Deafness Notification Report 2021

Rīpoata Whakamōhiotanga Turi

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

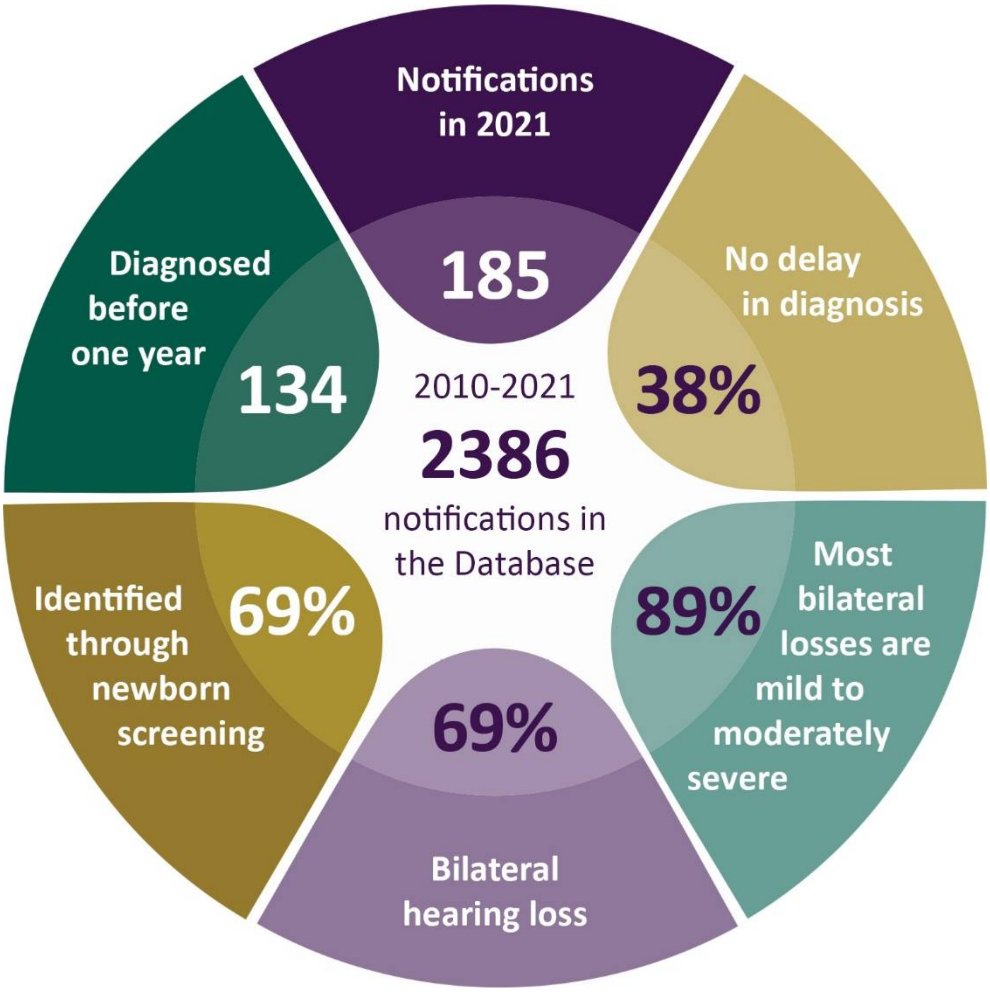
Janet Digby, Levare Limited

December 2022

Enable NEW ZEALAND

Summary

Whakarāpopoto



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

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The Deafness Notification Database

Te Pātengi Raraunga Whakamōhiotanga Turi

- **Our sincere thanks to the mātua (parents)/kaitiaki (caregivers) and rangatahi (young people) who consented to share details of their child's/their own hearing loss, and to the many clinicians around the motu for sending us notifications. Nga mihi maioha ki a koutou.**
- **By entrusting us with their data, we have been able to understand more about those children and young people diagnosed with hearing loss in Aotearoa New Zealand and the nature of their hearing losses. This, in turn, is being used to inform clinicians, decision makers and whanau to help those diagnosed to date and in the future.**
- **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 three additional groups of children and young people; those born overseas, those with unilateral hearing losses and those whose hearing losses are acquired after birth.**

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 2021.

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 2021 calendar year.

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

Where 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[i].

"Ka mua, ka muri"

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

Whānau of children who have had their hearing screened through the Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP) sign a consent that includes sharing information with this Database, while others sign a separate consent presented by the audiologist or audiometrist.

The analyses contained in this report generally pertain to 2386 children and young people notified with a hearing loss diagnosed between the start of 2010, when the DND was relaunched, and the end of 2021 where notifications were provided before our March 2022 cut-off date.

i Further information about consent processes can be found in the section on Notifying Cases on page 76.

Since 2010, the Database has included children and young people 18 years or younger, born in Aotearoa 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)[ii] for pure tone audiometry and 30dB HL or greater over four frequencies for ABR.

The database has included tamariki born overseas, those with acquired hearing losses and those with hearing losses which include one ear (unilateral) only since 2010.

This report and the DND generally exclude children with Auditory Processing Disorders. For those interested, comprehensive *New Zealand Guidelines* were published by the New Zealand Audiological Society 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[iii].

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

- Making notifications to the Database – see Appendix A on page 76 if you are an audiologist or audiometrist and wish make notifications.
- *History of the Database and changes to the inclusion criteria* - see Appendix B: History of the Database, *on page* 76.
- *Terminology used in this report to describe hearing losses* - see Appendix E: Terminology used in this report, *on page* 80.
- *The completeness of notifications* – see Appendix C: Completeness of notifications, *on page* 79.
- Commonly used terms can be found in the Glossary, which begins on page 82 of this report.

Acknowledgements

We extend our sincere and heartfelt thanks to the 185 parents (mātua), caregivers (kaitiaki) and young people (rangatahi) who consented to share details of their child's/their own hearing loss for the Database in 2021.

As a result of this willingness to share basic diagnostic information, service providers can be better informed about current and likely 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 in future.

The time taken by audiologists and audiometrists to make notifications and to do this in such a careful and considered way is also greatly appreciated. It is clear from how this is done, including by departments which are under strain, that diagnosing clinicians care deeply about the wellbeing of both their patients and their whānau.

This report has been funded by Enable New Zealand, through a contract with the Ministry of Health (MOH). The reports' current authors would like to thank the Ministry of Health 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 (Te Rarawa, Ngāi Takoto) of the University of Auckland and Dr Andrea Kelly

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

iii 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.

of Auckland District Health Board. Their input into these reports is significant and greatly appreciated. Ngā mihi nui ki a kōrua.

Contact details

In 2019, readers of this report were approached for feedback on the future direction of these reports. A summary of the results is here.

Feedback from that survey resulted in several changes to recent reports, including the addition of key points at the beginning of each section.

Further feedback on this report is always 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 before the deadline for 185 children and young people diagnosed during 2021, most of whom were born in Aotearoa New Zealand. The year was (again) extraordinary because of the COVID-19 pandemic; the resulting challenges to service provision were significant though generally well-managed to reduce delays in screening, diagnosis and intervention for children and young people, and their whānau.**
- **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. Twelve percent of tamariki (children) and rangatahi (young people) notified to the Database between 2010 and 2021 had one or more confirmed 'additional disabilities' at the time their hearing loss was notified, though later diagnosis of these is common. The most common types are syndromic, medical and neurodevelopmental in nature.**
- **A little over two thirds of notifications to the DND are for children and young people with bilateral hearing losses (69%) with the rest being for those with unilateral hearing losses.**
- **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 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.**
- **Almost 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 eighty-five children and young people diagnosed during 2021, and whose hearing losses met the criteria for inclusion, had their information notified to the Database by 18th March 2022, this year's cut-off date for notifications[i, ii]. There are now 2386 cases included in the main dataset that forms the basis for analysis within this year's report.

These notifications were received from a total of 46 audiologists and audiometrists, with notifications from 19 of the 20 district health boards (DHBs).

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.

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

To maximise the number of notifications to the Database, ongoing 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 rangatahi with hearing loss.

Number of notifications

Figure 1 shows the number of notifications that met the criteria for the main dataset in each year[ii].

Since 2010, these totals may differ from the number of notifications received by the cut-off date for each year's report[iii].

For example, by March 2020, 188 notifications had been received for the 2019 notification year. Since then, an additional 19 notifications have been received for children and young people diagnosed during that year, as shown in the graph below.

One reason for late notifications is that 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.

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

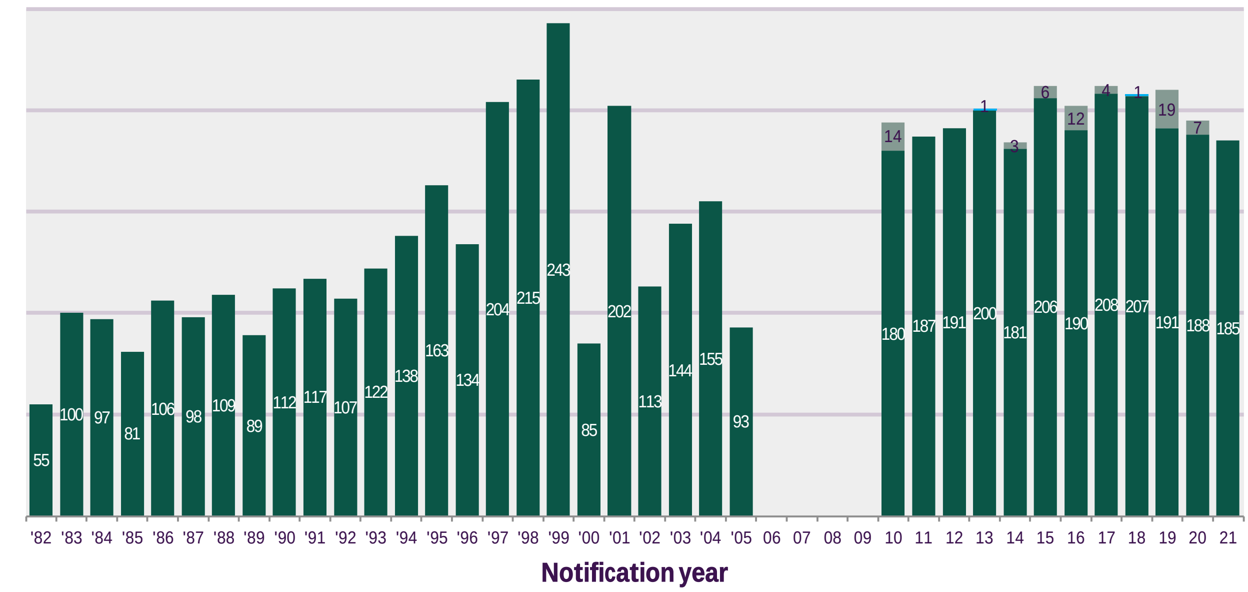

Figure 1: Notifications by year 1982-2005 (numbers included in each year's report) and 2010-2021

(*number of records contained in the database as at the time of publication in dark green with subsequent additions in green and subtractions in blue*)

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i 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.

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 indicate 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 in this current series 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 within this report. Specific changes are described in detail in the reports in which these were first made. Previous DND reports can be found on the New Zealand Audiological Society website.

Another extraordinary year

Before detailing further findings relating to notifications received for the 2021 calendar year, it is important to again acknowledge the extraordinary and challenging nature of the year resulting from the COVID-19 pandemic.

Continued lockdowns meant services, including those focused on tamariki and rangatahi who are hard of hearing, were again significantly interrupted, particularly in Tāmaki Makaurau (Auckland).

Alert Level Changes

There were several alert level changes during 2021 and the end of the year saw the introduction of a new system to indicate the level of restrictions, the COVID Protection Framework.

Alert Level 4 Newborn hearing screening continued to be provided before discharge for babies born in hospital/birthing centres during 2021 as an essential service as outpatient facilitated screening was generally unavailable.

Outpatient and audiology appointments were prioritised during that time for acute cases. Non acute appointments were paused[i].

Acute cases that were prioritised for diagnostic audiology included: babies with meningitis meningoencephalitis, or meningococcal septicaemia; babies with cytomegalovirus (CMV), Down Syndrome or other syndromes associated with hearing loss those with head/brain trauma; and those referred from newborn hearing screening who were eight weeks of age or older, particularly those with a bilateral screening referral.

Alert Level 3 Across the DHBs there were differences in timing on the restart of normal services. For many DHBs, outpatient services did become available with priority placed on babies who had commenced screening or who were six weeks or older. Some of the big metropolitan DHBs (such as Auckland and Waitematā) resumed normal testing and others only offered testing for acute and prioritised cases.

Alert Level 2 Services, including general outpatient screening, returned to normal with safeguards in place for staff and whānau. During this time DHBs worked quickly to develop localised strategies to support as many parents as possible to attend outpatient appointments safely, and to catch up on those babies who had missed their first screening or follow-up appointments. The timeliness of catch-up was particularly important for those older babies who were now less likely to sleep (be settled enough) for an aABR screening.

These changes were implemented despite the additional challenges associated with the fact that some members of the screening workforce are older and did not wish to continue screening due to COVID-19 restrictions.

COVID-19 Alert Level and Protection Framework Changes during 2021	
COVID Alert Level system	
14th February	Level 3 Lockdown begins in Auckland, rest of New Zealand moves to Alert Level 2
22nd February	Level 2 Lockdown begins in Auckland, the rest of New Zealand moves to Alert Level 1
28th February	Auckland moves to Alert Level 3. The rest of New Zealand moves to Alert Level 2.
12 March	Auckland moves to Alert Level 2 at 6am. The rest of New Zealand moves to Alert Level 1.
23 June	Wellington moves to Alert Level 2 at 11:59pm.
29 June	Wellington moves to Alert Level 1 at 11:59pm. All of New Zealand is now at Alert Level 1.
17 August	All of New Zealand moves to Alert Level 4.
31 August	All of New Zealand south of Auckland moves to Alert Level 3.
2 September	Northland moves to Alert Level 3. All of New Zealand (except Auckland) is now at Alert Level 3.

i Priority cases were further refined when the Delta variant arrived in Aotearoa New Zealand.

COVID-19 Alert Level and Protection Framework Changes during 2021 *(cont)*	
7 September	New Zealand (except Auckland) moves to Alert Level 2.
21 September	Auckland and Upper Hauraki move to Alert Level 3.
25 September	Upper Hauraki moves to Alert Level 2.
3 October	Raglan, Te Kauwhata, Huntly, Ngāruawāhia, Hamilton City and some surrounding areas move to Alert Level 3 for 5 days.
5 October	Alert Level 3 restrictions in Auckland are eased.
7 October	Waikato Alert Level 3 boundary is extended to include Waitomo District, including Te Kuiti, Waipa District and Ōtorohanga District.
8 October	Northland moves to Alert Level.
19 October	Northland moves to Alert Level 2 at 11:59pm.
27 October	The parts of Waikato at Alert Level 3 move to Step 1 of Alert Level 3.
2 November	Upper Northland moves to Alert Level 3. The parts of Waikato at Alert Level 3 Step 1 move to Alert Level 3 Step 2.
9 November	Auckland moves to Alert Level 3 Step 2.
11 November	Upper Northland moves to Alert Level 2.
16 November	Parts of Waikato move to Alert Level 2.
COVID-19 Protection Framework	
From December 2nd 2021	Northland, Auckland, Taupō and Rotorua Lakes Districts, Kawerau, Whakatane, Ōpōtiki Districts, Gisborne District, Wairoa District, Rangitikei, Whanganui and Ruapehu Districts begins at Red level, the rest of North Island, South Island at Orange.
30 December 2021	Northland - Red Rest of North Island, South Island - Orange

Screening and audiology services

The National Screening Unit, in collaboration with DHBs and the New Zealand Audiological Society's Paediatric Technical Advisory Group (PTAG), retained and expanded the National COVID-19 Strategy to support newborn hearing screening and diagnostic audiology provision across all alert levels.

The scope of services provided at each alert level was managed to minimise risks to staff and whānau and their babies. Delivery of equitable services to ensure all babies had the opportunity to have their hearing tested remained a priority.

Where newborn hearing screening or diagnostic audiology services could not be delivered, the DHBs were asked by the Ministry of Health's Universal Newborn Hearing Screening and Early Intervention Programme to have plans in place to track babies who did not commence or complete the pathway and that supported 'catch-up' for babies not seen, once services could recommence.

Anecdotally, while some DHBs reported higher levels of screening declines during Alert Level 4, many also reported fewer Did Not Attends because of the extra clinics being put in place, which better met need.

Screening catch-up was achieved through extra outpatient clinics and for some DHBs, having audiology complete screening using OAE. Approaches did vary, but for those DHBs outside of metro Auckland most achieved this within a month. For the metropolitan Auckland area this impact was more significant, but each adopted a strategy based on the wider options developed with the PTAG. This is thought to have helped them manage to continue to deliver levels of service that meant coverage remained at a reasonable level and made catch-up more manageable.

While COVID-19 has proven challenging in many ways for both whānau and hearing kaimahi/staff, it created some opportunity for service innovations, a number of which continued in 2021 after being established in the first year of the pandemic.

B4SC screenings were paused during early childhood and school closures for children in the general population. In addition, B4SC screening staff and their public health nurse colleagues were, in some areas, seconded from regional public health to the COVID response meaning screening was stopped or reduced considerably, in some cases for a significant period of time. Some audiologists noticed a significant reduction in referrals from these services.

Data provided by the New Zealand Audiological Society confirm that early 2020 saw a reduction in the number of audiology roles being advertised around the country (for both private clinics and DHBs), as the first lockdown hit. Numbers of advertisements for both DHBs and private clinics bounced back somewhat during 2021 and again in 2022 though the increase in advertisements reported in 2022 is also likely to reflect the introduction of a new contract NZAS has with one company which runs regular advertisements each week[2].

Acknowledgements

The National Screening Unit has asked that PTAG and the coordinators, audiologists and service managers who provided their expertise and advice to the NSU be gratefully acknowledged for their mahi during 2021. The ongoing success of the programme during the year was thanks to their efforts, their support and advice, and effective collaboration to find solutions that worked for the DHBs, that maintained quality in the programme and that worked towards the collective goal of enabling all babies in NZ to have their hearing tested.

In addition, the Screening Unit singled out the extraordinary work of screeners and audiologists in the metro Auckland area during 2021. The resilience of these teams, and their willingness to keep going during such a long period of adversity was impressive and demonstrated considerable tenacity and commitment by staff. This meant the UNHSEIP could continue to deliver, ensuring babies were able to have their hearing assessed as soon as possible.

The resilience of the metro Auckland DHBs has been specifically acknowledged as impressive during the very long lockdown experienced in this area.

The authors of this report would like acknowledge the contributions of Kylie Bolland (Chair of the New Zealand Audiological Society's Paediatric Technical Advisory Group) and Dr Samantha Everitt of the National Screening Unit for this section of the report.

Education

The following update was provided by the National Director Learning Support Delivery, within the Ministry of Education, Dr David Wales.

"At Alert Levels 3 and 4, children and young people were advised to learn from home, although at Level 3 the children of essential workers were able to attend school/kura or early learning centres if they were unable to be supervised at home.

