losses* - see Appendix E: Terminology used in this report, *on page* 69.
- *The completeness of notifications* – see Appendix C: Completeness of notifications, *on page* 68.
- Commonly used terms can be found in the Glossary, which begins on page 71 of this report.

Acknowledgements

Our sincere thanks to the 191 parents (mātua), caregivers (kaitiaki) and young people (rangatahi) who consented to share details of their child's/their own hearing for the Database.

As a result of this willingness to share basic diagnostic information, 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 and to do this in such a careful and considered way is greatly appreciated. It is clear from how this is done, including by departments which are under-staffed and under considerable strain, that diagnosing clinicians care deeply about the wellbeing of both their patients and whānau.

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 relaunched 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 2010-2019:

- 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.

Contact details

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

Feedback from that survey resulted in a number of changes to the 2018 report, including the addition of key points at the beginning of each section.

Further feedback on this report is welcome. Questions and feedback about the DND reports should be directed to its primary author, Janet Digby. Janet can be contacted by email here.

Notifications

Ngā Whakamōhiotanga

- **Notifications were made for 191 children and young people diagnosed during 2019, 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. Fifteen percent of tamariki (children) and 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 (71%) with the rest being for unilateral hearing loss. Almost a third of children and young people diagnosed with a unilateral hearing loss in 2010 had a bilateral hearing loss by 2018.**
- **Research suggests that, as with more severe hearing losses, mild and unilateral hearing losses (UHL) are also associated with poorer outcomes.**
- **Māori are more likely to have bilateral hearing losses and more mild and moderate hearing losses than their European counterparts. Māori also have more 'mixed' hearing losses and less permanent conductive losses than their European counterparts.**
- **One in five of those whose information was notified to the DND have an immediate family member with a permanent hearing loss.**

General information

One hundred and ninety-one notifications pertaining to cases first diagnosed during the 2019 calendar year, and meeting the criteria for inclusion, were received by 13th March 2019, this year's cut off for new notifications[i, ii]. There are now 1970 cases included in the main dataset.

These notifications were received from a total of 65 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].

i Reports prior to 2006 contained information about diagnoses notified in each calendar 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 included in the dataset on which most of this report's analysis is based.

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. With 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.

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 tamariki and rangitahi with hearing loss.

Because only a small number, and likely a small proportion of cases met the criteria for the high frequency category in previous years[2], we have not described this group in this year's report, and we will not be seeking these notifications in future.

Number of notifications

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

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

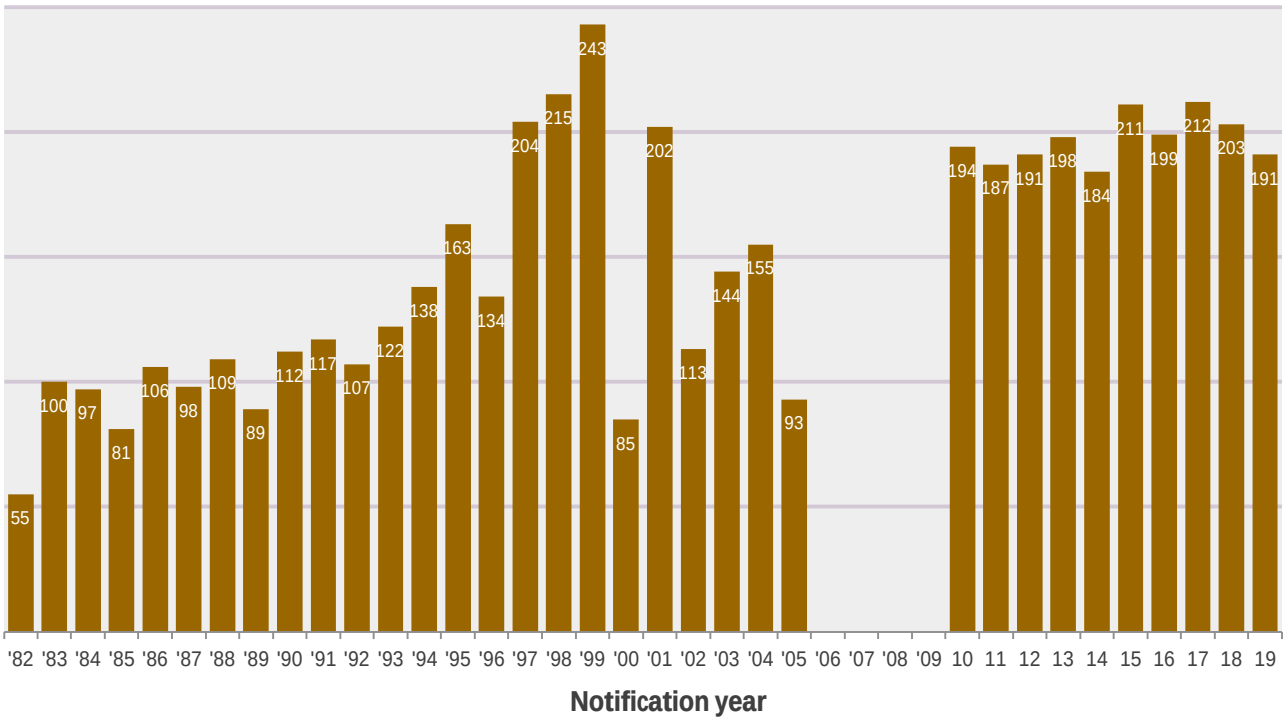

Figure 1: Notifications by year 1982-2005 and 2010-2019

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 that 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. Reports from 2010 show the total number of notifications that met criteria for inclusion that had been received by the cut-off date each year, in the March following the calendar year for each report.

iv 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.

v Data from 2010 to 2019 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.

Gender

Of the 1970 cases (2010-2019) contained in the main dataset, 45% of these are listed as female (n=884) and 55% male (n=1086). This represents a ratio of 1: 1.23[i].

This gender difference was particularly noticeable in 2016 notifications with only 39% of notifications recorded as female, and 61% male[ii].

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].

Hearing Australia's data on those under the age of 21 who have hearing aids or cochlear implants[iii]. show a similar pattern, with higher numbers of hearing loss among males (52.2%) than females (47.8%) in 2019[5, iv].

This pattern is seen in all Australian states in 2019, except for South Australia and ACT, in which the ratios of male to female is almost 1:1, and for those aged 21-25 years of age, for which fewer than half of cases were male (45.4%)[5].

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 2010-2019 period. During that time, an average of 5% of children and young people notified have been born overseas, with the birthplace of an additional 6% being uncertain.

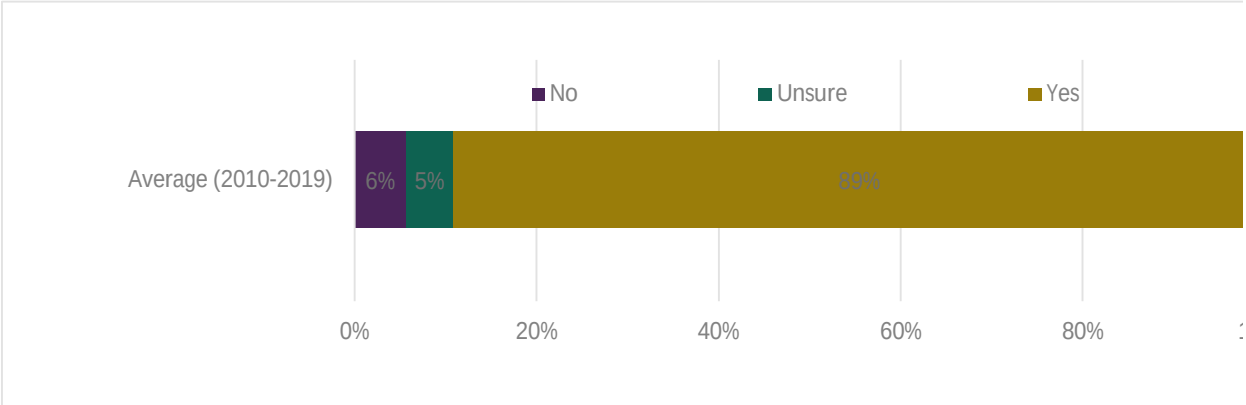

Figure 2: Proportion of cases born in New Zealand (2010-2019)

i 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.

ii Historical figures change slightly from previous reports as late notifications are added to the Database.

iii This source reports on children and young people, under the age of 26 who received services from Australian Hearing (now Hearing Australia) in 2014.

iv 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 at least in part 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 191 notifications to the Database in 2019, 3% were known to be born outside New Zealand, with birthplace listed as uncertain in a further 1% of cases.

DHB representation

Table 1 contains the percentage of 2019 notifications from each DHB and compares these with the percentage of the population under the age of 20 from the 2018 Census[i].

The third column in that table shows the percentage of notifications received for 2010-2019 from each district health board – this can be compared with their relevant percentage in the population for those under the age of 20[ii].

Please note that this table in last year's report mistakenly contained 2016 percentages in the first column and, therefore, incorrect figures in the third column.

Tamariki notified to 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)[iii].

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

- the size of each DHB population within the age range for the Database;
- 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 These schools have now been replaced with a single national entity; Ko Taku Reo.

District Health Board	Percentage of notifications received in 2019 (under 19 years)	Percentage of population under the age of 20 (Statistics New Zealand, 2018 Census[7])	Percentage of notifications received 2010-2019 (under 19 years)
Auckland	4%	9%	7%
Bay of Plenty	7%	5%	7%
Canterbury	11%	11%	13%
Capital and Coast	12%	6%	9%
Counties Manukau	12%	13%	13%
Hawke's Bay	6%	4%	3%
Hutt Valley	5%	3%	4%
Lakes	2%	3%	2%
Midcentral	3%	4%	3%
Nelson Marlborough	6%	3%	4%
Northland	5%	4%	6%
South Canterbury	2%	1%	2%
Southern	7%	6%	6%
Tairāwhiti	2%	1%	2%
Taranaki	1%	3%	3%
Waikato	10%	9%	8%
Wairarapa	3%	1%	1%
Waitematā	5%	13%	6%
West Coast	1%	1%	1%
Whanganui	0%	1%	1%

Table 1: Percentage of notifications (2019) compared with the estimated percentage of population under 20 years of age by district health board (2018 Census) and the proportion of notifications by DHB (2010-2019).

A recent New Zealand research project, aiming to improve understanding of what happens to children and young people following diagnosis, requested follow-up information from notifying clinics seven years following the initial diagnosis and notification of a hearing loss to the DND.

At least some data was received on 163 of the 194 children and young people notified to the DND in 2010 (84%)[i]. This study found that only 56% of children/young people were still in the care of the notifying clinic (often the district health board's audiology service) 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.

Seventy-two of the 163 children and young people (44%) notified to the database in 2010 were no longer in the care of the notifying clinic by 2017. Of these, the notifying clinic:

- had no information about fifty-nine children and young people;
- had some information on where twelve children and young people had moved to or who was now looking after them; and
- had not seen two of the children since the original diagnosis in 2010.

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.

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 person 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 that is searchable) while others do not.

These figures demonstrate the importance of clinic information systems and communications between clinics to ensure tamariki are not lost to follow-up.

Additional disabilities

Introduction

Increasing estimates of the global burden of childhood disability[i] (Olysanya *et al.*, 2020), suggest that more than one in 10 children and adolescents are affected by epilepsy, intellectual disability, vision or hearing loss. When other conditions such as developmental delay and cerebral palsy are included this figure will increase[8].

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.

As outlined in Nelson and Bruce's 2019 review paper on this topic (2019)[9]:

- the population of children and young people who are hard of hearing and who have one or more additional disabilities are difficult to characterise due to the range of conditions included and the type and severity of the various disabilities;
- specific aetiologies including hereditary syndromes, maternal infections, prematurity and meningitis indicate a higher likelihood of specific 'concomitant' disabilities, including those which are intellectual or developmental, Autism Spectrum Disorder, learning disabilities, ADD, ADHD, emotional disabilities, speech and language impairments and vision issues;
- individual children may have one or several disabilities and each can vary in both presentation and degree;
- the presence of disabilities makes 'compensation for loss of hearing much more difficult';
- early identification has been found to positively impact outcomes across domains of children with additional disabilities although it is common for these children to begin to receive early intervention at later ages than those without additional disabilities; and
- there is a great deal yet to be discovered about prevalence, how to accurately diagnose and assess progress in young people in this group and provide them with optimally effective interventions.

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

i Children with such additional disabilities are sometimes referred to as being 'deaf plus' or Deaf with Disabilities (DWD). The authors of this report are yet to come across a term that is inclusive given the broad range of conditions and differences that are included in this section. Suggestions for a better term are most welcome.

Overseas 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 2. 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, and because our data showing the proportion of children with an additional disability are 'point in time' figures from the time of the hearing loss diagnosis.

Outcomes

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].

Source	Date	Location	Details	Rates
Nelson and Bruce[11]	2019	United States	Review paper	25-51% of d/Deaf or hard of hearing (DHH) students in the United States, with higher rates reported among those with severe and profound sensorineural hearing loss (SNHL)
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 United Kingdom, United States, 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	United Kingdom	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	United Kingdom	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis[16]	1997	United Kingdom	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 United States children with a hearing loss had an additional disability

Table 2: Additional disabilities, selected overseas rates for comparison

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[18].

DND data

A wide definition of additional disability is used within the DND. Of the 1970 records in the Database for 2010-2019, the majority (78%) have no 'additional disability'. Fifteen percent have a confirmed additional disability and a further 9% are listed with a possible although as yet unconfirmed additional disability. Just 1% of cases (n=23) contained no data on whether an additional disability was known to be present. 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.

