



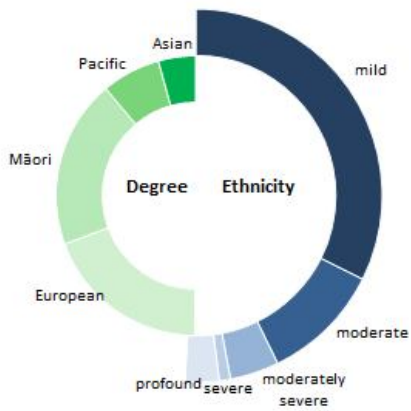
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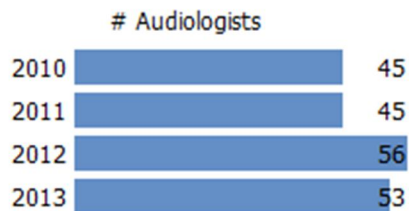
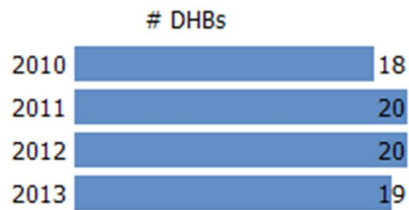
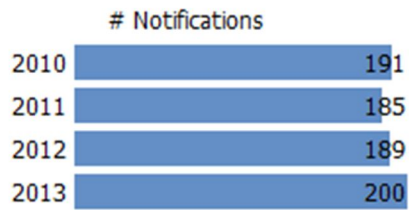
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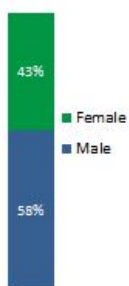
## Characteristics of 2013 Notifications



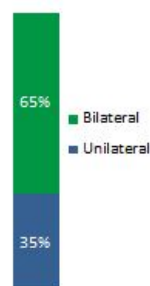
Groups more likely to be identified earlier
born in New Zealand
profound hearing loss
loss suspected to have been present at birth
bilateral hearing losses
Groups more likely to be identified later
born overseas
mild hearing losses
acquired hearing losses
unilateral hearing losses



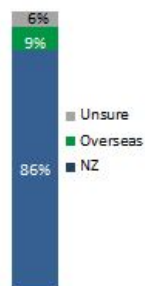
Gender



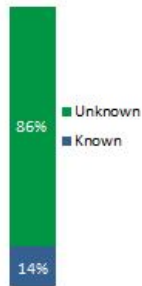
Unilateral/Bilateral



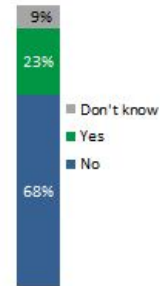
Birthplace



Aetiology



Family History



## For notifying audiologists

The authors of this report would like to extend a huge thank you to all audiologists who have provided notifications to the database for the 2013 calendar year. We understand you are not compelled to provide this information and we know how busy you are. Thank you for contributing to our understanding of hearing loss among New Zealand's children and young people.

Audiologists (including non NZAS members) are strongly encouraged to make future notifications to the database by following this link:

<https://www.surveymonkey.com/s/DeafnessNotificationDatabase>

We would appreciate it if you could keep the following points in mind when making future notifications:

1. **If you have any questions at all, please contact Janet Digby:** [janet@levare.co.nz](mailto:janet@levare.co.nz) or by telephone (09) 4456006. If in doubt about whether a case meets the criteria, please notify the case.
2. **Send us your notifications as soon as possible following diagnosis**  
Traditionally, the administrators of the Deafness Notification Database (DND) have attempted to collect all notifications in the year the diagnosis was made, e.g. a newly diagnosed case from 2004 was to be notified to the database in 2004 and information from this notification was to be included within the 2004 report. However, not all notifications have historically been provided in the year in which the diagnosis was made.  
  
***We strongly encourage all audiologists to get their notifications into the database as soon as possible following diagnosis and always before the end of January the following year. i.e. 2014 notifications should be provided by the end of January 2015.***  
  
This ensures these reports contain accurate information about those children notified during each calendar year. We understand that, with cases diagnosed late in the year, not all families may have consented to provide information about their child or young person to the database. We are considering a later deadline for 2014 notifications in 2015, and we will alert audiologists of any changes when a decision is made.
3. **Submit notifications online, no paper forms please**  
Notifications to the database can only be made online – please do not submit paper forms for inclusion.
4. **Read questions carefully and provide as much information as possible**  
Please read the online form carefully when making your notifications and provide as much specific information as possible in the spaces provided.
5. **Complete audiometric data**  
Please provide audiometric data for 0.5, 1.0, 2.0 and 4.0 kHz wherever possible.
6. **Suspicion and confirmation of hearing loss**  
Please provide information on the suspicion and confirmation of hearing loss as requested in the notification form.

**Age at suspicion:** This is the age at which the hearing loss was first suspected. This may relate to the age the child was referred from the newborn hearing screening programme.

**Date at diagnosis:** 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.

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# The 2013 Report

## Introduction

Welcome to the fourth annual report describing notifications to the re-launched Deafness Notification Database (DND). This report includes diagnoses made throughout New Zealand during the 2013 calendar year.

Since the DND was re-launched in 2010, the following definition has been used to determine which cases are included in the DND, and therefore in the analysis for this report:

*Children and young people 18 years or younger, born in New Zealand or overseas, with -*

- *a permanent hearing loss in one or both ears<sup>i</sup>*
- *an average loss of 26dBHL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0 kHz)<sup>ii</sup>*

Historical information about the database's inclusion criteria can be found in Appendix A: History of the database, on page 41 of this report.

Steps have been taken to ensure key data contained within this report are comparable with previous deafness notification data. However, in some cases, individual questions have been amended to make these more specific and/or to reflect improved understanding in a particular area (such as family history) and as a result a number of longitudinal comparisons are not possible.

Please note unless otherwise specified, analyses within this report describe characteristics of the full number of 2013 notifications for which data were provided.

Bars and lines in graphs depicting data from the 2010 to 2013 time period are coloured in the theme-colour for each year's report (turquoise for 2010, purple for 2011, orange for 2012 and dark green for 2013) while externally sourced data is contained in tables in grey.

This year we have added a glossary of commonly used terms which can be found on page 49 of this report.

## Contact details

The authors of the report hope that ongoing changes made to the way information is analysed and presented will improve the value of these reports over time. We ask that readers get in touch to provide us with feedback on this report to help guide the development of future reports.

Feedback on this report and any questions about the DND should be directed to the primary author of the report, Janet Digby. Janet can be contacted at: [janet@leware.co.nz](mailto:janet@leware.co.nz) or by telephone, (09) 445-6006.

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<sup>i</sup> 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.

<sup>ii</sup> Based on feedback from the audiological community, high frequency hearing losses which would not meet the original criteria but which would meet the 26dB HL average based on audiometric data from 2.0kHz, 4.0kHz, 6.0kHz and 8.0kHz have been collected as a trial from July 2011. We will continue to trial inclusion of this special group within the database. A limited analysis of data from high frequency hearing losses notified in 2013 can be found in Appendix B: High frequency hearing losses, on page 40.

## Completeness of notifications

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

There may be certain types of cases, such as those which are mild and/or unilateral, that are under-represented within notifications, and as a result inferences made from the data contained in this report should be taken as indicative only. (See the section titled *Number of notifications* on page 9 for further information.)

Hearing losses among Māori are more likely to be underrepresented in the DND as disparities in access to, and within, the health system have been found for this group<sup>1</sup>.

Despite these limitations, we can use a number of methods to provide some indication of the number of new diagnoses of hearing loss annually among children and young people. These methods are listed in the appendix which begins on page 46. Based on these analyses it is likely that the database has been receiving notifications for between 50% and 70% of all cases diagnosed each year, since 2010.

We hope that, as time passes, further efforts can be made to increase the proportion of notifications received, improving the ability of the database to inform the Ministry of Health, Ministry of Education, clinicians and other service providers, about the number and nature of new diagnoses of hearing loss among New Zealand children and young people.

## Acknowledgements

Thank you to the 200 families who provided consent so that we could collect details about their child's hearing loss. As a result of your willingness to share basic information about your child's diagnosis, service providers will be better informed about current and future demand for services, including the skills required to better serve the needs of families.

The time taken by individual audiologists around the country to make notifications and request consent from families is also very much appreciated, as are efforts of those who have completed the analysis for reports prior to 2006, which had its own unique challenges.

This report has been funded by [accessible](#), through a contract with the Ministry of Health. We would like to thank the Ministry of Health for funding the database from 2012. Without this support, people working with children who are deaf or hearing impaired would not have up to date information to help them better understand the number and nature of new diagnoses in New Zealand.

We would also like to acknowledge the New Zealand Audiological Society for funding towards the re-launch of the DND in 2010 and 2011.

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

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

# Notifications

## General information

Two hundred notifications pertaining to cases first diagnosed during the 2013 calendar year, and meeting the criteria for inclusion, were received by the 31<sup>st</sup> of January 2013<sup>i</sup>. These notifications were received from a total of 49 audiologists, with notifications from 19 of the 20 district health board areas. This is up slightly from the 191 notifications received in 2012, which were received from 19 [DHBs](#) and 49 audiologists.

It not possible to ascertain how long, on average, audiologists took to make each individual notification as some online forms were left open for a number of hours or even overnight. However, it is clear that many individual notifications took less than five minutes to enter using the online form, as was the case in previous years.

Of those children and young people whose hearing loss was notified to the database, notifications peaked at the end of the notification period (November to January) and also in August. This may be the result of the general shortage of audiologists nationwide and the timing of their holidays, or due to other reporting pressures, which are considerable.

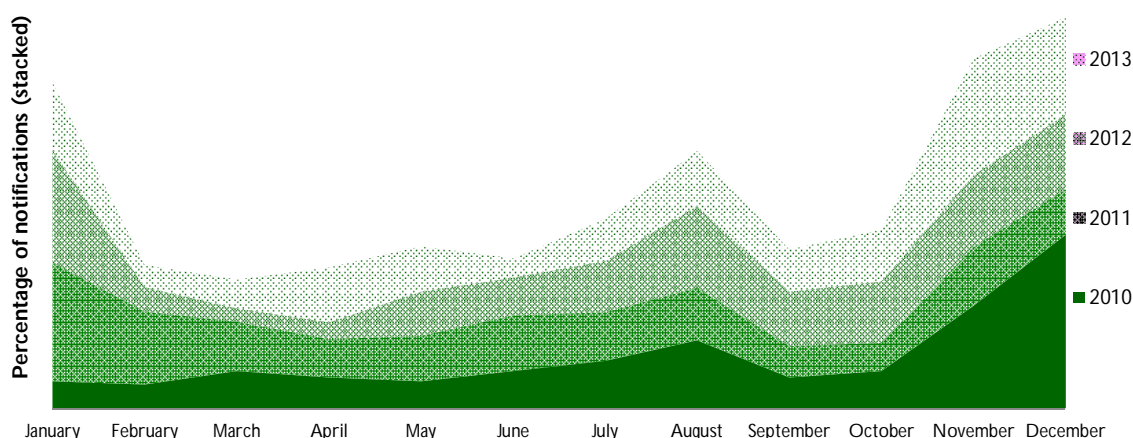


FIGURE 1: STACKED AREA CHART SHOWING NOTIFICATIONS BY MONTH (2010-2013)

Slightly more of the cases notified were male (57%) than female (43%). The ratio of boys to girls had been falling slightly since the database was re-launched, from 1.18:1 in 2010, to 1.125:1 in 2011 and 1.122:1 in 2012, but has now risen to 1.35:1.

In overseas research, boys are commonly found to have higher rates of hearing loss than girls. This figure is at the upper limit of the proportion of male cases found in this research, which ranges between 52% and 58% (1:1.08 and 1:1.38) of the total reported in various jurisdictions within 2011's Comprehensive Handbook of Pediatric Audiology<sup>2</sup>.

A statistical analysis was conducted to find any differences in our database between the severity profile, type and distribution of hearing loss by gender. This analysis did not discover any gender differences.

<sup>i</sup> Reports prior to 2006 contained information about diagnoses notified in a given year, rather than diagnosed within that year. As a result the number of notifications was variable, and increased in years where there were greater efforts made to encourage audiologists to send in notifications. For example, in 2004 there were an additional 288 retrospective notifications received from the Children's Hearing Aid Fund (CHAF) audit.