Advisors continued to keep in touch with families by phone, Zoom or social media where meeting in person was not possible.

NZSL tutoring and teaching took place online during Levels 3 and 4 via individual and whole class online learning sessions.

Advisors on Deaf Children: preliminary data available to the Ministry of Education suggest there was no noticeable reduction in the number of children of new requests for support identified with hearing loss through the UNHSEIP growing from 153 in 2020 to 189 in 2021 and the total number of children being supported by services growing overall. *[See the section titled Ministry of Education from page 69 for more information].*

There are no data yet on whether the COVID-19 pandemic may have resulted in later referral for educational support among tamariki and rangatahi who are hard of hearing, including because these children were not in formal learning environments during a good part of 2020, or because difficulties experienced were seen as secondary to other concerns during what was psychologically and financially an exceedingly difficult year for so many.

Anecdotal evidence suggests that some families were anxious about sending their children back to school following lockdowns and some kept their children home after services resumed. In such cases, this may have had also had negative effects on their education and/or hearing aid use."

Gender

Background

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[i] show a similar pattern, with higher numbers of hearing loss among males (52.2%) than females (47.8%) in 2019[5, ii], although ACT and Southern Australia have a ratio approaching 1:1 and those aged 21-25 years of age contain a predominance of females.

Local data

Of the 2386 cases (2010-2021) contained in the main dataset, 45% of these are listed as female (n=1072) and 55% male (n=1314). This represents a ratio of 1: 1.24.

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 within any notifications.

This gender difference was particularly noticeable in 2016 and 2020, which approached or reached a ratio of 60 males for every 40 females notified[iii].

Birthplace

Tamariki born outside Aotearoa New Zealand have been formally included in the Database since 2010.

Figure 2 shows the proportion of cases notified by birthplace for the 2010-2021 period. During that time, 6% of children and young people notified have been born overseas, with the birthplace of an additional 5% being uncertain.

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 1-3% in 2017-2020. 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 because of newborn hearing screening.

Of the 185 notifications to the Database in 2021, 4% were known to be born outside Aotearoa New Zealand, a drop on the figure reported last year though that figure has fallen with the inclusion of late notifications. Lack of certainty around birth-place was listed in a further 1% of cases.

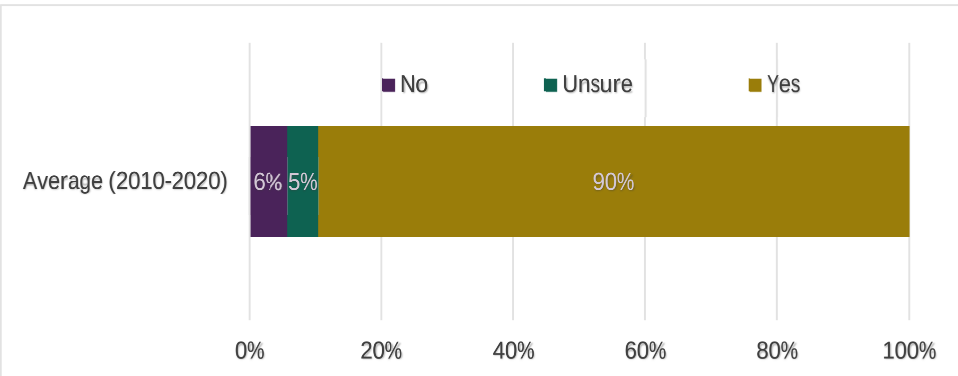


Figure 2: Proportion of cases born in New Zealand (2010-2021)

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

ii 0.1% of cases were of unknown gender.

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

DHB representation

Table 1 contains the percentage of 2021 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 the table shows the percentage of notifications received for 2010-2021 from each district health board – this can be compared with their relevant percentage in the population for those under the age of 20[ii].

Tamariki notified to the Database are more likely to be of Māori ethnicity than their proportion in the general population would predict. As a result, DHBs with more than 20% of their population identifying 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 what was Kelston Deaf Education Centre (KDEC) (Auckland) or van Asch Deaf Education Centre (VADEC) (Christchurch) and is now Ko Taku Reo .

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 capacity and commitment among staff to making notifications to the Database.

A recent local study[iii], described in previous DND reports, found that only 56% of tamariki/rangatahi were still in the care of the notifying clinic (often the DHB's audiology service) seven or eight years after their diagnosis. For those who were still in the care of the notifying clinic, 31% had not been seen by that clinic for at least two years. Of the 163 children and young people about whom follow up information was provided; the notifying clinic held no information about fifty-nine children and young people.

We understand from speaking with audiologists on the Paediatric Technical Advisory Group (PTAG), that it is possible that DHBs who provided the notifications to the DND 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. This means the audiologist may not know where the child or young person was now receiving care.

Some district health board audiology services have the ability to search for individuals outside their catchment (e.g. there is a database for those in the South Island that is searchable), while others do not. This may be improved by the new 'regional teams' focus.

These figures demonstrate the importance of functional clinic information systems and of communication between clinics to ensure tamariki and rangatahi are not lost to follow-up when families move between areas.

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

ii Please note, these percentages are rounded.

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

District Health Board	Percentage of population under the age of 20 (Statistics New Zealand, 2018 Census[7])	Percentage of notifications received in 2021 (under 19 years)	Percentage of notifications received 2010-2021 (under 19 years)
Auckland	9%	5%	6%
Bay of Plenty	5%	6%	7%
Canterbury	11%	13%	13%
Capital and Coast	6%	5%	9%
Counties Manukau	13%	19%	13%
Hawke's Bay	4%	3%	3%
Hutt Valley	3%	4%	4%
Lakes	3%	2%	2%
Midcentral	4%	0%	3%
Nelson Marlborough	3%	3%	4%
Northland	4%	3%	6%
South Canterbury	1%	2%	2%
Southern	6%	7%	7%
Tairāwhiti	1%	4%	3%
Taranaki	3%	2%	3%
Waikato	9%	11%	8%
Wairarapa	1%	1%	1%
Waitematā	13%	5%	6%
West Coast	1%	1%	1%
Whanganui	1%	3%	1%

Table 1: The estimated percentage of population under 20 years of age by district health board (2018 Census) compared with Percentage of notifications (2021) and the proportion of notifications by DHB (2010-2020).

Additional disabilities

Introduction

A disability is any condition that makes it more difficult for a person to do certain activities or effectively interact with the world around them (socially or materially).

Increasing estimates of the global burden of childhood disability[i] from 2020 suggest that more than one in ten 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 and this can place an additional burden on families when compared with those tamariki and rangatahi without additional disabilities.

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;

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.

- 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 for children with additional disabilities though it is common for these children to begin to receive intervention at later ages than those without; 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).

Overseas data

While it is difficult to compare reported rates of additional disabilities between groups of children 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.

Source	Date	Location	Details	Rates
Nelson and Bruce[10]	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[11]	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[12]	2012	United Kingdom Review	Review of twelve 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[13]	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.*[14]	2002	United Kingdom	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
Fortnum and Davis[15]	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[16]	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.

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[17].

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 several 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 provide an indication of 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 Database – the one used at the start of this section.

Of the 2386 records in the main dataset, including all children and young people diagnosed with hearing loss in 2010-2021, the majority (77%) have no 'additional disability' listed. Ten percent are listed with a possible although as yet unconfirmed additional disability. Twelve percent have one or more confirmed additional disability(ies). One percent of notifications had no additional disability information provided.

2021 data

Of 2021 notifications, only 12% 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 16% of cases there was uncertainty regarding whether the child or young person had an additional disability[i].

The majority of those who were listed as having an additional disability had one additional disability listed, while smaller numbers had two, three, four or even five noted.

There are now higher numbers of cases within the database compared with previous reports. This is because those who are listed in other parts of the notification form as having atresia and microtia are now included within the 'yes' category.

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. However, 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 at the time of the hearing loss diagnosis.

Additional disability	Number of tamariki	Percentage
Yes	260	11%
Unsure whether AD exists, no confirmed diagnosis	216	9%
No additional disability	1887[ii]	79%
No data	23	1%
Total	**2386**	**100%**

Table 3: Proportion of cases by additional disability status (2010-2021)

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.

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.

ii This figure is lower than in last year's report as those who have been listed as having atresia in the UNHSEIP part of the form have been included as having an additional disability in this year's figures.

Column four of Table 4 shows the total proportion 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.

Notification Year	Cases with a known additional disability	Cases with a possible additional disability	Cases with additional disability (2002-2005) and total confirmed and possible (2010-2019)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	11%	10%	22%
2011	13%	5%	18%
2012	14%	11%	25%
2013	10%	11%	21%
2014	13%	8%	21%
2015	9%	10%	19%
2016	6%	9%	15%
2017	10%	10%	20%
2018	12%	10%	22%
2019	15%	8%	23%
2020	14%	14%	28%
2021	12%	16%	28%
Average 2010-2021	**12%**	**10%**	**22%**

Table 4: Proportion of cases with a known additional disability (2002-2020)

Factors influencing rates of additional disabilities included in the DND

Previously, the authors of this report believed that the earlier identification of tamariki with hearing loss was the likely reason behind the drop in the proportion of those with confirmed additional disabilities reported at the time of diagnosis of the hearing loss. The rationale suggested 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, 19] and that tamariki with hearing loss in Aotearoa New Zealand are not all routinely assessed by a paediatrician.

More recent notifications to the DND (shown in Table 4) show the general downward trend from 2012-2016 has reversed since 2017, growing from 20% in that year, to 28% for 2020 and 2021.

When examining these data, we can see that rates of additional disabilities being present at the time of notification are higher among those diagnosed over the age of two years old, as expected.

Even with average age at diagnosis falling, we can see growth in the proportion of cases with a possible additional disability, with these figures at their highest levels since the Database was

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.

relaunched in 2010. We are unsure why this might have been the case, but perhaps one factor could be that many parents were spending considerably longer with their tamariki than usual due to COVID related school closures, meaning issues they noticed resulted in more prompt identification of additional disabilities compared with before the pandemic.

Immunisation rates

Recently there has been concern regarding immunisation rates, which have fallen from their peak in 2016. These rates are particularly low for Māori tamariki and those who live in income poverty[20].

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."[21]

Since the pandemic began, further reductions in the numbers of children receiving immunisations have been reported, resulting in record low coverage rates. Overall coverage rates have fallen from nearly 80% in 2017 to around 65% in June 2022. Rates of immunisation among Māori and Pacific[22]; just 47% of Māori aged 18 months had full immunisation coverage over the past year, a drop of 26% since the start of the pandemic[23].

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[24, i]. Some children and young people have more than one 'additional disability' listed on their notification form.

In an attempt to better describe the range of additional disabilities seen among children and young people whose data are contained in the Database, we have developed a new approach to grouping these responses by type and we have applied this to all records, as seen in Table 5: Number of cases by type of additional disability (2010-2021)

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.

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 a higher rate of confirmed additional disabilities when compared with their peers who are diagnosed under the age of two (13% vs 8%).

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 parents, caregivers or medical professionals. For example, for 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. Before the implementation of newborn hearing screening, other conditions were often identified first, followed by a diagnosis of hearing loss.

i 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.

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.	75	29%
Medical	Medical conditions and issues, such as cardiac problems, bladder issues, renal issues and lung issues. (Please note that atresia and microtia is not included as an additional disability.)	80	29%
Neurodevelopmental	Issues with the growth and/or development of the brain or central nervous system, such as ADHD, autism, developmental delays, and intellectual disabilities.	73	26%
Sensory	Issues relating to the sensory system that 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	45	16%
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	38	14%
Medical-developmental	Medical conditions and issues related to development such as hydrocephalus and cleft palate	28	10%

Table 5: Number of cases by type of additional disability (2010-2021)[i]

Bilateral and unilateral loss

Proportion of unilateral and bilateral hearing losses

The proportion of 2010-2021 cases in the Database thought to be bilateral/unilateral was 69:31 (see Figure 3, page 21)[ii].

Influences on this proportion

Immunisation coverage (including for conditions such as mumps) in Aotearoa New Zealand rose significantly as described in the previous section. More recently, concerns about falling immunisation rates have been raised, with particular concern expressed about rates for Māori and those living in poverty[20]. Mumps is one cause of unilateral hearing loss.

The number of cases resulting from changes in immunisation is 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[25].

Differences between the proportions of bilateral and unilateral notifications in each severity category are shown in Figure 3 on page 21.

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[26, 27]. About a third of all

i Table 5 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, but both within the same category.

ii 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 rangatahi contained in the Database.

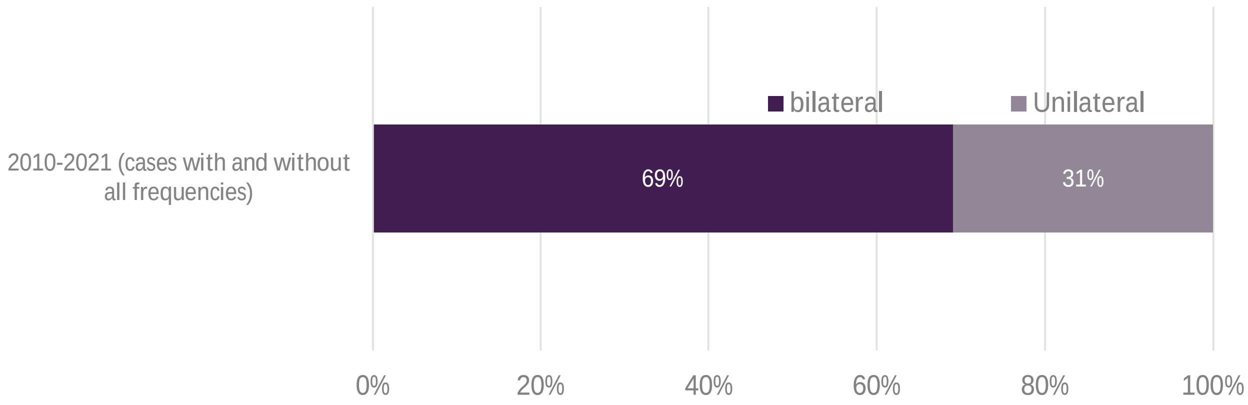


Figure 3: Proportion of bilateral and unilateral cases (2010-2021)

notifications to the Database since 2010 (31%) are for children and young people diagnosed with unilateral hearing loss. Over time, a proportion of those children will go on to have their hearing loss include their other ear.

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[28, 29, 30].

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[31, 32, 33, 34, 35].

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 less support[36].

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*.[37] 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 2.0 kHz) that is greater than 15 dB HL, 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[iv], and that environmental causes are also commonly implicated. As a result, aetiologic assessment following diagnosis, including complete otologic 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.

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[38].

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[39]. Prevalence rates

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.

iv Progressive hearing losses are common in such cases as described here.

rise with age to between 3.0 and 6.3% among children 6-19 years of age, according to Ross *et al.*[40]

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[41, 42, 43].

Here in Aotearoa New Zealand, a recent analysis of data provided for 163 of the 189 notifications to the DND in 2010[44], described in the 2019 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.'[45] 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[46].

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[47].

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[48].

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[37, i].

Single-sided deafness

Definition and management

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 in the previous subsection of 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[49,50]. The boundaries for these 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 that focused on adult research (2016) concluded that no recommendations for the management of unilaterally deaf *adults* could be made based on the current evidence[51].

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 unilateral hearing loss, who may receive a bone conduction hearing aid (e.g. if there is a permanent conductive hearing loss due to aural atresia).

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

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.

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].

DND data

The data contained in Table 6 show the proportion of total notifications each year that met the DND's definition for SSD[ii]. The proportion of 2010-2021 cases[iii] that were SSD is 5.45%.

Children and young people in this category[iv] are not eligible for publicly funded cochlear implants except in the case of meningitis but can opt for privately funded implants or receive implants if they are covered by ACC[52]. It is likely a good number will not have a robust auditory nerve[53], meaning implantation is not valuable for them.

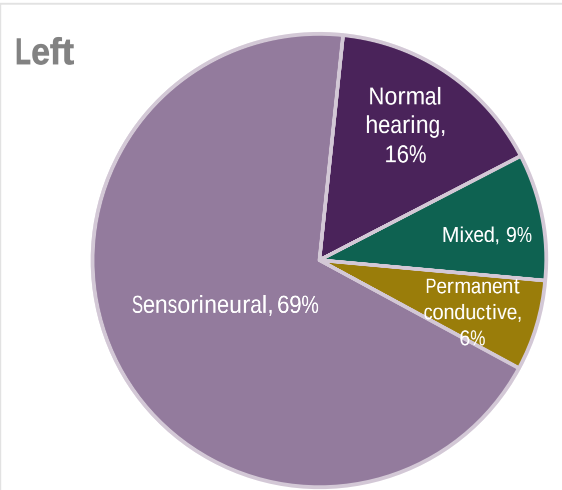
Notification Year	Proportion of cases with single sided deafness
2010	6%
2011	4%
2012	8%
2013	10%
2014	8%
2015	5%
2016	5%
2017	6%
2018	4%
2019	5%
2020	4%
2021	2%
Average 2010-2021	**5%**

Table 6: Single-sided Deafness Cases by Year (2010-2021)

Types of hearing loss

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

The 2013-2021 data for this question are contained in Figure 4. 'ANSD' (Auditory Neuropathy Spectrum Disorder) is offered as an option within sensorineural hearing loss (SNHL) and is not split out in the Figure.



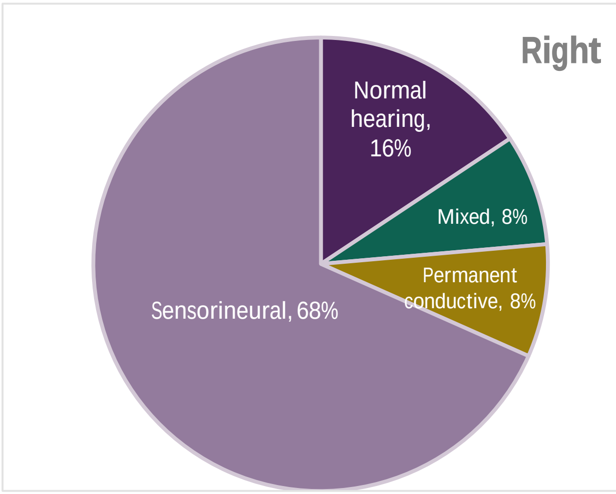


Figure 4: Type of hearing loss (2013-2021)

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 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.

iii Based on determinations including interpolated data.

iv Where the worse ear has a severe hearing loss or worse from 1kHz to 8Khz

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

The most commonly reported type of hearing loss contained in notifications was sensorineural (68% in the left ear and 68% in the right), followed by normal hearing (16% in the left ear and 16% in the right). 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.

Auditory Neuropathy Spectrum Disorder

Three percent of children's right and left ears were recorded in the ANSD category.

Prevalence of ANSD among those children with permanent hearing loss may be approximately 10%, according to a 2015 review by Rance[54]. Among those from the Avon newborn hearing screening programme in England,[55] 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 population, also supported by our lower proportion of severe and profound hearing losses when compared with other jurisdictions examined.