Additional disability	Number of cases	Percentage
Yes	226	11%
Unsure whether AD exists, no confirmed diagnosis	181	9%
No additional disability	1540	78%
No data	23	1%
Total	**1970**	**100%**

Table 3: Proportion of cases by additional disability status (2010-2019)

2019 data

Of 2019 notifications, 15% of children and young people were known to have one or more disabilities *in addition* to their hearing loss at the time the notification was made. In a further 9% 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 2019 with either a confirmed or possible additional disability (at the time of diagnosis) has increased to levels not seen since 2012.

Comparison with previous data

The proportion of tamariki notified with one or more additional disabilities is not directly comparable to data reported prior to the 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 4 shows the total proportions of confirmed and unconfirmed 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 tamariki and those who live in poverty[19].

Nikki Turner, director of the Immunisation Advisory Centre noted in early 2020 that "There are two reasons why we are having coverage problems. The first is the historic immunity gaps particularly in adolescents and young and mid-life adults. The second is lower coverage in our infant immunisation programme, particularly for tamariki Māori and children from low-income families."[20]

Previously, the authors of this report believed that the earlier identification of tamariki with hearing

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.

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) and total confirmed and possible (2010-2019)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	11%	10%	21%
2011	13%	5%	18%
2012	14%	11%	25%
2013	11%	11%	22%
2014	13%	8%	21%
2015	9%	10%	19%
2016	7%	10%	17%
2017	10%	8%	18%
2018	11%	9%	20%
2019	15%	9%	24%
Average 2010-2019	**11%**	**9%**	**20%**

Table 4: Proportion of cases with a known additional disability (2002-2019)

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. 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 immunisation coverage, particularly between 2007 and 2013[i, 21] and that tamariki with hearing loss in New Zealand are not all routinely assessed by a paediatrician.

More recent notifications to the DND (shown in Table 4) suggest the general downward trend from 2012-2016 may have reversed. There are a number of possible contributing factors to changing data and it is not possible at this time to determine the cause of this change.

Most common types of additional disabilities

There is a wide variety of reported conditions contained within notifications, including those related to a specific syndrome, cerebral palsy, general or global developmental delays, intellectual disability, and vision problems[22, ii]. Some children and young people have more than one 'additional disability' listed on their notification form.

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.

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 developed a new codeframe to group these responses by type and applied this to all records, as seen in Table 5.

This table shows a series of types of disability, a description of 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	61	27%
Medical	Medical conditions and issues, such as cardiac problems, bladder issues, renal issues and lung issues.	58	26%
Neurodevelopmental	Issues with the growth and/or development of the brain or central nervous system, such as ADHD, autism, developmental delays and intellectual disabilities	59	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	42	19%
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	32	14%
Medical-developmental	Medical conditions and issues related to development such as hydrocephalus and cleft palate	17	8%

Table 5: Number of cases by type of additional disability (2010-2019)

Table 6 shows the number of children/young people who are listed as having each additional disability code. For example, those listed with two additional disability codes include 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.

Rates of additional disabilities and the effect of age at diagnosis

When we examine cases of hearing loss diagnosed among children under and over the age of two years there is a clear difference in the proportion with confirmed additional disabilities. Those over the age of two at diagnosis have almost double the rate of confirmed additional disabilities when compared with their peers who are diagnosed under the age of two, as shown in **Error! Reference source not found.**.

This difference is likely to be due to the time it takes to confirm additional disabilities and because these conditions may take time to become noticeable to caregivers and medical professionals. For example, in a child whose hearing loss is identified as a direct result of universal newborn hearing screening, this may be the first condition that has been identified.

Number of additional disability codes / category	Number of children/young people	Percentage
No additional disability	1545	78.4%
One additional disability code	185	9.2%
Two additional disability codes	31	1.6%
Two additional disability codes	6	0.3%
Three additional disability codes	1	0.1%
Unconfirmed AD	181	9.2%
No data	21	1.1%
Total	**1776**	**100%**

Table 6: Number of cases by number of additional disability code types (2010-2019)

Bilateral and unilateral loss

Proportions of unilateral and bilateral hearing losses in the Database

The proportion of 2010-2019 cases that were thought to be bilateral/unilateral was 71:29 (see Figure 3, below)[i].

Other influences

Immunisation coverage (including for conditions such as mumps) in New Zealand rose significantly from 45% in 2007 to 92% in 2012[23]. More recently, concerns about falling immunisation rates have been raised, with particular concern expressed about rates for Māori and those living in poverty[19].

The number of cases resulting from changes in immunisation are likely to be small, and so the impact on numbers of cases of hearing loss diagnosed that have been notified to the Database will likely not be visible.

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[24].

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

i From 2015 these reports have contained data for cases that contained completed audiometric data for all eight datapoints, as well as data for those which are interpolated. The interpolated data includes a good deal more cases (1970 vs 1235) and so we will focus on this figure from now on in these reports, as it is likely to be a more accurate reflection of all rangitahi contained in the Database.

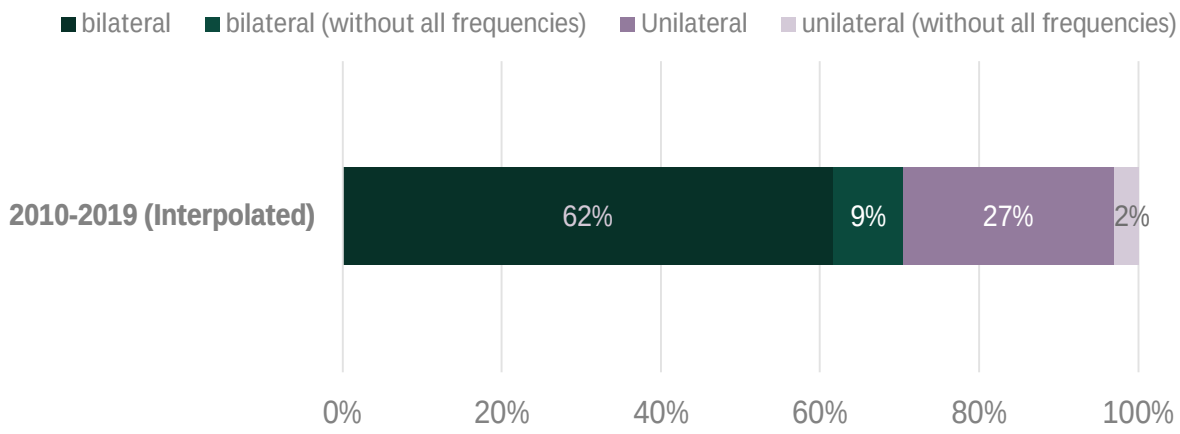

Figure 3: Proportion of bilateral and unilateral cases (2010-2019)

Unilateral hearing losses

Unilateral hearing loss 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[25, 26].

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 commonly minimised the implications of unilateral hearing loss in children[27, 28, 29].

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[30, 31, 32, 33, 34].

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[35].

To reflect the now 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].

Bagatto *et al*.[36] completed a review paper in 2019 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.

A New Zealand study 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.

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.

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[37].

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[38]. Prevalence rates rise with age to between 3.0 and 6.3% among children 6-19 years of age, according to Ross *et al.*[39]

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[40, 41, 42].

Here in New Zealand, a recent analysis of data provided for 163 of the 189 notifications to the DND in 2010[43], described in last year's report showed that 32% of those children or young people with a unilateral hearing loss ended up with a bilateral hearing loss by the time the follow-up data was provided. This is not easy to characterise as not all children and young people's data pertained to 2017/2018; some data provided related to information collected much earlier than that, at their last appointment with the clinic, for example.

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.'[44] 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.

A supplement was produced in 2013 stating 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[45].

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[46].

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[47].

Management

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 2019 review[36, i].

Single sided deafness

Background

Severe or profound unilateral hearing loss can be referred to as single-sided deafness (SSD). This category is effectively a subgroup of the unilateral hearing loss category referred to elsewhere in this report.

Different case definitions for 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[48, 49]. The boundaries for these

i 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.

degrees of loss also differ depending on the jurisdiction.

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[50].

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].

Notification Year	Proportion of cases with single sided deafness
2010	4%
2011	4%
2012	7%
2013	10%
2014	8%
2015	6%
2016	5%
2017	6%
2018	4%
2019	4%
Average 2010-2019	6%

Table 7: Single Sided Deafness Cases by Year (2010-2019)

DND data

The proportion of 2010-2019 unilateral hearing loss cases[ii] which met the DND's criteria for SSD is 21%.

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

Types of hearing loss

A question about the type of hearing loss was added to the notification form part way through 2013. This asks audiologists to describe the type of loss in each ear[iv]. Options provided are: 'sensorineural', 'mixed', 'permanent conductive', 'normal hearing[v]', 'other' and 'don't know'.

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

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 16% in the left). See Figure 4 for full detail. Please note that the cases with normal hearing in one ear relate to those children and young people with a unilateral hearing loss, indicating they have normal hearing in one ear.

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[51].

i These average thresholds have been chosen considering the ASHA (American Speech-Language-Hearing Association) codeframe for severity, 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 hearing 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.

iv 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.

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

Among those from the Avon newborn hearing screening programme in England[52] 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.

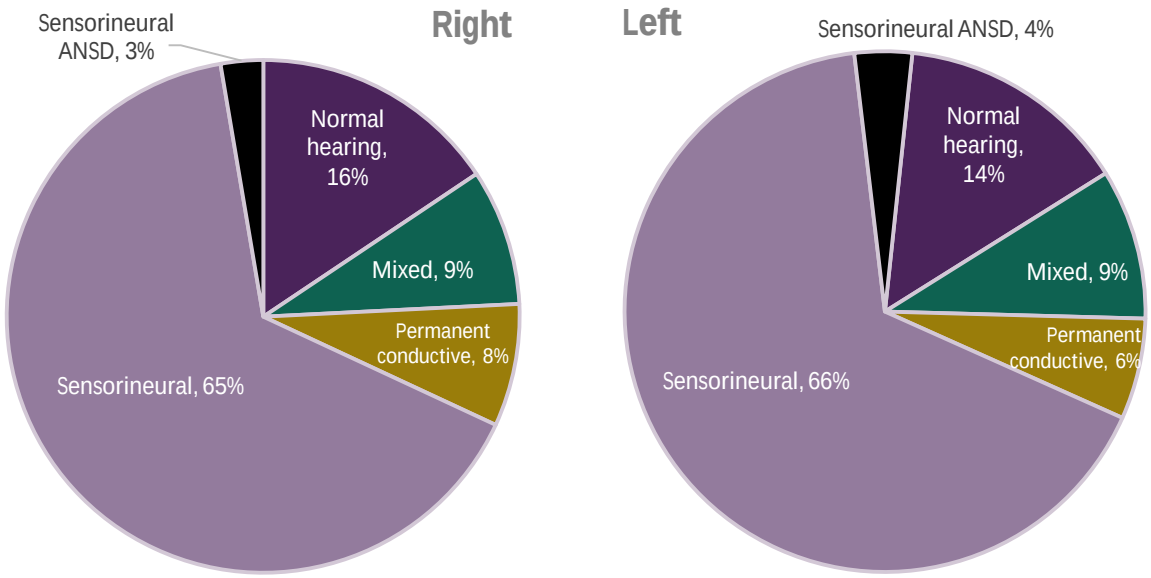

Figure 4: Type of hearing loss (2010-2019)

An analysis of the types of hearing loss among 2010-2016 notifications, included in a previous report,[53] 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)[i].

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.

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 had not been 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?[ii] (or had a permanent hearing loss if they have died).' The results for this question are shown in Figure 5.

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

ii The DND reports prior to 2005 showed 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 in an attempt to gain more specific responses about the nature of the family history.

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

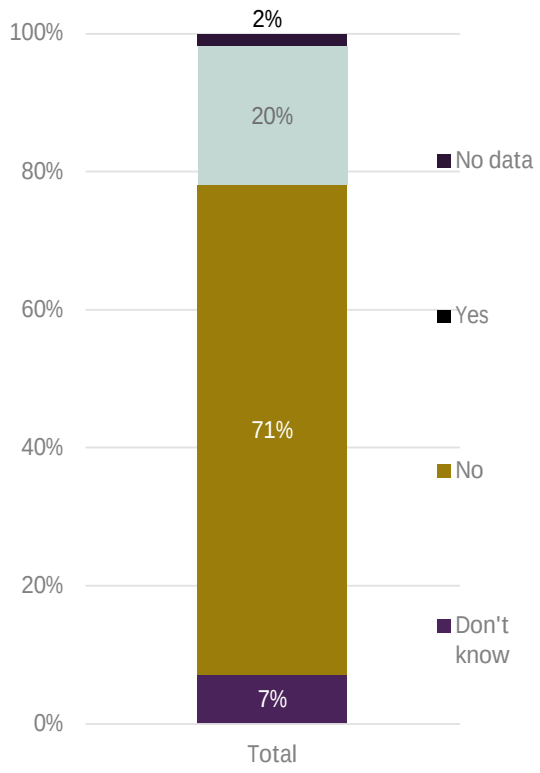

Figure 5: Immediate family member with hearing loss (2015-2019)

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 2015-2019 in Figure 5.

Full dataset

Figure 5 shows data from 2015-2019 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 64 to 78% during that time, with between 17 and 22% listed as having one.

Ethnicity

Mātāwaka

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

Representation

Background

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

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 a participant may 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 for ethnicity, allowing only one ethnicity code per participant.

For further information on ethnicity coding in the Database, please refer to *Appendix B: Notifications and ethnicity*, on page 66.

Full dataset

Of the 1970 notifications in the main dataset (covering 2010-2019 notifications) all but 24 (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 0.52% in 2017-2019.

The majority of notifications (89%) contain one code, and a smaller proportion (8% and 1%) contain two or three codes respectively.