## Number of notifications

Notifications are collected through an online survey form, to reduce data entry errors and make it as easy as possible for audiologists to notify cases. Efforts have been made to publicise the database to all audiologists working with children and young people, in an attempt to collect as many notifications as possible.

Figure 2 shows the number of notifications meeting the criteria in each year. You can find information and how the inclusion criteria have changed over time from page 41 onwards. Please note that the 2001 to 2005 figures included in previous reports have been revised and the figures now show the number of notifications which met all inclusion criteria at the time and were included in the Database's annual reports<sup>i</sup>.

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

- notifications have been reported based on calendar years throughout the period of operation of the database i.e. 1982-2005 and 2010-2013;
- the period from 1982 to 2005 contains notifications to the original National Audiology Centre/ADHB administered database;
- no data are provided for 2006 to 2009 as the database was not operating during this period; and
- data for 2010 to 2013 relate to notifications provided to the newly re-launched database. Notifications were removed from the main analysis for the reasons stated below.

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

- slight losses (those not meeting the 26dBHL average across four frequencies in at least one ear);
- cases where the child or young person was reported as having mild hearing loss with normal bone conduction thresholds<sup>ii</sup> (assumed to be a transient conductive hearing loss unless a permanent conductive hearing loss was specifically stated, e.g. due to ossicular fixation);
- notifications with significant missing information; and
- notifications where consent had not yet been provided by the parent/caregiver.

Figure 2 illustrates the variability in the number of valid notifications provided to the original database, particularly in the last six years of its operation<sup>iii</sup>.

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

- Recently, the Auckland District Health Board kindly allowed access to their dataset<sup>iv</sup> (1982-2005) so that new notifications could be checked against those received previously to ensure no duplicates were being included in the current analyses. Duplicates were identified based on National Health Index (NHI) and by name, using fuzzy matches to detect potential duplicates which couldn't be identified based on the NHI, due to missing

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

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

<sup>iii</sup> Greville completed an analysis of the data in 2005, and noted that data reported in previous reports contained a number of duplicates, presumably from previous year's notifications; these have been removed for this analysis.

<sup>iv</sup> Access to this dataset allowed the authors to confirm that the notifications included in the database prior to 2005 included a number of duplicates, and we can confirm that the number of notifications reported before 2005 was artificially inflated as a result. There is no way of understanding which cases were included in each of the previous database's annual datasets, so the figures below are those figures reported in annual reports at the time.

information or data entry errors in the ADHB dataset. A small number of small changes to pre-2012 data appear in this year's report (compared with 2010 and 2011 reports) as a result of access to this information.

- Eleven additional notifications which were submitted in late for the 2010 year have now been included in the dataset, as they met the criteria for inclusion and some allowance was made for audiologists being slow to respond to the 2010 re-launch of the DND. This inclusion of late notifications has not been permitted since and will not occur in the future.
- Occasionally, an audiologist will report to us that a diagnosis previously notified to the database and which at that time met the criteria for inclusion has been revised and no longer meets the criteria for the database. These cases are then removed from the database.

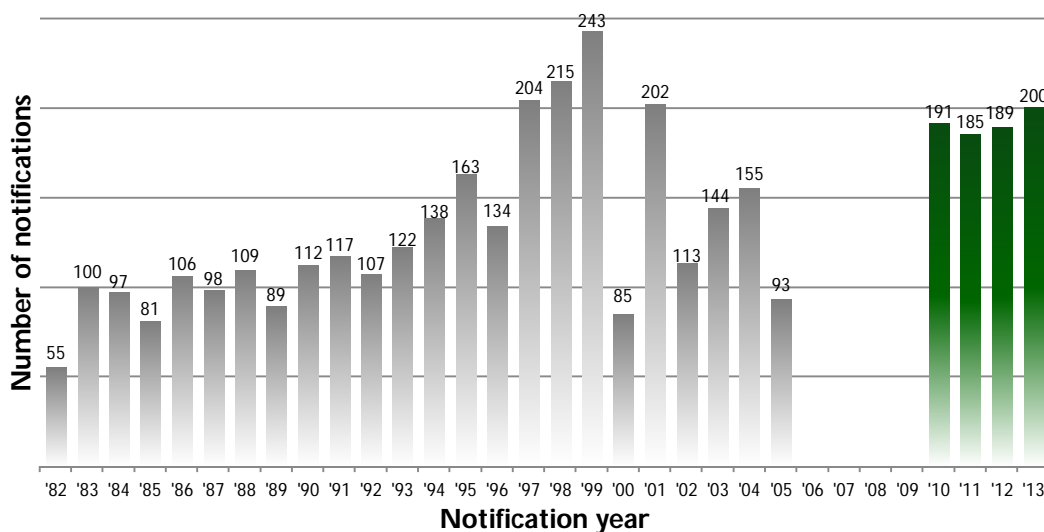


FIGURE 2: NUMBER OF NOTIFICATIONS BY YEAR 1982-2005 AND 2010-2013<sup>i,3,4,5,6,7</sup>

### Birthplace

Was this child or young person born in New Zealand? (2013)

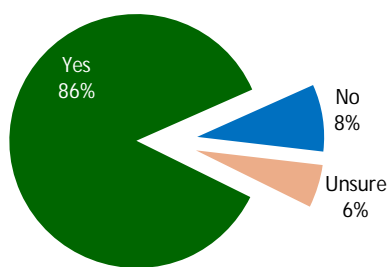


FIGURE 3: PROPORTION OF 2013 CASES BORN IN NEW ZEALAND

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

As shown in the figure, of the 200 cases included in the main analysis in 2013, 8% were known to be born outside New Zealand. Birthplace was uncertain in a further 6% of cases reported to the database in 2013.

Between 3% and 9% of cases in the database were born outside of New Zealand between 2010 and 2013.

<sup>i</sup> The lower number of notifications which met the criteria in 2005 and were described in the 2005 DND report was attributed by the author to removal of duplicate entries.

## Regional representation

Table 1 contains the percentage of 2013 notifications from each district health board (DHB) and compares this with the percentage of the population under the age of 20 from the 2013 Census<sup>i</sup>.

In addition to the natural fluctuations in the number of hearing losses diagnosed among children and young people in a given year, other factors influencing notification levels are likely to include:

- the size of the population within the age range for the database and the prevalence of hearing losses within that population;
- the date the child or young person was diagnosed, and whether it is appropriate to ask for consent for the database at the time of diagnosis, or whether this is best done at a later appointment which may be after the cut-off date for notifications;
- the number of FTE audiologists employed by each district health board;
- workload of these audiologists; and
- the level of commitment among staff to making notifications to the database.

It is worth noting that, historically, clinicians believe there is a preponderance of deafness in Auckland and Christchurch as many families have traditionally moved from the regions to these areas so their children could be schooled at [KDEC](#) (Auckland) or [van Asch](#) (Christchurch). In addition, it is interesting to note that the DHBs reporting higher numbers of notifications than their population would suggest are almost all those DHBs with a higher proportion of Māori and/or Pacific populations (e.g. Counties Manukau, Northland Bay of Plenty, Tairāwhiti).

District health board	Percentage of notifications received 2013 (under 19 years)	Percentage of population under the age of 20 <sup>ii</sup> (2013 New Zealand Census)
Auckland	5%	9%
Bay of Plenty	9%	5%
Canterbury	13%	11%
Capital and Coast	9%	6%
Counties Manukau	19%	14%
Hawke's Bay	2%	4%
Hutt	3%	3%
Lakes	4%	2%
Midcentral	5%	4%
Nelson Marlborough	3%	3%
Northland	9%	4%
South Canterbury	1%	1%
Southern	6%	6%
Tairāwhiti	4%	1%
Taranaki	1%	2%
Waikato	6%	9%
Wairarapa	1%	1%
Waitemata	5%	13%
West Coast	0%	1%
Whanganui	1%	1%

**TABLE 1: PERCENTAGE OF NOTIFICATIONS (2013) COMPARED WITH THE ESTIMATED PERCENTAGE OF POPULATION UNDER 20 YEARS (2013 CENSUS) OF AGE BY DISTRICT HEALTH BOARD**

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

<sup>ii</sup> Statistics New Zealand (2013): Estimated subnational population (DHB, DHB constituency), by age and sex, at 30 June 2013 (2013 boundaries).

## Other disabilities

The presence of one or more additional disabilities can have a significant impact on both outcomes for children and young people with hearing loss and on the level of support they might require, particularly from special education services.

Of cases notified for the 2013 period, 12% were thought to have disabilities *in addition* to hearing loss at the time the notification was made. In a further 12% of cases there was uncertainty regarding whether the child or young person had an additional disability<sup>i</sup>.

The most commonly reported conditions specified were those related to a specific syndrome (5 children), general or global developmental delays or intellectual disability (5), vision problems<sup>ii</sup> (4 children), and physical abnormalities (4 children).

The proportion of children notified with 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 yet been confirmed.

When the 'unsure' figure is added to the proportion of cases with an additional disability, the figure is more consistent with those reported before the re-launch of the database.

Notification Year	Proportion of cases with a known additional disability	Proportion of cases with a possible additional disability	Proportion of cases with additional disability (2002-2005) Total confirmed and possible (2010-2013)
2002	-	-	29%
2003	-	-	21%
2004	-	-	23%
2005	-	-	18%
2010	12%	10%	22%
2011	14%	5%	19%
2012	16%	11%	27%
2013	12%	12%	24%

TABLE 2: PROPORTION OF CASES WITH A KNOWN ADDITIONAL DISABILITY

Earlier identification of children with hearing loss is likely to result in lower levels of reported additional disabilities as these are reported at the time of diagnosis of the hearing loss. This is because children may have not yet been diagnosed with these conditions, or they have conditions have not yet developed (e.g. vision impairments are more common in older children and diagnoses of autism spectrum disorder are typically not made in the first year of life).

Other possible reasons for downward pressure on the proportion of children reported with additional disabilities include:

- Children with hearing loss in New Zealand may not be routinely assessed by a pediatrician, hence additional disabilities may be under-diagnosed; and
- Immunisation coverage in New Zealand has risen significantly since vaccination for children became a PHO Performance Programme (PPP) indicator in January, 2006 (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

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

<sup>ii</sup> No local data is available on the rates of vision problems among deaf and hearing impaired populations in New Zealand, but some professionals recommend routine referral for ophthalmological assessment for children diagnosed with significant bilateral hearing impairment.

2007 to 91% in September 2013<sup>8</sup>. Such improvements have reduced rates of meningitis in New Zealand and this may have an impact on the proportion of children with hearing loss with additional disabilities, although the numbers are likely to be small<sup>i</sup>.

### Overseas additional disability data

While it is difficult to compare reported rates of additional disabilities among hearing impaired children, as the definition for hearing loss and for disabilities differ and are not always described within scientific papers, a selection of rates from various jurisdictions are described below. The first paper listed shows the huge variability, presumably the result of such definitional differences.

New Zealand DND figures are similar to Australian estimates of the proportion of deaf children with an additional educational need, although this is unlikely to be a fair comparison due to differences in how additional disabilities are defined.

Source	Date	Location	Details	Rates
<b>Ear Foundation for National Deaf Children's Society<sup>9</sup></b>	2012 (review date)	United Kingdom	Review of 12 papers from 2002-2012 containing prevalence rates thought to be relevant to the UK, US, Australia, New Zealand	Most common additional disabilities: <ul style="list-style-type: none"> <li>• visual impairment (4-57% depending on the definition)</li> <li>• neurodevelopmental disorders (2-14%)</li> <li>• speech language disorders (61-88%)</li> </ul>
<b>Fortnum <i>et al</i><sup>10</sup></b>	2002	UK	Sample of 17,169 children with hearing loss	27.4% with additional disabilities
<b>Fortnum and Davis<sup>11</sup></b>	1997	UK	Trent region study of permanent congenital hearing impairment	38.7% of children found to have one or more additional clinical or developmental problems, although this study used a wide definition of additional needs.
<b>Holden, Pitt and Diaz<sup>12</sup></b>	1998	United States	60% of deaf and hearing impaired children in the United States in the 1996/97 year	20-40% of all US children with a hearing loss had an additional disability
<b>LOCHI<sup>13</sup></b>	2013	Australia	Study examining 260 children in Australia born with hearing impairment	18% of children within their sample have one additional disability, 10% with two and 9% with three or more
<b>The Consortium for Research into Deaf Education<sup>14</sup></b>	2011/12	UK	Annual national survey of educational staff	21% of deaf children (including unilateral and bilateral and mild to profound losses) had an additional special educational need in addition to their hearing impairment

TABLE 3: ADDITIONAL DISABILITIES, SELECTED OVERSEAS RATES FOR COMPARISON

<sup>i</sup> It is difficult to compare the number of cases of meningitis over time as this information was not collected prior to the re-launch of the database in 2010 and as there is no specific question related to meningitis in the current database. Further information on meningitis cases can be found on page 17.