One factor contributing to variations in reported prevalence of ANSD could be differences in whether auditory nerve hypoplasia or aplasia are included[56]. In Aotearoa some of these cases may be included in the SNHL category.

An analysis of the types of hearing loss among 2010-2016 notifications, included in a previous report,[57] 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].

A repeat of the type of loss by ethnicity for 2010-2020 data also found higher proportions of mixed losses in this group, and lower proportions of this type of hearing loss among those children and young people listed as Asian.

Hearing loss present at birth

Of all 2010-2021 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 response to this part of the form was provided, the audiologist indicated they were 'unsure' in 39% of cases, with the hearing loss likely to have been present at birth in 47% and not 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.

2010-2021 data continues to show a similar pattern, with European children and young people less likely (43%), and those listed as Māori and/or Asian being the most likely (50% and 59%) to have been listed as having a hearing loss thought to be present at birth.

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

Family hearing history

The question in the DND relating to family history is 'Does an immediate family member (only a mother, father or sibling) have a permanent hearing loss?[i] (or had a permanent hearing loss if they have died).' This question was introduced part way through 2014.

The results for this question are shown in Figure 5[ii]. That figure shows data from 2015-2021 notifications – years containing responses to this question for all cases. 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 65% to 81% during that time, with between 13% and 22% listed as having one (or more).

When 2021 figures are examined in isolation, they show the highest proportion of children diagnosed with no family history of hearing loss (81%)[iii].

This year the likelihood of children and young people having an immediate family member (only a mother, father or sibling) with a permanent hearing loss close was examined for each of the largest ethnic groups.

Further analyses shown in Table 7 shows that:

- Asian children and young people are the least likely to have a close family member with a permanent hearing loss (7%); and
- Māori children and young people in the database are the *most likely* to have close family member(s) with a permanent hearing loss (23% respectively).

See also the section in this report on Aetiology which begins on page 36.

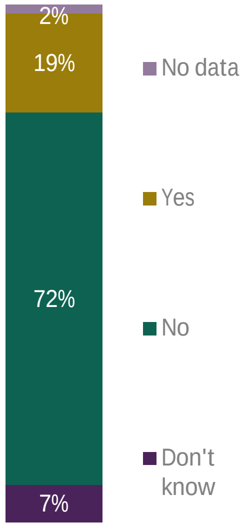


Figure 5: Immediate family member with hearing loss (2015-2021)

Changes in Connexins are known to be the most common genetic cause of hearing impairment among those without syndromes in many populations. A systematic review of the published literature including 571 studies found different distributions of Connexin in Asian than in

Ethnicity	No	Yes	Don't know
Asian	87%	7%	7%
European (this includes NZ European)	76%	19%	5%
Pacific Peoples (includes Cook Island Māori)	70%	20%	9%
Māori	67%	23%	10%

Table 7: Likelihood of close family member with permanent hearing loss (2015-2021)[iv]

European populations[58]. No studies have been undertaken to establish what groups in Aotearoa New Zealand have the highest prevalence of hearing loss which results from genetic changes.

i 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 grand-parent, and then about each specific relative. Between 13% and 24% of cases reported a 'family history of hearing loss' between 2010 and 2013.

ii 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. 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 2015-2020 in Figure 5.

iv Figures without data have not been included in this table. In each case they comprise 2% or less in each row.

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 within this group based on the size of their population under 20 years of age. Those in this group are less likely to have a hearing loss present at birth.**
- **Disparities across the health system have been well-documented for Māori in terms of their access to, and through, the health system. Research on equity for hearing services is limited but shows similar patterns.**
- **Hearing losses among Māori children and young people may not be notified to the Database as consistently as other groups, including as they are more likely than their European counterparts to have a less severe hearing loss. Even considering this, the number of notifications from those of Māori ethnicity are higher than expected based on their population and this pattern is confirmed by other sources.**
- **Overall, Pacific and Asian children and young people are notified to the Database in proportions roughly equivalent to their relative population size for this group.**

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 76.

Full dataset

Of the 2386 notifications in the main dataset (covering 2010-2021 notifications) all but 28 (<1%) contain at least one ethnicity code. The number of notifications containing no ethnicity codes has dropped from an average of 1.74% in 2010-2015 to 0.75% in 2016-2021.

Most notifications (89%) contain one code, and a smaller proportion (9%, 0.8% and 0.04%) contain two, three or four codes, respectively.

Multi-coded 2018 Census data are included for comparison in Figure 6. As individuals may identify

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.

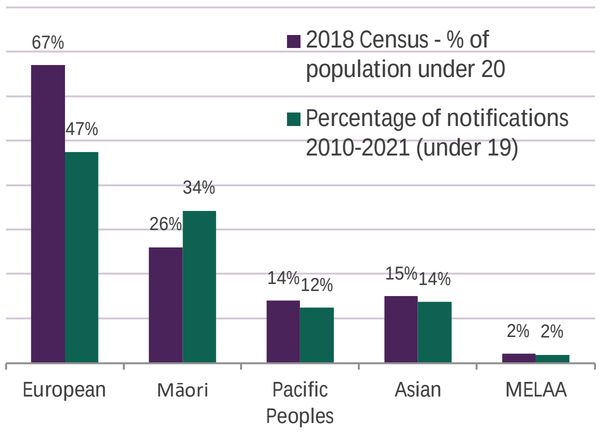


Figure 6: Notifications by ethnicity (2010-2021) compared with Census data (2018)[59]

(or be identified by their parents) as belonging to 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 notifications from 2010-2021, which includes those under the age of 19[i].

Please note that MELAA figures for bilateral and unilateral hearing losses reported in this figure relate to a very small number of cases (n=41).

The European ethnic group was still the largest in the Census by a significant margin, at 67% of the population under 20 years of age but only makes up 48% of notifications to the Database.

Those of Māori ethnicity are over-represented in the Database, comprising 34% of notifications and 26% of the population under 20 years of age.

Those children and young people of Asian or Pacific ethnicities are being diagnosed in approximately the same proportions as would be expected by their population under 20 years of age.

Note that 29% of notifications from Auckland and Waitematā DHBs (2010-2021) are listed as Asian, more than double the overall proportion for the country as a whole. These DHBs report that Asian children are overrepresented in their diagnoses when compared their relative population size.

Unilateral and bilateral hearing losses

Of 2010-2021 cases, including those with interpolated audiometric data, 69% are recorded as bilateral, while the remaining 31% are unilateral.

Figure 7 shows a comparison of the percentage of bilateral and unilateral notifications for each ethnic group during the 2010-2020 period. These data include not only cases where data has been interpolated, but also those with one or more frequencies remaining missing after interpolation. 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[69].

Differences can also be seen when comparing bilateral losses among Māori tamariki[ii] notified between 2010 and 2021 (76%), with those who are European[iii] (65%), and those described as both Māori and European (71%)[iv].

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). Data from children and young people included in the main dataset (2010-2020) show that 11% of Māori tamariki compared with 18% of European children were not thought to have a hearing loss present at birth.

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

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

iii 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.

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

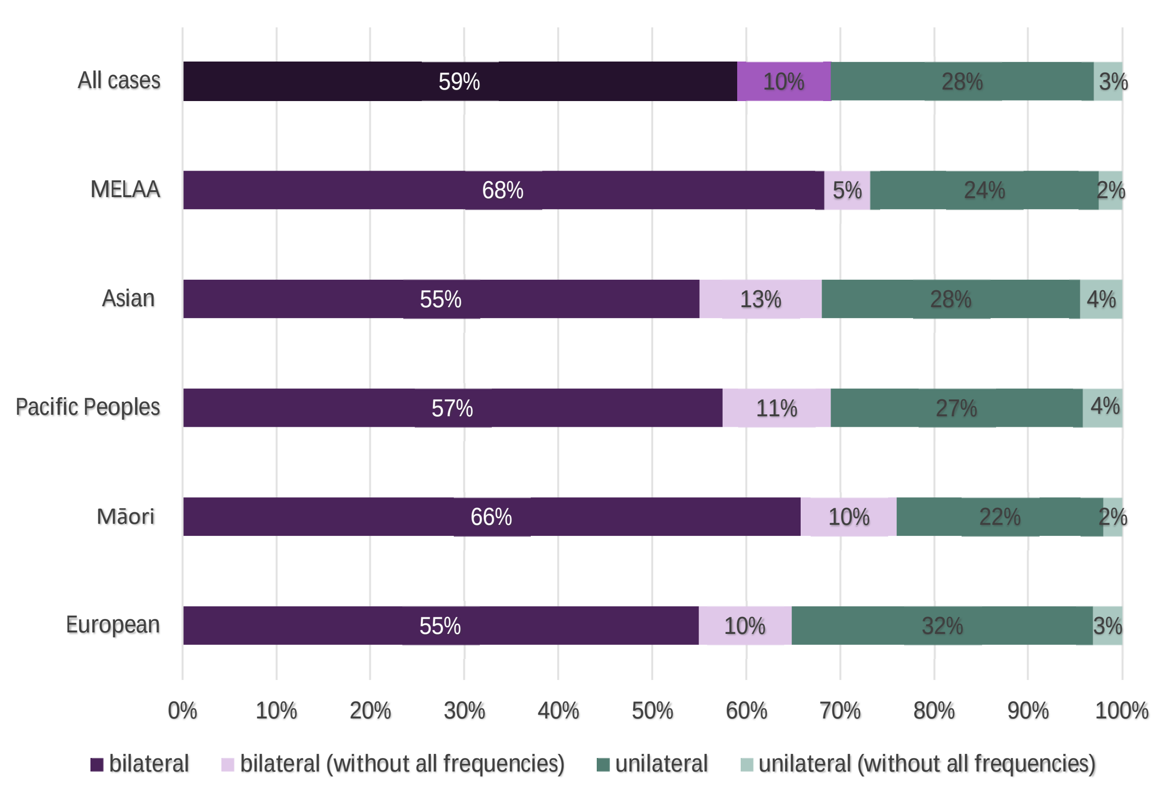


Figure 7: Proportion of unilateral and bilateral hearing losses by ethnicity (2010-2021)[i]

Hearing loss among Māori

Prevalence

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

As mentioned above, while the proportion of notify-cations from those of European ethnicity are con-siderably lower than one would expect based on the size of their population, and notifications from those of Māori ethnicity are higher than expected.

Several sources reinforce the higher prevalence of hearing loss comparing 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"[60].
- Greville (2001) found higher prevalence of temporary and permanent hearing loss among Māori children[61].
- 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[62].
- Household Disability Surveys:
  - » 1991-2006 Surveys[63] suggest Māori had higher rates of hearing disability (tamariki and adults) and higher rates of unmet need for technology and equipment when compared with non-Māori[64]. (For information about the limitations of these data please see the 2011 DND Report[65].)
  - » The 2013 Survey continues to suggest Māori had higher unmet need for technology and equipment when com-pared with non-Māori[66] but also that they now have *lower* rates of hearing disability compared with their European counter-

i Based on interpolated data and manual checks to determine bilateral/unilateral status
ii In this report the New Zealand Māori ethnic group is referred to as Māori.

parts[67], 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[68].

- Findings from Digby *et al*. (2014) indicated young Māori have higher rates of permanent hearing loss than their European peers, based on the previous and post re-launch DND datasets, which included notifications from 1982-2005 and 2009-2013[69].
- 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%)[70] and this pattern still holds with 2020-2021 B4SC data showing 4% referral rates for Māori, compared with 3% for NZ European children and young people as shown on page 52[ii].

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[71] 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 New Zealand's Universal Newborn Hearing Screening and Early Intervention programme (UNHSEIP) does not target or identify all mild hearing losses[iii, 72]. Māori have higher proportions of these hearing losses when compared with other ethnic groups including Europeans. The B4 School Check targets mild and greater hearing losses[73].

Unequal health access and outcomes for Māori

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[74, 75, 76, 77, iv].

Disparities documented in many areas of health demonstrate Māori have poorer access 'to, and through' the health system[71, 78, 79], that they receive a poorer and slower service, and are less likely to receive appropriate levels of care[80], 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 and there is some indication that engagement with and recognition of Māori has actually been dismantled in some areas[81, 82, 83].

Both the Waitangi Tribunal 2575 inquiry (Stage One)[84] and the New Zealand Health and Disability System interim report[85] 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[v]. The Treaty guaranteed Māori their full rights and benefits as citizens.

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

ii It is important to note that high referral rates for Māori may relate to higher rates of ear disease, as referral doesn't only relate to permanent hearing loss.

iii "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.

iv 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*.

v 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.

The Tribunal's Stage One report acknowledged that while the health sector is not able to influence all the social determinants of health, persistent inequalities constitute health sector Treaty breaches. It recommended 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 asserted DHBs and other health agencies were 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[86].

A number of district health boards (now districts within Te Whatu Ora) have in recent years re-asserted their commitment to achieving equity for Māori, including Northland [87] and Auckland ,[88] and 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 Europeans, despite the overrepresentation of these groups in industries where noise exposure is higher, and a higher prevalence of hearing loss overall[89].

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[90].

As described by Pokorny *et al.* (2022) referral rates for Māori do not reflect their increased rates of hearing loss and ear disease[149]. Māori appointment attendance rates remained 64% lower in their analysis than non-Māori even after adjusting for socio-economic deprivation, waiting times and telephone contact.

The latest data from the Atlas of Healthcare Variation (Surgical Procedures) suggests that public grommet insertion rates are low in some areas compared with the national average, particularly in 0–4-year-old Māori and Pacific[91]. (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 recorded as Māori are less likely to have their screening completed than their European counterparts[62].

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[i] differ, as do waiting times.

Asian tamariki

It is important to note that (as with Pacific Peoples) Asian New Zealanders are far from a homogenous group – this group contains children and young people from many different countries and ethnicities.

Among the differences experienced by this group when considered as a whole, compared with other ethnic groups commonly found in Aotearoa, they have:

- a higher proportion of severe and profound hearing losses;
- a lower likelihood of having not attended appointments or rescheduled these (for any reason);

i DHBs see most tamariki and rangatahi with hearing loss.

- a lower likelihood of experiencing waits to see a hearing professional or accessing services in their area; and/or
- a tendency to have good access to and through other parts of the health system, as demonstrated by their high vaccination rates[92].

Deprivation

Pōharatanga

- **Deprivation scores in the New Zealand Index of Deprivation are drawn from Census data and indicate the level of deprivation for each of many small areas in Aotearoa New Zealand. As a whole, 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 around the motu with one or more disabilities are also more likely to live in areas of higher deprivation than those without. No such correlation exists in the United Kingdom, where disability allowances are much higher.**
- **Our DND data show that children and young people notified to the Database who are of European ethnicity are much less likely to be living in the most deprived areas than those of Māori and/or Pacific ethnicities.**
- **As income and poverty are significant determinants of health, professionals seeing children with hearing loss can expect to see poorer health among these families, but particularly for those identified as Māori and/or Pacific. This is likely to result in greater barriers to engagement with hearing and other services.**

Overview

International data demonstrates prevalence of congenital hearing loss is lower in countries with higher incomes. Lower levels in higher income countries are thought to be due to lower infection rates and better access to preventative measures and healthcare services[93].

The New Zealand Child Poverty Monitor reports note that Aotearoa 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[94].

Tamariki with disability and deprivation

Child Poverty Action Group (NZ, 2015) report that children with disabilities in Aotearoa New Zealand are at a greater risk of living in low-income households than those without such disabilities[95].

Statistics New Zealand reports that overall, 11% of children under the age of 15 have a disability.

Once adjustments are made for differences in age profiles by population, Māori and Pacific[96] groups are also more likely to be living with low incomes.

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 use some medical services less frequently[97].

However, Child Poverty Action Group (NZ) also note that such differences are not inevitable and cite the United Kingdom's much higher disability allowances, which is thought to be the reason there is no correlation between childhood disability and poverty in that country[98].

Introduction to the New Zealand Deprivation Index

Here in Aotearoa 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).

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 Aotearoa New Zealand.

The variables used to determine the deprivation score for a specific meshblock (small area) are contained in Table 8.

Deprivation data provided by Te Whatu Ora | Health New Zealand has been included in our DND analyses since the 2016 report[i]. Data for this report are based on information provided by the Ministry of Health and is based on NZDep2013 as NZDep2018 does not currently have domicile code mapping.

These 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[99]. The deprivation scores allocated to the primary addresses associated with each National Health Identifier are used in this analysis. As at the time of writing, these are provided to us by Te Whatu Ora | Health New Zealand[ii].

Of the 2386 tamariki in the main dataset, 98% had deprivation data available[iii].

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 dataset are much more likely to live in high deprivation areas than lower deprivation areas when compared with the population at large, and with children generally.

- Only 7% of children in our dataset (2010-2021) are living in NZDep areas scoring a 1 on the index (the lowest deprivation areas), compared with 10% in the New Zealand population at large. In comparison, 19% of children included in the dataset are living in NZDep areas scoring a 10 (highest deprivation areas), almost double the 10% found in the New Zealand population at large.

i Though recent reports now include deprivation data for the full dataset.

ii Please note that NZDep scores relate to the addresses at which tamariki were living at the time the Ministry of Health provided the deprivation score of their area from the NHIs provided - it does not relate to specific individual's level of deprivation.

iii Data were unavailable for tamariki whose: NHI was not valid, those who had no NHI listed, those whose notification came after the deprivation scores were provided by Te Whatu Ora, and those who live outside New Zealand. For those whose NHI was not valid or missing, NHIs were sought but a small number were not provided, or not provided until after the analysis for this year was completed.

- Those 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.

Figure 8 shows the distribution of cases by deprivation status, split by ethnicity.

The 2016 report showed comparisons for those children and young people notified to the Database during 2010-2016 for children 0-5 years of age, and those 6-17 years of age[100,i]. DND distributions for these age groups both skewed more towards the higher deprivation scores than the national distribution for tamariki of the same age[ii]. This was particularly the case for tamariki aged 6-17, which contains a preponderance of those living in the four most deprived area groupings when compared to the national figures[iii].