Multi-coded 2018 Census data is included for comparison in Figure 6 for the first time. As individuals may identify (or be identified by their parents) with more than one ethnicity, the totals add to more than 100%. This figure shows the total response count for ethnicity from the 2018 Census (for those under the age of 20) and compares this to the ethnicity breakdown for

i The term European is used in this report to mean all those of European descent. However, the vast majority of notifications to the Database are for those born in New Zealand and can be considered New Zealand European, rather than having been born in Europe.

ii Ideally, we would like to ask notifying clinicians to provide more detailed information on ethnicity, but ethnicity coding is not that easy to get right without training and as we are relying on the help of these clinicians to provide notifications we don't want to make notifying cases more onerous than they already are.

iii The MELAA category relates to people 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.

deafness notifications from 2010-2019, which includes those under the age of 19[i].

The European ethnic group was still the largest in the Census by a significant margin. The proportion associated with this group is static when compared with the 2013 Census.

Those of Māori ethnicity are over-represented in the Database, comprising 32% of notifications and 26% of the population overall.

Pacific Peoples are under-represented in the Database, with 14% of the population and only 9% of notifications.

Last year, those listed with Asian ethnicity were also over-represented in the data, with 18% of notifications coming from this group, while only 12% of the population categorised as 'Asian'. With the new 2018 Census data, we can see that the proportion of notifications from this group is roughly proportionate with the size of this population among under 19s[ii].

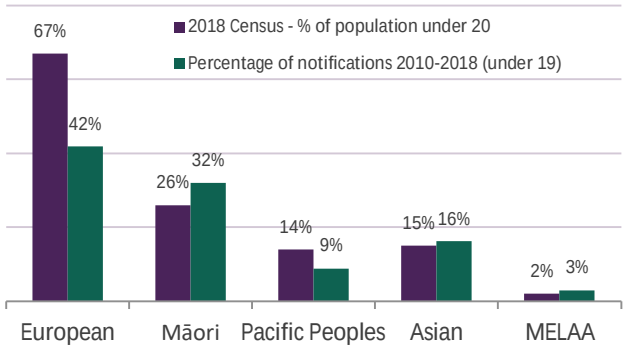

Figure 6: Notifications by ethnicity (2010-2019) compared with Census data (2018)[54]

Unilateral and bilateral hearing losses

Of 2010-2019 cases, including those with inter-polated audiometric data, 71% are recorded as bilateral, while the remaining 29% are unilateral.

Figure 7 shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group during the 2010-2019 period. 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 also on analysis of the now larger sample) supports the conclusions from the 2014 paper by Digby *et al.*, which found a higher proportion of bilateral hearing losses among young Māori when compared with their European counterparts[64].

This difference can also be seen when comparing bilateral losses among Māori tamariki[iii] notified between 2010 and 2019 (79%), with those who are European[iv] (66%), and those described as both Māori and European (73%)[v].

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

Hearing loss present at birth

Of all 2010-2019 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 42% of cases, with the hearing loss likely to have been present at birth in 44% and unlikely to have been present at birth in 14% of cases.

Analysis of 2010-2016 cases described 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.

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

ii Those in this ethnic group comprise 15% of the population and 16% of notifications.

iii 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.

iv European refers to an ethnicity of which individual children or young people are predominantly of European descent; that they or their forebears originated in Europe.

v These figures now include interpolated data, and those whose hearing loss was bilateral without all datapoints included on the notification form.

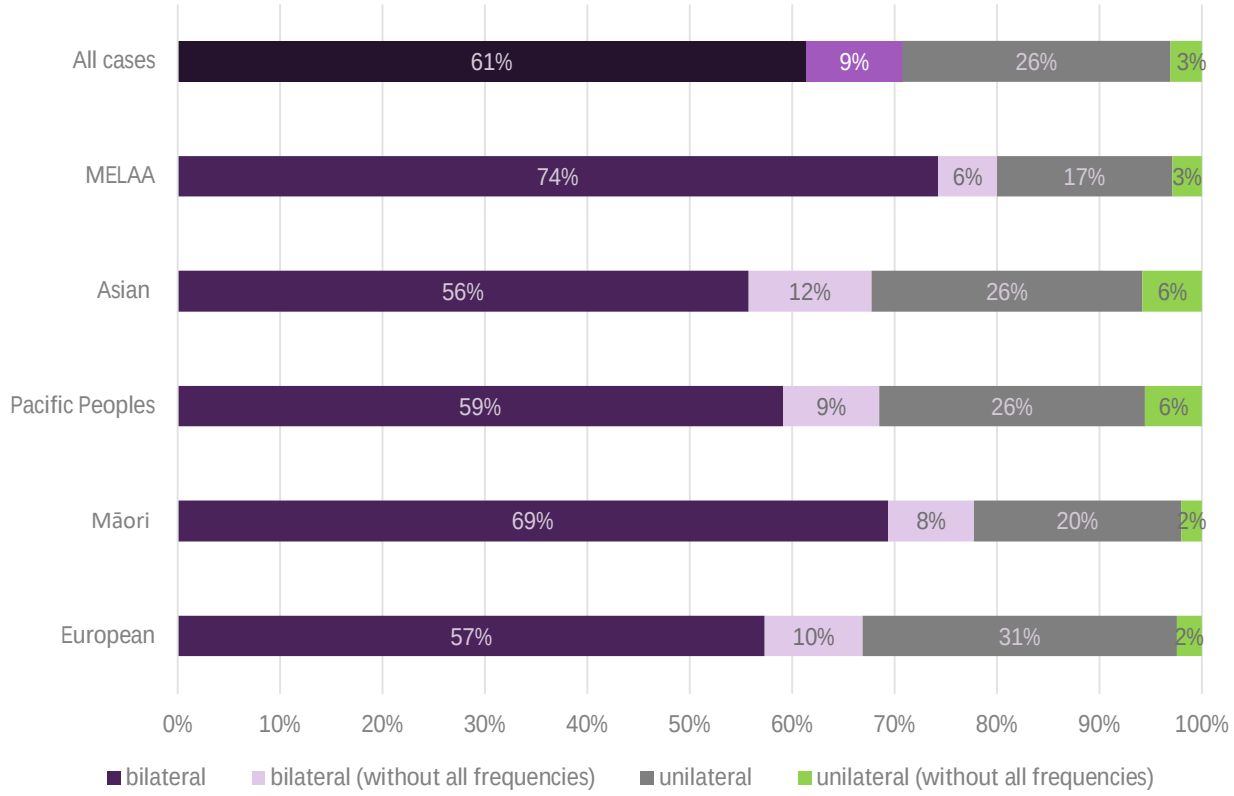

Figure 7: Proportion of unilateral and bilateral hearing losses by ethnicity (2010-2019) based on interpolated data and manual checks to determine bilateral/unilateral status

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).

Hearing loss among Māori

Prevalence

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

As mentioned above, the proportion of notifications from those of European ethnicity are considerably lower than one would expect based on the size of their population, 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:

- Whakarongo Mai (1989) concluded that while the full extent of hearing impairment among Māori was not known because of information gaps, "a number of local and detailed studies demonstrate convincingly that hearing loss occurs excessively among Māori people" [55].
- Greville (2001) found higher prevalence of temporary and permanent hearing loss among Māori children[56].
- 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[57].
- Household Disability Surveys -
  - 1991-2006 Surveys[58] suggest Māori had higher rates of hearing disability

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

(tamariki and adults) and higher rates of unmet need for technology and equipment when compared with non-Māori[59]. (For information about the limitations of this data please see the 2011 DND Report[60].)

  - The 2013 Survey continues to suggest Māori had higher unmet need for technology and equipment when compared with non-Māori[61] but also that they now have *lower* rates of hearing disability compared with their European counterparts[62], although this seems to relate to the lower age profile for Māori (younger people have fewer disabilities).
  - No Disability Survey was completed in 2018, with the Māori Social Survey being completed following the 2018 Census and alternating with the Disability Surveys after subsequent Censuses[63].
- 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[64].
- B4 School Check data:
  - Data from the B4 School Check[i] analysed by Searchfield *et al*. (2011), show higher rates of referral from hearing screening for Māori tamariki (9%) compared with non-Māori (5%)[65] and this pattern has continued with 2018/19 data showing 3.5% European children referring on their hearing screen, compared with 7.1% of Māori children. 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%[66]. Post-screening diagnostic results are not available. Rates were similarly high for Māori when compared with Europeans since 2010/11.

Reasons for under-representation

Despite a good number of 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 - it is probable that less severe (especially mild) hearing losses are less likely to be identified; and
- disparities in access to, and within, the health system[67] suggest fewer cases may be found or notified when compared with those in the European population.

The risk of underrepresentation is higher for older Māori children and young people whose hearing was not screened as newborns and for those children and young people who develop a hearing loss after birth.

It is worth keeping in mind that screening programmes, including New Zealand's Universal Newborn Hearing Screening and Early Intervention programme (UNHSEIP) do not target or identify all mild hearing losses[ii, 68]. The B4 School Check targets mild and greater hearing losses[69].

Unequal access and outcomes

The health status of Māori, as with other indigenous populations, has been undermined by New Zealand's colonial history which has seen resources taken from Māori, and further marginalisation through cultural oppression and the introduction of new social systems based on European norms and values[70, 71, 72, 73, iii].

Disparities documented in many areas of health demonstrate Māori have poorer access 'to, and

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

ii "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.

iii Such causes are not dissimilar to those reported by indigenous peoples in other countries. An introduction to this topic can be found in King *et al*'s 2009 paper in The Lancet.

through' the health system[67, 74, 75], that they receive a poorer and slower service, and are less likely to receive appropriate levels of care[76], resulting in poorer health outcomes.

Despite relatively strong national policy frameworks recognising Māori health needs and engagement in health, these frameworks have not been successfully implemented[77] and there are signs that 'engagement with and recognition of Māori' have recently been dismantled[77, 78, 79].

Both the Waitangi Tribunal 2575 inquiry (Stage One)[80] and the New Zealand Health and Disability System interim report[81] identified the ongoing failure of the Crown to deliver health equity for Māori and called on the Crown to abide by its obligations under te Tiriti o Waitangi/the Treaty of Waitangi[i]. The Treaty guaranteed Māori their full rights and benefits as citizens.

The Tribunal's Stage One report acknowledges that while the health sector is not able to influence all the social determinants of health, persistent inequalities constitute health sector Treaty breaches. It recommends that the principles derived from te Tiriti by the Royal Commission on Social Policy *(partnership*, *participation* and *protection)* be extended to include *equity* and *options*. It also asserts DHBs and other health agencies are not doing enough to reduce inequalities.

A recent review (2020) of two decades of qualitative research into the experiences of Māori within the public health system categorised barriers into three groups: organisational structures, staff interactions and practical considerations[82].

A number of district health boards have in recent years reasserted their commitment to achieving equity for Māori, including Northland District Health Board[83] and Auckland District Health Board,[84] which referenced the important role of eliminating institutional racism in achieving equity.

Hearing service disparities

There has been limited research on inequalities within hearing services.

Thorne *et al.* (2008) found considerably lower rates among ACC claims for Māori (and Pacific Peoples) relative to New Zealand Europeans, despite the overrepresentation of these groups in industries where noise exposure is higher, and a higher prevalence of hearing loss overall[85].

An article by McCallum *et al.* (2015) in the *New Zealand Medical Journal* examined 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[86].

The latest data from the Atlas of Healthcare Variation (Surgical Procedures) suggests that grommet insertion rates are low in some areas compared with the national average, particularly in 0-4 year old Māori and Pacific children. (It is worth noting there are differing views about the efficacy of grommets as a treatment for middle ear disease. Regardless, it is unlikely that differences in otologic treatment practices would be applied based on ethnicity.)

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[57].

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

Such investigations are particularly important as there is no service specification for audiology services nationally, meaning that services offered by district health boards[ii] differ, as do waiting times.

i A summary of policies and legislative statutes that underpin government's commitment to Māori, including within health, and those in selected other countries with indigenous populations can be found in Ferdinand *et al*. (2020), which can be found in the references of this report.

ii DHBs see most tamariki and rangatahi with hearing loss.

Deprivation

Pōharatanga

- **Deprivation scores are drawn from the latest 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 of higher deprivation than those without. This pattern is consistent with data from the United States and from regional comparisons based on income. However, no such correlation exists in the United Kingdom, where disability allowances are much higher.**
- **Our DND data shows that 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 ethnicities.**

Overview

International data demonstrates that rates of congenital hearing loss are higher for those living in developing regions and lower in countries with higher incomes. Lower levels of hearing loss in higher income countries are thought to be due to lower infection rates and better access to preventative measures and healthcare services[87].

The New Zealand Child Poverty Monitor notes 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[88].

Tamariki with disability and deprivation

Overall, 11% of children under the age of 15 have a disability. In New Zealand, Māori and Pacific have higher than average disability rates once adjustments are made for differences in age profiles by population[89].

Child Poverty Action Group (2015) report that children with disabilities in New Zealand are at increased risk of living in low-income households[90].

This pattern is also found in the United States, where Boss *et al.* (2011) evaluated disparities in socio-economic status among hearing impaired children nationwide through the 1997-2003 National Health Interview Survey. It found that families of children with hearing impairment live closer to the poverty level and less frequently use some medical services[91].

However, Child Poverty Action Group also note that such differences are not inevitable and cite the United Kingdom's much higher disability allowances, which they see as the reason that there is no correlation between childhood disability and poverty there[92].

Introduction to NZDep

Here in New Zealand, we are fortunate to have Deprivation 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 that are the least deprived, and scores of 10 allocated to the 10% of areas that are the most deprived[93]. 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 does not relate to the individual's specific level of deprivation.

Deprivation data provided by the Ministry of Health has been included in our DND analyses since the 2016 report. Data for this report is based on NZDep2013 as NZDep2018 does not currently have domicile code mapping.