## Bilateral and unilateral loss

Unilateral hearing losses are known to impact on educational performance and a significant proportion of these hearing losses progress over time<sup>15</sup>. As a result, cases of unilateral loss, where these losses are greater than 26dBHL in the hearing impaired ear, have been included in the DND since its re-launch in 2010.

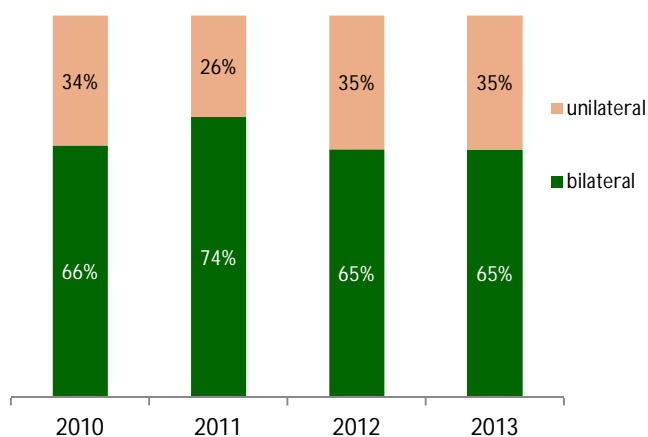


FIGURE 4: PROPORTION OF BILATERAL AND UNILATERAL HEARING LOSSES (2010-2012)

Although unilateral hearing losses were not included in the DND before 2006, a number of these cases were notified to the database each year and these numbers were provided in the annual reports. Comparisons with previous DND data (prior to 2005) are not possible as unilateral hearing losses were not within the criteria for the database, and although a number were reported, this group is incomplete within this older dataset.

Figure 4 shows the overall proportion of unilateral and bilateral hearing losses by year. Differences between the

proportions of unilateral notifications in each severity category are shown in Figure 12 on page 35.

As immunisation coverage (including for conditions such as mumps) in New Zealand has risen significantly from 45% in 2007 to 92% in 2012<sup>16</sup> it will be interesting to see whether a drop in the proportion of newly diagnosed unilateral hearing losses can be detected over time. Falling rates of measles, mumps and meningitis may contribute to such a decline, although as mentioned previously the number of cases involved is likely to be very small.

## Types of hearing loss

Information on the types of hearing loss notified is now being collected for each hearing impaired ear. Part way through the 2013 year, we began asking audiologists "Bearing in mind the maximum thresholds of BC testing... Do you think it is most likely that this hearing loss is...", for each ear.

Options provided were; sensorineural, mixed, permanent conductive, [ANSD](#) (Auditory Neuropathy Spectrum Disorder), Other and Don't know. Please note that the ANSD group have sensorineural hearing losses where ANSD has been confirmed; i.e. this group is effectively a subgroup of the sensorineural category.

The most commonly reported type of hearing loss within notifications to the DND, which included this information, was sensorineural (76% in the right ear and 78% in the left), followed by mixed losses (11% in left ears and 9% in right ears), and permanent conductive losses (8% in left ears and 12% in right ears). Two percent of all cases were recorded as ANSD (3% of the sensorineural category, while 1 to 2% of cases were listed as uncertain type.

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

# Ethnicity

## Representation

All but one of the 2013 notifications contained one or more ethnicity codes. Of those with one or more codes, 90% of respondents' selected one code for their child's ethnicity, while 9% selected two codes and 1% selected three. Thus, a small number of notifications specify more than one ethnic group.

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

The majority of notifications provided to the database since its re-launch in 2010 relate to children and young people of New Zealand European and/or Māori<sup>i</sup> ethnicity. Multi-coded Census data for 2013 was not available at the time this report was finalized. As the 2006 Census data is now somewhat dated, we will defer a comparison with the proportion of young people in the population until next year's report.

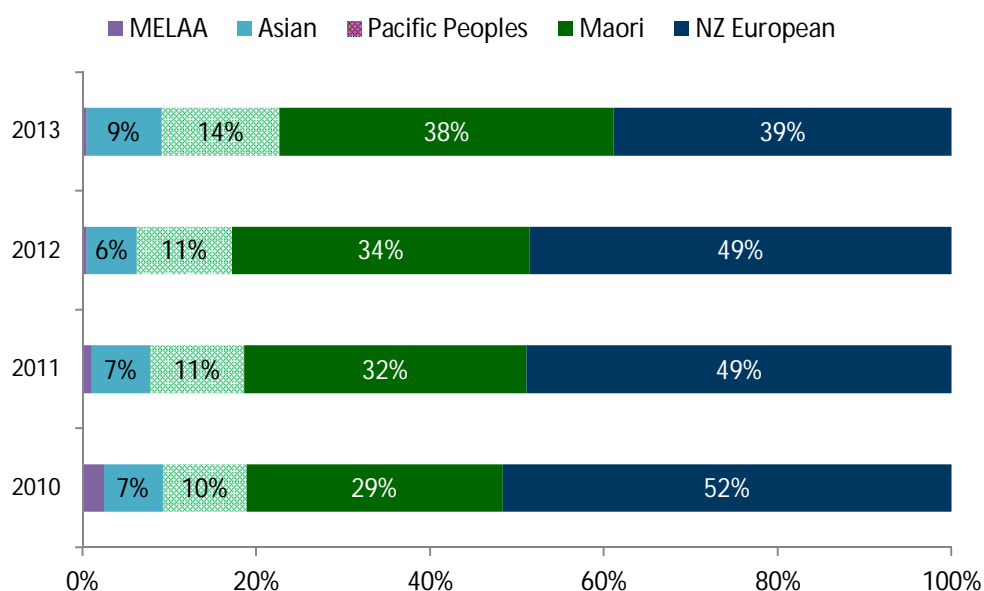


FIGURE 5: ETHNICITY OF 2010-13 CASES

## Ethnicity differences

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

- The Household Disability Surveys (1991-2006<sup>17</sup>) – these suggest Māori may have higher rates of hearing disability (children and adults) and higher rates of unmet need for technology and equipment when compared with non-Māori<sup>18</sup>. *For more about the limitations of this data please see the 2011 DND Report<sup>19</sup>.*
- Referral rates from the [B4 School Check](#)<sup>ii</sup> (2011) analysed by Searchfield *et al*, show higher rates of referral from hearing screening for Māori children (9%) compared with non-Māori

<sup>i</sup> In this report New Zealand Maori ethnic group is referred to as Maori.

<sup>ii</sup> The B4 School Check aims to screen all children before they reach school, and to identify and provide intervention to those children identified with targeted conditions. Part of this Check involves screening children for hearing loss. This screening should be completed on all children not already under the care of an ENT specialist or audiologist following their fourth birthday. Those not screened before they reach school should be screened after their arrival at school. This screening involves audiometry, usually conducted by a [Vision Hearing Technician](#). If the child passes



(5%)<sup>20</sup>. It is important to note that high referral rates for Māori may indicate higher rates of ear disease as these figures do not just relate to permanent hearing loss.

- Universal newborn hearing screening: While only limited programme data is available to describe diagnoses resulting from newborn hearing screening, Māori children were being referred at higher rates (2.4% compared with their New Zealand European counterparts 1.2%) and diagnosed at higher rates (13.8% compared with 10.8% for their New Zealand European counterparts) between April 2012 and December 2012<sup>21</sup>.

DND reports have historically shown that the greatest number of notifications pertain to Māori and New Zealand European children and young people, and that milder degrees of hearing loss are more commonly reported among Māori<sup>22,23</sup>.

A recent analysis of DND data (Digby et al, under review) indicates significant differences in hearing loss prevalence and severity profile do exist, with young Māori more likely to be notified to the database, and less likely to be reported as having severe or profound losses than their NZ European peers. This information can be used to inform policy and practice in both screening and clinic settings to identify hearing loss early and allow engagement with intervention services. This is particularly important as Māori health is 'characterised by systematic disparities in health outcomes, exposure to the determinants of health and health system responsiveness'<sup>24</sup>.

Although Māori are well represented among hearing loss notifications, this group may still be underrepresented in DND statistics because of their greater chance of having a less severe (mild or moderate) hearing loss. It may also be that disparities in 'access to, and within, the health system'<sup>25</sup> for Māori may mean not all cases are found or notified.

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

### Unilateral and bilateral losses

A total of 749 children and young people with completed ethnicity information were notified and met the criteria during the 2010 to 2013 period. These data contain some records (less than 10%) which had multiple codes for ethnicity, and so appear in more than one group. Over the whole group, 68% of cases notified are recorded as having bilateral hearing losses, while the remaining 32% have unilateral hearing losses.

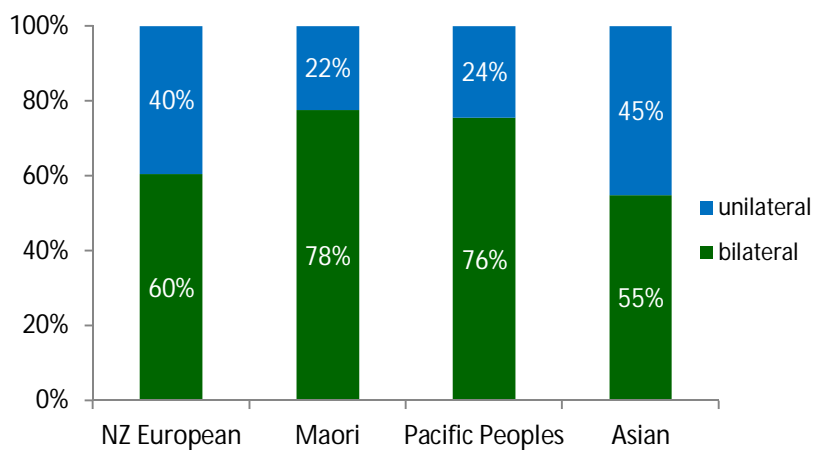
Figure 6 below shows a comparison of the percentage of notifications for this group, split by ethnicity, which are bilateral and unilateral in nature. MELAA has been excluded from this figure as due to the small sample sizes, particularly in 2012 and 2013.

This data supports the recent paper by Digby *et al* (under review) which confirms a larger proportion of bilateral hearing losses among young Māori when compared with New Zealand European counterparts.

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this test, no further referrals are required. Should the child refer on audiometry, tympanometry should be conducted.





**FIGURE 6: PROPORTION OF UNILATERAL AND BILATERAL HEARING LOSSES BY ETHNICITY (2013)**

As Figure 6 shows, the proportion of bilateral to unilateral hearing losses is quite different across ethnic groups, with lower proportions of bilateral hearing loss reported among Asian New Zealanders and higher proportions among those of Pacific Island and Māori ethnicity. Future DND reports will examine the significance of differences in the rates of bilateral loss between those of New Zealand European and Pacific Island and Asian ethnicities.

## Aetiology

All but four of the 200 cases which met the [inclusion criteria](#) for the 2013 period, contained information relating to the cause of the hearing loss.

As seen in Figure 7 below, the proportion of hearing losses where the cause was thought to be known has decreased significantly through the period 2010 to 2013, when compared with earlier figures. At least some of this difference is thought to be the result of changes in the cause information requested, as the notification form has been made more specific, asking for confirmed, and not suspected cause.

Another reason for the lower proportion of cases with a known cause, reported in the re-launched database, is that more children in the later period are being diagnosed earlier with hearing loss, due to newborn hearing screening. Now that more babies are being diagnosed with hearing loss, genetic testing is less likely to have been performed at the time the hearing loss is diagnosed. In addition, hearing losses may be identified before a full picture of the other issues is known, perhaps reducing the likelihood that hearing losses which are part of a syndrome are identified.

It is worth noting that additional genetic causes of hearing loss are being identified over time. Had New Zealand not implemented universal hearing screening, the proportion of cases with a known cause would have likely risen over time as additional genetic causes are discovered<sup>26,27</sup>, however genetic testing is typically not occurring early or in all cases of identified hearing loss.