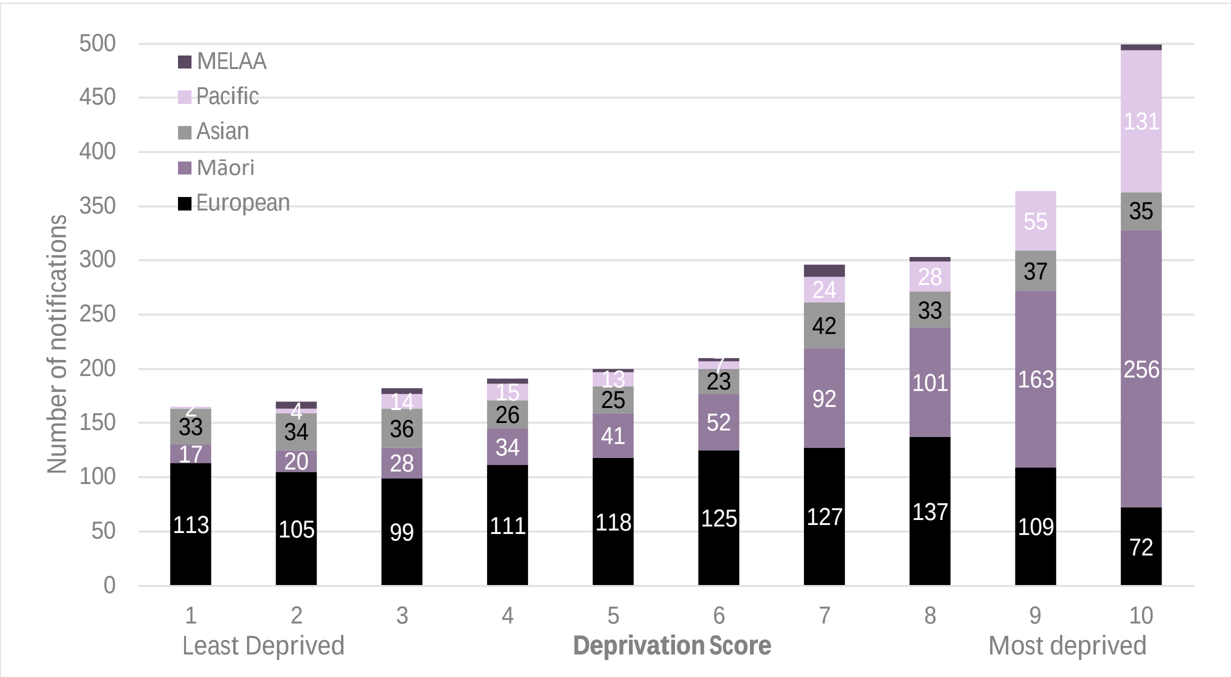


Figure 8: Deprivation scores (NZDep2013) of tamariki in the DND by ethnicity (2010-2021)[iv]

To further illustrate the differences between ethnic groups in the Database we have grouped the 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% (with scores of 1-3). A visual representation of this analysis can be found in Figure 9.

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 and experiencing the effects of financial hardship.

Professionals should keep in mind that income and poverty are significant determinants of

i The founders of the New Zealand Deprivation Index kindly shared data on the national deprivation distribution (NZDep2013) of tamariki in relevant age groups, so we could compare this with the distribution for children and young people whose information was notified to the Database. We are grateful for these data. 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 To make the bulk of the figures easily visible in this graph we have omitted those for children and young people listed as MELAA. These are in ascending order of deprivation: 0, 7, 5, 5, 3, 3, 11, 4, 0, 5).

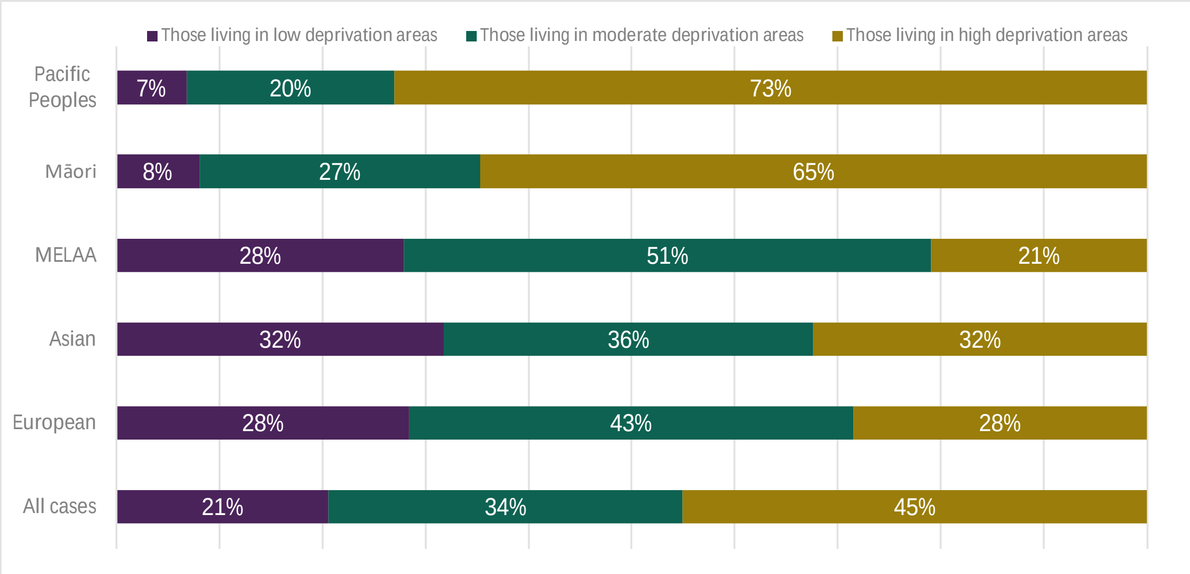


Figure 9: Deprivation scores grouped by deprivation and ethnicity (2010-2021 cases)

health[101]. As a result, the families they see are more likely to experience poorer overall health[101] (including greater barriers to accessing health services[102] and lower housing stability[103]) and higher rates of stress and mental health issues among both adults[104], young people and children[105, 106] than those in less deprived areas. These factors are likely to result in greater barriers for families to engaging with services, including audiology and ENTs.

The majority of families in areas of high deprivation will be of Pacific, Māori and/or MELAA ethnicities. Children and young people of Pacific ethnicity are 2.6 times as likely than those who are European to live in an area with high deprivation.

Aetiology

Ngā pūtake

- **Almost all (99%) of the records in the Database contain information about whether the aetiology (cause) of the child or young person's hearing loss was known at the time of the notification, and nine in ten cases have an unknown cause.**
- **The aetiology of hearing loss is either genetic or non-genetic in nature. The proportion of hearing losses that have a confirmed genetic cause is increasing.**
- **Children and young people recorded as European are more likely to have a known aetiology when compared with their Māori and Pacific and Asian counterparts.**
- **The proportion of hearing losses among children and young people with a known cause has been falling since the relaunch of the Database in 2010 and particularly from 2014, likely 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 or cause 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.

Changes in genes can influence the ear's structure, how the brain makes sense of sounds, or both. As the cochlea is complex it requires many instructions to develop and function. The child may have inherited changes to their DNA (e.g. missing bases or changes, of from one or both parents) or the change could have occurred only for the child with hearing loss (sporadic). Sometimes genetic and external factors together result in hearing loss[107].

The American College of Medical Genetics and Genomics estimated in 2014 that 30% of genetic deafness is syndromic[108]. In non-syndromic deafness with a genetic cause, the most common genetic mutations found are in the GJB2 and Pendrin genes.

Autosomal recessive inheritance is observed in about 80% of non-syndromic cases[109]. The Otoferlin gene has been implicated in cases of ANSD[110].

The proportion of hearing losses with a confirmed genetic cause is increasing over time[111, 112], 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 (around 247 as at 2021[107]), many cases still remain unexplained[113].

'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 four years[114]. A 2021 study by Batissoco *et al.* reported that the frequency of genetic aetiologies reaches 60% in developed countries and is expected to be

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

lower in countries such as Brazil though these rates are likely to increase in developing countries with improving healthcare.

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[114].

In tamariki, mumps is thought to be the most common cause of unilateral acquired sensorineural deafness, which is usually sudden in onset and profound in severity[115].

Cytomegalovirus (CMV) is a the most common congenital, non-genetic cause of hearing in overseas studies, causing 10-20% of cases in those under the age of five[116]. Typically, this infection is benign and innocuous, presenting as cold systems, but that is not the case for those who are pregnant and have no antibodies. It is difficult to predict which children with congenital CMV infection will develop hearing loss and whether or not the loss will continue to deteriorate[117]. General knowledge about CMV and how to prevent infections, which are particularly common among those who work and/ or live with young children, is not widely shared.

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[118].

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[119].

DND data

Almost 99% of the 2386 records in the dataset (2010-2021) 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 with aetiological information, 89% are of unknown cause, with the remaining 10% of cases listed as having a hearing loss with a known cause. The proportion with a known cause has been falling over time, but has lifted considerably in 2021, as can be seen in Figure 10. The reason for this change is not known, though the number of cases for which aetiology was not provided was lower this year, with five of the 185 cases notified falling into this category.

Keep in mind that the Database collects information at the point of diagnosis or soon after, meaning aetiological investigations which may have been done after that time for children and young people in the Database, cannot be included in the notification forms.

A key reason for the generally increasing proportion of cases without a known cause compared with historic levels 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 recent measles epidemic[120] is not yet known. It is worth

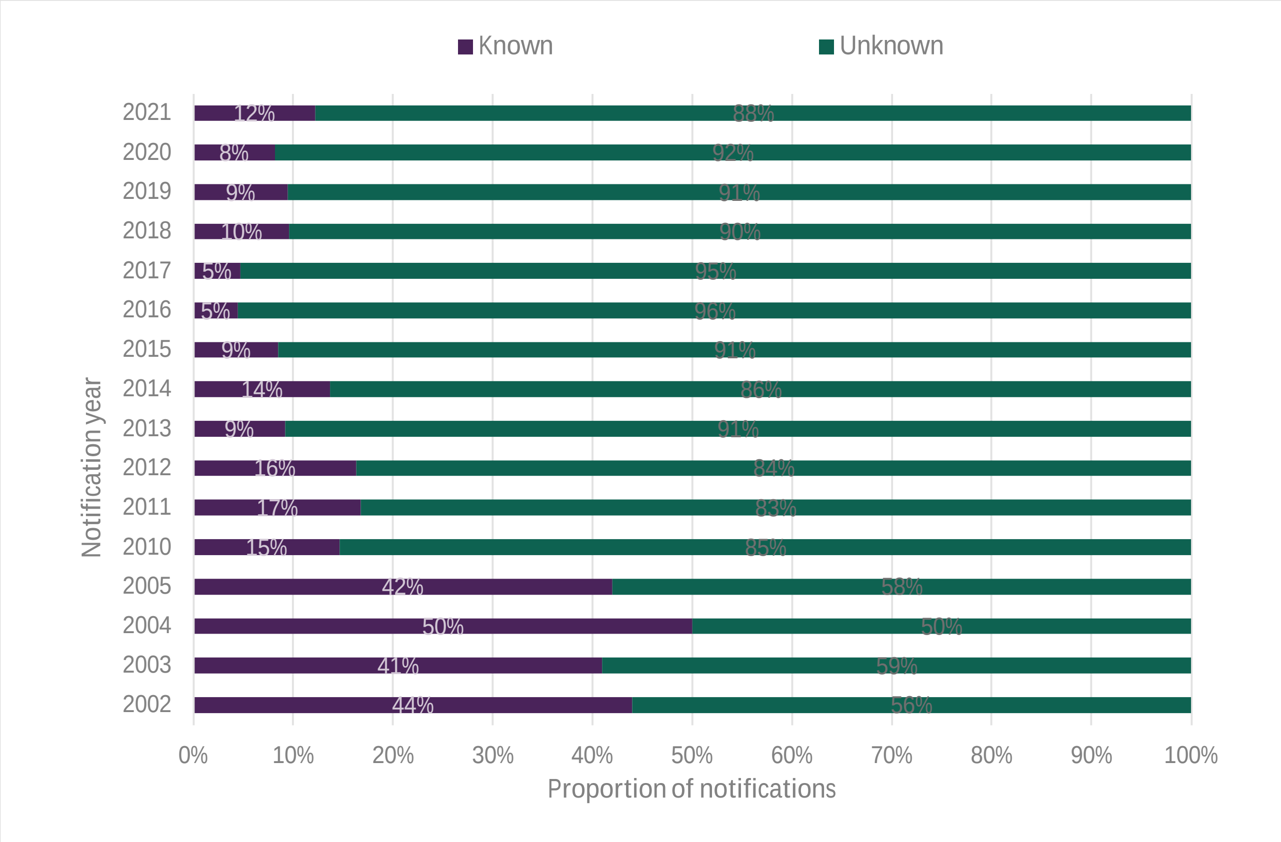

Figure 10: Proportion of hearing losses of known and unknown cause notified 2002-2005 and 2010-2021

noting that the current concern regarding mumps incidence in Aotearoa 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[121].

Cytomegalovirus

The importance of cytomegalovirus (CMV) in causing deafness among tamariki in Aotearoa New Zealand is not yet understood. A surveillance study in New Zealand is underway.

A systematic review conducted in 2008 found that approximately 14% of children with congenital CMV infection develop a sensorineural hearing loss of some type, and 3-5% develop one which is bilateral and moderate to profound in nature. This paper estimated that 15-20% of cases are attributable to congenital infections from this virus[122].

A 2014 analysis of data on 178 infants with congenital CMV infection found that those identified based on clinical suspicion had more severe disease at birth and sequalae than those identified at newborn screening[123].

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%)[124, 125].

Aetiology and ethnicity

In Aotearoa New Zealand during the 2010-2021 period, those children and young people with bilateral hearing losses which were recorded as severe or profound in severity were more likely to have a known aetiology than those categorised as mild.

When analysing these data by ethnicity, 14% of those listed as European have a known aetiology, compared with 10% of Māori, 10% for Pacific Peoples and only 5% for those of Asian ethnicity. This shows a significant difference between New Zealand European and other groups.

For each of these groups, as with the total, the proportion of children and young people whose hearing loss has a known aetiology had previously been dropping over time when compared with pre-2014 levels. This shift is presumably the result, at least in part, of reducing average ages of identification.

Aetiology types

Children and young people with syndromes

Those with hearing loss of known genetic cause can be split into those with syndromic and those with non-syndromic hearing losses.

Among the 2386 children and young people in the Database, thirty-three specific syndromes had been confirmed, affecting 78 children and young people. This number represents 3.3% of the 2,386 children and young people in the main dataset.

The most common syndromes identified were Down Syndrome (also referred to as Trisomy 21), which was identified at the time of the notification for 22 children and young people, Pierre Robin Syndrome/Goldenhar Syndrome and which were present in 13 children and young people.

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.

In an attempt to further describe conditions seen in children and young people, these have been categorised and included them in the section *Most common types of additional* disabilities on page 19.

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.
- For very young infants and those with disabilities, behavioural methods for identifying hearing loss may be unreliable, hence objective methods are used to diagnose these children. Prior to implementation of objective newborn hearing screening across Aotearoa New Zealand, the average age of tamariki at the time of diagnosis was, understandably, very high. Parents were the group most likely to first suspect their child's hearing loss.
- Since nationwide implementation of newborn hearing screening, the proportion of children and young people born in Aotearoa New Zealand whose hearing losses have been identified before the age of one has increased greatly from 24 in 2010, to well over 100 in recent years.
- The most recent data available from the UNHSEIP (from 2017) shows an estimated 94% of the eligible population had their hearing screened and 85% were screened by the B4 School Check during the 2020/2021 year.
- There are two peaks for identification of hearing losses among New Zealand tamariki – those identified from newborn hearing screening, mostly before the age of one year, and a smaller peak for those diagnosed around the time the child starts school, often associated with the B4 School Check.
- In the Database, those born overseas, those with mild, acquired and/or unilateral hearing losses along with those who are Pacific or MELAA have overall had a greater likelihood of having their hearing loss identified later. Pacific children and young people have seen particularly large reductions in median age at diagnosis over recent years.
- In the Database, tamariki Māori have a slightly later median age of diagnosis compared with Europeans; during 2021 their median age at diagnosis is three months, a month later than those in the European group.
- Understanding how the system is performing for Māori is not easy as they have higher proportions of mild and moderate hearing losses that are often diagnosed later, and more bilateral hearing losses that are often diagnosed earlier. In addition, inequities in the social determinants of health, and access to and through the health system, disadvantages tamariki Māori.
- Since 2013, newborn hearing screeners have been the most likely group to first suspect hearing losses among children and young people in Aotearoa New Zealand, with 59-68% of recent notifications now resulting from a screening referral. Seventy four percent of the 122 children notified in 2021 as a result of a newborn screening referral were diagnosed by the internationally recommended age of three months.

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 Aotearoa New Zealand and diagnosed in 2021.

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

	2010	2015	2021
Most likely to suspect	Parent or caregiver (37%)	Newborn hearing screener (46%)	Newborn hearing screener (68%)
Second most likely to suspect	VHT (17%)	Parent or caregiver (18%)	Parent or caregiver (6%)
Third most likely to suspect	Medical professional (10%)	VHT (10%)	VHT (6%)

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

Some changes can be seen in the groups most likely to first suspect a hearing loss in 2020 and 2021, perhaps because at various times the country, or parts of it, were in lockdown.

The proportion of cases first suspected by parents or caregivers has generally remained below historic levels reported in the original Database.

This group have gone from being most likely to first suspect a child or young person's hearing loss – in more than a third of cases (37% in 2010 and 2011) – to being first in 6-10% of cases during 2016-2021. 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 cases than any other group, 68% in 2020.

Evidence exists that behavioural methods relied upon some years ago for identifying a hearing loss were not an accurate method of screening for hearing loss in infants and some children with additional disabilities [126, 127, 128].

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. This change involved a move towards use of objective methods such as newborn hearing screening.

Age at diagnosis

Figure 11, shows the number of children whose hearing loss is identified based on the age of the child[ii] for selected years 2010 to 2021. There is now a notable and growing peak in the number of notifications during the first year of life – this is undoubtedly in large part the effect of the universal newborn hearing screening programme.

One hundred and thirty-four tamariki received a diagnosis during their first year of life in 2021, the highest number to date. One hundred and twenty-two of these cases were listed as having their diagnosis made as a direct result of newborn hearing screening. This year's figure is considerably higher than the 24 children diagnosed before their first birthday in 2010.

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

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

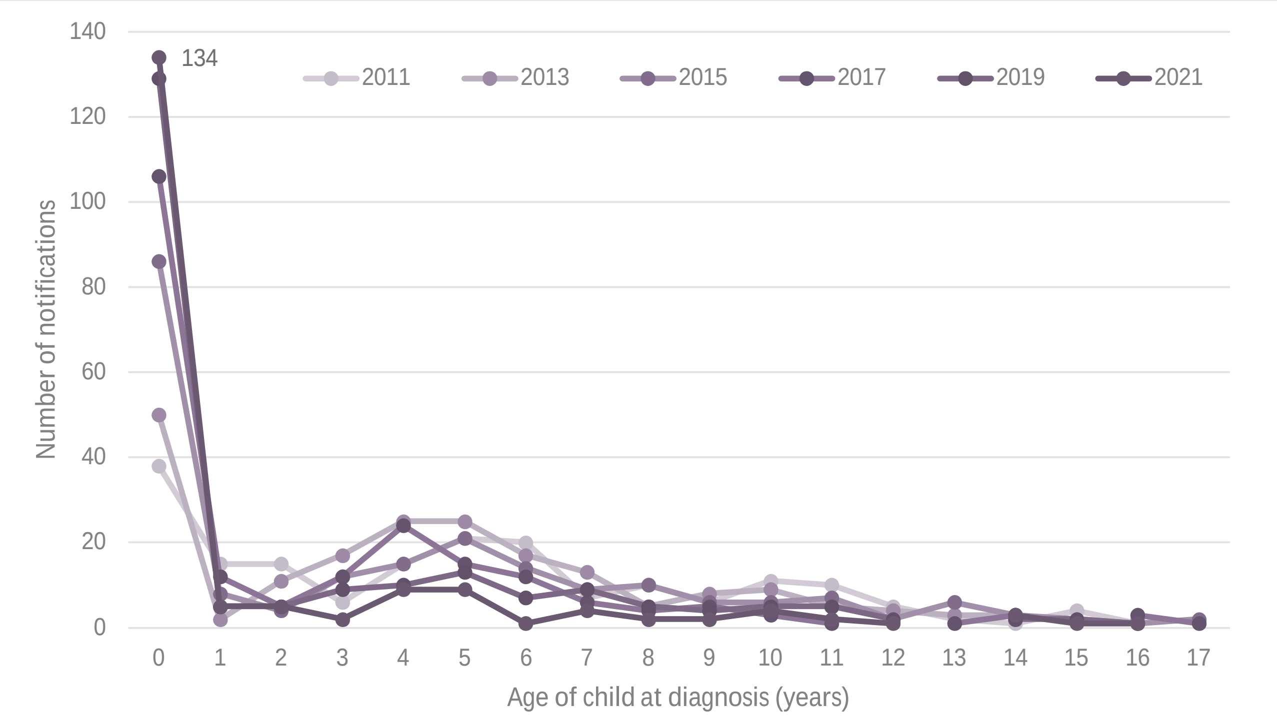

Figure 11: Number of children diagnosed by age in years (selected years, 2010-2021)

This is a positive trend, as it indicates more tamariki are having their hearing loss diagnosed early. A further, smaller peak in diagnoses has been seen, generally for four and five-year-olds, though this is a smaller peak in 2021 than seen in previous years; this peak is very likely to correspond to the B4 School Check[i, 129].