Of the 1970 tamariki in the main dataset, 97% had deprivation data available. Data were unavailable for tamariki whose: NHI was not valid, those who had no NHI listed and who live outside New Zealand. For those whose NHI was not valid or missing, NIHIs were sought but not provided, or not provided until after the analysis was completed.

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

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 when compared with the population at large.

Tamariki who live in the most deprived areas are also 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 founders of the New Zealand Deprivation Index kindly shared data on the national deprivation distribution (NZDep2013) of tamariki

i As at the date of extraction.

in relevant age groups, so we could compare this with the distribution for children and young people whose diagnosis was notified to the Database[94].

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[iii].

Figure 8 shows the distribution of cases by deprivation status with splits by ethnicity. 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.

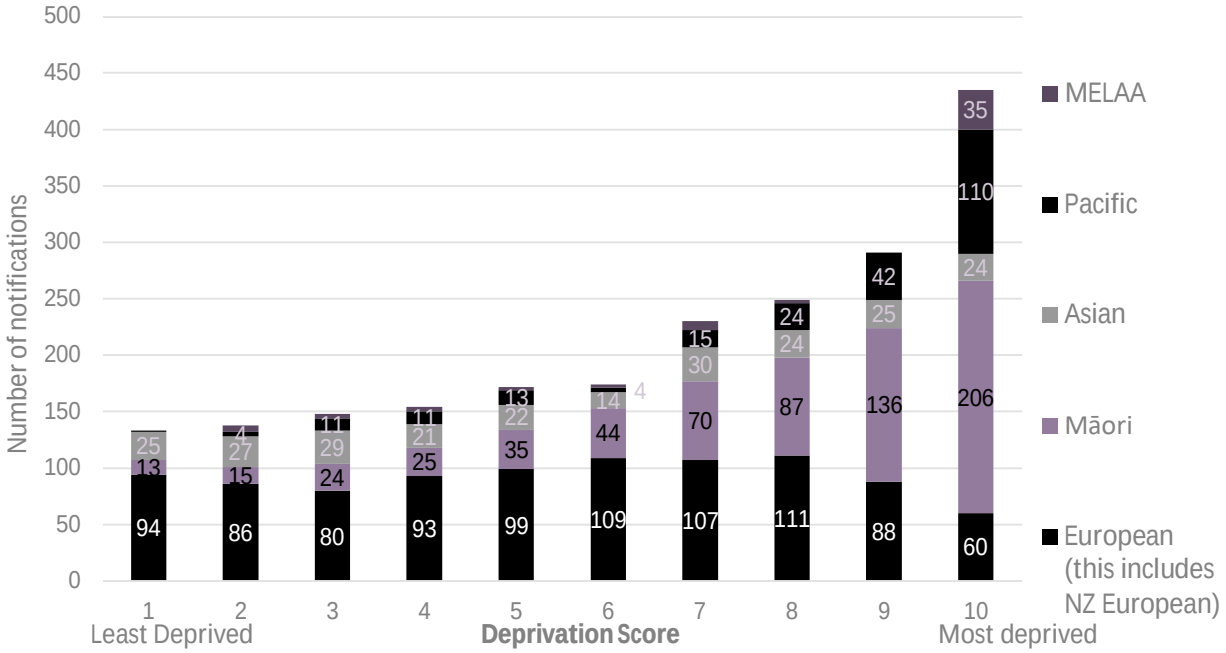

Figure 8: Deprivation scores (NZDep2013) of tamariki in the DND by ethnicity (2010-2019)[iv]

To further illustrate the differences between ethnic groups in the Database we can examine proportion of tamariki who are living in the most deprived 30% of areas (with scores of 8-10 on the scale), the middle 40% (with scores of 4-7) and the least deprived 30% (8-10).

As shown in Figure 9, 65% of Māori children, 75% of Pacific children and 58% of MELAA children in the Database are living with their whānau in the most deprived areas, compared with only 28% among Europeans.

Implications

These data demonstrate that audiologists and other hearing professionals working with young people who are hard of hearing are likely to see a high proportion of families living in 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 A logistic regression was conducted for 2010-2016 notifications to see whether a linear or non-linear relationship existed between tamariki having other known disabilities and level of deprivation. No association was found (p=0.7801).

iv 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.

Figure 9: Grouped deprivation scores by ethnicity (2010-2019 cases)

Professionals should keep in mind that income and poverty are significant determinants of health[95]. As a result, the families they see are more likely to experience poorer overall health[95] (including greater barriers to accessing health services[96] and lower housing stability[97]) and higher rates of stress and mental health issues among both adults[98], young people and children[99, 100] than those in less deprived areas.

They also indicate that the majority of families in areas of high deprivation will be of Pacific, Māori or MELAA ethnicities.

Aetiology

Ngā pūtake

- **The aetiology of hearing loss is either genetic or non-genetic in nature. The proportion of hearing losses which have 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 91% 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 29 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[101]. 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[102].

The proportion of hearing losses with a confirmed genetic cause is increasing over time[103, 104], 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[105].

'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[106].

Non-genetic aetiologies resulting in an early onset of hearing loss include prematurity and infections during pregnancy, such as cytomegalovirus (CMV). 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[106].

In tamariki, mumps is thought to be the most common cause of unilateral acquired sensorineural deafness and is usually sudden in onset and profound in severity[107].

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[108].

[i] The term 'genetic defects' is used in the paper referenced 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[109].

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[110].

New Zealand data

A little over 99% of the 1970 records in the dataset (2010-2019) 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, 91% are of unknown cause, with the remaining cases listed as having a known cause.

When we examine these data by ethnicity, we can see that 14% of those listed as European have a known aetiology, compared with 10% of Māori and 10% for Pacific Peoples. For each of these groups, as with the total, the proportion with a known aetiology is dropping over time, presumably the result, at least in part, of reducing average ages of identification.

As seen in Figure 10, 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.

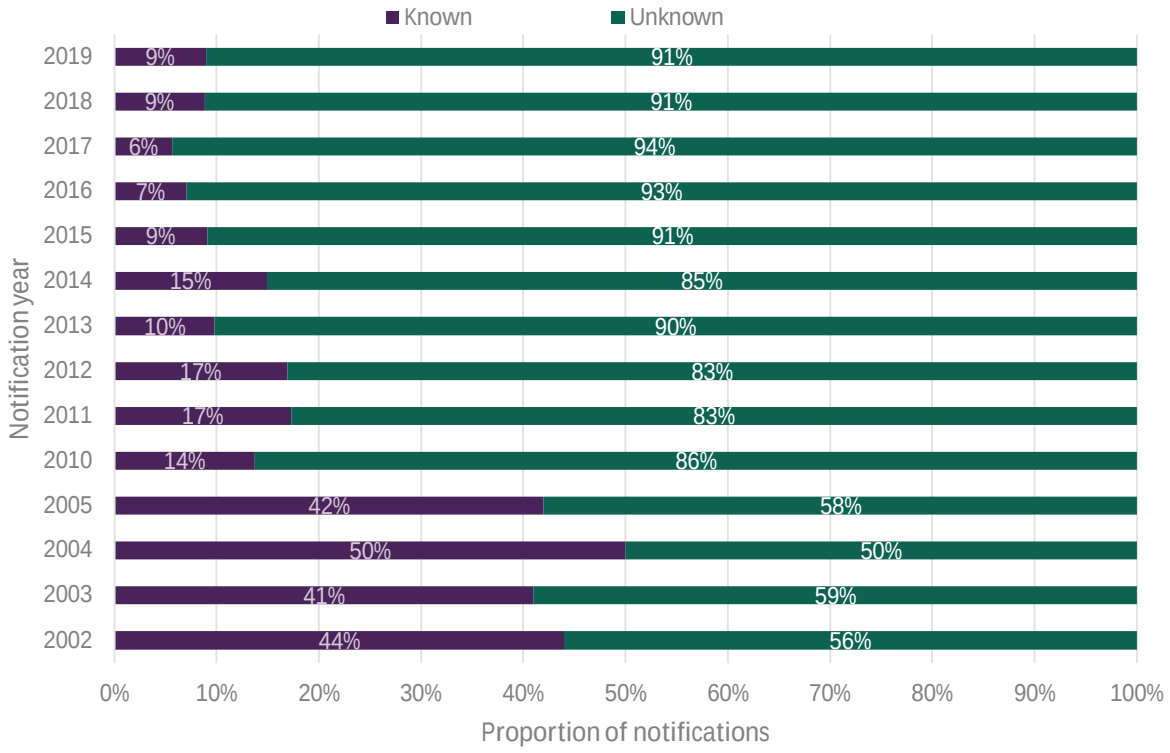

Figure 10: Proportion of hearing losses of known and unknown cause notified to the DND by year diagnosed (2002-2005 and 2010-2019)

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.

Mumps, measles and meningitis were previously often considered by audiologists as possible causes of hearing loss; however, this had become less common as a result of generally increased immunisation coverage, although these rates have recently fallen. The impact of the current measles epidemic[111] 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[112].

The importance of 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%)[113, 114].

In New Zealand during the 2010-2019 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 included them in the section *Most common types of additional* disabilities on page 16.

Children and young people with syndromes

Among the 1970 children and young people in the 2010-2019 dataset, twenty-nine specific syndromes had been confirmed, affecting 68 children and young people. This number represents just under 3.5% 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 21 children and young people, Pierre Robin Syndrome/Sequence and Goldenhar Syndrome and which were present in eight and seven children/young people respectively.

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. 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[115].

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[116].

Identification of hearing losses

Te tautuhi i ngā take i turi ai

- **Hearing loss can be present at birth or can develop at any time. The DND contains information about the age at which children have their hearing loss identified, and also the age at which a hearing loss was first suspected.**
- **Behavioural methods for identifying hearing loss among infants are generally not reliable for very young children and those with disabilities, so prior to implementation of objective newborn hearing screening across New Zealand, the average age of tamariki at the time of diagnosis was very high. Parents were the group most likely to first suspect their child's hearing loss.**
- **The most recent data shows that an estimated 94% and 91% of the eligible population had their hearing screened by the UNHSEIP (during the reporting period for 2017) and the B4SC (2018/2019). The eligible population for the B4SC is those enrolled with Primary Health Organisations (PHOs), and not all children are enrolled.**
- **Since implementation of newborn hearing screening nationwide, the proportion of children and young people born in New Zealand whose hearing losses have been identified before the age of one has increased greatly from 24 in 2010, to more than 100 in 2017-2019.**
- **Since 2013, newborn hearing screeners have been the most likely group to first suspect hearing losses among New Zealand children and young people, with 56-57% of all notifications now resulting from a screening referral[i]. Sixty five percent of the cases notified in 2019 were diagnosed by the internationally recommended age of three months.**
- **There are two peaks for identification of hearing losses among New Zealand tamariki – from newborn hearing screening and from the period around the time the child starts school.**
- **Those born overseas, those with mild, acquired and/or unilateral hearing losses have a greater chance of having their hearing loss identified later.**

Who first suspected the child's hearing loss?

Information on who first suspected the child or young person's hearing loss was recorded for 95% of tamariki born in New Zealand and diagnosed in 2019.

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

The proportion of 2019 cases first suspected by parents or caregivers is significantly below historic levels reported in the original Database, in which between 34% and 52% of cases were first suspected by parents during the 2000-05 period.

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%

i The percentage shown in Figure 11 of 56% differs slightly from the 57% shown in Table 14 because these are data provided by notifying audiologists from two different parts of the notification form, answering two slightly different questions.

in 2010) – to being first in only 10% of cases in 2019.

	2010	2014	2019
Most likely to suspect	Parent or caregiver (37%)	Newborn hearing screener (39%)	Newborn hearing screener (56%)
Second most likely to suspect	VHT (17%)	Parent or caregiver (22%)	VHT (10%)
Third most likely to suspect	Medical professional (10%)	VHT (13%)	Parent or caregiver (10%)

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

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 disabilities, even those used by paediatric audiologists or hearing screeners, were not an accurate method of screening for hearing loss in infants and some children with additional disabilities[117, 118, 119].

than half of all cases of hearing loss notified to the Database.

Evidence exists that behavioural methods, previously relied upon for identifying a hearing loss among babies and those with additional

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 recent 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.

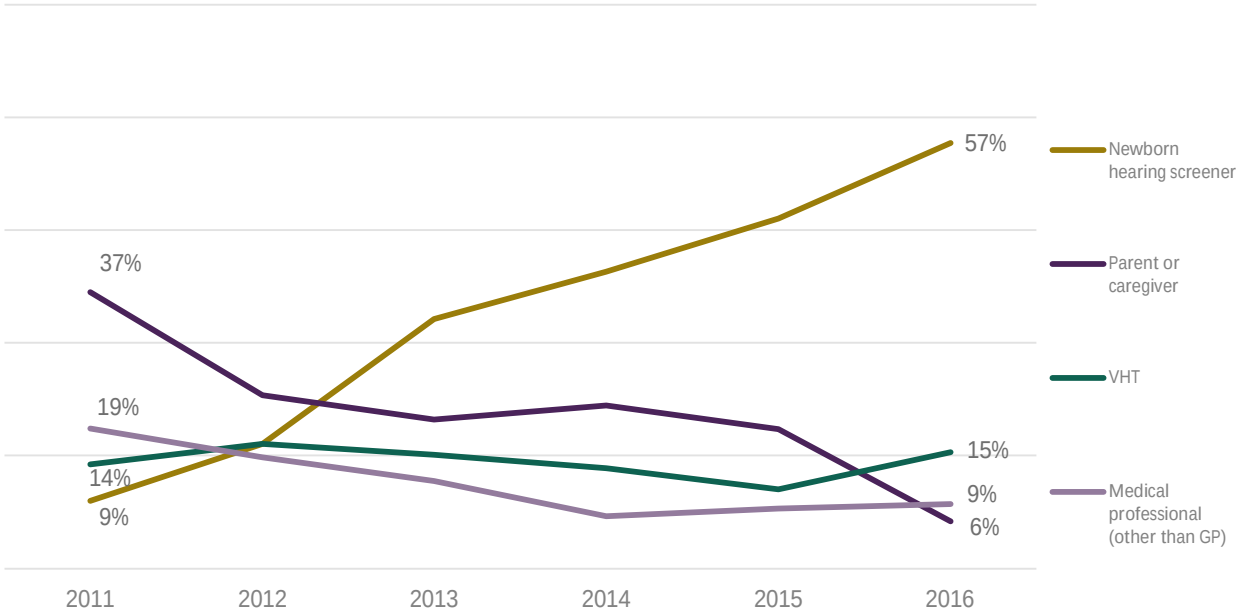

Figure 11: Top Five groups to first suspect hearing loss among notified cases born in New Zealand (2010-2019)

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.