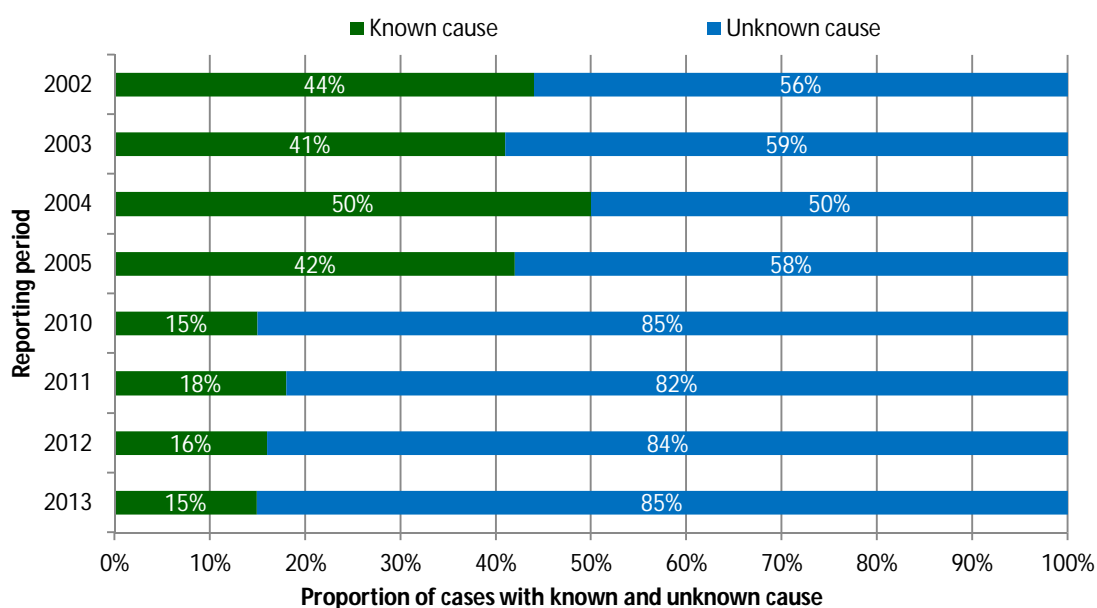


FIGURE 7: PROPORTION OF HEARING LOSSES OF KNOWN AND UNKNOWN CAUSE NOTIFIED TO THE DND BY YEAR

There has been a drive among the New Zealand based ENT specialist community in the last few years to increase the proportion of hearing losses which undergo aetiological investigations, such as genetic testing.

Although practice varies, ENT specialists generally refer young people/families of children with hearing loss, where there is no clear explanation of the cause of the hearing loss, for genetic testing. Over time, more genes and mutations are being added to those for which testing is available. The most common mutations found are in the GJB2 and Pendrin genes. ENT specialists request the tests and counsel patients about the results. If there are multiple or unusual mutations ENT specialists refer to genetic services<sup>28</sup>.

Table 4 below shows the aetiological breakdown where this was recorded as 'known'. Fewer cases notified for the 2013 period contained information about the cause of the hearing loss, even though the proportion of cases listed as being of known cause remained similar. This may be, at least in part, due to the rising number of children and young people whose hearing loss is diagnosed before the age of one year.

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

TABLE 4: NUMBER OF CASES OF KNOWN AETIOLOGY BY TYPE (2010-2013)

Of the cases of hearing loss diagnosed in 2013 and notified to the DND, one was listed as being the result of meningitis, while another was listed as being caused by a viral illness. This compares with three cases in 2012, four cases in 2011, and six cases in 2010 listed as being the result of meningitis<sup>29</sup>. However, it is worth noting that this information is not specifically requested and so may be incomplete.

Overseas, [aetiology](#) is reported as more likely to be investigated in cases of bilateral hearing loss, and where the hearing loss is more severe in nature, compared with unilateral cases or cases which are milder in terms of their severity<sup>30</sup>. This year's DND data shows that aetiology is (only slightly) more likely to be known in cases of bilateral hearing loss. This result may be different if the database tracked notified children, and their aetiological results, over time.

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

It is worth noting that identification of one aetiology does not exclude another aetiology. For example, the A1555G mitochondrial mutations may predispose a patient to hearing loss, and this hearing loss is expressed when certain antibiotics are used<sup>31</sup>.

Further related information, on the family history of cases, can be found in *Appendix C: Family History* on page 44.

# Identification of hearing losses

## Who first suspected the hearing loss?

Information on who first suspected the child's or young person's hearing loss was recorded for all but seven of the 167 children and young people confirmed as being born in NZ and who were diagnosed in 2013.

Table 5 shows the top three groups which first suspected hearing losses among notified cases since the re-launch of the database in 2010. Other groups who commonly suspected hearing losses first in 2013 included: medical professionals (other than the GP) - 11%, and educators and teachers – 5%.

2010	2011	2012	2013
<b>Parent or caregiver (49%)</b>	Parent or caregiver (42%)	Parent or caregiver (33%)	Newborn hearing screener (34%)
<b>VHT (22%)</b>	Medical Professional (21%)	VHT (23%)	Parent or caregiver (20%)
<b>Medical Professional (12%)</b>	VHT (15%)	Newborn hearing screener (23%)	Vision hearing screener (16%)

**TABLE 5: THREE GROUPS MOST LIKELY TO FIRST SUSPECT A HEARING LOSS 2010-2013 (BORN IN NEW ZEALAND)**

Strong evidence exists that behavioral methods for identifying a hearing loss, even those used by paediatric audiologists or hearing screeners, are not an accurate method of hearing screening in young children<sup>32,33</sup>. The challenges parents face in trying to identify a hearing loss in their young child, particularly when their hearing loss is not so severe as to prevent speech and language development, are considerable.

Therefore, it is pleasing to see that there has been a noticeable change in the groups most likely to first suspect a hearing loss among children and young people, over the last three years, towards those using objective methods. For the first time in 2013, newborn hearing screeners, not parents, are most commonly the first to suspect hearing loss.

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 responsible for the reported increase in the role of newborn hearing screeners in first suspecting the hearing loss in 2012 and 2013, given that the UNHSEIP coverage rates had not at that time increased significantly from 2011 levels. However, the growing role of newborn hearing screeners is undeniable.

The proportion of 2013 cases first suspected by parents or caregivers is now lower than at any time since the database was re-launched in 2010. It is also below historic levels in the original DND database, which reported between 34% and 52% of cases first suspected by parents in the 2000 to 2005 period.

Further information about those first suspecting the child's hearing loss is contained in Figure 8, below. This figure shows the proportion of cases first suspected by each of three groups, since 2010 for those children and young people born in New Zealand.

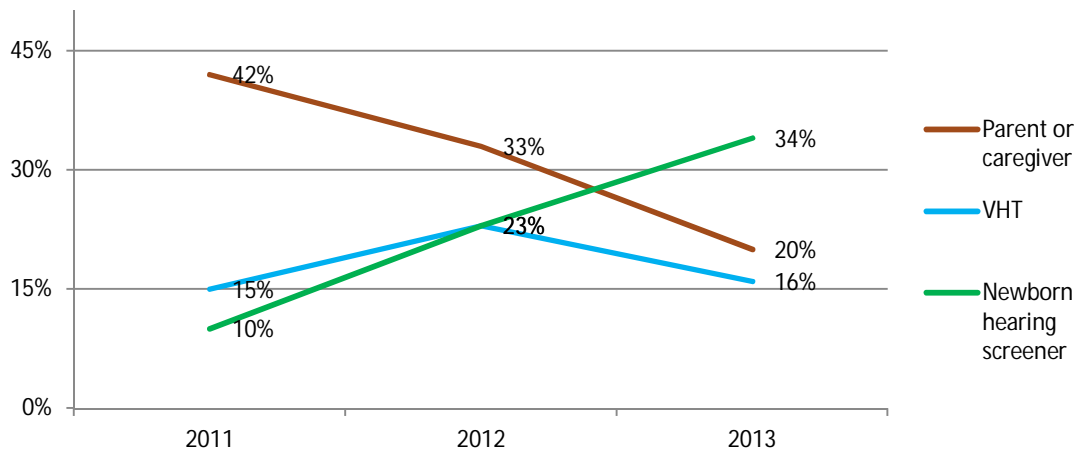


FIGURE 8: FIRST SUSPICION OF HEARING LOSS FOR CHILDREN BORN IN NEW ZEALAND (2010-13)

### Age at diagnosis

Figure 9 shows the number of cases identified by the age of the child. For the first time, by the end of 2013, the majority of children having their B4 School Check will have been screened soon after birth by the newborn hearing screening programme.

There is a notable peak in the number of notifications during the first year of life - this is undoubtedly the effect of the universal newborn hearing screening programme. The peak for diagnosis in the first year of life is now twice as high as it was in 2010, when the database was re-launched. This shows a positive trend, as more children are being diagnosed early.

A further peak can be seen for four, five and six year olds; this is likely to correspond to the B4 School Check<sup>34</sup>. (See page 14 or the glossary for further information on the B4 School Check.)

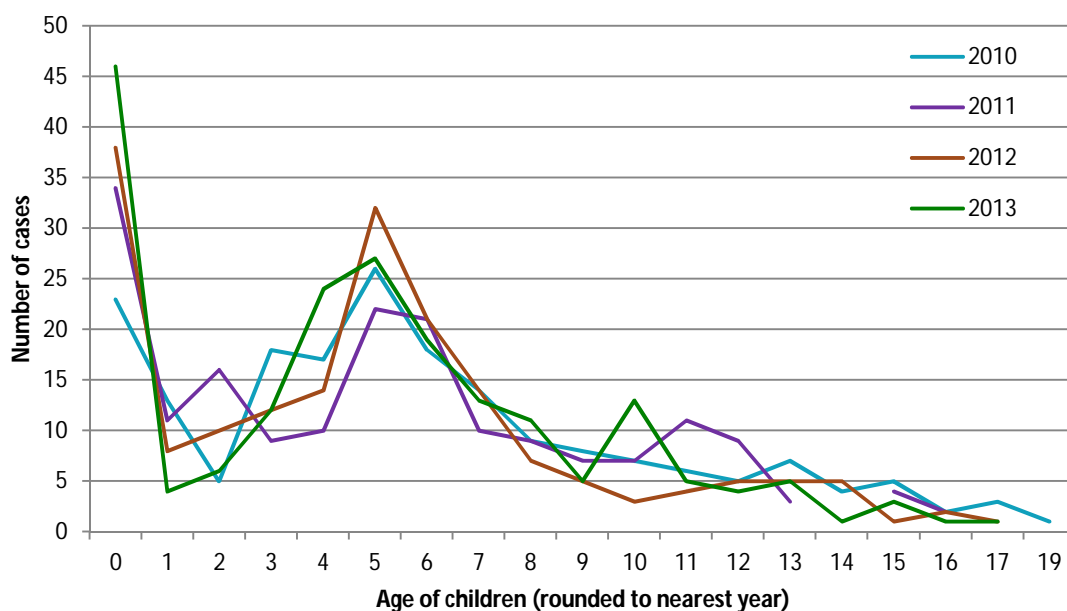


FIGURE 9: NUMBER OF CHILDREN DIAGNOSED BY AGE (2010-2013)

### Overall age at identification

There are a number of issues with reporting the average age at identification (diagnosis) for all groups of children. While this may have some meaning as it describes the average age at which providers will begin working with children to provide interventions of some type, the average

relates to all newly diagnosed children, as it is not possible to separate out children with hearing losses which are late onset (such as progressive and acquired hearing losses).

Keeping this in mind, the average and median ages at diagnosis for children with all degrees of hearing loss are described in **Error! Reference source not found.**

Table 6 shows that while the average age at confirmation is dropping, year on year, the reduction is quite slow and seems to have been influenced by the increase in the number of notifications around 5 years of age for 2012 and 2013 as well as the increases at 10 years of age for 2013 and at 10-11 years for 2011. Those born in New Zealand have a more marked drop in the average age than the full sample, which includes those born overseas and a small number where the place of birth is unknown.

	2010	2011	2012	2013
<b>Average all cases</b>	68	62 <sup>i</sup>	61	61
<b>Average born in New Zealand</b>	65	57	56	54

**TABLE 6: AVERAGE AGES OF DIAGNOSIS FOR ALL CASES (2010-2013)**

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

For the purpose of comparison with previous data, the average age at diagnosis is presented, but the average age for 2013 has also been split by further subgroups in Table 7 below, to add meaning to this measure.