The number of tamariki being identified at between the ages of four and six has fallen from an average of 30-36% in the years 2010-2014 to 10-23% in 2016-2021. This drop may reflect that some children who were previously being identified by childhood hearing screening at or around school age are now being identified through newborn hearing screening.

It is worth noting that New Zealand had, historically, a very high average age of identification when compared with similar jurisdictions prior to the implementation of universal newborn hearing screening nationwide.

Coverage rates for the B4SC had been thought to be high in previous Ministry data, though revised figures show the proportion of children not checked is significant, and has risen to 15% in 2020-2021.

Figures from 2021 show 10% of children were first diagnosed with hearing loss between four and six years of age. [See the section on the B4 School Check which begins on page 50 for further information.]

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 can be useful for comparisons with measures used before 2006 and as a general indicator of the trend in age at diagnosis.

It is important to remember that such averages relate to all newly diagnosed tamariki, as it is not possible to separate out those with hearing losses that are late-onset (such as progressive and acquired hearing losses). In addition, this overall average age includes all children diagnosed in the notification period, for whom specific confirmation age data was available[ii]. This includes a small and shrinking number of young people born

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

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

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 and described on the notification forms provided to the Database are described in Table 10[i]. The analysis shows there has been a fall in the overall average age of confirmation over time.

This drop is particularly noticeable for children and young people born in Aotearoa New Zealand and those whose hearing loss was thought to have been present at birth. *(See the section on Delays in Diagnosis which begins on page 55 for more information.)*

The groups who are more and less likely to be identified later can be found in Table 11 below.

Average	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019	2020	2021
All cases	63	57	62	60	60	53	44	37	37	42	38	38
Born in New Zealand	61	52	57	53	53	48	37	32	33	40	31	19
Hearing loss thought to be present at birth	34	25	19	21	15	20	11	11	8	12	9	6

Table 10: Average ages of diagnosis for all cases in months (2010-2021)

Tamariki more likely to be identified later	*Tamariki more likely to be identified earlier*
born overseas unilateral and/or mild hearing losses acquired hearing losses, e.g. late onset, progressive and trauma related live in areas with a deprivation score of 8, 9 or 10 (the most deprived areas)	born in Aotearoa New Zealand bilateral hearing losses, particularly bilateral profound, severe or moderately severe hearing loss hearing loss thought by the clinician to have been present at birth

Table 11: Early and late average ages of identification (2010-2021)

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.

Children under the age of four are more likely to be missing some severity data[ii], meaning some 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 regardless, and as not all mild hearing losses present at birth are detected as a result of newborn hearing screening. The notification form does not include information about the proportion of losses that are thought to be progressive in nature.

i 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.

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

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2010-2021)	Total number of cases
mild	54	688
moderate	37	366
moderately severe	28	116
severe	23	57
profound	9	98

Table 12: Average age at diagnosis, in months, for *bilateral* hearing losses by degree (ASHA codeframe) using interpolated data with manual checks (2010-2021)[i]

Children diagnosed before three months of age

The highest proportions of severe/profound hearing loss are found among children diagnosed before three months of age, particularly for those with a unilateral hearing loss.

Those diagnosed after the age of three months are less likely to have more severe hearing losses (moderately severe or greater) diagnosed than those diagnosed before three months.

Those with mild hearing losses form a greater proportion of diagnoses for those diagnosed after 90 days.

Both unilateral and bilateral cases are less likely to have moderate hearing loss when diagnosed after 90 days.

See *Severity profile by age at diagnosis* which begins on page 66 for further information.

Age at diagnosis and ethnicity

Table 13 shows the average and median identification ages (2010-2021) for each ethnic group[ii], for all children and young people notified, where ethnicity information was provided.

With the exception of those children and young people of Asian ethnicity, all other large groups have later average and median age of diagnoses than those in the European group.

Please note that differences in the characteristics of hearing losses among each ethnic group, such as degree of loss and the proportion of cases present at birth, will influence these figures, meaning they are not a strict reflection of how systems are performing for each group.

Median ages in months have tipped into very low territory during this and the 2020 report due to the high numbers of newborn notifications. This is impressive given the challenges clinics have faced during the pandemic. However, taken alone these median ages do not help the reader conceptualise the "tail" that exists in terms of children and young people who had their hearing loss diagnosed later, reflecting both losses that were acquired or progressive in nature and those diagnoses that were delayed.

A number of the previous series of 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[iii].

Children and young people in all ethnic groups show improvements in average age at diagnosis when looking across the 2010-2021 period.

Asian tamariki

Children and young people in this ethnic group seem to have benefited quickly from the implementation of newborn hearing screening when compared with others, though this benefit seems to have stalled rather than continued, with the 2021 average age in months at detection being the highest of all groups, at 28 months.

It is worth noting however that the median age at diagnosis for Asian tamariki is still among the lowest of all ethnic groups, at two months.

This suggests there is still a group of children and young people in this category who are diagnosed later, preventing the average from falling as it has in some other groups. However, this group also had the highest proportion of cases that were suspected to be present at birth, at 59%.

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

ii 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.

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.

Ethnic Groups	Average months at diagnosis (2010-2021)	Median months at diagnosis (2010-2021)	Median months at diagnosis (2021)
European	48	39	2
Māori	47	41	3
Pacific Peoples	54	46	3
Asian	34	4	2
MELAA	52	24	1[i]
All groups	48	39	2

Table 13: Average and median months at diagnosis by ethnicity (2010-2020 and 2021)

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.

Māori tamariki

Māori tamariki and rangatahi were identified at an average age of 47 months over the full period, very similar in average the 48-month average age of their European counterparts. Māori particularly have seen a big reduction in the *median* age of diagnosis, moving from an average age of 55 months in 2010 to an average age of 21 months in 2021.

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 severity69.

These opposing effects make it difficult to understand how the system is performing to detect hearing losses early among Māori tamariki and rangatahi. 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, or of improvements in the health system's ability to reduce inequalities for Māori, although we have no evidence to support these suggestions.

Other Groups

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

MELAA children and young people have a high average age at identification over the years, at 52 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.

The average age at detection over the 2010-2021 period has been of particular concern for Pacific children, at 54 months, although recent years have seen a drop from a high of 84 months in 2012 to 16 months in 2021. This is an enormous shift and will make a real difference to the lives of these tamariki and their 'aiga, as it enables early intervention, and or monitoring to begin.

Pacific children have also seen pleasingly large reductions in median age at diagnosis during the last few years. These reductions may in part be related to changing characteristics within the cohorts identified over time, or they may reflect better system performance for this group.

Children and young people of Asian ethnicity experienced a swift reduction in their average age at diagnosis after implementation of newborn hearing screening and this average remained lower than historic levels for this group. However, recently the average has not fallen as low as for some other groups. The median age at diagnosis for this group has fallen however and is now lowest equal at two months, matching the median for New Zealand European children.

[i] Note this group is very small, containing three children and young people who are identified as MELAA.

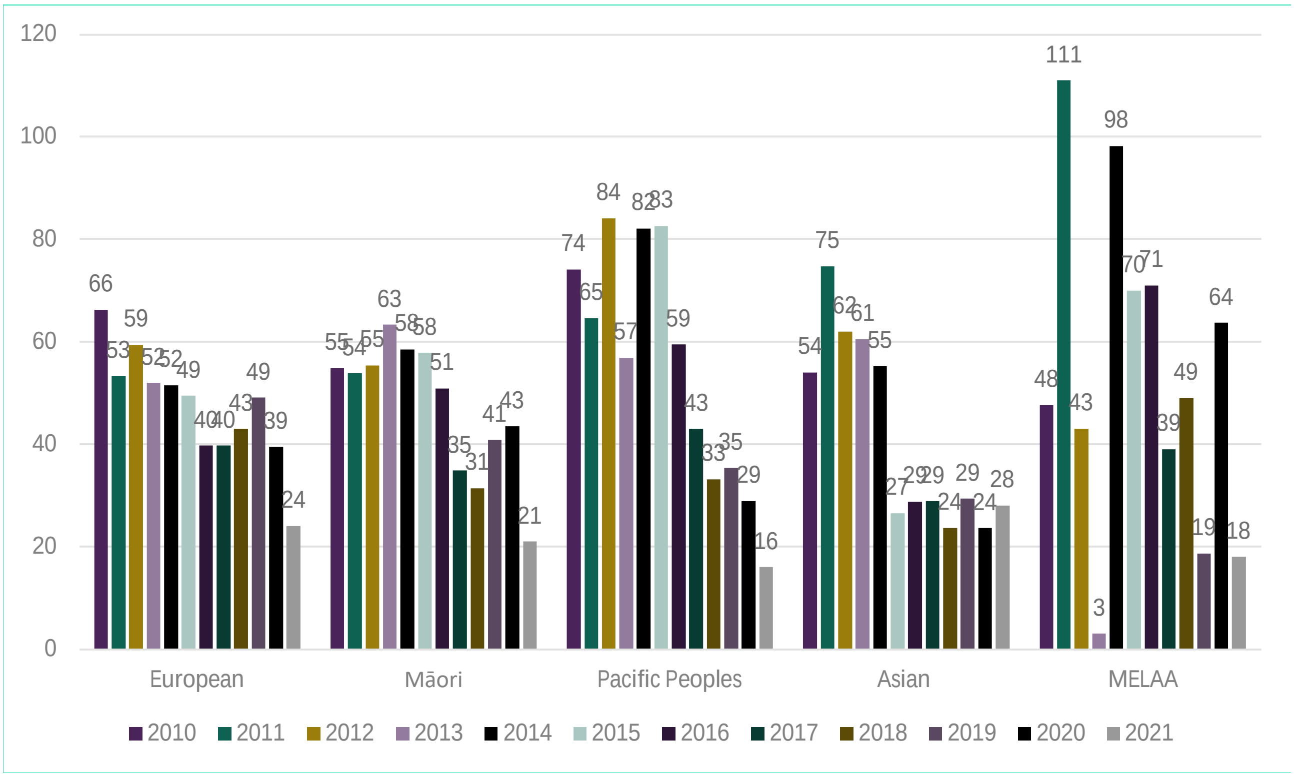

Figure 12: Average age of diagnosis by ethnicity in months (2010-2021)

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[130, i, ii].

The policy and quality standards for the UNHSEIP note that while children with mild hearing losses below 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'[130].

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

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

Screening status

Table 14 shows the screening status of Aotearoa New Zealand-born children notified to the Database (and therefore diagnosed) in the period 2010 to 2021.

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

Please note that this table includes those diagnosed at varying ages because there are some rangatahi in each year who 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

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 found in newborn hearing screening programmes, as referred to by Neumann *et al.* in the *International Journal of Neonatal Screening* January 2019 and by Matulat and Parfitt in the same journal in September 2018.

iii 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.

	Was universal newborn hearing screening (using aABR or aOAE) offered to this family after this child or young person's birth?	2010	2016	2019	2020	2021
No	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	0%	1%	2%
	No, this service was not available at the time	67%	12%	7%	4%	1%
Unsure	Unsure whether screening was offered to this family	7%	3%	6%	5%	2%
Yes	Yes, a screening programme was in place, but the child was directly referred to audiology due to atresia	0%	5%	3%	5%	3%
	Yes, screening was offered but this child was not screened	1%	3%	2%	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%	5%	3%	4%	4%
	Yes, this child was screened and passed	1%	16%	19%	15%	13%
	Yes, this child was screened and referred but passed the resulting diagnostic test*	0%	1%	5%	2%	2%
	Yes, this diagnosis is a result of a referral from screening	18%	52%	52%	60%	69%
Other	Other	0%	0%	0%	0%	1%
	No data	1%	1%	1%	2%	1%

Table 14: Screening status of children born in Aotearoa New Zealand and diagnosed during selected years[i]

no UNHSEIP monitoring reports have been produced since 2017, and as audiological assessment data were incomplete prior to that, the performance of the UNHSEIP, including the extent of loss to follow-up, is unknown.

The most recent NSU UNHSEIP Summary Report62, 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 did not compare favourably with our Australian neighbours, who were screening 97% of babies by one month of age in 2020[131].

Since the 2017 NSU UNHSEIP Summary Report, there have been significant improvements in the mechanism for collecting newborn hearing screening data and now all screening data are submitted electronically from three different sources. A UNHSEIP data warehouse is under development to combine data from the different sources to enable accurate national monitoring reporting. It is anticipated that a UNHSEIP 2020 Monitoring Report for all 20 districts will be available in early 2023.

Birth prevalence

The implementation of newborn hearing screening has afforded Aotearoa New Zealand 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[ii], 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[62].

i Please note that some figures in this table have been rounded and so not all sum to 100%. These figures are slightly different from those reported in previous years, due to small numbers of retrospective notifications, a small change in the codeframe this year to include a small number of cases which don't fit the codeframe and the inclusion of the proportion of cases which didn't contain data for this question.

ii 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 Aotearoa 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 Aotearoa New Zealand when the UNHSEIP achieves high coverage and low loss to follow-up in all regions.

These 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.

During 2021, a total of 122 of notifications were for babies born in Aotearoa New Zealand who were diagnosed as a direct result of newborn hearing screening. This has risen considerably from the 28 identified in this way during 2010, while newborn hearing screening was still being rolled out around the motu.

Please note that this table now includes a new category for those children and young people screened and referred from newborn hearing who passed the subsequent diagnostic testing, and then were diagnosed later.

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

New Zealand's UNHSEIP was implemented to reduce the length of time between birth or when a hearing loss develops and the start 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 previously common 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.

Our UNHSEIP's 1-3-6 goals are:

- 1 - ≥ 95 percent of babies to be screened by one month of age;
- 3 - ≥ 90 percent of audiology assessments to be completed by three months of age;
- 6 - initiation of appropriate medical, audiological, and early intervention education services by six months of age.

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, continues to 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 has changed over time.

There has been a pleasing overall reduction in the average age at diagnosis for cases referred from newborn hearing screening in Aotearoa New Zealand (therefore born in Aotearoa New Zealand), from fourteen months in 2010, to four months in 2021.

Of the 122 cases notified in 2021 that were identified as a direct result of newborn hearing screening in Aotearoa New Zealand, 74% were diagnosed by the internationally recommended age of three months[i]. This is the highest proportion reported to date with the previous highest figure being 73% in 2018.

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

	2010	2011	2012	2013	2014	2015	2016	2017	2018	2019	2020	2021
Average months at diagnosis	10[ii]	8	5	7	5	6	5	5	3	5	4	3

Table 15: Age at diagnosis for children referred from and diagnosed as a direct result of the newborn hearing screening programme (2010-2021)

Identification of false negatives

The DND likely provides the only method for identifying potential false negatives from the newborn hearing screening programme[132, iii].

In 2020, 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 2020 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. We have no information on which, if any, of these factors might account for any false negatives in the Aotearoa New Zealand environment.

Twenty-three of the tamariki who were born in New Zealand and identified with hearing loss during 2021 had been screened previously as part

i 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%.

ii Please note that this figure is different to the one previously reported as conflicting data existed in a number of 2010 where they were listed as conflicting information existed regarding their place of birth.

iii 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.

of the UNSHEIP and passed this screening. This figure 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 23 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.

	2010	2014	2018	2020	2021
Total cases identified by year who were screened previously (i.e., are not currently referrals from the UNHSEIP) and who passed this screening	2	20	32	25	23
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 Aotearoa New Zealand	2	10	18	10	4

Table 16: Potential false negatives and cases previously referred from hearing screening, selected years, tamariki born in Aotearoa New Zealand only

The first of these groups have known acquired hearing loss, while the second is those with hearing losses where the diagnosing clinician believed this was not present at birth[i] (it is possible Aotearoa New Zealand has a greater prevalence of progressive hearing losses because of our high rate of CMV124).

Of the four 2021 cases identified as *potential* false negatives in Table 16, the age of identification for these tamariki ranged from four, to seven and a half years of age.

B4 School Check

Background

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

There is no national reporting that helps us understand the efficacy of the hearing screening in the B4 School Check. As a result, key information is unknown, including the proportion of children who:

- are referred from the hearing screening who go on to receive diagnostic assessment,
- complete this assessment as a result of this screening,
- begin intervention,
- benefit from this screening in terms of improved outcomes.

Lower screening coverage suggests it is likely that groups under-served by our health services (such as Māori and Pacific) are not benefiting equally from this screening programme when compared with New Zealand Europeans. Without any basic measures of programme efficacy, we can't confirm the degree of inequity or its causes.

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

Outcome	Description	2010/11	2012/13	2014/15	2016/17	2018/19	2020/21
Pass Bilaterally	The child was screened and passed.	71%	70%	76%	77%	74%	74%
Referred	The child was screened and referred to a relevant service.	6%	5%	5%	5%	5%	3%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	9%	7%	5%	5%	5%	5%
Under care	The child is already under the care of a relevant service.	1%	3%	3%	3%	3%	2%
Decline	The hearing check was declined by the caregiver.	5%	4%	1%	1%	1%	1%
Not Checked[i]	The child did not receive a hearing check.	9%	10%	9%	10%	12%	15%
Population	**Derived from the B4SC Database**	**52,681**	**64,658**	**65,651**	**66.312**	**66,675**	**66,705**

Table 17 B4 School Check Hearing Screening data (those tamariki screened in alternating years)[ii]

Recent data changes by Ministry of Health

B4 School Check hearing screening data for alternating cohorts from selected years are shown in Table 17 (see previous reports in this series for data from other years).

Please note that these data include children born between 08/07/2005 and 07/07/2016, i.e. having their fifth birthday during the 2010/2011 - 2020/2021 financial years[iii].

The data source used by the Ministry of Health has shifted to include more children in the denominator and so these data are not comparable with previously reported data contained in DND reports[iv]. The number 'not checked', and the denominators included in the table below have increased, significantly for the most recent years, as a result of this change, while earlier years have seen these figures drop.