Age at diagnosis

Figure 12, below, shows the number of children whose hearing loss is identified by the age of the child[i] for 2010 and the current year. 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 2019 (n=107) as it was in 2010 (n=24), when the Database was re-launched, although it peaked in 2018 at 113.

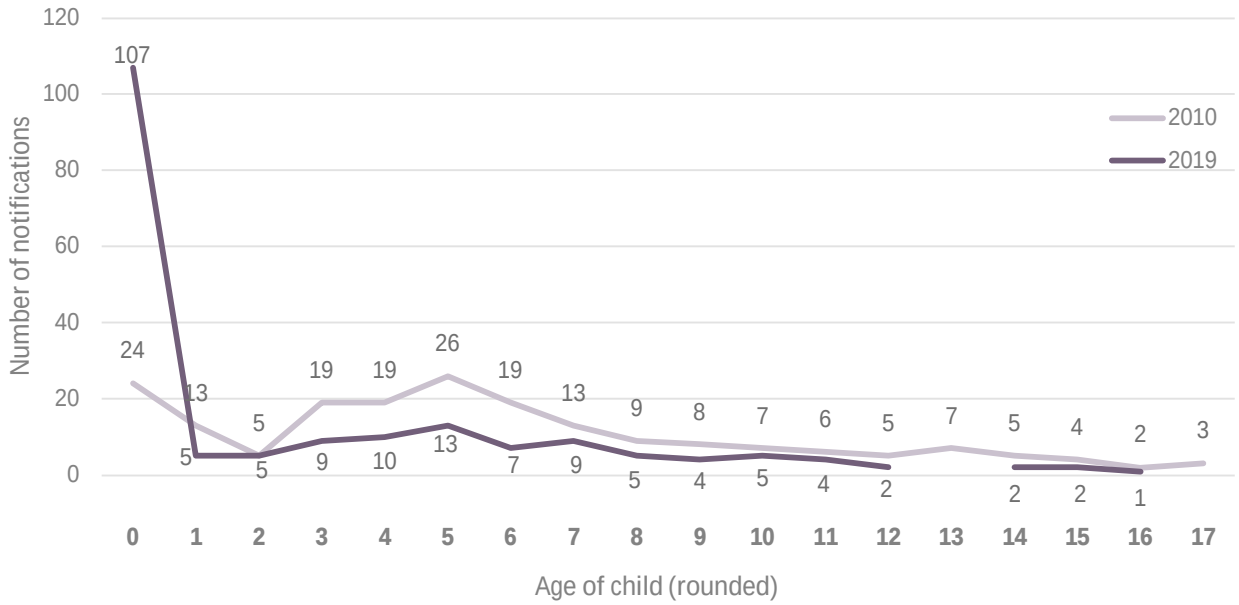

Figure 12: Number of children diagnosed by age (2010 and 2019)

This is a positive trend, as it indicates more and more tamariki are being diagnosed early. A further, smaller peak can be seen for four, five and six-year-olds; this is likely to correspond to the B4 School Check[ii, 120]. 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 45 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. That suggestion was supported by our very high average age of identification when compared with similar jurisdictions.

Overall age at identification

Caution: There are several issues with reporting the average age at identification (diagnosis) for all groups of tamariki. However, describing data in this way may be useful for comparisons with measures used before 2006. It also describes the average age at which providers will begin working with tamariki to provide interventions of some type. It is important to remember that such averages relate to all newly diagnosed tamariki, as it is not possible to separate out those with

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

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

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

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

Keeping these considerations in mind, the average ages at diagnosis for children diagnosed as described on the notification forms provided to the Database are described in Table 10[ii]. The analysis 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. This is particularly important given the long tail of delay which exists.

	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019
Average all cases	65	57	61	60	60	53	44	37	36	40
Average born in New Zealand	62	53	56	54	53	47	37	32	32	37

Table 10: Average ages of diagnosis for all cases in months (2010-2019)

The average age at diagnosis is presented in Table 10 for comparison with previous data, and 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
unilateral hearing losses	bilateral hearing losses, particularly bilateral profound, severe or moderately severe hearing loss
mild hearing losses	
acquired hearing losses, e.g. late onset, progressive and trauma related	hearing loss thought by the clinician to have been present at birth

Table 11: Early and late average ages of identification (2010-2019)

Age at diagnosis by severity of hearing loss

Table 12 shows the average age at diagnosis (identification 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.

A number of records in the DND contain incomplete severity information, so we have included those determined to be bilateral using both data from the audiologist and interpolated datapoints.

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 32 months in 2019. Please note that these changes may relate to cohort differences as well as overall

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

ii 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.

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[i], 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 notification form does not include information about the proportion of losses, which are thought to be progressive in nature.

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2010-2019)	Total number of cases
mild	59	602
moderate	38	322
moderately severe	28	85
severe	24	41
profound	11	74

Table 12: Average age at diagnosis, in months, for *bilateral* hearing losses by degree (ASHA codeframe) using interpolated data with manual checks (2010-2019)[ii]

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

Table 13 shows the average identification ages (2010-2019) for each ethnic group, for all children and young people notified, where ethnicity information was provided. Please note that differences between ethnic groups, such as degree of loss and the proportion of cases present at birth, will influence these figures.

When viewing data on ethnicity, please keep in mind that Table 13 is based on multi-code data, hence a 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.

All ethnic groups show improvements in age at identification between 2019 and the full 2010-2019 period.

Ethnic Groups	Average months at diagnosis (2010-2019)	Median months at diagnosis (2010-2019)	Number of cases (2010-2019)	Median months at diagnosis (2019)
European	50	44	931	34
Māori	50	48	659	11
Pacific Peoples	61	56	235	26
Asian	37	6	248	3
MELAA	57	36	35	2
All groups	50	46	1928	9

Table 13: Average months at diagnosis by ethnicity (2010-2019)

The average age at detection over the 2010-2019 period is of particular concern for Pacific children, at 61 months, although recent years have seen a drop from a high of 83 months in 2012 to 34 months in 2019. These differences may be related to changing characteristics within the cohorts over time. Pacific children have also seen large reductions in median age at diagnosis, from 56

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

ii 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.

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

months between 2010-2019 to a median of 26 months in 2019.

MELAA children and young people also have a high average age at identification over the years, at 57 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.

Māori children and young people have been identified at an average of 50 months over the full period, the same as their European counterparts. Their average age at diagnosis has dropped from a high of 65 months in 2013 to 36 months in 2019. Māori particularly have seen a big reduction in the *median* age of diagnosis, moving from 48 months for the full period to 11 months in 2019.

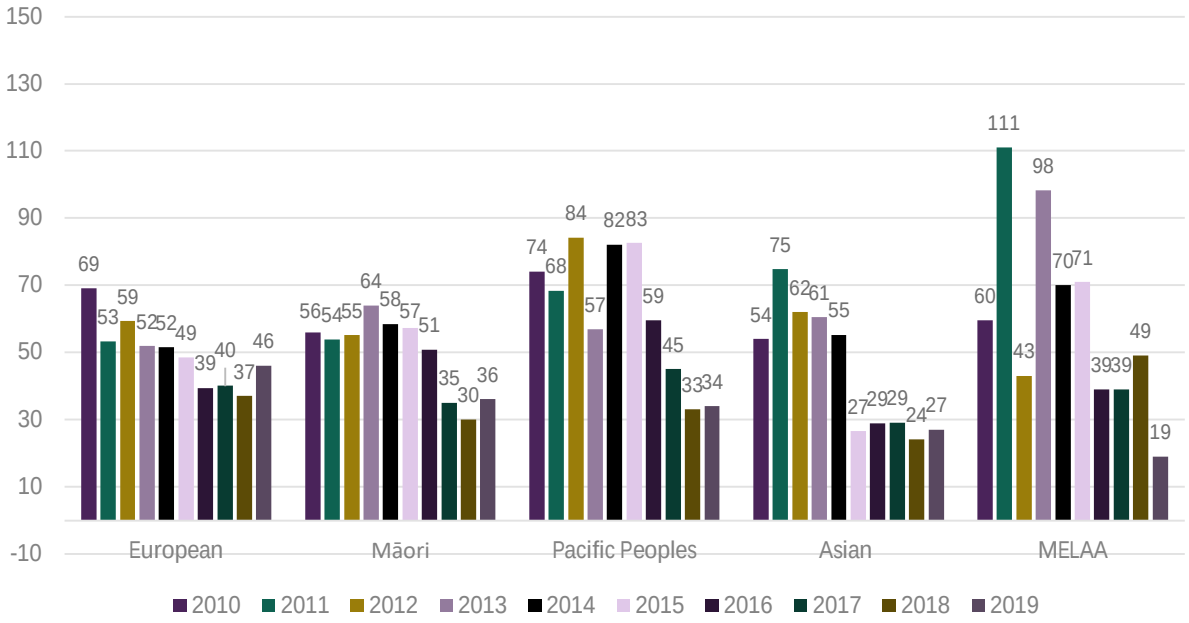

Figure 13: Average age of diagnosis by ethnicity in months (2010-2019)

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 severity hearing losses than their European peers, losses that are on average identified later than those that are of greater severity[64].

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, or of improvements in the health system's ability to reduce inequalities for Māori, although we have no evidence to support these suggestions.

The authors of this report hope future analyses will shed further light on the types of hearing losses that are common among each ethnic group, so we can better understand the reasons for their later average age at diagnosis and reduce inequities.

Newborn hearing screening

The target condition for the Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP) includes any hearing loss greater than 35 dB eHL at 500 Hz and greater than 30 dB eHL at any frequency in the range 1–4 kHz, in either ear[121, i, ii].

The policy and quality standards for the UNHSEIP note that children with mild hearing losses below

i The target permanent congenital hearing loss includes conductive impairment associated with structural anomalies of the ear but does NOT include temporary impairment attributable to non-structural middle ear conditions.

ii This is a common threshold in found in newborn hearing screening programmes, as referred to by Neumann *et al.* in the *International*

this threshold may not be 'candidates for amplification, these children should still be monitored audiologically, as they may be at risk for progressive hearing loss and the deleterious effects of additional temporary conductive hearing loss'[121].

It is worth noting that Māori tamariki are more likely to have mild or moderate hearing losses than their European counterparts.

Implementation

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

Screening status

Table 14 shows the screening status of New Zealand-born children notified to the Database (and therefore diagnosed) in the period 2010 to 2019.

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 those diagnosed at varying ages because some children in most years were not 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 incomplete, and as no monitoring reports have been produced since 2017, the true extent of loss to follow-up in the UNHSEIP cannot be ascertained.

The most recent NSU UNHSEIP Summary Report[57], included data for babies screened from 1 January to 31 December 2017 and these data were summarised in the 2018 DND report. At that time, 94% of babies born during 2017 completed screening during the period, with 89% completing within the target of one month of age. This does not compare favourably with our Australian neighbours, who are screening 97% of babies by one month of age[122].

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	2019
No	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	5%	4%	1%	0%
	No, this service was not available at the time (at the time of diagnosis)	68%	54%	37%	12%	7%	7%
Unsure	Unsure whether screening was offered to this family	7%	6%	5%	2%	6%	6%
Yes	Yes, a screening programme was in place, but the child was directly referred to audiology due to atresia	0%	1%	1%	3%	2%	4%
	Yes, screening was offered but this child was not screened	1%	1%	1%	3%	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%	4%
	Yes, this child was screened and passed	1%	6%	13%	16%	17%	21%
	Yes, this diagnosis is a result of a referral from screening	18%	28%	38%	53%	61%	57%

Table 14: Screening status of children born in New Zealand and diagnosed, selected years[ii]

Journal of Neonatal Screening January 2019 and by Matulat and Parfitt in the same journal in September 2018.

i Implementation of New Zealand's 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.

ii 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.

Referrals from the UNHSEIP

The UNHSEIP has provided much needed local data to help 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[i], at around 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[57].

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 97 of the 2019 notifications (57%) 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 dropped slightly on 2017 and 2018 figures.

Sixty three percent of children diagnosed in 2019 had their hearing loss identified by the internationally recommended age of three months[ii], a drop on the 73% in 2018 and 67% in 2017.

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 not known.

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 or subjective testing.

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 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 for cases referred from newborn hearing screening in New Zealand (therefore born in New Zealand), from fourteen months in 2010, to five months in 2018.

Of the 97 cases notified in 2019 that were identified as a direct result of newborn hearing screening in New Zealand, 75% were diagnosed by the internationally recommended age of three months.

Table 15 shows the changes in the average age at diagnosis since 2010 for cases referred from newborn hearing screening.

i Overseas, a number of comparable newborn hearing screening programmes (such as those in the United Kingdom 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 We are using a more accurate method for calculating this figure now, based on all records where a specific date of diagnosis is provided. As a result, it isn't directly comparable to previous figures. Using the previous method, this year's proportion of cases diagnosed by three months would have been 75%.