<b>Groups more likely to be identified later</b>	<b>Groups more likely to be identified earlier</b>
born overseas - 103 month average	born in New Zealand – 54 month average
mild hearing losses - 75 months	profound hearing loss - 18 months
acquired hearing losses, e.g. late onset, progressive and trauma related - 79 month average	hearing loss suspected to have been present at birth - 21 month average
unilateral hearing losses - 81 months	bilateral hearing losses - 50 months

**TABLE 7: EARLY AND LATE AVERAGE AGES OF IDENTIFICATION (2013)**

### **Age at diagnosis by severity of hearing loss**

Table 8 shows the average age at diagnosis (confirmation of hearing loss) for children and young people with *bilateral* hearing loss in each of the American Speech Language Association (ASHA) severity categories. As expected, mild and moderate hearing losses are identified later than more severe losses. Please note that a reasonable number of records in the database contain incomplete severity data and also that the table below only includes cases of bilateral hearing loss, so these

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

<sup>ii</sup> 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.

figures do not describe the age at diagnosis for the full 2013 cohort. Those cases with incomplete severity data are more likely to have been diagnosed as babies and as a result are more likely to have an earlier diagnosis than the average found in the full sample.

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

Degree of hearing loss (ASHA, Clark, classification system)	Average months at diagnosis (2011)	Average months at diagnosis (2012)	Average months at diagnosis (2013)
<b>Mild</b>	81	78	75
<b>Moderate</b>	79	68	65
<b>Moderately severe</b>	53	42	NA (not enough cases)
<b>Severe</b>	48	NA (not enough cases)	NA (not enough cases)
<b>Profound</b>	39	22	18

**TABLE 8: AVERAGE AGE AT DIAGNOSIS, IN MONTHS, FOR BILATERAL HEARING LOSSES BY DEGREE CONTAINING SEVERITY DATA (2013)<sup>i</sup>**

The greatest variability in the age at diagnosis is for mild and moderate hearing losses, although there are a number of relatively late diagnoses for children and young people with a profound hearing loss, including bilateral cases. The database does not include information about the proportion of losses which are thought to be progressive in nature.

### Age at diagnosis and ethnicity

Figure 10 shows the average identification ages by ethnic group, for all children and young people notified, where ethnicity information was provided. Please note this graph shows only those born in New Zealand, as there is a significant difference between those born in NZ (identified earlier on average) and those not born in New Zealand (identified later on average). In addition, keep in mind that these data are not priority coded, hence a small number of cases can be in two or more ethnicity groups at one time. MELAA data are contained in this graph but there were no children within this group notified in 2013 - this group is always very small.

While Māori are more likely to have bilateral hearing losses (which are on average identified earlier than unilateral losses), they are also more likely to have mild and moderate hearing losses (which are on average identified later than severe and profound losses) than their NZ European counterparts (Digby et al, under review). This makes it difficult to understand how effectively the system is working to detect hearing losses early among Māori children and young people. We will look closely at data in 2014 to see whether we can draw any conclusions about these data.

In addition, the proportion of cases reported as Māori within the database has grown since 2010 – this could be due to a greater focus on accurately coding ethnicity in some areas, although we have no data to confirm this possibility.

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

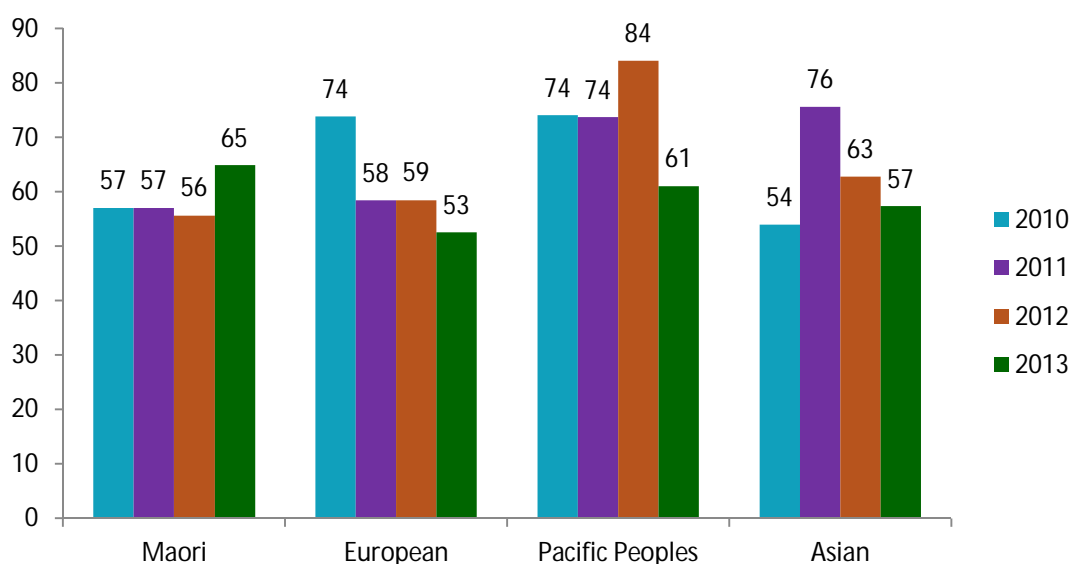


FIGURE 10: AVERAGE AGE OF DIAGNOSIS BY ETHNICITY IN MONTHS (2010-2013)

A number of previous DND reports (1995-2005) noted that Māori and/or Pacific children were identified later than New Zealand European children, although this difference was not reported in every DND report<sup>i</sup>.

### Newborn hearing screening

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

As with last year, all district health boards were screening babies for the full calendar year, and therefore the full notification period. Data in this section of the report relate only to those children born in New Zealand.

### Screening status

Table 9 shows the screening status of NZ-born children notified in the period 2010 to 2013. Please note that this table shows children diagnosed at varying ages, hence it includes children who were born before the [UNHSEIP](#) was fully rolled out. As expected, the proportion of children being diagnosed as a result of a referral from the UNHSEIP is increasing, and the proportion of notifications not offered screening is falling.

Of particular interest were the eight children (5% in the table above) who were referred from their newborn hearing screen and for whom follow-up did not occur at the time, but who were later diagnosed with a hearing loss, and those children who were screened and passed this screening.

Please note that the children in the database range from 0 to those under 19 years of age. Almost half (48%) of the cases notified for 2013 (n=82) were not screened as no screening service was available in their area at the time of their birth.

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

<sup>ii</sup> Northland, Waitemata, Auckland, Counties Manukau, Wairarapa, Nelson/Marlborough, Southern and West Coast.



Was universal newborn hearing screening (using aABR or aOAE) offered to this family after this child or young person's birth?		2010	2011	2012	2013
<b>No</b>	No, a screening programme was not in place, but the child was directly referred to audiology due to atresia	3%	4%	4%	3%
	No, this service was not available at the time (at the time of diagnosis)	69%	54%	55%	48%
<b>Unsure</b>	Unsure whether screening was offered to this family	7%	4%	6%	6%
<b>Yes</b>	Yes screening was offered but this child was not screened...	1%	1%	1%	0%
	Yes, the child was screened and referred but follow-up did not occur at the time, and so this is a delayed diagnosis	0%	4%	2%	5%
	Yes, this child was screened and passed	1%	7%	5%	7%
	Yes, this diagnosis is a result of a refer on the screening test	19%	27%	27%	32%

TABLE 9: SCREENING STATUS OF CHILDREN BORN IN NEW ZEALAND, 2010-2013

Loss to follow-up is a significant issue for newborn hearing screening programmes internationally. As assessment data from the UNSHEIP is still incomplete, the true extent of loss to follow-up in the UNHSEIP cannot be ascertained. However, we can look for differences in loss to follow-up within the DND.

During the 2010 to 2013 period, 5% of children and young people of Māori ethnicity were diagnosed late because, although they referred from their hearing screen, this was not followed up at the time. This is a concern in its own right, but also as it is more than double the proportion (2%) experienced by New Zealand European children and young people.

Overseas studies have also found differences in rates of loss to follow-up between various groups. One such study, from the Massachusetts EDHI programme, found children who were non-white, covered by public insurance, whose parents smoked during pregnancy and those who lived in outlying areas, were at particular risk of being lost to follow-up before their diagnosis<sup>35</sup>.

Also of some concern, is that 19 of these cases were for children under the age of five at the time of diagnosis, i.e. they were born after the start of implementation of screening in their area but not offered screening, although pleasingly, this number has dropped since 2012 when it was 24 cases. This could be because some DHBs are not attempting to achieve universal coverage in their area.

In addition, one child was reported as not being screened, despite the fact that newborn hearing screening was offered by the DHB at the time of the child's birth.

### Referrals from the UNHSEIP

Overseas, a number of comparable newborn hearing screening programmes (such as those in the UK and Australia) seem to be converging at a birth prevalence of approximately 1.0 to 1.1 per thousand babies for bilateral hearing losses, and approximately an additional 0.5 per thousand unilateral hearing losses<sup>36, 37, 38, 39</sup>. This suggests that, if and when the UNHSEIP achieves high coverage and low loss to follow-up, we may expect approximately 90 diagnoses directly from the programme each year, based on a figure of 62,000 births per year<sup>i</sup>. This number may be greater if the prevalence of hearing losses in New Zealand is higher than in similar jurisdictions.

A total of 53 of the 2013 notifications related to children born in New Zealand diagnosed as a direct result of newborn hearing screening. Although the number of notifications resulting from newborn hearing screening plateaued at 27% in 2011 and 2012, this number has increased somewhat in 2013, and now sits at 32%, nearly a third of notifications.

<sup>i</sup> This is an approximation of the number of births reported in 2010.

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

	2010	2011	2012	2013
<b>Number of diagnoses resulting from universal newborn hearing screening</b>	28	44	45	53
<b>...As a proportion of total notifications</b>	16%	27%	27%	32%

**TABLE 10: DIAGNOSES RESULTING FROM NEWBORN HEARING SCREENING IN NEW ZEALAND<sup>i</sup>**

The latest National Screening Unit monitoring report, including data from the nine month period from 1 April to 31<sup>st</sup> December 2012, reports that:

- approximately 83% of babies born across the country during this period completed their newborn hearing screening;
- 78% of those babies completing audiology in the period had their audiology assessment completed by three months of age;
- of the 672 (1.7%) of babies referred from screening, audiological data was provided to NSU for 57% of these; and
- 42 babies were recorded as identified as having a permanent congenital hearing loss. This number excludes permanent conductive (n=4) and temporary conductive hearing losses identified through the UNHSEIP<sup>40</sup>.

Notifications from two large DHBs (Auckland and Waitemata) are thought to be underrepresented in the 2013 database. This is likely to have reduced the number of potential DND notifications resulting from UNHS that were notified in 2012 and 2013.

#### **Key screening goals – age at diagnosis**

The UNHSEIP was implemented in New Zealand to reduce the age of intervention for children born with hearing loss, as this approach has been successful overseas in improving outcomes. Screening programmes achieve this by significantly reducing the age at diagnosis for hearing losses present at birth, compared with identification approaches reliant on risk factors. Key aims of newborn screening programmes include the screening of children by one month of age, diagnosis of hearing loss by three months and the start of intervention by six months of age. These are known as the 1-3-6 goals, and are commonly used in newborn hearing screening programmes internationally.

Measuring the proportion of children with hearing losses identified before the benchmark of three months of age, as a result of a referral from newborn hearing screening, will be an important measure of the success of the New Zealand newborn hearing screening programme. The annual DND reports should provide useful data to show how the overall age at identification changes over time.

There has been a pleasing reduction in the average age at diagnosis of cases referred from newborn hearing screening in New Zealand (therefore born in New Zealand), from 10 months in 2010 to 8 months in 2011 and 5 months in 2012, although this has risen slightly to 6 months in 2013.

Of the 53 cases notified in 2013 and identified as a result of newborn hearing screening within NZ, 41 (77%) were diagnosed by the internationally recommended age of three months (in practical terms within this data this means less than around 3 years and 5 months). This is a pleasing improvement from 59% in 2011 and 73% in 2012. The target age of 3 months for diagnosis has still not been met across all cases, however.

<sup>i</sup> Please note that the table shown in the 2011 report contained data for all cases, whereas this table contains data only for children born in NZ

A number of cases demonstrated that referral processes are not working as quickly as they should in all instances:

*“There was a delay between referral from NBHS [newborn hearing screening] to diagnostic ABR. We did not receive the referral until 06/05/2010 – [the] baby was over 3 months old.”*

Of the 12 children diagnosed after 3 months of age, one or more reasons for the delay were reported in all but one case. More than half of these cases had the diagnostic delay attributed to the ‘audiologist having difficulty getting a confirmed diagnosis’. Reasons provided are listed below:

- audiologist having difficulties getting a confirmed diagnosis (n=7);
- parents not attending appointments (n=5);
- waiting time to see a hearing professional (n=2);
- follow-up in the system did not occur as scheduled (n=2); and
- multiple screenings in the UNHSEIP (n=1).