The Ministry of Health's aim using the B4SC database is to provide a more comprehensive dataset as a single source for both the numerator and denominator for a more accurate representation of hearing outcomes by ethnicity, including the estimate of those not checked. Having said that, while this denominator is an improvement on the one used previously, as it includes both children enrolled with PHOs and well as those who come directly to the attention of the B4SC programme, there will still be some children who are not included. Ethnicity information provided

i The number not checked is calculated by finding the difference between the total count of children turning 5 in the financial year and those with hearing outcomes.

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

iii These figures exclude children who, sadly, have a date of death against their record.

iv The data source now used is the Before School Check Database and includes records of children having their 5th birthday during the equivalent financial years. The Ministry of Health reports this is a change from previous reports (prior to 2019/20) so as to align the numerator and denominator better using the same date of birth range as well using the same data source for both the numerator and denominator. Previous reports used the PHO enrolled population, which has the limitation of excluding children who are unenrolled. The B4SC database is a national information system for capturing and storing information about children receiving their B4SC. The B4SC database receives input from the National Enrolment Service (NES) of children between 0 and 7 years of age. While the B4SC database could potentially miss children not enrolled with a PHO, it also contains records of some families who come into contact with the B4SC Program directly, e.g. via Early Childhood Education Centres (ECEs). The aim of using the B4SC database is to provide a more comprehensive dataset as a single source for both the numerator and denominator for a more accurate representation of hearing outcomes by ethnicity including the estimate of those not checked. The Ministry also notes that the "B4SC data used combines the records of children who have been assigned to a provider and those not yet assigned. However, unassigned data were only available from 2017 onwards. Hence, the number not checked prior to 2017 may be slightly underestimated and the pass rate may be overestimated." Also, the dataset could potentially include children who have moved overseas, as there is currently no systematic way of excluding these records.

previously by the Ministry was 'priority' coded, meaning children whose parents identify them as belonging to more than one ethnicity have this collapsed into one code, based on a specific algorithm. This year, for the first time we include multi-coded ethnicity data which aligns to how we describe ethnicity for children and young people within our own database.

Outcome	Description	*All cases*	Māori	Pacific	Asian	MELAA	NZ European
Pass Bilaterally	The child was screened and passed.	***74%***	66%	63%	75%	76%	79%
Referred	The child was screened and referred to a relevant service.	***3%***	4%	5%	3%	2%	3%
Rescreen	The child was unable to complete the screen, so a rescreen has been booked, normally in around 6 months.	***5%***	7%	8%	4%	4%	4%
Under care	The child is already under the care of a relevant service.	***2%***	3%	3%	2%	2%	3%
Decline	The hearing check was declined by the caregiver.	***1%***	1%	0.4%	0.2%	0.1%	0.4%
Not Checked	The child did not receive a hearing check.	***15%***	18%	21%	15%	17%	11%

Table 18 B4 School Check Hearing Screening data by ethnicity (2020-2021)[i,ii,133]Error! Bookmark not defined.

Insights

Programme coverage

The proportion of tamariki overall who were listed as 'not checked':

- has increased in recent years (see Table 17) regardless of the denominator used (see previous DND reports for figures using the enrolled PHO population as the denominator); and
- is considerably higher among *non*-New Zealand European groups, particularly children recorded as Māori or Pacific (See Table 18).

Please note that the figures in the tables shown demonstrate a reduction in the number of children screened over time even though they only include data from half of the 2021 year. (Next year's data on the B4 School Check coverage rates will describe what happened during the period containing the longest lockdown for education providers.)

Referral and rescreen rates for Māori and Pacific tamariki are also higher than those for children listed as New Zealand European, Asian or MELAA. For example, the new data from Ministry of Health shows the overall referral rate for tamariki completing the hearing screening completed as part of the B4 School Check is 3% (2020/2021). As with previous years, Māori and Pacific tamariki have higher referral rates (4% and 5%), with New Zealand European, Asian and MELAA tamariki having lower rates than the average (3%, 3% and 2%).

Children listed as NZ European, Asian or MELAA are almost half as likely to be booked for a re-screen (4%) when compared with those listed as Māori and Pacific (7% and 8%).

Having more accurate data through using the revised denominator for coverage calculations is helpful to inform efforts to reduce inequalities in access to B4 School Check screenings.

Diagnoses resulting from the B4 School Check

This year, an analysis has been completed for tamariki diagnosed between four and six years of age, an approximation of those whose diagnosis is most likely to be the result of the B4SC.

Figure 13 shows a marked fall in the number of diagnoses among those in this age group at diag-

i An 'other' ethnicity category is included in the B4SC data provided by the Ministry of Health. As this is a very small group (n=642) we haven't included it within this table. The children in this category are listed as 'not checked' in 21.3% of cases.

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

nosis, over time. This may be the result of increasing UNHSEIP coverage after the roll-out of this programme was complete in 2011, when all DHBs were screening for the full calendar year. Increasing coverage and efficacy of the UNHSEIP would result in fewer children being identified later[i].

It could also be in part due to the increasing number of tamariki 'not checked' in recent years, or other factors which may have resulted in a reduction of children referred from this screening programme going on to have their hearing loss diagnosed.

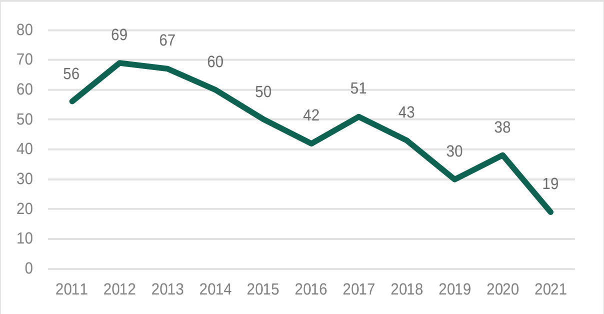

Figure 13: Number of children diagnosed at ages 4,5 and 6 by year (2010-2021)

In the last two years, the number of tamariki in this 'not checked' group has increased further, with re-analysed data with improved denominators showing 'not checked' figures were likely to be (on average) around 15% in 2021 rather than the 5% previously reported. This upward trend in the proportion of children not checked was evident even before the pandemic created additional and significant coverage challenges.

This is unsurprising considering how much time Auckland particularly was in lockdown during 2021, meaning children were not in early childhood or school settings to enable hearing screening to be completed as part of the B4 School Check. In addition, some B4SC staff and colleagues were seconded into the COVID response, and some audiologists noticed a significant reduction in referrals from this programme during the period. *[See also* Another extraordinary year *which begins on page 10.)*

There is a 'mop-up', to catch any children and young people who didn't complete the B4SC before they reached school. Anecdotally, this may not have been consistently applied around the motu. The Ministry's B4 School Database only contains information on children up to five years and seven days in age and not all results from this database are transferred into the ENROL (Education) Database, meaning it is not always easy to identify children who haven't had their check, so this can be addressed.

Recent research

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[ii,134].

The Welcome to School Study data (2017) suggested that in some areas there was likely to be a considerable number of children not enrolled with a PHO who were not included in the reported figures below, and this conclusion has now been confirmed[135, iii].

This study 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[135]. It found that although 75% of children had developmental delays and 64% had 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 Aotearoa New

i It is interesting to note that the B4SC database (Ministry of Health) only includes tamariki until they reach five years seven days old. As a result, this dataset does not provide the longitudinal data relating to children screened, rather it provides a snapshot of data until just after children turn five years of age.

ii 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. 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 B4 School Check at that time was 10%, the same as in 2018-19.

iii In addition, some children who were not enrolled with a PHO were actually screened making it difficult previously to understand the overall coverage rate for the hearing screening completed within this Check.

Zealand children and therefore it is inappropriate in the Aotearoa New Zealand context[136].

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 seven months, down from 26 months in 2010. This is undoubtedly, 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 months or even years between when their hearing loss is first suspected and when it is diagnosed and intervention can begin.**
- **Across 2010-2021:**
  - **Children and young people born overseas, Māori and Pacific children, those with mild hearing losses and those living in the most deprived areas are among those groups more likely to experience diagnostic delays. Asian children are more likely to have a short delay or no delay at all in getting their diagnosis when compared to all other ethnic groups.**
  - **'Audiologists having difficulty getting a confirmed diagnosis' was the most commonly mentioned reason for delays in diagnoses between 2010 and 2021. Such delays can be the result of conductive overlay or the child being unwell.**
- **2021 data demonstrates increases in delay for children and young people listed as New Zealand European and/or Asian, making these delays longer than for Māori and/or Pacific for the first time since the relaunch of the database in 2010. It also shows that this year "waiting time to see a hearing professional" is the most commonly listed cause for delay.**

Diagnostic delays

There are many variables that are correlated with 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 socio-economic status, plus factors related to the hearing loss itself, such as its severity.

One important variable influencing outcomes that hearing professionals can influence is how quickly the child's hearing loss is diagnosed; calls for earlier identification of babies with a hearing impairment have been made for nearly 80 years[137].

Early diagnosis seeks to maximise benefit during sensitive periods of neurological and linguistic development and limit children from falling behind their peers[138, 139, 140, 141, 142].

There are several ways to limit such delays, inclu-ding early and regular screening of children and young people for hearing loss. This screening in Aotearoa New Zealand includes the UNHSEIP, which aims to identify hearing loss in the newborn period and allow early intervention to begin and the B4 School Check, which aims to identify hearing losses among four year olds, before they reach school.

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 type of 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 21 months in 2020[i].

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

Additional efforts 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 22 on page 63):

- 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 requires a sustained and collaborative effort, and 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 whānau.

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."[143]

Delays and staffing

Broader contextual issues such as availability of ear and hearing-care professionals influence delays in diagnosis as well as issues with intervention and follow-up.

Internationally there is a health workforce shortage, and the shortage of allied healthcare workers is also a growing concern, exacerbated by an aging population requiring care in many developed countries[144], including Aotearoa New Zealand.

Kamenov *et al.* (2021) analysed data from ORLs in 138 countries, audiologist data from 102, SLTs from 124 and teachers of the deaf from 86 countries. This analysis found an enormous shortage of ear and hearing care professionals, and urged immediate action to ensure sufficient and equitable access to services[145].

For the period covered by this report (2021), hearing care for children and young people in Aotearoa New Zealand was generally provided by the public health system, through district health boards. At times district health boards, particularly those outside the main centres, had struggled to fill vacancies for audiologists. This resulted in long waiting times, which are thought to be associated with lower attendance levels[146]**Error! Bookmark not defined.**.

The initial emergence of COVID-19 was thought to have reduced recruitment challenges for the public sector, as the private sector, which traditionally has paid higher salaries, hired fewer staff. As the pandemic progressed, and the private sector began hiring again, some observers believe the number of audiologists moving into the public sector has dropped back to more normal levels. This latest shift seems to have reduced capacity of the public sector to provide diagnostic, intervention and monitoring services to tamariki and rangatahi.

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

Length of diagnostic delays

Average and median delays 2010-2021

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 length of delays is calculated based on the date of diagnosis and the age of the child at the time the hearing loss was first suspected, which is given in years and months. In many cases, particularly with older children, there isn't a precise date for the child or young person's age at the time of first suspicion. As a result, calculated delay periods reported within this report are in whole months, rather than days which are available for things like age at diagnosis. Exact date of diagnosis data was collected for every notified case from 2011.

The average delay in 2021, 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 seven months, the same as the figure for 2020. This is an impressive result given the significant implications for services of the COVID-19 pandemic during 2020.

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

The proportion of children and young people whose cases were notified to the Database in 2021 and who had no delay listed or a delay of one month by the time of diagnosis has grown considerably from 38% in 2011 to 61% in 2021[iii].

The 'long tail' of delays

Further information about the 'long tail' for delays shown in the 2020 report showed a compressed distribution in general as more and more children have a relatively short delay. Children and young people who are Pacific and /or Māori show higher average delays while European and Asian New Zealanders showed lower median and average delays within that analysis.

Year	Delay in months
2010	26
2011	16
2012	10
2013	12
2014	12
2015	11
2016	9
2017	9
2018	7
2019	10
2020	7
2021	7

Table 19: Average delay in months by year, 2010-2021[iv]

It also seems likely these differences over time are (in part) related to over-representation of these groups in areas that are the most deprived (scores 8-10) meaning they, on average, will have additional barriers to both good health and health system access.

Children and young people listed as Pacific Peoples have similarly higher rates of average age at identification and longer delay and are even more over-represented in the areas of New Zealand that are most deprived. Keep in mind that the Pacific Peoples category contains children and young people from a large and diverse group of Pacific communities.

Groups at increased risk of diagnostic delays have generally included children and young people:

- with a hearing loss *not* thought to have been present at birth; and

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 Aotearoa New Zealand.

ii 2010 and 2011 coincided with the completion of the nationwide roll-out of newborn hearing screening. 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 This is a more accurate set of figures than those provided in the 2020 report, with additional context for how these figures are calculated provided in the first two paragraphs of this section.

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

- 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;
- who are listed as being of Māori and/or Pacific Peoples or MELAA ethnicity/ies; and

In addition, those living in an area that scores an 8, 9 or 10 on the deprivation index are underrepresented in those with a zero- or one-month delay.

Shorter delays for some

Those children and young people recorded as Asian had a significantly lower average age at diagnosis than those from other ethnic groups, and a shorter average delay (See page 44 and Table 20). Children in this group are significantly more likely to have delays of zero or one month than are those from other ethnic groups. Keeping in mind that the 'Asian' group is also far from homogenous, this overall difference is likely to be a reflection of their:

- higher proportion of severe and profound hearing losses;
- lower likelihood of not attending appointments or have rescheduled these (for any reason) and to experience waits to see a hearing professional (see the next section for more information);
- higher likelihood of living in areas of the lowest deprivation (scores 1, 2 and 3 on the deprivation scale) and lower likelihood of living in areas of the greatest deprivation (8-10 on the deprivation scale), meaning as a group they will be less likely to have poorer health and will face fewer barriers accessing the health system; and
- tendency to have more successful access to and through other parts of the health system, as demonstrated by their high rates of participation in other health promotion efforts, including COVID-19 vaccination[147].

Ethnicity	European	Māori	Pacific	Asian	MELAA
Average (months, 2010-2021)	10.5	10.7	11.1	6.9	8.4
Median (months, 2010-2021)	2	2	3	1	3
Average (months, 2021)	9.8	6.1	5.2	8.5	0.7

Table 20: Average and median months of delay by ethnic group (2011[i]-2021)

A pleasing change this year

Figure 14 shows changing average delays for children and young people identified since 2011.

Most groups, including Māori and Pacific tamariki and rangatahi have overall seen steady declines in average delays from first suspicion of a hearing loss to diagnosis. During 2021, average delays for Māori and Pacific tamariki and rangatahi have fallen further which is pleasing. Both these groups have seen further declines in average delays despite the challenges brought by COVID-19 hit our shores in early 2020.

Interestingly, European and Asian groups have seen rises in the average age at identification during 2021, as can be seen in that figure. We are unsure of the reasons behind this change.

i We have used 2011 data as the starting point for this series as during 2010 we weren't collecting specific dates of diagnosis, making delay calculations less accurate.

Delay causes

2010-2021 cases

The notification form asks hearing professionals notifying cases for the reason(s) for the delay. Not all notification forms included one or more reasons for the delay listed, including some for which there was a length of delay specified.

The analysis in Table 21 examines the reasons for delay 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[i].

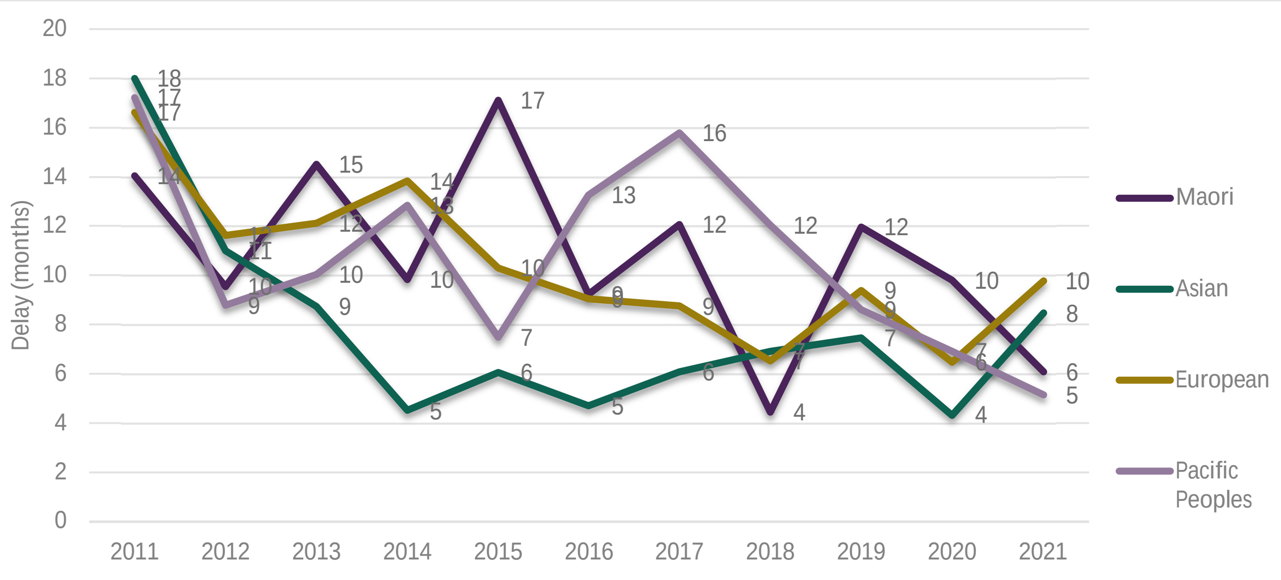

Figure 14: Average delay in months by ethnic group (2011-2021)

While 'did not attend' or DNA are commonly used to describe the cause of delays within health services, this term is viewed as unhelpful by some, as it is seen to allocate the burden to families to attend, rather than place it on services to successfully engage families.

When delays in diagnosis are examined for 2010-2020, several patterns emerged:

- Māori and Pacific families and those living in areas of higher deprivation were considerably more likely than European or Asian groups not to attend appointments or to have delayed these for any reason;
- European and Māori families were 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.
- children and young people living in the least deprived areas (1, 2 and 3 on the scale) were significantly less likely to have 'Parents did not attend appointments/delayed or rescheduled these (for any reason including distance, ill family member, cost, declined offer(s) of appointments)' as a reason for the delay; and
- in terms of reasons provided for delays, Māori were significantly less likely than other groups to have no reasons listed for their delay.

Table 21 now shows both the most common reasons for delay across the 2010-2021 period and also the average delay for children with and without each of these reasons listed. This demonstrates that those tamariki whose delay was attributed (at least in part) to 'follow up lost in the system' is associated with an average delay of 25 months. Where this reason was not listed, this delay is 13 months. Parents and professionals suspecting

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

something other than hearing loss, and the audiologist having difficulties getting a diagnosis were also associated with very significant delays.