	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019
Average months at diagnosis	14	8	6	7	5	6	5	5	3	5

Table 15: Age at diagnosis for children referred from and diagnosed as a result of the newborn hearing screening programme (2010-2019)

Identification of false negatives

The DND likely provides the only method for identifying potential false negatives from the newborn hearing screening programme[123, i].

In 2019, no cases notified to the Database were explicitly identified as having wrongly passed their New Zealand based newborn screening, meaning we have no confirmed false negative cases for this year. This is not to say that one or more babies diagnosed in 2019 were not incorrectly passed at their newborn hearing screening, just that none were recorded as such in the notifications.

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

Thirty-five of the tamariki who were born in New Zealand and identified with hearing loss during 2019 had been screened previously as part of the UNSHEIP 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 35 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 16.

The first of these have known acquired hearing loss, while the second is those with hearing losses where the diagnosing clinician believed this was not present at birth[ii] (it is possible New Zealand has a greater prevalence of progressive hearing losses because of our high rate of CMV[113]).

Of the fourteen 2019 cases identified as *potential* false negatives in Table 16, the age of identification for these tamariki ranged from almost three, to eight years of age.

	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019
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	11	20	28	28	32	31	35
Number of cases from regional screening programmes, or from the UNHSEIP, that 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 New Zealand	2	5	4	7	10	19	18	18	17	14

Table 16: Potential false negatives and cases previously referred from hearing screening, 2010-2019, born in New Zealand only

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 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.

B4 School Check

The B4 School Check (B4SC) 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.

There is no national reporting that helps us understand the efficacy of the hearing screening within the B4 School Check. As a result, the information about the proportion of children who refer on the hearing screen who go on to receive diagnostic assessment, who is diagnosed as a result and when and what type of intervention they receive, or their outcomes, is unknown.

B4 School Check hearing screening data for alternating cohorts from selected years are shown below (see previous reports in this series for data from other years). The proportion of eligible children not screened[i] in 2018-19 was 9% in 2018-19, compared with 10% in 2015/16.

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, 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[124].

Please note that the data used for that paper were from 2014/15. The proportion of eligible children who were listed as 'not checked', 'decline' or 'under care' by the B4SC at that time was 10%, the same as in 2018-19.

Outcome	Description	2011/12	2013/14	2015/16	2017/18	2018/19
Pass Bilaterally	The child was screened and passed.	65%	72%	80%	84%	80%
Referred	The child was screened and referred to a relevant service.	5%	5%	5%	5%	5%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	8%	6%	5%	6%	6%
Under care	The child is already under the care of a relevant service.	3%	3%	4%	4%	3%
Decline	The hearing check was declined by the caregiver.	5%	3%	1%	1%	1%
Not Checked	The child did not receive a hearing check.	16%	12%	5%	1%	5%
Population	**Derived from the PHO enrolled population.**	**65,692**	**65,335**	**62,581**	**61,005**	**61,757**

Table 17 B4 School Check Hearing Screening data (those tamariki screened in alternating years from 2010-2019)[ii, iii, 66]

i This comprises those already under care of a relevant service, those for whom caregivers declined a hearing check, and those who did not receive a hearing check.

ii The Ministry of Health notes that the population used is the PHO enrolled population. They 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.

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

However, it is also worth noting that the denominators for the B4SC comprise children who are enrolled with a PHO. Welcome to School Study data suggests that in some areas there may be a significant number of children not enrolled with a PHO and therefore not included in the reported figures below[128]. Ministry of Health data indicates that 97% of children under the age of 5 are enrolled[125]. There was an overall enrolment rate of 94% as at October 2019, while PHO enrolment is lower among Māori at 91%[126].

Some children who are not enrolled *are* screened making it difficult to understand the overall coverage rate for the hearing screening completed within this Check.

The overall referral rate for tamariki completing this screen is 5% (2018/2019). As with previous years, Māori and Pacific tamariki have higher referral rates (7.1% and 9.6%), and Asian and MELAA tamariki lower rates than the average (4.4% and 4.9%). The lowest referral rate was for European tamariki, at 3.5%.

This year the proportion of tamariki not checked rose to 4.5%, considerably higher than the 1.0% reported by the Ministry last year.

A recent study (Welcome to School, 2017) focused on the health and development of students starting school in Tāmaki (an area in Auckland) in which 90% of the tamariki are Māori and/or Pacific[127]. It found that although 75% of children had developmental delays and 64% below average language skills, very few parents reported concerns about their child's development at the B4 School Check or school entry. This suggests that the B4 School Check Parental Evaluation of Developmental Status (PEDS) questions may not work well for all New Zealand children and therefore that it is inappropriate for the New Zealand context[128].

These findings have implications for Māori and Pacific whānau whose tamariki have a hearing loss. There are signs that current screening protocols/instruments may exacerbate rather than narrow pre-existing inequalities for these groups of children (due to thresholds set for referral, for example). In addition, systems and practices that are Euro-centric and create inequities may reduce the chance that hearing losses are identified promptly when they develop outside the two- or three-points during childhood at which hearing is currently screened.

Delays in Diagnosis

Ngā takaroa ki te whakatau māuiui

- **Delays in diagnosing hearing loss among children and young people are a known contributor to poorer outcomes. Such delays can be reduced by hearing professionals, researchers, advocates and decision-makers in a number of ways.**
- **The average delay between first suspicion of a child or young person's hearing loss and its confirmation is now ten months, down from 26 months in 2010. This is undoubtedly, at least in large part, due to nationwide implementation of the newborn hearing screening programme.**
- **Even this much improved average delay remains too-long and some children and young people are waiting years before their hearing loss is diagnosed and intervention can begin. Children and young people born overseas, those with unilateral and/or mild or moderate hearing losses and those living in higher deprivation areas are more likely to experience delays. Māori and Pacific children are more likely to experience longer delays compared with their European counterparts.**
- **Forty seven percent of the children and young people diagnosed in 2019 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 mentioned reason for delays in diagnoses between 2010 and 2019. Such delays can be the result of conductive overlay or the child being unwell.**

Diagnostic delays

There are many variables that impact on a hard of hearing child's communication and learning outcomes. These include child specific factors like cognitive ability, family factors such as the level of maternal education, and factors related to the hearing loss such as its severity.

One important variable which hearing professionals are able to influence is how quickly the child's hearing loss is diagnosed, and calls for earlier identification of babies with a hearing impairment have been made for nearly 80 years[129].

Early diagnosis seeks to maximise benefit during sensitive periods of neurological and linguistic development and limit children from falling behind their peers[130, 131, 132, 133, 134].

There are a number of ways to limit these delays, including early and regular screening of children and young people for hearing loss. Newborn hearing screening programmes commonly use the 1-3-6 goals, which aim for 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, to target these reductions.

This approach has proven overall to be successful overseas, and in New Zealand, reducing the average age at diagnosis for all bilateral notified cases where the child was born in New Zealand, from 45 months in 2004 (prior to implementation of a national programme for screening newborns) to an average of 27 months in 2019[i].

i These figures are not found elsewhere in the report as they represent only children born in New Zealand and diagnosed with a bilateral hearing loss, to match criteria for inclusion in the Database prior to 2005.

However, significant disparities remain, including how the benefits of interventions like newborn hearing screening are distributed among the population, including for Māori tamariki.

Additional approaches are needed to further limit diagnostic and therefore interventional delays in order to improve outcomes.

There are a number of types of changes which can be the focus of work to reduce diagnostic delay within hearing services (see Table 20):

- service culture, resourcing, and employment;
- individual clinical practice;
- systems, policies and processes, including IT infrastructure;
- education of the public and other groups about hearing loss and when to seek help.

Change will require a sustained and collaborative effort, but hearing professionals demonstrate, including through the care and time they take to provide notifications to this Database, that they are committed to providing an ever-improving standard of care to children, young people and their families.

Some of this change will require hearing professionals and services acknowledging their *"responsibility for differential quality of care, including between Māori and non-Māori, reducing a culture of blaming Māori for the state of their health and acknowledging Pākehā privilege within health services*[135]*."*

Length of diagnostic delays

Average delays

Those notifying cases to the Database were asked to provide information about the length of delay in identifying a child or young person's hearing loss.

The average delay in 2019, between first suspicion and confirmation of the child or young person's hearing loss, *including* those born overseas, and mild, acquired or unilateral hearing losses[i] was ten months, up from six months in 2018.

Ten months remains a significant average delay between first suspicion of a hearing loss and its confirmation, although average delays in the last five years are greatly improved on 2010 and 2011 figures[ii].

Groups at increased risk of diagnostic delays include those:

- children and young people who were born overseas;
- with a mild to moderately severe bilateral hearing loss;
- with a unilateral hearing loss and those who the audiologist expects will receive a single hearing aid, e.g. due to asymmetry;
- with a hearing loss not thought to have been present at birth.

Year	Delay in months
2010	26
2011	16
2012	10
2013	12
2014	12
2015	11
2016	9
2017	9
2018	6
2019	10

Table 18: Delay in months by year, 2010-2019[iii]

Higher proportions of Māori[iv] and Pacific tamariki have delays of three, six months, and two years

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

ii 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. For example, the severity profile of cases can differ from year to year, as can the proportion of children with acquired or progressive hearing loss.

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

iv A 2017 examination of 2010-2016 notification data showed that the average delay for Māori was 7.5 months higher than those for New Zealand European tamariki. There was also a significant association

or more when compared with their European counterparts.

Just under half (47%) of rangitahi notified to the Database in 2019 experienced a delay under one month (including those with no delay[i]) in receiving their diagnosis. This is a drop from last year when the same figure was 52% but shows an improvement on the 43% average for 2010-2019.

Delay causes

Cases from 2010 to 2019

The notification form also asked notifying hearing professionals for the reason(s) for the delay, where one or more were provided. Not all notification forms included one or more reasons for the delay listed, including some which demonstrated a delayed diagnosis existed.

The analysis in Table 19 examines the reasons for delay (2010-2019) where one or more reasons were listed and *where the delay was reported to be greater than one month*, measured from the time the hearing loss was first suspected until the time when the hearing loss was diagnosed[ii]. It shows the most commonly cited reasons for delays in diagnosis, and a selection of approaches to reducing the various types of delay are included in Table 20.

Rank (most mentioned)	Reasons for delay
1st	Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)
2nd	Parents did not attend appointments/ delayed or rescheduled these (for any reason including service failed to engage family)
3rd	Waiting time to see hearing professional or accessing services in their area
4th	Parents or educators suspected something other than hearing loss (e.g. speech delay, developmental delay)
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

Table 19: Most common reasons listed for delays in diagnosis (2010-2019) for cases with a diagnosis delayed by more than one month

A previous examination of 2010-2016 notification data showed Māori tamariki were 1.6 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 provided reasons for this delay, by a factor of 1.32. When delays in diagnosis of one month or more are examined further (still a significant delay for many tamariki) a number of patterns emerge:

- European and Māori families are more likely to have suspected something other than hearing loss, or to have had no concern about hearing mentioned as a reason for delay than other groups;
- Māori families are considerably more likely not to attend appointments or to delay these for any reason (30% of cases among Māori have this reason listed for delays compared with 10% among European families);
- Pacific families are also more likely to have experienced diagnostic delays due to non-attendance – 23% of Pacific children have delays of this type;
- Asian and MELAA families are less likely to have this reason listed among those for the delay (6% and 8% of cases respectively).

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. Details can be found in the 2017 report.

i This is based on the child's age at suspicion and date of diagnosis. 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.

ii Delays for children and young people born overseas are included in this table.

Focus area	Approaches to reducing delays
Resources	• secure greater funding for public sector audiology services to reduce waiting times for clients and whānau through: o advocacy to demonstrate the value of audiology services and the importance of effective IT infrastructure; o collaborative work to collate existing evidence for the value of audiology and hearing services and new research to better understand the long-term benefits of audiology services for the paediatric population in New Zealand; • advocate for the introduction of a service specification for audiology services to define a minimum set of services available within each district health board and reduce geographical disparities.
Clinical	• efficient clinical practice to complete assessments over fewer appointments (Following 2016's Diagnostic and amplification protocols,[136] which can be found on the National Screening Unit (NSU) website; • active paediatric certificates required for those diagnosing children under the age of three; • clinical staff to engage with professional development and mentoring opportunities and inter-professional and other support networks; • close collaboration with ENT services to minimise delays for children with middle ear conditions.
Employment	• employment of staff holding the NZAS Paediatric Certification for those diagnosing children under the age of three; • employment of staff who have an understanding of what it means to practice in culturally safe ways for those in the local population, including Māori.
Service: understanding and planning	• understand the client population, evaluate and monitor in-service attendance and clinical outcomes, including monitoring unmet need, and implement improvement plans to equalise outcomes; • utilise feedback on service efficacy from monitoring and evaluation sources (e.g. NSU re the UNSHEIP).
Service: systems, policies and processes	• consider more attempts to contact families before discharging from service, strong channels of communication between referring and receiving DHBs and robust processes to ensure children who leave the service are received by a new service; • introduce, improve or integrate systems and processes for scheduling follow-up and seeing this occurs in a timely way, including through effective systems and IT infrastructure; • ensure prompt referral from newborn hearing screening and resulting assessment and reduce delays to see clinicians; • strengthen relationships between community-based screeners and audiology services to expedite referral processes where needed and also draw on the relationships already existing to encourage engagement; • offer services closer to home for families to reduce disparities for rural or semi-rural families (e.g. community-based clinics or outreach).
Service: reducing engagement barriers	• include other teams to support family engagement and effective prioritisation to maximise paediatric outcomes and reduce inequalities through Primary Health Organisations (PHOs) and public health teams; • consider increasing scheduled time for appointments (particularly for new clients and refinement of communication with families) and offering flexible appointments (particularly for those who are unable to take leave from work, including those outside of normal business hours); • build or strengthen cultural safety by working individually and as a team to understand different cultural frames and what this means for the way services are organised, offered to whānau and how tamariki and their whānau are treated. [There are excellent resources on this topic, including ones focused on improving access to healthcare for Māori[137], a statement on cultural safety from the Medical Council[138], and this paper focused on the difference between cultural safety and cultural competency[139].]; • remove or mitigate cost barriers for patients associated with attendance, e.g. offering assistance with travel and other costs. [Public transport options may be insufficient or impossible, particularly for new mothers[82].]; • actively work to reduce rates of non-attendance (DNA rates); • connect families with additional support options such as volunteer support networks; • work to increase the chance whānau and rangatahi see the same clinician and other staff members at their visits – this could be examined in conjunction with hubs where multiple services are available at once, and coordinate appointments with visiting families
Education: Improve understanding of hearing losses among tamariki	• provide parent/whānau education so they can identify signs of a possible hearing loss, better understand screening, and understand what to do, including materials specifically designed for Māori whānau; • clear guidance on pathways for parents so they know what to do if they suspect their tamariki may have a hearing loss; • education for the public on hearing loss and the value of screening, early diagnosis and intervention; • education for teachers and other education professionals on hearing loss and when a child or young person should see an audiologist or other hearing professional for an assessment; • education for medical professionals on hearing loss, when to refer to audiology, the purpose and timing of hearing screening and what this screening does.