Two 2013 cases mentioned multiple newborn hearing screenings as the reason or part of the reason for the delay in diagnosis – the target is that screening is completed before the child reaches the age of 1 month:

*“Was screened 3x by NBHS with last screening at age 4 months old before referred to audiology. 1x Audiology appointment rescheduled because mum was sick.”*

*“Screening was not completed until baby was 2 months, then the family had to travel from [one area] to [another area] for 2 Audiology appt. to get confirmed diagnosis.”*

More information about delays causes in all groups can be found in the section on *Delay causes*, beginning on page 30.

### **Identification of false negatives**

The DND probably provides the only method for identifying potential [false negatives](#) from the newborn hearing screening programme<sup>41</sup>.

Cases included in the potential false negative category (below) 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.

Eleven of the children identified with hearing loss during 2013 had been screened previously and passed this screening. This is not necessarily a concern, as many children develop hearing losses after birth, accounting for approximately half of all cases of hearing loss.

Of these 11 cases, there are two groups which may be useful to remove to help us identify potential false negatives. The first of these is acquired hearing losses, while the second is those with hearing losses where there is some uncertainty - they were either suspected to have been present at birth or the diagnosing audiologist was unsure whether the hearing loss was likely to have been present at birth<sup>1</sup>. As the second of these groups is based on a relatively subjective assessment by the clinician, these cases may or may not be cause for concern.

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<sup>1</sup> Audiologists completing the notification form were asked to answer ‘yes’, ‘no’ or ‘unsure’ to the question ‘Was the hearing loss thought to have been present at birth?’ However, the answer to this question provides only a rough indication, as we cannot know whether the hearing loss was indeed present at birth.

	2010	2011	2012	2013
Total cases identified by year who were screened previously (i.e. are not currently referrals from the UNHSEIP) and who passed this screening	3	11	10	11
Number of cases from regional screening programmes, or from the UNHSEIP, which were not acquired loss, and cases where the loss was thought to have been present at birth <sup>i</sup>	3	5	5	6

**TABLE 11: POTENTIAL FALSE NEGATIVES AND CASES PREVIOUSLY REFERRED FROM HEARING SCREENING**

To narrow our focus further, we can examine the cases identified in 2013 which passed their newborn hearing screen, and in whom the hearing loss was thought to have been present at birth (as opposed to those that could have been or were not) and *not* an acquired hearing loss. The cases in this group were most likely to be false negatives. This year there were no children in this group. This may mean there were no false negatives identified for screening during 2013, or just that none were identified as such and notified to the database.

Please note that, while there was a Ministry of Health initiated recall of 3,422 babies in 2012, 2,064 of whom had potentially been incorrectly screened, there are no 2013 notifications specifically identified as having been false positives associated with this recall. However, only 901 children had been rescreened by November 28, 2012<sup>42</sup> so there may be false negative cases associated with these screening problems which have not yet had their hearing loss identified due to the low rescreening rate to date.

<sup>i</sup> Please note this label was incorrect in the 2012 report.

# Delays in diagnosing hearing loss

## All cases for which delay information was provided

Audiologists were asked to provide information about the length of delay and reasons for the delay, where one existed. Cases where there was no identification delay or where this delay was one month or less represented 36% of all 2013 cases, leaving 64% with delays of more than one month.

The average delay in 2013, between first suspicion of the hearing loss and confirmation of the loss, *including* those born overseas and those with mild, acquired or unilateral hearing losses was 12 months.

While this is a significant average delay between first suspicion of a hearing loss and confirmation of this loss, average delays in 2012 and 2013 are an improvement on 2010 and 2011 figures. This is likely to be due to the introduction of, and improvements within, newborn hearing screening programmes throughout the country.

Year	Delay in months
2013	12
2012	9
2011	18 <sup>i</sup>
2010	22 <sup>ii</sup>

TABLE 12: DELAY IN MONTHS BY YEAR (2010-2013)

## Limited dataset

In an attempt to make comparisons with the length of delay in previous years we have removed cases of acquired hearing loss, those born overseas and those with unilateral hearing losses to conform to the criteria in place for the database before 2006.

Please note that this calculation is now being done using codeframes used in the original dataset from 1996 to 2005, to aid comparability. When the appropriate cases are removed (those with less than moderate or greater hearing loss) the average age at confirmation for 2013 data is 43 months as, shown in Table 13, below<sup>iii</sup>.

Interpretation of these data is difficult, due to the much smaller sample size included in this calculation, and due to potential differences in the way delay and average age in months at diagnosis is calculated now, compared with in the previous dataset. However, it is still worth noting that these data show a downward trend in average delay and average ages at confirmation, although the average delay is still significant.

<sup>i</sup> Revised from the 20 months reported in 2011.

<sup>ii</sup> Revised from the 20 months reported in 2010.

<sup>iii</sup> Some previous notification reports have calculated the proportion of cases with a significant delay of six months or more between first suspicion and confirmation of the hearing loss. As the number of newborns identified with hearing loss grows, and as the goal for identifying these losses is before 3 months of age, applying this six month threshold for determining whether a delay exists no longer seems appropriate.

Year	Average age in months at confirmation of hearing loss <sup>i</sup>	Delay in months
2013	43	9
2012	44	5
2011	54	12
2010	58	11
2005	33	10
2004	45	14
2003	46	11
2002	35	11

TABLE 13: DELAY COMPARISONS WITH PREVIOUS DATA (EXCLUDING ACQUIRED, UNILATERAL, BORN OVERSEAS AND MILD LOSSES) USING 1995-2005 CODES FOR DEGREE OF LOSS

## Delay causes

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

In 2013, 45% of notifications contained one or more reasons for the delay in identifying the child or young person's hearing loss. Of these, more than half had two reasons listed for the delay, while almost half two or more reasons listed.

The most frequently mentioned cause of delay relating to children and young people identified in 2013 related to parents not attending appointments (for any reason).

Reasons for delay	Number of cases where option selected 2010	Number of cases where option selected 2011	Number of cases where option selected 2012	Number of cases where option selected 2013
<b>Audiologist had difficulties getting a confirmed diagnosis (e.g. child unwell)</b>	28	37	35	30
<b>Parents did not attend appointments (for any reason)</b>	12	18	29	35
<b>Waiting time to see hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource)</b>	11	26	27	28
<b>Difficulty getting a referral to audiology (e.g. GP or other health professional dismissed parent concern and no referral was made)</b>	8	13	8	6

TABLE 14: REASONS FOR DELAY BETWEEN SUSPICION AND CONFIRMATION OF HEARING LOSS 2010 TO 2013

Other common/important reasons for delay included: follow-up lost in the system (n=8), parents moved a number of times making follow-up difficult (n=5).

One parent decided that the child's diagnostic appointment following referral on their newborn hearing screening was unnecessary, although this was compounded by the travelling distance to receive an assessment:

*"Mum felt child failed newborn hearing screenings due to congestion at the time these took place and did not want to travel 2.5 hours for the diagnostic ABR appointment so declined further involvement"*

<sup>i</sup> These figures have been recalculated this year – previous calculations of average age contained in this table in previous years did not exclude cases which were not moderate or greater based on the better ear.

*from audiology. Child was later tested following the diagnosis of her younger sibling via newborn hearing screening with subsequent confirmation of her sibling's hearing loss using VRA and the fitting of hearing aids."*

Two audiologists recorded that they or the family suspected the child had been incorrectly passed at their [B4 School Check](#).

It is important that parents understand screening is not a diagnostic assessment and that a screening is only attempting to identify children with a high index of suspicion at a particular time; it doesn't preclude that a hearing loss may develop after the child is screened.

### **Multiple reasons for delays**

The following quotes illustrate the multiple reasons for the delay in diagnosis for some cases during 2013:

*"Child only shifted to [area] a few years ago. He was born in the [another area] then the family moved to [a third area]. In [a third area], the family tried to access SLT services but was told that he didn't qualify as his speech wasn't that bad. No hearing screen has been conducted that I am aware of until now."*

*"Family referred by paediatrician at [age], DNA. Seen by speech and language therapist and early intervention team but no re-referral made despite deaf sounding speech. Referred by mobile ear clinic with documented fail on audiogram in 2009, waited 6 months instead of being referred directly to audiology. Seen by ENT Registrar in 2010 without a hearing test taking place. F/Up scheduled with hearing test arranged in ENT clinic, didn't attend appointment. Re-referred to ENT again in 2012 and seen after another 6 month wait in 2013."*

*"Child was screened and passed with [an] underlying BC at pass levels. ENT was monitoring and took a long time to do EUA [Investigation of ears under anesthetic] as they assumed his conductive hearing loss was glue ear. EUA found that left ear was atretic and right EAM was too narrow to insert grommets. Hence permanent conductive hearing loss."*

# Severity

## Audiometric data

Audiometric data were requested for both right and left ears. Audiologists notifying cases to the database were asked to provide air and bone conduction thresholds from the pure tone audiogram. In cases where the young age of the child meant the audiologist was unable to obtain audiometric data, audiologists were asked to estimate thresholds from the ABR using correction factors from the National Screening Unit's 2009 Universal Newborn Hearing Screening and Early Intervention Programme National Policy and Quality Standards<sup>i</sup>.

Examining the four data points for each ear shows that these data-points were provided for 156 and 155 of the 200 cases notified to the database, for right and left ears respectively. Notifying audiologists are being encouraged to provide more audiometric data for cases being notified.

Audiologists were approached about a number of cases, and were able to complete some missing information. Of the cases which still contained missing data, data is more commonly reported for 0.5 kHz and 2.0 kHz and less likely to be reported for 4.0 kHz and 1.0 kHz frequencies.

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

Where a significant air bone gap was present, bone conduction thresholds at the appropriate frequencies were also collected and bone conduction ABR correction factors of -5 for 0.5 and 2.0 kHz were provided to audiologists in the online notification form<sup>ii</sup>.

In 2013, 70% of cases notified contained data taken from the behavioural pure tone audiogram, with the remaining 30% were based on the ABR.

As shown in Figure 11, below, the proportion of cases with pure tone audiometry data is dropping slowly, from 79% in 2010 to 70% in 2013. This is likely to be an indication that fewer children being assessed are old enough to have their hearing assessed behaviourally. We hope to see this figure drop further in future years as newborn hearing screening programme coverage rates continue to increase and hearing loss is diagnosed at younger ages.

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

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



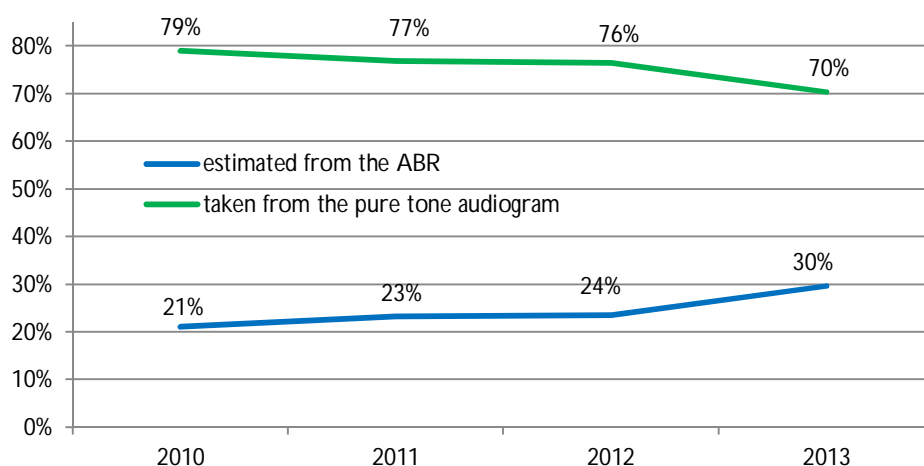


FIGURE 11: PROPORTION OF THRESHOLDS FROM ABR VS PTA (2010-2013)

## Classifications

A large number of classification systems are used to categorise hearing loss severity, locally and in overseas jurisdictions. Differences between these systems make it difficult for meaningful direct longitudinal and geographical comparisons of the proportion of children in a particular severity category<sup>i</sup>. 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 15 shows some of the differences between local and overseas severity classifications (these systems use an average of the pure-tone thresholds at 0.5 kHz, 1.0 kHz, 2.0 kHz and 4.0 kHz).

Audiologists in New Zealand are commonly using Clark's 1981 (ASHA) classifications within their clinical practice, as per the New Zealand Audiological Society practice guidelines.