Rank (most mentioned)	Reasons for delay	Average length of delay for cases with reason listed, and not listed
1st	Audiologist had difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell)	21 months, 11 months
2nd	Parents did not attend appointments/ delayed or rescheduled these (for any reason including service failed to engage family)	19 months, 12 months
3rd	Waiting time to see hearing professional or accessing services in their area	14 months, 13 months
4th	Parents/child/carers or educators (not health professionals) suspected something other than hearing loss or had no concern (e.g., speech delay, developmental delay, selective hearing, passed screening test)	23 months, 13 months
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	25 months, 13 months

Table 21: Most common reasons listed for delays in diagnosis (2010-2021) for cases with a diagnostic delay of one month or more[i] and average length of delays for cases with and without these reasons listed

At the other end of the scale, waiting time to see a hearing professional is only associated with a month greater delay when compared with those who did not have this reason listed as a cause of the delay.

Recent analyses of audiology data by Waikato DHB as part of their Equity Project were included in last year's report. Māori with bilateral moderate or greater hearing losses were diagnosed later than non-Māori. Factors contributing to delays among Māori were middle ear issues, delayed referrals from screening and in one case a DNA for an audiology appointment[148].

A recent paper in the *New Zealand Medical Journal* reminds us that not all efforts to increase attendance at appointments for hearing services audiology clinics will equalise attendance rates by ethnic group[149]. It notes that non-attendance rates of 21-38% have been reported in audiology and ORL services in Aotearoa New Zealand.

This paper describes a retrospective audit at Counties Manukau and found that there were no differences in attendance rates between those who had participated in telephone consultation and those who had not.

> *"Pacific and Māori children were 68% and 64% less likely to attend appointments after adjusting for socio-economic deprivation level, waiting time and telephone consultation compared to NZ European children. Longer waiting times were significantly associated with decreased attendance rates."*

This analysis found that attendance was found to be associated with ethnicity and waiting times, with those families waiting the longest time being less likely to attend, as the authors note had been previously reported. Telephone consultation did not improve attendance rates overall nor for ethnicity subgroups.

The authors of this study noted that while the catchment area for their clinic contains high proportions of Māori and Pacific whānau, these groups are not well represented in the audiology workforce. They suggest approaches to improve cultural safety could assist, as could finding ways to introduce the clinician when making 'cold' calls to whānau.

i 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.

This year's cases

In 2021, 34% 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 to the highest levels since 2010 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[ii].

Children and young people whose hearing loss was diagnosed as a direct result of a referral from the newborn hearing screening programme had an average delay to diagnosis during 2021 of one month, an impressive result given the considerable impact of lockdowns during the year on services .

"Waiting time to see a hearing professional or accessing services" was the most commonly mentioned cause of a delay in children's diagnoses for 2021, with 19 cases noted as being affected by this type of delay. This is also unsurprising given the service disruptions during the year caused by the pandemic. (This reason for delay was the third most commonly mentioned in 2019, the year immediately preceding the start of the pandemic.)

This was followed by "audiologist had difficulties getting a confirmed diagnosis" in 14 cases, and "parents or caregivers not attending appointments/delayed or rescheduled these", also with 14 diagnoses delayed for this reason.

Comments provided by audiologists shedding further light on diagnostic delays are provided below.

COVID-19 delays

In 2021, seven cases where one or more reasons for a delayed diagnosis was provided specifically mentioned COVID-19 as a reason for this delay. This is one fewer than in 2020 and seems to be a very good result.

Comments provided elaborated on this cause, which delayed screening and diagnostic appointments:

"Approx. 1 week delay for final ABR appt due to COVID level 4 lockdown restrictions"

"Bit of a delay due to outbreak of COVID-19 and consequent level 4 restrictions"

Other causes

Waiting time to see a hearing professional featured in a number of comments:

"Newborn hearing screening process took until baby was over 2 months old."

"Needed to wait for theater booking. ABR was done in conjunction with grommet insertion and cleft lip repair."

"Equipment stolen from the contracted Audiologist who does our ABRs. Had to be rescheduled."

Difficulties for the audiologist in getting a diagnosis included two cases where three ABR sessions were required.

"Three ABR sessions required as baby did not sleep well during the day."

"Baby did not sleep for 3 x ABR's. Seen for ABR under GA when possible for age."

Two cases seem to indicate some delays in getting a confirmed diagnosis within the private sector:

"This child had been referred elsewhere but was told after several months that there was no capacity to see them. Family called DHB enquiring about waiting times, booked into available cancellation slot at short notice." (Diagnosed at 21 months of age)

"Saw another private audiologist who didn't obtain complete results, wanted to delay repeat testing for 6 weeks. Came to see me in the interim." (Diagnosed at 52 months of age)

Attendance rates

Fourteen children and young people had their diagnosis delayed as a result of non-attendance at appointments. COVID-19 is likely to contribute to the number of whānau delaying non-urgent hearing care appointments in recent years, including because they could not or did not feel comfortable engaging through telehealth options[150].

Cases where the whānau or young person did not attend the appointment have typically been referred to as DNAs (Did Not Attend). More recently, it is becoming more common for clinics to refer to these delays as being the result of services not attracting patients or whānau, relabelling these cases as "Did not attract". This puts

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

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

the onus on the service to do what's needed so whānau/patients can attend appointments, to reduce delays in diagnosis and the start of intervention. This work also has implications for service efficiency.

"Nelson Marlborough Health general manager of Māori health and vulnerable populations Ditre Tamatea said it was time for the health sector to take responsibility for the attendance rates and change "did not attend" to "did not attract"."[151]

As mentioned in previous reports, reducing rates of non-attendance has at times been an area of focus in some district health boards, not always in a sustained way, as resources, support and ongoing funding for continued efforts are not always prioritised. Significant improvements have been achieved for periods of time as a result of increased focus on reducing DNA rates.

Successful processes have been implemented in Capital and Coast, which saw a drop of almost 50% in DNA rates for specialist appointments among Pacific patients over a five-year period, and Come Hear in Taranaki saw drops of 100%.

Common factors successful in reducing barriers to health service access include removing cost barriers, addressing transport and childcare issues[152], knowing the client population, personal engagement, a non-judgemental approach[153], strengthening cultural safety, and flexibility in service arrangements[154].

Marewa Glover from the Massey University School of Public Health said in 2017 that it *"cost money and time to go to appointments...People are struggling to pay their bills and feed their kids...If people can't pay their power, they certainly are not going to have money to go to appointments."*[155]

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

It has also been suggested 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[156]. This points to the need for strong collaboration between audiology and ENT services and the need for early bone conduction testing as indicated by relevant protocols.

Diagnoses from newborn hearing screening

When only children and young people whose diagnoses were the direct result of a referral from newborn hearing screening are considered, there are considerably fewer diagnostic delays reported.

"Waiting time to see a hearing professional or accessing services" was the most commonly mentioned cause of delay for this group during 2021, with five cases noted as being affected.

This was followed by "audiologist had difficulties getting a confirmed diagnosis" in two cases, and "parents or caregivers not attending appointments/ delayed or rescheduled these, also with two diagnoses delayed for this reason.

In one case, the cause of the delay was 'the screening incident' which resulted in incorrect screening of 2064 babies in six district health board audiology services between 2009-2012[157]:

"Irregularities in NBHS - child passed NBHS and was offered rescreen due to incident however was not rescreened." (diagnosed at 11.5 years of age)

Of the ten tamariki whose 2020 diagnosis was a direct result of a referral from the UNHSEIP and whose diagnosis was later than three months of age, *one or more* reasons for the delay were reported in six cases:

- 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=5);
- audiologist having difficulties getting a confirmed diagnosis (n=1);

Without early ABR testing for tamariki referred following their hearing screening, it can be more difficult to obtain a diagnosis until they can be tested using Visual Reinforcement Audiometry (VRA). Depending on the child, this approach can[i]. begin to be used from six months to two years of age

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

Approaches to reducing delay

Table 22 shows the most commonly cited reasons for delays in diagnosis, and a selection of approaches to reducing the various types of delay are included.

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,[158] 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 existing relationships 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[159], a statement on cultural safety from the Medical Council[160], and this paper focused on the difference between cultural safety and cultural competency[161].]; • 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 mothers86.]; • 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 22: Approaches to reduce diagnostic and interventional delays and reduce inequalities for tamariki with hearing loss.

Severity

Taumaha

- **Audiometric data is now much more likely to be estimated from the ABR than from the pure tone audiogram as children are being diagnosed at younger average ages.**
- **Many different frameworks categorise severity of hearing loss around the world. Here in Aotearoa New Zealand, the Clark (ASHA) framework is most commonly used by hearing professionals.**
- **New Zealand DND data show a relatively higher proportion of children and young people with mild and/or moderate hearing loss, and fewer with severe/profound hearing loss than in other similar jurisdictions we have examined. Several factors are likely to contribute to this, including the higher numbers of milder degrees of hearing loss found among Māori and Pacific children and young people.**
- **Asian children and young people have the greatest proportion of severe and profound hearing losses when compared with other ethnic groups, with almost triple the rate of profound hearing losses found among Māori.**

Audiometric data

Audiometric data are requested for both the right and left ears of all tamariki and rangatahi 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) policy and quality standards[i, ii].

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.[iii].

As shown in Figure 15, below, the proportion of cases for which the thresholds were determined through ABR is rising, from 21% in 2010 to 75% in 2021. This change is due to reducing numbers of tamariki being old enough to have their hearing assessed behaviourally, a result of the UNHSEIP. As UNHSEIP coverage levels have not been reported since 2017 we do not know whether the continued increase in the rate of children having their thresholds estimated from the ABR corresponds with further improvements in coverage, though this seems likely.

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

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

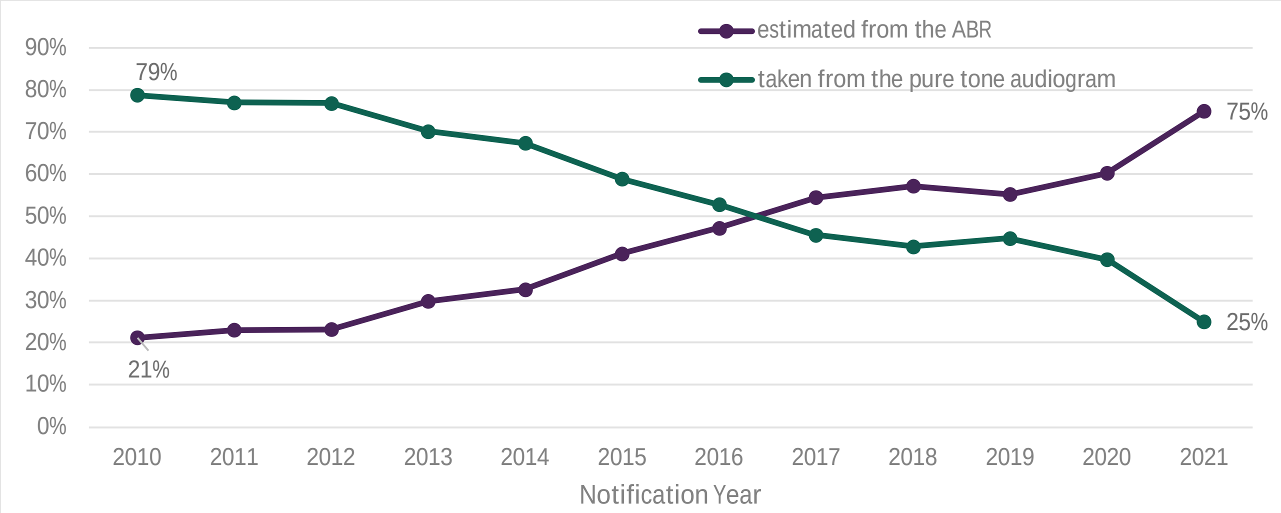

Figure 15: Proportion of cases containing thresholds from ABR and the Pure Tone Audiogram, by year, 2010-2021

Classifications

In Aotearoa New Zealand, the Clark (ASHA) code-frame is most 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 80.

Degree of loss	Clark 1981 (ASHA)[162]
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 23: Clark's 1981 ASHA severity codeframe

Calculating severity for notifications

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[i].

Table 24 shows the proportion of cases in unilateral and bilateral categories in each severity (degree) grouping. Please note that the labels in the 2020 report were reversed.

Degree of loss using ASHA severity codeframe	Unilateral 2010-2021	Bilateral 2010-2021
Mild	47%	54%
Moderate	18%	27%
Moderately severe	10%	7%
Severe	8%	4%
Profound	17%	7%
Sample size	n=624	n=1370

Table 24: Comparison of severity distributions for children with bilateral and unilateral hearing losses, 2010-2021, using interpolation and manual checks

Further information about interpolation and its use in this report can be found in Appendix G which begins on page 81.

By categorising notifications using the DND severity codeframe (1996-2005) and applying

i 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.

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.

exclusion criteria from the original database[i], a longitudinal comparison of the proportion of rangatahi in each group was included in the 2019 report, using data reported between 2001 and 2004 and more recent data. We noted that the severity profile of cases had changed with more mild losses in the more recent data.

Severity profile by age at diagnosis

Last year's report showed the severity profile of children and young people diagnosed before three months of age versus those diagnosed later, split by whether they have unilateral or bilateral hearing loss.

This year a similar analysis was conducted to examine the proportion of children diagnosed by degree of hearing loss, based on age at diagnosis using below six months of age as the category separator.

These analyses found:

- higher proportions of severe/profound hearing loss are found within children diagnosed under three and six months of age; and
- those with mild hearing losses form a greater proportion of diagnoses for those diagnosed above three and six months of age, particularly for those with unilateral hearing loss.

As cases diagnosed among those less than six months of age are generally identified through newborn hearing screening, and this screening doesn't target or detect all mild hearing losses, this severity profile may not reflect prevalence of these hearing losses in this age group, which is thought to be higher among Māori[ii, 72].

Degree of loss (ASHA severity categories)	Children with unilateral loss (worse ear average thresholds)		Children with bilateral loss (better ear average thresholds)	
	Diagnosed under six months of age	Diagnosed above six months of age	Diagnosed under six months of age	Diagnosed above six months of age
mild	30%	50%	43%	56%
moderate	18%	18%	31%	27%
moderately severe	13%	10%	8%	10%
severe	12%	7%	5%	3%
profound	28%	15%	13%	4%

Table 25: 2011-2021 Degree of loss for children and young people, by age at diagnosis

Severity profile differences between bilateral and unilateral hearing losses

As shown in previous reports, there are differences between the severity profile of bilateral hearing losses (which contain less severe and profound losses) and those with unilateral hearing losses (which show more children with severe and profound losses)[iii, iv].

This is particularly the case when the comparison is made between the ear with hearing loss in

i The original Database excluded cases of unilateral hearing losses, tamariki born overseas and those with acquired hearing losses.

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 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 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.

iv 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.

unilateral cases and the better ear in cases of bilateral loss[i]. Other reasons for these differences may relate to:

- unilateral hearing losses for children and young people 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 hearing losses are progressive[ii];
- bilateral hearing losses are more likely to be identified more quickly and therefore have less time to progress;
- low and mid-frequency congenital hearing losses, 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.

Ethnicity and severity profiles

Within 2010-2021 cases for children and young people with *bilateral hearing losses*, severity profiles are somewhat different between ethnic groups as can be seen in Figure 16. Numbers for the MELAA group are very small and change a great deal from year to year so should be treated with caution.

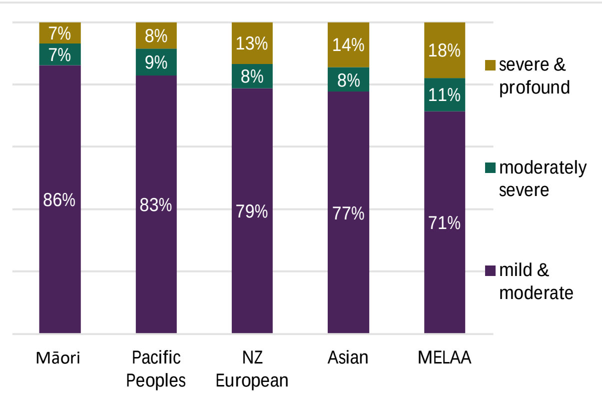

Figure 16: Degree of hearing loss by tamariki by ethnicity for bilateral hearing losses (better ear, 2010-2021)

Māori tamariki

Both historically and in recent years, DND reports have shown that European and Māori children have the greatest number of diagnoses, and that milder degrees of hearing loss are more commonly reported among Māori[65, 163]. These findings have been confirmed by analysis of 1982-2005 data[69, iii] and 2010-2016 data[iv].

A previous analysis of cases that were listed only as Māori or European (rather than both) was also completed for those with bilateral hearing losses, 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.

Last year, notifications for children and young people diagnosed between 2010 and 2020 with mild hearing loss were examined by ethnicity and this showed Māori were significantly more likely than expected to have these losses, and European less likely.

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

Other ethnic groups

Pacific children and young people, like their Māori counterparts in the Database, also have a higher likelihood of mild or moderate hearing losses than their Pākehā (New Zealand European) counterparts.

Children and young people from the Asian ethnic group are most likely to have severe or profound hearing losses. They have almost triple the rate of profound hearing losses than those who are recorded as being of Māori ethnicity.

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

ii It is worth noting that as the average age for identifying hearing loss reduces because 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.

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

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

Comparisons with international data

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

Despite differences in cohort, 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 than other jurisdictions in these comparisons.

Details can be found in the reports noted, comparing:

- 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 may have higher hearing loss prevalence overall, and there is a smaller proportion of severe and profound hearing losses than other similar countries.

Factors that may be contributing to the generally small proportion of more severe hearing losses are listed below:

- This may be, at least in part, due to the fact that Māori have a different severity profile to other ethnic groups.
- 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 recorded proportion of more severe losses may be smaller.
- Some cases with audiometric data points in the severe and profound range did not contain complete audiometric data and these have not been included in this table, meaning severe losses (and other degrees too) may be under-represented[i].
- Often children diagnosed with hearing loss have a sloping hearing loss and the better thresholds reduce the average degree of hearing loss.
- As noted previously, vaccination programmes had reduced rates of meningitis in Aotearoa New Zealand and this reduction was expected to have led to a reduction in rates of (more severe) hearing loss[164]. 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[165], although mumps, for example, almost always causes single-sided deafness.

Recent research suggests those children with milder degrees of hearing loss who were previously unaided, can have poorer phonological memory and morphosyntactic skills, raising questions about leaving mild hearing loss untreated[166], although research focusing on mild hearing losses remains 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[167, 168], and that those families with children who have cochlear implants are managing and promoting device use more than those with hearing aids[169].