Table 20: Approaches to reduce diagnostic and interventional delays and reduce inequalities for tamariki with hearing loss

Current year's cases

In 2019, 38% of all cases had one or more reasons for delay listed[i]. 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 overall average age at identification and rising number of cases with no delay reported.

When 2019 cases with one or more reasons for delay are examined, the most common were:

- audiologist having difficulties getting a confirmed diagnosis (n=14);
- parents did not attend appointments/ delayed or rescheduled these - for any reason including service failed to engage the family (n=10);
- difficulty getting a referral to audiology (n=3);
- child or young person had other medical issue(s) which took precedence (n=2);
- follow up lost/referral in the system did not occur or was delayed (n=2);
- parents or educators suspected something other than hearing loss (e.g. speech delay, developmental delay) (n=1);

In addition to selecting from one or more pre-coded reasons for delay, notifying hearing professionals also had the ability to comment further regarding the reason(s) for delayed diagnoses.

Selected comments relating to tamariki diagnosed in 2019 with delays of more than one month are included below:

- *"Referred by [vision hearing technician] from B4 school screening. This resulted in diagnosis. [Had] DNA'd follow up after newborn hearing screening referral." [98-month delay]*
- *"Had test done by private audiologist who found 'normal' results." [91-month delay]*
- *"Since his birth, all hearing tests have come back saying his hearing is "borderline". Due to lack of other hearing loss symptoms, nothing was done – mum." [17-month delay]*
- *"Screened and infant ABRs at another DHB in 2018. x2 ABR yielded inconsistent results so it was decided to wait for behavioural results. [Child] then transferred to [name of DHB] and VRA confirmed sloping bilat SNHL." [Eight-month delay]*
- *"Was identified following [newborn hearing] screen as requiring surveillance audiology at 18 months due to perinatal asphyxia and ventilation administered and prolonged NICU stay, but either not referred or referral missed." [Five-month delay]*

DNA rates

A good number of the comments provided by audiologists and audiometrists in the notification forms indicate that non-attendance at appointments is a key cause of delays in diagnosis.

High DNA (Did Not Attend) rates result in poorer outcomes for individuals who do not receive timely diagnosis and/or intervention. They also have implications for service efficiency.

Reducing rates of non-attendance has been a focus in many district health boards, not always in a sustained way, as resources, support and ongoing funding for these efforts are not always prioritised.

Significant improvements have been achieved as a result of a focus on reducing DNA rates.

For example, Capital and Coast DHB efforts resulted a drop of almost 50% in DNA rates for specialist appointments among Pacific patients over a five-year period. Conversations with patients with a history of not attending revealed many feared for their job if they asked for time off, while others noted difficulties with transport or childcare as the reason. Each Pacific patient now receives a call two days prior to the appointment and transport is sometimes arranged[140].

Another example of success can be found in Taranaki hearing services. In 2016, Come Hear was

i Seventy-four percent of those had one reason listed for the delay, and 26% had two or more reasons for the delay listed.

launched by Taranaki DHB's Audiology and Māori Health teams to improve outcomes for Māori children by reducing DNA rates at DHB audiology clinics from 20-31% to less than 10% within six months. By January 2017, this rate had dropped to 0%[141].

Common factors successful in reducing barriers to health service access include removing cost barriers, knowing the client population, personal engagement, a non-judgemental approach[142], high levels of cultural safety, and flexibility in service arrangements[143].

Marewa Glover from the Massey University School of Public Health said in 2017 that it "cost money and time to go to appointments. The more obvious reasons are financial. People are juggling a lot of demands… People are struggling to pay their bills and feed their kids. People have to make choices… If people can't pay their power, they certainly are not going to have money to go to appointments[144]."

Māori and Pacific whānau often have higher rates of non-attendance and are also more likely to live in areas of high deprivation than European whānau.

There is also the suggestion that higher rates of middle ear issues among Māori (and Pacific) children may require multiple appointments when there is an underlying SNHL and that this can result in delays in diagnosis[145]. This points to the need for a strong collaboration between audiology and ENT services and the need for early bone conduction testing as indicated by protocols.

Waikato District Health Board is currently examining the reasons for delayed diagnoses among Māori tamariki with a view to reducing these. Initial indications suggest rurality can mean babies are less likely to have their hearing screened as an inpatient in the first days of life, and that this can mean screening is delayed for this group of whānau[145].

Delays attributed to newborn hearing screening

Of the sixteen tamariki whose 2019 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);
- 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);
- child or young person had other medical issue(s) which took precedence, e.g. feeding issues, medically fragile (n=2);
- 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);
- 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).

One important consideration for newborn hearing screening referrals is the importance of prompt referral from the UNHSEIP to audiology, and the 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), typically this is possible at six months to two years of age[i].

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

Severity

Taumaha

- **Audiometric data is now much more likely to be estimated from the ABR as children are being diagnosed at younger average ages.**
- **Many different frameworks categorise severity of hearing loss around the world. Here in New Zealand the Clark (ASHA) framework is most commonly used by hearing professionals.**
- **New Zealand DND data show a relatively higher number of cases with mild and/or moderate hearing loss, and a smaller number of cases with severe/profound hearing loss than in other jurisdictions where comparisons have been done. A number of factors are likely to contribute to this, including the higher numbers of milder degrees of hearing loss found among Māori.**

Audiometric data

Audiometric data are requested for both the right and left ears of all tamariki and young people notified to the Database. Those notifying cases 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 14, below, the proportion of cases for which the thresholds were determined through ABR is rising, from 21% in 2010 to 57% in 2018. This strongly suggests that over time fewer tamariki are old enough to have their hearing assessed behaviourally.

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

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

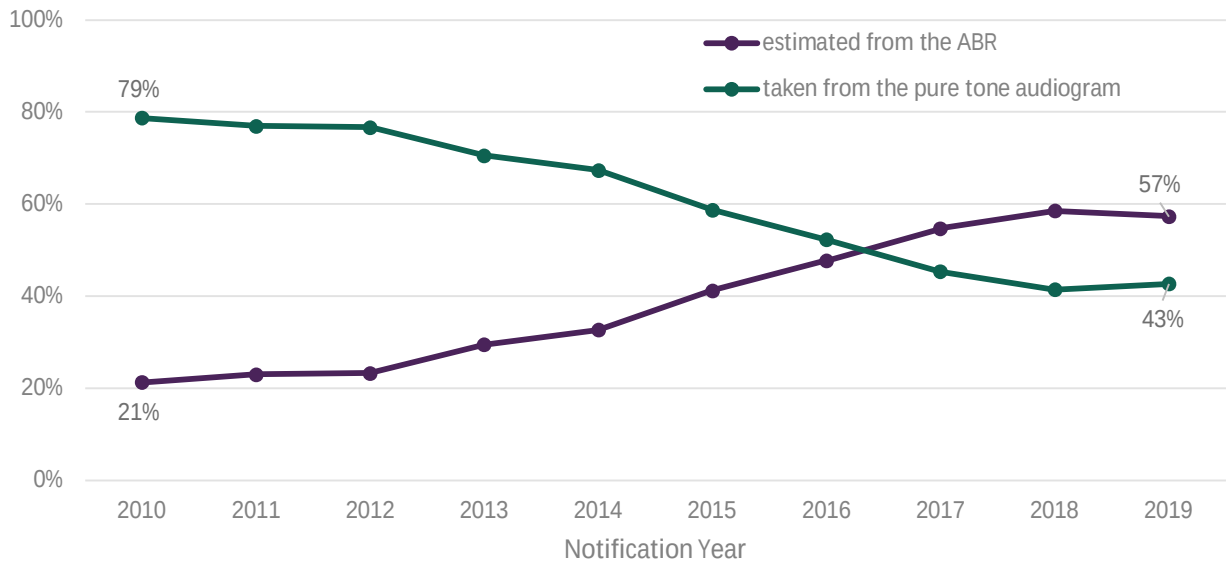

Figure 14: Proportion of cases containing thresholds from ABR (as opposed to being taken from the PTA), by notification year 2010-2019

Classifications

In New Zealand, the Clark (ASHA) codeframe is most commonly used clinically. 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 69.

Degree of loss	Clark 1981 (ASHA)[146]
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 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 DND has requested full audiometric data from those notifying cases, in an attempt to allow meaningful comparisons with overseas data.

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 table includes all notifications from 2010-2019.

Further information about interpolation and its use in this report can be found in Appendix G which begins on page 70.

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.

Degree of loss using ASHA severity codeframe	Bilateral 2010-2019	Bilateral 2010-2019 (interpolated and manual checks)	Unilateral 2010-2019	Unilateral 2010-2019 (interpolated and manual checks)
Mild	61%	54%	51%	45%
Moderate	26%	29%	17%	15%
Moderately severe	6%	8%	8%	8%
Severe	3%	4%	5%	7%
Profound	4%	6%	19%	24%
Sample size	n=760	n=1147	n=470	n=506

Table 22: Comparison of severity classifications by methodology, 2010-2019

Severity profile differences between bilateral and unilateral hearing losses

Most previous reports have contained a graph showing the severity profile for tamariki notified to the Database whose losses were bilateral and compared these with those whose losses were unilateral. Cases selected required all four data-points to be completed for each hearing-impaired ear.

For 2017 and subsequent reports, 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 15 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).

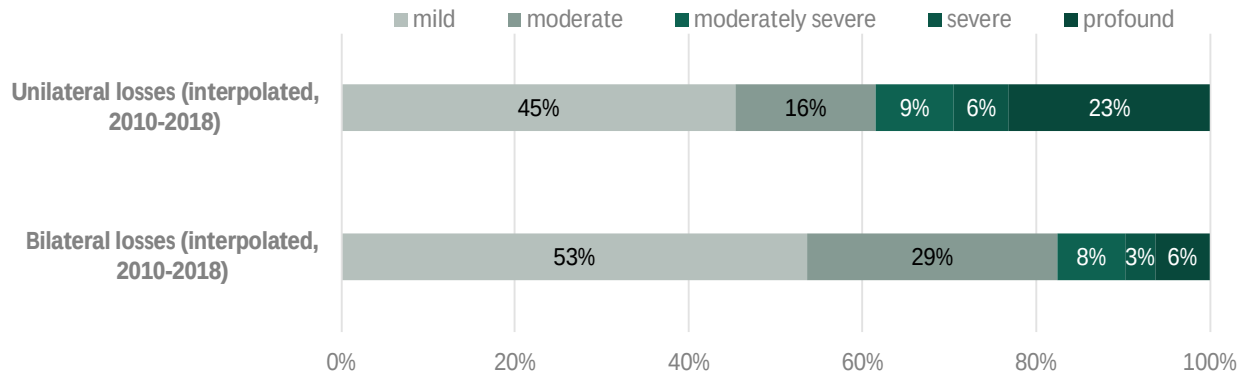

Figure 15: Unilateral and bilateral hearing losses by degree (2010-2019)[i]

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[ii]. Clearly, these differences lessen when comparison is made with the worse ear in bilateral cases.

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

ii Usually for those with bilateral hearing loss it's the better ear audiogram which is used to determine severity for statistical purposes.

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[i];
- 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 rangatahi in each group is possible using data reported between 2001 and 2004 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[ii].

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)

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 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[iii].
- 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 had reduced rates of meningitis in New Zealand and this reduction was expected to have led to a

i It is worth noting that as the average age for identifying hearing loss reduces as a result of newborn hearing screening, the severity distribution at the time of diagnosis for hearing losses should be shifting towards the lower severity categories because progressive hearing losses will not have had time to worsen before diagnosis.

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

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

reduction in rates of (more severe) hearing loss[147]. However, more recently, coverage rates have fallen. Regardless, any 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[148], 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 children, and that milder degrees of hearing loss are more commonly reported among Māori[60, 149].

These findings have been confirmed by analysis of: 1982-2005 data[64, i] and 2010-2016 data[ii].

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 indicate that young Māori have fewer severe and profound hearing losses than their European counterparts.

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, 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:

- United Kingdom, Finland and United States data with New Zealand data 2010-2012 (2012 report);
- Colorado data with New Zealand data 2010-2013 (2013 report);
- Australian data with New Zealand data from 2010 to 2015 (2014 report);
- Colorado data with New Zealand 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 ethnic groups.