	1996-2005 NZ DND	1982-1996 NZ DND	Clark 1981 (ASHA) <sup>43</sup>	Jerger and Jerger (ASHA) <sup>44</sup>	World Health Organisation <sup>45</sup>	CDC <sup>46</sup>	Proposed code from Davis and Davis (2011)
<b>Normal</b>			-10- 15dBHL		≤25dBHL		
<b>Slight</b>			16-25dHBL	0-20dBHL	26-40dBHL		
<b>Mild</b>	26-40dBHL	30-55dBHL	26-40dHBL	20-40dBHL		21- 40dBHL	30-39 dBHL
<b>Moderate</b>	41-65dBHL		41-55dHBL	40-60dBHL	41-60dBHL	41- 70dBHL	40-69 dBHL
<b>Moderately Severe</b>		56-85dBHL	56-70dHBL				
<b>Severe</b>	66-95dBHL		71-90dHBL	60-80dBHL	61-80dBHL	71- 90dBHL	70-94 dBHL
<b>Profound</b>	>95dBHL	≥86dBHL	≥91dBHL	≥81dBHL	≥81dBHL	≥91dBHL	95+ dBHL

TABLE 15: COMPARISON OF AUDIOMETRIC SEVERITY CLASSIFICATION SYSTEMS

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

### Calculating severity for notifications

While the New Zealand DND collected some audiometric data for a number of years until the end of 2005, this information was insufficient to allow comparisons to be made easily with data from other jurisdictions. From 2010, the re-launched database has requested full audiometric data from audiologists notifying cases, in the hope that more meaningful comparisons can now be made with overseas data.

As the original database (1982-2005) did not keep 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.

### Severity for recent notifications

Table 16 shows the severity of hearing losses diagnosed for the first time in 2013, which is calculated in two ways. The first of these is using data containing all 8 data points, while the second contains extrapolations. Remaining graphs in this report contain severity data from records containing all eight data points only.

While only cases where all 8 audiometric data points are present are able to be included in most severity calculations, interpolation of data has been used in some cases, to provide a more complete picture of the severity of hearing losses diagnosed in 2013. Extrapolation is only used where three of the four data points are provided for one ear, and where surrounding data-points are provided.

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

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

- proportion of less severe hearing losses is higher among bilateral cases when compared with those pertaining to only one ear;
- number of bilateral hearing losses for which severity can be calculated rises from 87 to 108 when extrapolation is used;
- proportion of mild bilateral losses drops when these cases are removed, increasing the proportion of moderate and greater hearing losses; and
- proportion of moderate and moderately severe losses rises for unilateral cases, with the sample rising by six cases to 68.

Degree of loss using ASHA severity codeframe	Bilateral 2013	Bilateral (extrapolated) 2013	Unilateral 2013	Unilateral 2013 (extrapolated)
<b>mild</b>	68%	59%	42%	39%
<b>moderate</b>	24%	28%	17%	18%
<b>moderately severe</b>	3%	5%	11%	12%
<b>severe</b>	1%	5%	26%	26%
<b>profound</b>	4%	3%	3%	4%
<b>Sample size</b>	87	108	62	68

TABLE 16: COMPARISON OF SEVERITY CLASSIFICATIONS BASED ON METHODOLOGY

### Severity profile differences between bilateral and unilateral hearing losses

A difference can be found in the severity profile of bilateral hearing losses (less severe and profound losses, compared with unilateral hearing losses which have more severe and profound losses, is seen in Figure 12.

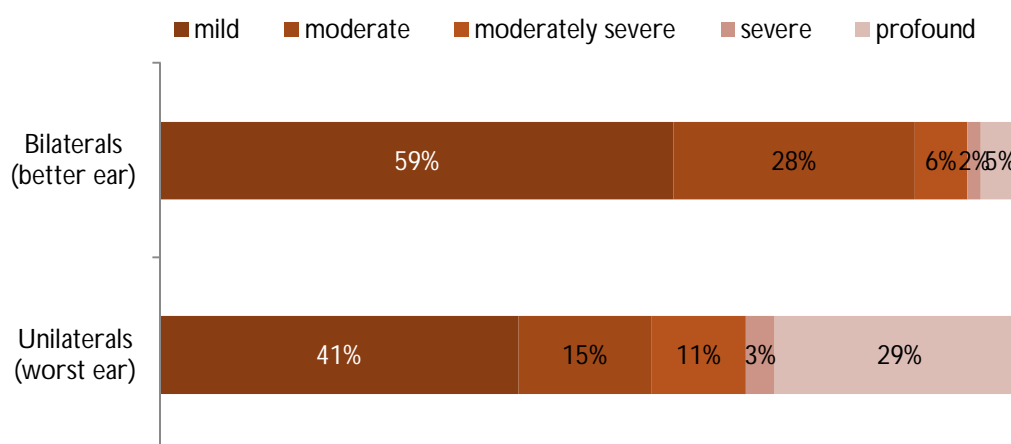


FIGURE 12: UNILATERAL AND BILATERAL HEARING LOSSES BY DEGREE (2010 - 2013)

This is particularly the case when the comparison is made between the ear with hearing loss in unilateral cases and the better ear in cases of bilateral loss. Clearly these differences lessen when comparison is made with the worse ear in bilateral cases.

Other reasons for these differences may relate to:

- Unilateral hearing losses within the database are, on average, found later than bilateral hearing losses and may have had more time to become more severe where these are progressive losses. Bilateral hearing losses are more likely to be identified more quickly and therefore have less time to progress;
- Low and mid frequency congenital hearing losses are more likely to be bilateral in nature and are more likely to be mild or moderate; and
- Differences in genetic and other causes of unilateral versus bilateral hearing losses.

### Comparisons with previous data

By categorising the notifications using the DND severity codeframe (1996-2005), a longitudinal comparison of the proportion of children in each group is possible using data reported between 2001 and 2005 and more recent data. Table 17 shows the proportion of hearing loss notifications in each category between 2010 and 2013 and compares this with data from 2001 to 2004<sup>i</sup>. The 2010 to 2013 figures shown here exclude those children born overseas, unilateral hearing losses and those with acquired hearing losses as reports prior to 2005 excluded these cases.

Proportion of cases notified by degree of hearing loss	2001	2002	2003	2004	Average 2001-2004	2010	2011	2012	2013
<b>Mild</b>	47%	47%	56%	43%	<b>48%</b>	59%	<b>60%</b>	<b>54%</b>	<b>66%</b>
<b>Moderate</b>	35%	39%	33%	34%	<b>35%</b>	33%	<b>28%</b>	<b>42%</b>	<b>28%</b>
<b>Severe</b>	10%	9%	6%	15%	<b>10%</b>	4%	<b>5%</b>	<b>1%</b>	<b>3%</b>
<b>Profound</b>	8%	5%	5%	7%	<b>6%</b>	5%	<b>3%</b>	<b>3%</b>	<b>3%</b>

TABLE 17: NOTIFICATIONS BY DEGREE OF HEARING LOSS USING 1996-2005 CLASSIFICATION SYSTEM, SELECTED CASES ONLY

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

Previously we found that the severity profile of cases seemed to be different from previous years – we noted that we would be watching future data to see whether the severity profile returned to a pattern which more closely matched those seen before 2005. A return to historical patterns is not evident, either with 2013 cases containing full audiometric thresholds, or when compared with data in Table 16, in an attempt to include more cases.

Findings this year show a very small proportion of severe and profound hearing losses, and the highest proportion of mild cases since the database was re-launched. Factors which may be contributing to the generally small proportion of more severe hearing losses are listed below:

- Information about individual children and young people are included in the dataset at the time of first diagnoses. A greater proportion of hearing losses are now being identified earlier, thanks to the introduction of newborn hearing screening. As a result, progressive hearing losses have not yet had the time to worsen, meaning the proportion of more severe losses may be less;
- 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<sup>i</sup>;
- Often children diagnosed with hearing loss have a sloping hearing loss and the better thresholds reduce the average degree of hearing loss; and
- As noted previously, vaccination programmes have reduced rates of meningitis in New Zealand and this reduction is expected to have led to a reduction in rates of (more severe) hearing loss<sup>47</sup>. However, the reduction in the number of more severe cases due to meningitis is likely to be small.

It is interesting to note that some overseas data, including those contained in Table 18 also indicate lower numbers in the severe category when compared with the profound category, even when the codeframes are standardised as they are in this case.

### Ethnicity and severity profiles

A recent analysis by Digby *et al* [under review] has shown a difference in the severity profile of hearing losses among Māori compared with their New Zealand European counterparts, with Māori having greater numbers of mild and moderate hearing losses. (Refer to page 15 for additional information on ethnicity differences.)

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

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

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

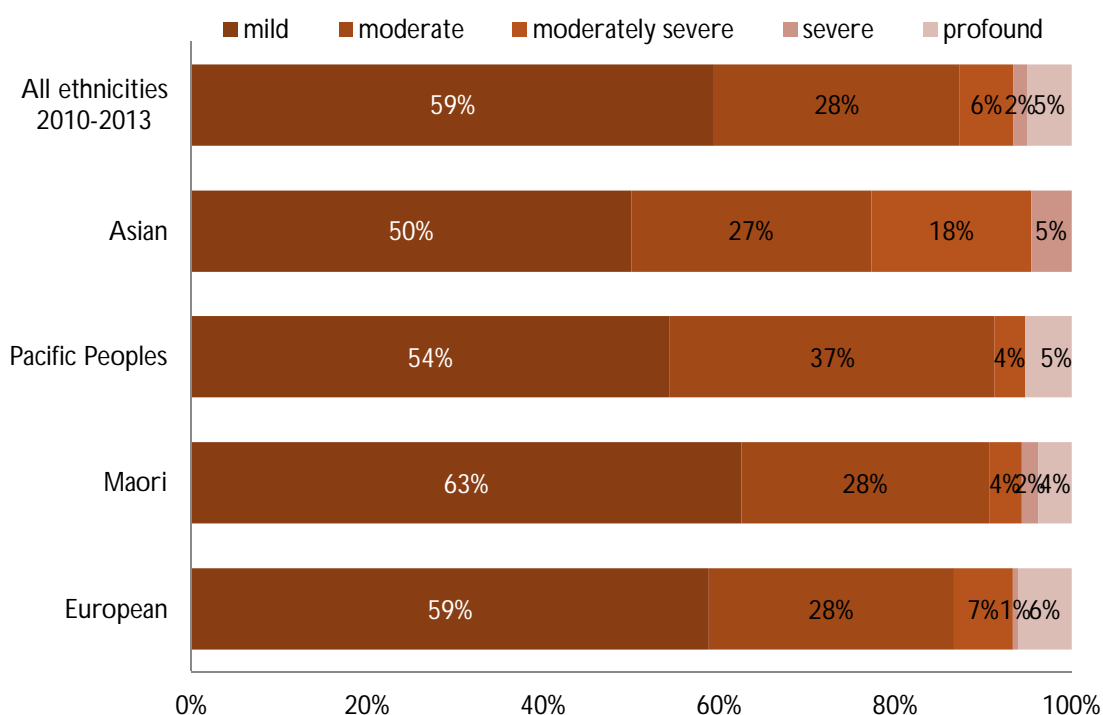


FIGURE 13: 2013 BILATERAL CASES BY DEGREE AND ETHNICITY (CASES CONTAINING COMPLETE SEVERITY DATA)

## Comparisons with international data

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

The 2012 Notification report contained a comparison of moderate and greater hearing losses among our database with those from the UK, Finland and the USA. This showed a greater proportion of moderate hearing losses, and fewer severe and profound losses than those analyses. This could in some part be due to the fact that our local data contains some records with only limited information. In addition, the overseas data excludes cases of mild hearing losses.

The table below compares our bilateral local data (2010-2013) with data from Colorado<sup>48</sup> (2006-2012 data from a largely European population). Local DND data have been recoded to match the severity codeframe used in Colorado. This shows a relatively high proportion of mild hearing losses, and fewer with severe and profound losses in the New Zealand sample.

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

TABLE 18: SEVERITY COMPARISON COLORADO AND NEW ZEALAND

# Hearing aids and cochlear implants

## Hearing aids

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

As has been the case with other data since 2010, the majority of children and young people with a hearing loss which was first diagnosed in 2013 are to be fitted with two hearing aids.