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

Intervention and support

Wawaotanga me te tautoko

- **The Ministry of Education provides services to students who are deaf and hard of hearing through groups such as Advisors on Deaf Children and other specialist educators. In 2021, they provided services to approximately 2,070 children under the age of eight, including 787 babies and young children identified as a result of the UNHSEIP.**
- **In the 2021 year, the Ministry of Education funded support for children and young people who are deaf and hard of hearing from birth to Year 13 through First Signs support (Deaf Aotearoa), birth to five years of age, Cochlear Implant Habilitation programmes, habilitation support, and Ko Taku Reo - Deaf Education NZ**
- **At the time of diagnosis, professionals notifying cases expected just two thirds of the children and young people diagnosed in 2021 would receive two hearing aids. In total, 1,913 children and young people received hearing aids provided through MOH funding during the year.**
- **Forty children and young people around the country received publicly funded cochlear implants during the 2021 calendar year.**

Ministry of Education

In 2021, the *Ministry of Education, Learning Support* provided service to approximately 2,070 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 babies, infants and children under the age of five identified as deaf and hard of hearing through the Universal Newborn Hearing Screening programme (UNHSEIP) and their families and whānau – number supported 787.
- Support for babies, infants and children under the age of five and their families identified as deaf and hard of hearing not through the Universal Newborn Hearing Screening programme (UNHSEIP) and their families and whānau – number supported 302.
- Support for school-aged children (Year 1 to Year 3, at school) identified as deaf and hard of hearing with moderate communication and learning needs – number supported 983.
- For the calendar year 2021 the Ministry of Education, Learning Support received 189 new requests for support for children identified with hearing loss through the Universal Newborn Hearing Screening Programme:
  - » 72% of children and their whānau were contacted within 10 working days of receipt of a request for support;
  - » 91% of children and their whānau began receiving support by one month following receipt of request for support;
  - » 100% of requests for support for children under six months of age began receiving support by six months of age.
- 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 - Deaf Education NZ

Authors note: The number of children receiving services from the Ministry of Education, particularly in the Year 1 to Year 3 age groups still seem high to the authors of this report when considered in the context of the number of children being diagnosed each year. Possible reasons for this are:

1) that the Database doesn't receive notifications for all cases diagnosed each year;

2) the way the number of children receiving support is calculated results in some double counting; and

3) the number of AoDCs providing support nationally is higher than historic levels, meaning there has been greater service capacity over the last few years.

Ko Taku Reo Deaf Education New Zealand

Aotearoa New Zealand has seen enormous changes in Deaf Education since its inception in 1880 with the Sumner School for the Deaf in Christchurch (later named van Asch College then Van Asch Deaf Education Centre); from a strictly oral approach that endured for almost a century, to now, when programmes and services are provided in a wide range of ways with all languages utilised (English, NZSL and Te Reo Māori).

Ko Taku Reo is New Zealand's provider of education services for Deaf and hard of hearing (DHH) children[i]. They have a large team of over three hundred specialist staff across Aotearoa New Zealand with specialist school provisions in Auckland and Christchurch.

Ko Taku Reo is a tri-lingual, tri-cultural organisation. With both Deaf and hearing staff, New Zealand Sign Language (NZSL) and English are used on a communication continuum throughout, from administration to the classroom.

Ko Taku Reo also reflects the importance of Māori culture and Te Reo Māori by adopting culturally sustaining pedagogy in celebrating diversity and respecting the preferred learning styles of the diverse range of DHH students nationwide.

The strategic focus of the 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.

Services provided through Ko Taku Reo include:

1. **Enrolled school**

   Ko Taku Reo currently have thirteen sites across Auckland, Christchurch and Wellington, with 120 students enrolled in total during 2022. Auckland has the greatest number (n=79), followed by Christchurch (n=37) and then Wellington (n=4). Students can access residential accommodation between 11 and 21 years of age at Kelston (Auckland) and Sumner (Christchurch).

2. **Outreach School Resource Teachers Deaf**

   Ko Taku Reo Outreach currently has 2,943 students receiving varying tiers of graduated educational direct and indirect support services. The Ko Taku Reo outreach service provides specialist teaching, advice and guidance, assistive technology and NZSL support to Deaf and Hard of Hearing students usually enrolled in their local mainstream school. This category includes children over the age of three years although most children receiving this support are between the ages of four and half and 21 years old on the condition of being enrolled in a school or ECE.

   Children in this category are not always **Ongoing Resourcing Scheme** (ORS) verified as this verification does not commence until children transition to school. ORS verified children make up approximately 10% of the DHH population receiving services nationally from Outreach.

   Funding for this service comes from ORS funding (0.1 and 0.2 FTEs) and Ko Taku Reo

[i] In 2019, the Kelston Deaf Education Centre in Auckland and the Van Asch Deaf Education Centre in Christchurch merged to become one national organisation: Ko Taku Reo Deaf Education New Zealand.

also has an allocation of RTDs under the moderate needs contract.

3. **Specialist support: funded, and teacher supplied by student's school**

   ORS verified children are school-aged children in mainstream schools and children in other specialist schools. These students have funding split with the ORS DHH specialist teacher time allocated to Ko Taku Reo, while teacher aide and other specialist support is funded from the MOE to the child's school of enrolment.

   For example, this funding can be used for teacher aids and other specialist support (occupational support, physical therapy, speech language therapy, Kaitakawaenga, etc.) for staff are employed by the MOE**.**

4. **NZSL@School**

   The purpose of the NZSL@School is to support schools in the creation of 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. In 2020, NZSL@School funding was provided to 101 students nationwide as top-up funding to increase the hours of their Communication/ Education Support Workers (C/ESW's). A further sixty-four students received support from an NZSL Tutor.

Continuing change

NZSL Hubs (Outreach) and Beacon School Projects (Outreach) are new services established by Ko Taku Reo and have been designed to meet the needs of students through extensive consultation with communities and whānau.

For more information on the outreach programme or other services, you can visit the Ko Taku Reo website.

Ngā mihi nui ki a koutou to Ko Taku Reo for providing data for this section of the report, the second-year data has been shared by Ko Taku Reo since the two Deaf Education Centres merged to form this new entity.

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 hearing loss, diagnosis of additional needs).

Of the 185 cases notified to the Database in 2021, 181 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 2021, are most likely to be fitted with two hearing aids (66%). This reflects the preponderance of bilateral losses notified to the Database.

Figure 17 shows three patterns with changes seen in more recent data compared with 2010-2013 levels:

- a reduction in the proportion of cases where the plan is to prescribe one or two hearing aids;
- the proportion of cases in which the professional notifying the case is unsure whether hearing aids will be provided has risen; and
- a fall in the proportion of children and young people expected to receive no hearing aids.

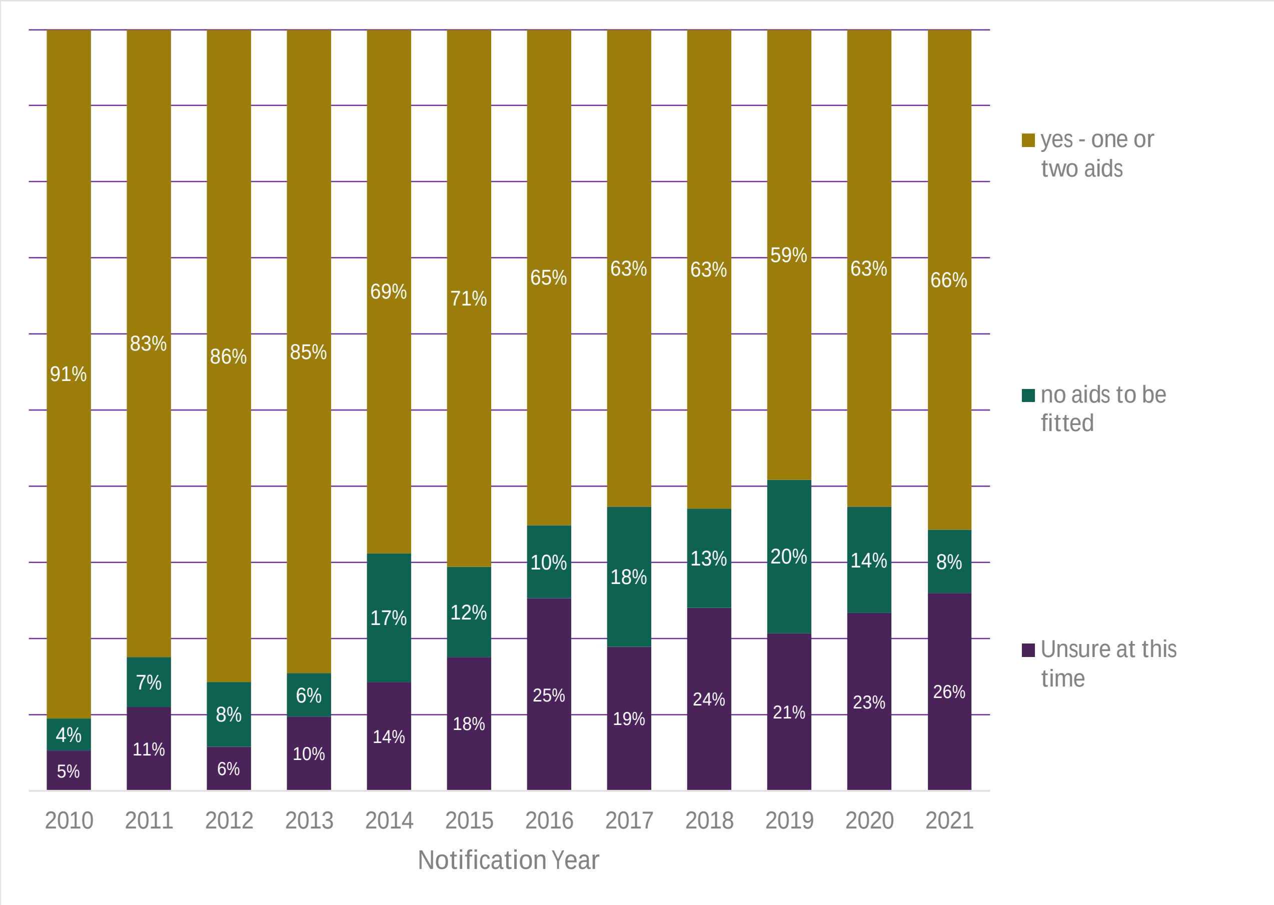

Figure 17: Hearing aids to be fitted by notifications (2010-2021)

When data for all children and young people notified from 2010 to 2021 were considered, the audiologist's intention was to:

- fit 75% of bilateral losses with one or two hearing aids, while 9% were not expected to receive any aids and the notifying clinician was unsure in 16% of cases; and

  fit 44% of unilateral hearing losses with one hearing aid, 20% two hearing aids[i], while 20% were not expected to receive any aids and the notifying clinician was unsure in 18% of cases[ii].

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 more European and less Māori children with zero or one hearing aid to be fitted, reflecting the proportion of bilateral hearing losses in these groups. *[See the 2016 report for more information.]*

An analysis was also conducted in 2016 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).

i The child or young person's second 'normal' hearing ear presumably had some hearing loss present though it didn't meet the criterion for the DND because it was lower than a 26dB HL average over .5,1.0,2.0 and 4kHz.

ii 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.

Public funding for hearing aids

To provide some context for these figures, data from the Ministry of Health's provider for Hearing Aid Services during the period covered by this report, are shown in Table 26[i].

These data show MOH funded hearing aids for tamariki under the age of nineteen, and those in fulltime education and under the age of 21 during the 2021 calendar year[ii, iii].

A total of 1,913 unique service users (tamariki and rangatahi) received hearing aid(s) during this period.

International research

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[170]. 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.

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Grand total
Māori	120	81	441	41	683
European	96	92	420	69	677
Pacific	85	42	206	40	373
Other	42	16	109	13	180
Total	343	231	1176	163	1913

Table 26: MOH Funding of Children's Hearing Aids, Calendar Year ending 31 December 2021, EnableNZ[iv, 171]

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[131].

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[172].

Visram *et al*. (2020) found that caregivers of eighty-one 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[173].

Prescribing and usage in Aotearoa New Zealand

A 2021 analysis by Waikato DHB found that for both Māori and Non-Māori with moderate or greater hearing loss, hearing aid fitting occurred on average approximately six weeks after diag-nosis, though medians for Māori were higher at 19 weeks, compared with 14 weeks for non-Māori[148].

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.

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

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

iv The current provider (EnableNZ) does not include repair or replace-ment requests, bone-anchored hearing aids, remote microphone (RM) systems, or funding for parts, molds or accessories in its data.

A New Zealand study followed up 163 of the 189 children and young people notified to the DND in 2010 seven-eight years later. Only 40% had been wearing their device(s) consistently since they were fitted.

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; in adult studies hearing aid use time correlates with severity of hearing loss.

Readers should also be aware 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. This information would be helpful to help us understand whether screening is resulting in appropriately early intervention for those tamariki and rangatahi who receive hearing aids.

Cochlear implants

As the DND notification form does not request specific information about cochlear implant referrals, the authors of this report thought it was useful to provide information about the number of cochlear implants provided to children and young people in Aotearoa New Zealand, and some background on the funding for these implants. This is below.

Funding from the Ministry of 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.

These implants are provided based on Ministry of candidacy criteria for children and young people who are assessed by the cochlear implant teams[i].

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 2021 calendar year there were 35 publicly funded cochlear implant devices provided in the Northern Region and 34 in the Southern Region, to children and young people under the age of 19. Please note this differs to figures in the table which relate to the number of children receiving implants, rather than the number of devices. This is a drop on previous figures, including those from 2020 in which 51 devices were provided in the Northern Region and 42 in the Southern.

Readers of these reports will notice these 2020 figures are lower than in recent previous years. A summary table showing this change can be seen in Table 27.

Number of children implanted by year	Southern region	Northern region
2016	33	38
2017	28	31
2018	33	32
2019	30	32
2020	29	26
2021	18	22

Table 27: Number of *children* receiving cochlear implants by year, split by cochlear implant programme (2016-2021)

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.

Children receiving cochlear implants	Southern Cochlear Implant Programme[174]		Northern Cochlear Implant Programme[175]	
	Ears	Children	Ears	Children
ACC cases	0	0	6	4
Public Funding - (1 Jan to 31 December)	32	16	28	17
Private procedures	1	1	0	0
Re-implants – recalled devices, failed integrity tests, or soft failures	1	1	1	1
Sequential or retrospective second cochlear implants (second ear for those under six already with one publicly funded ear - 1 January to 30 June)	0	0	0	0
	34	**18**	**35**	**22**

Table 28: Publicly funded cochlear implants provided in Aotearoa New Zealand during (2021)[i]

Some uncertainty exists about whether the reductions seen are due to the impact of COVID-19, including lockdowns, though some surgeries were delayed due to restrictions. Future data will provide a useful indication of whether COVID-19 was a significant factor.

i In some years the number of cochlear implants provided exceeds the number of profound or severe cases notified to the Database.

While the DND may be missing some notifications for children in the severe and profound categories, there are a number of other reasons why this figure is low compared with the number of children

implanted during the same period. One is that some children who are notified to the Database as having less severe hearing losses develop more significant losses over time, something which is not tracked by the Database.

Appendices

Ngā āpitihanga

Appendix A: Making notifications to the Database

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

Audiologists and audiometrists are asked to make future notifications to the Database by following this link.

Audiometrists are warmly 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 diagnosed during each calendar year.

   Resources for clinicians making notifications can be found here – these include a PDF version of the notification form, background information about the Database and previous Database reports.

2. **Consent:** Babies screened by the 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.

   Other children and young people diagnosed need be notified where a consent has been signed by the parent or caregiver, or for older rangatahi, by the young person diagnosed. This form should be kept on file by the diagnosing clinic.

Questions: 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 mahi on, and support for, the Database in its original 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 the period in which this Database was operating, it was managed by the National Audiology Centre on behalf of the Ministry of

Health, and later by the Auckland District Health Board.

The Database provided the only source of information from which the prevalence of permanent hearing loss among tamariki could be estimated, and from which the characteristics of new cases of hearing loss 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, at which time implementation of the national Universal newborn hearing screening and early intervention programme began.

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 newborn 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 also allowed communication with its members to call for notifications.

The authors of this report 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 original relaunch work done by the New Zealand Audiological Society, Janet Digby, 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)176."*

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, acquired and unilateral losses, and 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 current criteria includes children and young people 18 years or younger[ii]:

- 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[iii],
- 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;

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

ii To align with the age range used for the paediatric cochlear implant programmes.

iii 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.

- 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 the paper forms to electronic formats), and to try to make it as easy as possible for hearing professionals to notify cases.

A revised consent process was also implemented on re-launch to ensure all information is collected with the consent of the family, later this was added to through amendments to the newborn hearing screening consent which also includes consent from whānau to have their child's data included in the Database. Data is backed up regularly and forms are submitted through a secure link.

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.

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 Aotearoa 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.

The authors believe it is now likely that the Database has been receiving notifications for between 70% and 85% of all new cases diagnosed each year.

As time passes, we will continue to work in an effort 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 Aotearoa 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[177]. This method uses all ethnicity codes a person or their parent/caregiver chooses for them.

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 ethnicity codes 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 other commonly used method is the priority coding method, where those with multiple ethnicity codes have these reduced to a single code using a predetermined hierarchy.

A recent study utilising large-scale data of multi-ethnic New Zealand children, adolescents, and adults examined individual and contextual demographic characteristics associated with discrepancies between administratively prioritised and self-prioritised ethnicity. It found administrative prioritisation via a predetermined algorithm were more than 50% different from those which were self-prioritised[178].

Previous coding in the DND

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.

Categories used

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), European and Asian.

While it would be greatly preferable to collect more detailed information on ethnicity, we understand this may not be available for all cases and we don’t want to have any deterrents in place that would prevent cases being notified because, either we are requesting more detail than is easily available to the notifying professional, or we are adding too much to the time taken to complete the form.

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 or 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. This is not always possible depending on the context for specific sentences.

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 29 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 Aotearoa 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)[179]	World Health Organisation[180]	CDC[181]	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 29: 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 dB HL, 61-90dB HL, 91dB HL+), but don't name the categories so these are not included in Table 29.

Appendix G: Use of interpolation

Table 24 on page 65 shows the severity of hearing losses notified between 2010 and 2020.

While the Database contains estimates for those children and young people for whom all eight data-points are available, we generally rely on interpolated datapoints, to provide a more complete picture of the severity of hearing losses reported among children and young people notified to the Database[i].

Interpolation is only used where two data points surrounding the interpolated point are provided. The key thresholds under analysis in this report are: 0.5 kHz, 1.0 kHz, 2.0 kHz and 4.0 kHz.

This means the points that may be interpolated are 1.0kHz and 2.0kHz. 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 regarding interpolation:

- 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 collaborate closely with teachers from the two Deaf Education Centres[182]. Implementation of changes proposed in the Wilson Report (2011) were completed in 2015, meaning AODCs now work 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 Aotearoa 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) and Districts: These were organisations established to provide health and disability services to populations within a defined geographical area. DHBs were disestablished in 2022 under the (Pae Ora Healthy Futures) Act 2022 and replaced with 19 districts in four Regions within Te Whatu Ora Health New Zealand.

Data warehouse: A data warehouse is a type of database the integrates copies of transaction data from disparate source systems and provisions them for analytical use

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 2019. Since 2020, 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 Resourcing Scheme: The Ongoing Resourcing 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 child or young person's hearing loss was first suspected.

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 and respond to indirect service
- referrals via audiology;
- provide improved access to the curriculum for deaf and hard of hearing students.

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 Aotearoa 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 2019. Since 2020, 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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