In addition, recent research suggests those children with milder degrees of hearing loss who were previously unaided, can have poorer phonological memory and morphosyntactic skills, suggesting issues with leaving mild hearing loss untreated[150], 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[151, 152], and that those families with children who have cochlear implants are managing and promoting device use more than those with hearing aids[153].

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

ii 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.

Intervention and support

Wawaotanga me te tautoko

- **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. In 2019, they provided services to approximately 1,850 children under the age of eight, including 723 babies and young children identified as a result of UNHSEIP.**
- **Deaf education centres based in Auckland (Kelston) and Christchurch (van Asch) provided services in the 2018 year to 2,853 deaf and hard of hearing students nationwide during 2019.**
- **At the time of diagnosis, professionals notifying cases expected most children and young people diagnosed with a hearing loss since 2010 would receive two hearing aids.**
- **Sixty-two children and young people around the country received publicly funded cochlear implants during the 2019 calendar year, and 1,617 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, they hope to share these data for future reports.

In 2019, 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 2019, Ministry of Education Learning Support provided support to approximately 1,800 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 723 babies, infants and children under the age of five identified as deaf and hard of hearing through the UNHSEIP and their families and whānau.
- Support for 285 babies, infants and children under the age of five and their families identified as deaf and hard of hearing not through the UNHSEIP and their families and whānau.
- Support for 867 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
- Ko Taku Reo (previously provided by Kelston Deaf Education Centre and van Asch Deaf Education Centre), for 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 in 2019. The two DECs have had a combined Board of Trustees since 2012, and have now merged into *Ko Taku Reo: Deaf Education New Zealand*.

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.

There are terms used in the education sector that may not be familiar to readers of these reports, who are often 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 71.

During 2019, the Deaf Education Centres provided services to tamariki across the country. These services can be broken down into the categories[154] set out in Table 24.

Hearing aids

In each notification form, audiologists/ audiometrists were asked "How many hearing aids are to be fitted?" The resulting data represent the clinician'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).

All of the 191 cases notified to the Database in 2019 contained information about whether hearing aids were to be fitted.

As has been the case with data since 2010, children and young people whose cases were diagnosed in 2019, are most likely to be fitted with two hearing aids (53%). This reflects the preponderance of bilateral losses notified to the Database.

Figure 16 shows a reduction in the proportion where the plan is to prescribe one or two hearing aids, 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 has risen, likely for the same reason.

When data for all children and young people notified from 2010 to 2019 are considered, the audiologist's intention was to:

- fit 78% of bilateral losses with one or two hearing aids, while 7% were not expected to receive any aids and the notifying clinician was unsure in 14% of cases; and
- fit 40% of unilateral hearing losses with one hearing aid, 22% two hearing aids, while 20% were not expected to receive any aids and the notifying clinician was unsure in 19% of cases[i].

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, including potentially one or more which are at higher frequencies than those collected for the DND, 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.

Type of support	Description	KDEC	vADEC
1. School Provision	This category includes school aged children and young people who are supported by KDEC and vADEC staff (Teachers of the Deaf) in regular schools. Most of these children are ORS verified as 'high' or 'very high' needs. This category used to be referred to as 'Deaf Units'.	n=89	n=29
2. Resource Teachers Deaf	This category includes children over the age of 3 years although most children receiving this support are over the age of 4.5 years. Children in this category are not always ORS verified as they can't be verified until they begin school. Funding for this service comes from ORS funding (0.1 and 0.2 FTEs) and the Deaf Education Centres have some allocation of RTDs under the moderate needs contract.	n=448	n=360
3. Specialist support: DEC funded, and teacher supplied by student's school	These ORS verified children (school aged children in mainstream schools and children in special schools) have funding which goes from the Ministry of Education to their schools, including KDEC or BLENNZ. For example, this funding can be used for teacher aids and other specialist support (occupational support, physical therapists, speech language therapist, Kaitakawaenga, etc.) where staff are employed by the MOE.	n=30	n=60
4. ASSIST	ASSIST stands for Assessment and Involvement of a Specialist Teacher. ASSIST professionals provide assessment services for children to determine their needs. Implementation of changes following the Wilson Report meant that children over the age of eight receive services from the Deaf Education Centres rather than from AODCs. Children can be just on ASSIST, they can be on ASSIST and receive DEC funding in their school, or they can be on ASSIST and receive support from an RTD. This category of service is relatively new, and so is still evolving.	n=1100	n=668 ASSIST students receiving other services = 251
5. NZSL@Schools	The purpose of the NZSL@School is to support schools create learning environments so that deaf children, whose primary face -to-face language is New Zealand Sign Language NZSL, achieve educationally at the same level as their hearing peers and are confident and secure in who they are as a deaf person. As a result, NZSL@School provides a range of support to schools, deaf students and parents/whānau, in addition to any other special education support deaf students receive, to help schools understand and provide learning environments that meet the learning, communication and cultural needs of deaf students who use NZSL.		69

Table 24: Children receiving support through the Deaf Education Centres (2019 year)

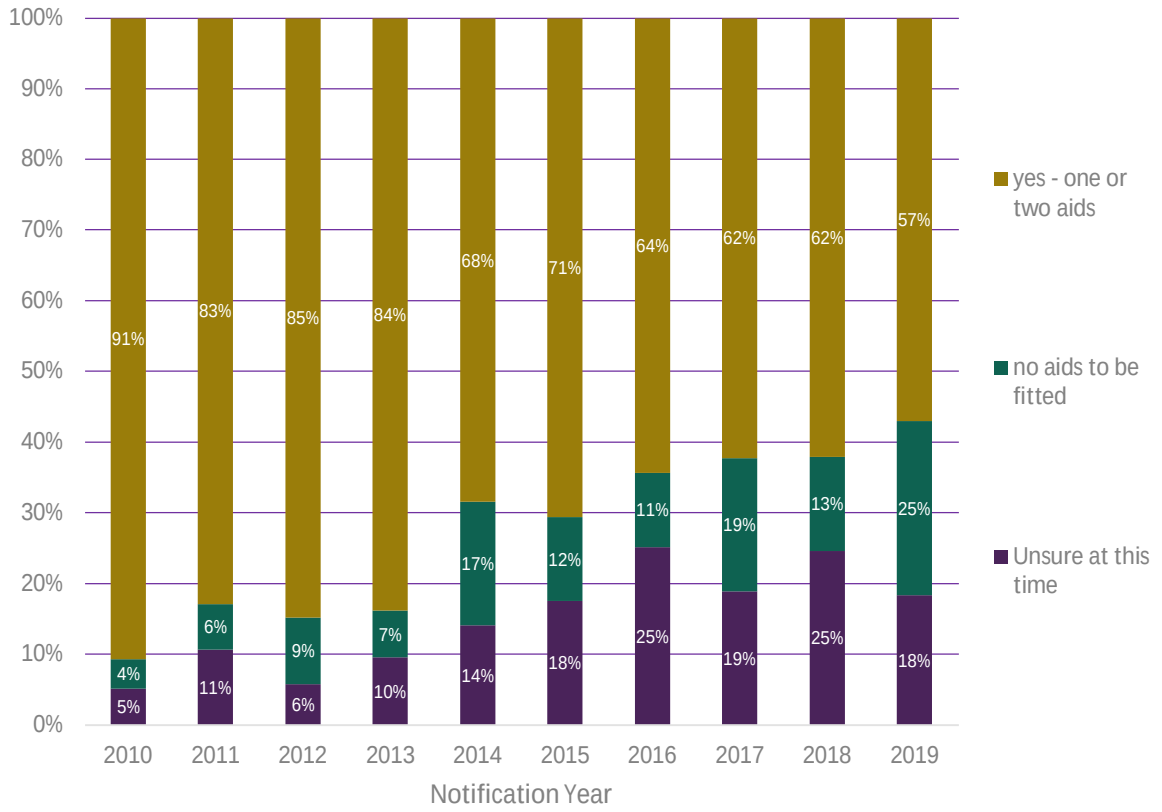

Figure 16: Hearing aids to be fitted by notifications (2010-2019)

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 provider for Hearing Aid Services during the period covered by this report, are shown in Table 25[i].

These data show MOH funded hearing aids for tamariki under the age of 19, and those in fulltime

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 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.

education and under the age of 21 during the 2019 calendar year[i, ii].

Enable New Zealand reports that some invoices may not have been presented in time for 2018 reporting and that as a result the 2019 figure may be inflated somewhat. 1299 children and young people received MOH funded hearing aids in 2018[iii].

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
Māori	108	76	312	42	538
European	91	82	360	91	624
Pacific	58	50	127	25	260
Other	41	23	109	22	195
Total	298	231	908	180	1617

Table 25: MOH Funding of Children's Hearing Aids, Calendar Year ending 31 December 2019, EnableNZ[155]

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 1617 unique service users (tamariki) received hearing aid(s) during this period.

A recent study in the United States 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[156]. Risks of oral language development delays were found to be moderated by early and consistent access to well-fitted hearing aids which provided optimised audibility.

Delays in hearing aid provision

While the Database doesn't ask for information on the reasons for delayed onset of intervention, some notifying clinicians do provide information on this in the final open-ended question which asks if there is anything they would like to tell us.

Reasons for delayed intervention include conductive overlays and medical management, as below:

"CROS fitting so indicated 2 devices. Delay from onset of loss to recommendation for amplification trial due to medical management."

"Conductive overlay delayed hearing aid management, unsure why has been left with mixed loss now when middle ears have essentially resolved."

Prescribing and usage

A New Zealand study 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;

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

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

iii Figures since 2018 don't compare well with those from 2017 and earlier. This is thought to be due to changes in what was counted before and after the change in provider.

- 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 self-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%;
  - » 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.

Forty six percent of children who were recorded as Māori had inconsistent, seldom or no device use, compared with 23% of Europeans. Please note that Māori are more likely to have milder hearing losses compared with their counterparts.

Readers should consider 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, or on the average age at first hearing aid fitting.

In Australia, the age at which children receive their first fitting with a hearing aid by birth year and the age of cochlear implantation shows a clear relationship between reducing ages of identification and earlier intervention, as a result of newborn hearing screening[122].

Munoz *et al*. (2019) surveyed parents with children under six on their experiences, from around the world. Hearing aid use was generally considered low by the authors, compared with the number of hours an infant is awake. Caregivers had positive views on information provided at the time of hearing aid fitting, but had ongoing challenges in hearing aid management.

Issues included a significant drop in the average number of hours the device was in use over time, a lack of loaner devices when theirs were in for repair, and lack of confidence and adherence to carrying out sound checks[157].

An upcoming paper from Visram *et al*. (2020) found that caregivers of 81 infants with a hearing loss in the United Kingdom revealed significant challenges in hearing aid management among very young children, with the authors suggesting that what is needed is specific behaviour change techniques to ensure intentions can be realised[158].

Cochlear implants

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

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

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

During the 2019 calendar year there were 49 publicly funded cochlear implants provided in the Northern Region and 51 in the Southern Region, to children and young people under the age of 19. These implants are provided based on Ministry of Health candidacy criteria for children and young people who are assessed by the cochlear implant teams[i].

Children receiving cochlear implants	Southern Cochlear Implant Programme[159]		Northern Cochlear Implant Programme[160]	
	Ears	Children	Ears	Children
ACC cases	3	3	2	2
Public Funding - (1 Jan to 31 December)	46	25	37	20
Private procedures	0	0	2	2
Re-implants – recalled devices, failed integrity tests, or soft failures	2	2	0	0
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
	51	**30**	**49**	**32**

Table 26: Publicly funded cochlear implants in New Zealand during (2019)[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

Ngā āpitihanga

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 (DND) was New Zealand's annual reporting system for new cases of hearing loss among tamariki 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. Dr Bill Keith and Oriole Wilson are acknowledged for their considerable work on and support for the DND in its previous form.

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 particular calendar year*, rather than those who were diagnosed in that year. 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 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 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)[161]."*

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 additional guidance has been provided to hearing professionals to clarify the type of cases that 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 re-launched 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 and these are documented.

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 now likely that the Database has been receiving notifications for between 65% and 80% of all new cases diagnosed each year.

As time passes, we hope to maintain or 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, in which every person identifying with a specific ethnicity is included in that specific grouping[162]. 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 27 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)[163]	World Health Organisation[164]	CDC[165]	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 27: 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 27.

Appendix G: Use of interpolation

Table 22 shows the severity of hearing losses notified between 2010 and 2019, calculated in two ways. The first of these is using data containing all eight data-points, while the second includes interpolation[i].

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

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

Key points:

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

i 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.

Glossary

Kuputaka

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[166]. Implementation of changes proposed in the Wilson Report (2011) were completed in 2015, meaning AODCs are now working with an 'Early Years' focus, on those 0-8 years of age.

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 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 provided educational programmes and services to Deaf and hard of hearing students in the northern part of New Zealand, roughly from Taupo northwards until 2020. Since 2019,

Ko Taku Reo has provided services nationwide, replacing van Asch and Kelston Deaf Education Centres.

Ko Taku Reo – Deaf Education New Zealand: New Zealand's provider of education services for Deaf and hard of hearing (DHH) children. Established in 2020, this organisation replaced the Kelston and van Asch Deaf Education Centres.

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.

Mātua: (noun) parents - plural form of matua (Source: Māori Dictionary).

Mahi: (verb) to work, do, perform, make, accomplish, practise, raise (money) (Source: Māori Dictionary).

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.

Rangatahi: (noun) youth/young person (Source: Māori Dictionary).

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: (verb) to be young, (noun) children – normally used only in the plural (Source: Māori Dictionary).

Tauira: (noun) student, pupil (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 provided educational programmes and services to Deaf and hard of hearing students, from roughly Taupō southwards until 2020. Since 2019, Ko Taku Reo has provided services nationwide, replacing van Asch and Kelston Deaf Education Centres.

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 was adapted from a helpful description found on the KDEC website, which no longer exists.

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