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

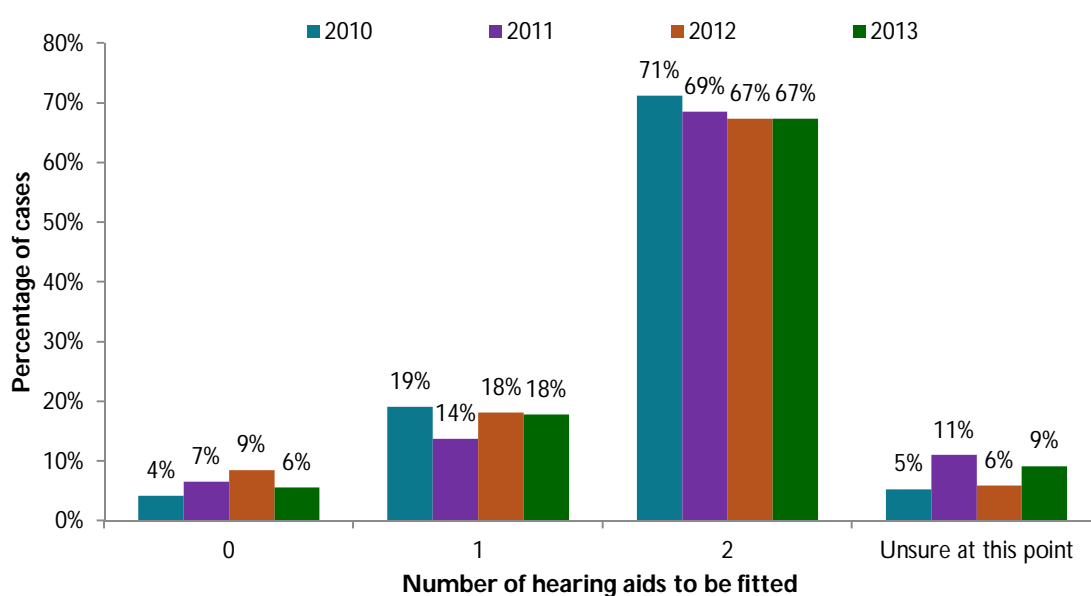


FIGURE 14: NUMBER OF HEARING AIDS TO BE FITTED (2010 TO 2013)

Since the database was re-launched in 2010, the number of children who are receiving hearing aids before 6 months of age has doubled, from 23 to 46. This is very likely to be, in large part, the result of the introduction of newborn hearing screening.

It is worth noting that some children with unilateral hearing losses were reported to be receiving more than one hearing aid. This is because although the average threshold for the better ear does not meet the 26dBHL average required for inclusion in the database<sup>i</sup>, one or more hearing thresholds are seen as sufficiently poor to warrant amplification in the better ear. This is indicative of one of the limitations related to classification systems, which average hearing thresholds across four frequencies and categorise children into broad severity groups.

In such cases, unilateral hearing losses, as they are usually defined, do indicate some level of asymmetry, but do not indicate that the child doesn't require help to improve their hearing in their better ear.

<sup>i</sup> Over the four audiometric frequencies: 0.5, 1.0, 2.0 and 4.0 kHz.

## Funding for hearing aids

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

This shows MOH funded hearing aids for children and young people<sup>49</sup> during the 2013 calendar year. A total of 1479 service users (adults and children) received hearing aids during this period, down slightly on the 1613 in the year ending June 2012. This data now corresponds to the reporting period for the DND.

Ethnicity	0-3 years	4-5 years	6-15 years	16-18 years	Total
<b>NZ European</b>	98	73	357	76	604
<b>NZ Maori</b>	75	78	302	19	474
<b>Pacific</b>	19	19	117	15	170
<b>Other</b>	36	22	152	21	231
<b>Total</b>	<b>228</b>	<b>192</b>	<b>928</b>	<b>131</b>	<b>1479</b>

TABLE 19: MOH FUNDING OF CHILDREN'S HEARING AIDS FOR THE CALENDAR YEAR ENDING 31ST DECEMBER 2013

## Cochlear Implants

Although we don't collect information about [cochlear implants](#) in the database, it is useful to provide some information about the number of cochlear implants provided to children and young people in New Zealand, and some background on the funding for these implants.

A single cochlear implant is funded by the Ministry of Health for all children and young people who meet the candidacy criteria. In cases where children have been deafened by meningitis, children receive two internal arrays and one processor.

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

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

During the 2014 calendar year there were 28 cochlear implants provided in the Northern Region and 21 in the Southern Region, to children and young people under the age of 19. These (unilateral) implants are provided based on clinical need meaning there is no waiting list for children who have been assessed by one of the cochlear implant teams and this assessment has shown that they may benefit from a cochlear implant.

Cochlear Implant Trust	Year ending 31 December 2013	Year ending 31 December 2012	Year ending 31 December 2011	Year ending 31 December 2010
Northern Cochlear Implant Programme <sup>50</sup>	28	13	16	17
Southern Hearing Charitable Programme <sup>51</sup>	21	18	17	18
<b>Total publicly funded new implants in NZ</b>	<b>49</b>	<b>31</b>	<b>33</b>	<b>35</b>

**TABLE 20: PUBLICLY FUNDED COCHLEAR IMPLANTS IN NEW ZEALAND**

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

While the DND may be missing some notifications for children in the severe and profound categories, there are a number of other reasons why this figure is low compared with the number of children implanted during the same period. One reason is that some children who were notified to the database as having less severe hearing losses develop more significant losses over time, something which is not tracked by the database. For example, The Northern Cochlear Implant Programme reported in 2011 that an increased and significant number of children and young people receiving cochlear implants over the last two years had progressive hearing losses. In such cases, the hearing losses would have been less severe at the time of initial identification and notification to the database.



# Appendix A: History of the database

## History of the DND

The DND was New Zealand's annual reporting system for new cases of hearing loss among children and young people from 1982 to 2005. This system included data on the number and age of children diagnosed with permanent hearing loss and annual reports describing collected notifications were released.

The data presented in reports before 2006 contains notifications provided to the database within a specific year; that is they pertained to children notified to the database in a calendar year, rather than those who are identified in that year<sup>52</sup>.

The database was managed by the National Audiology Centre on behalf of the Ministry of Health and later by the Auckland District Health Board.

The database provided the only source of information from which the prevalence of permanent hearing loss could be estimated, and from which the characteristics of new cases of hearing loss among children and young people could be understood.

In 2006, the Auckland District Health Board discontinued its contract to provide services associated with this database. No new provider was sought by the Ministry of Health. Prior to this, the National Audiology Centre held national contracts for a number of projects, including the collection and reporting of deafness data.

Between 2006 and 2009, a number of groups expressed concern that information on the number and nature of new hearing loss diagnoses among children in New Zealand was no longer being collected.

The DND was seen to have even greater importance from 2007, the start of implementation of the Universal Newborn Hearing Screening and Early Identification Programme (UNSHEIP). Information from the DND was seen as providing an important measure of changes in the age of identification and as the only way to identify potential false negatives within the screening programme.

In 2010 the DND was re-launched, with audiologists around the country encouraged to notify diagnosed hearing losses through a new online form. This re-launched database was funded by the New Zealand Audiological Society with help from Janet Digby.

We are delighted that the Ministry of Health began funding the DND from the start of 2012. The database is now managed through a contract with **accessible** and will build on the work funded by the New Zealand Audiological Society.

## Inclusion criteria

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

“Children under 18 years with congenital hearing losses or any hearing loss not remediable by medical or surgical means, and who require hearing aids and/or surgical intervention. They must have an average bilateral hearing loss (over four audiometric frequencies 500-4000Hz), greater than 26dBHL in the better ear (Northern and Downs classification 1984)<sup>53</sup>.”

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

The criteria for inclusion were modified for the 2010 re-launch of the database, based on feedback from a small working group<sup>i</sup>.

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

In addition, this database now includes children:

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

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

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

### Notifying cases

Although the Database was restarted by the New Zealand Audiological Society, efforts have been, and continue to be made, to publicise the database to non-members of the Society in an attempt to collect as many notifications as possible.

Notifications are collected through an online survey form, to reduce data entry errors and to try to make it as easy as possible for audiologists to notify cases. A revised consent process was also implemented on re-launch to ensure all information is collected with the consent of the family. Data is backed up regularly and information is sent through a secure link. Standardised methods for data analysis are now being used.

### Future renaming of the database

During 2012, feedback on the name of the database was sought from parents of deaf children, Advisors on Deaf Children (AODCs), and audiologists, on a possible change to the name of the database. This feedback did not provide a clear path for renaming the database.

Some individuals and groups felt that changing the name to a broader title, such as the Hearing Loss Notification Database, would have merit, as it would acknowledge the range of types and severity range of hearing losses included. Others felt changing the name of the database could cause confusion and reduce the number of notifications in the short term.

The name of the database (Deafness Notification Database) remains open for consideration. A new name may better reflect the purpose and nature of the database, particularly as changes to the inclusion criteria mean cases of unilateral hearing loss are now included in the database.

If any reader of this report has any ideas on what the database might be called in future, these will be gratefully received by Janet Digby, email: [janet@levare.co.nz](mailto:janet@levare.co.nz).

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

<sup>ii</sup> While cases of unilateral hearing loss were technically excluded from the database until 2005, there were still large numbers of notifications sent to the administrators of the database, although these were not included in the main analysis. Professionals consulted in the development of the re-launched database unanimously believed this group should be included in the database, at least in part as there is strong evidence that this group is at risk of poorer educational outcomes.

## Appendix B: High frequency hearing losses

Based on feedback from the audiological community, high frequency hearing losses which would not meet the original criteria have been collected, as a trial, from July 2011. After receiving eight completed notifications from July until December 2011, ten notifications were received for the 2012 year and 14 for 2013.

As these cases are not included in the main analyses within this document, below is a limited analysis of data from high frequency hearing losses notified in 2013. A small number of notifications which were recorded as high frequency losses actually met the criteria for the main dataset and so were included in this dataset and analysed as such.

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

- Only one of the 14 cases in this category in 2013 had a known aetiology listed, and this case was listed as an acquired hearing loss caused by ototoxic medication.
- 43% of these 2013 cases were reported as being of Māori ethnicity, as were 63% in 2011 and 60% of these 2012;
- years of birth for this sample ranged from 1995 to 2013 with six children aged 4-5 years old at diagnosis;
- 12 of the children/young people notified in this category were born in New Zealand, with birthplace of the remaining two being uncertain;
- 11 of the children and young people notified to this category were to receive one or two hearing aids;
- in one of the cases, the audiologist reported delays were at least in part due to the audiologist having difficulties getting a confirmed diagnosis (e.g. conductive overlay, child unwell), and in three the waiting time to see a hearing professional (e.g. DHB waiting lists to see audiologist, no audiology staff at the DHB, limited staff resource) was recorded as the reason for the delay in identification.

The figures below show the audiometric data for the ten children or young people with high frequency hearing losses, contained within this category. Please note that not all children and young people in this category had hearing loss in both ears, and not all audiometric data points were provided for all children.

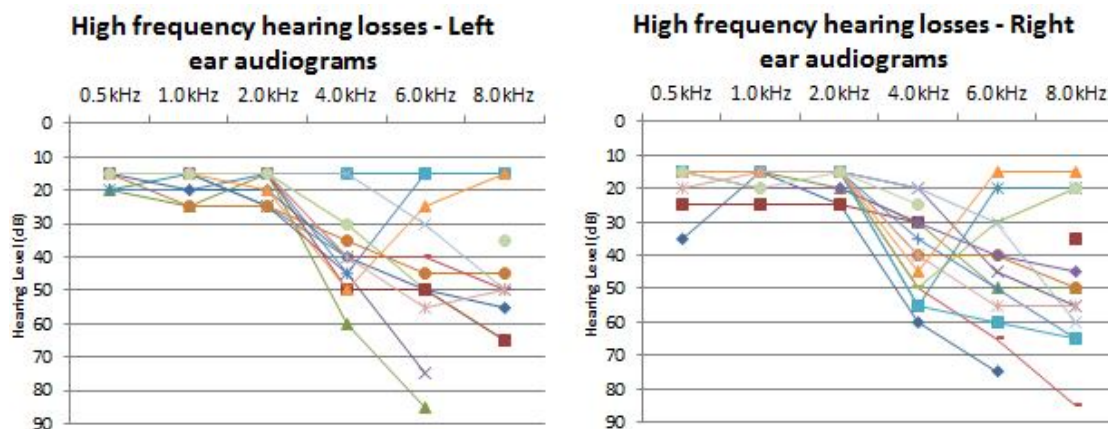


FIGURE 15: 2013 AUDIOGRAM DATA FROM HIGH FREQUENCY HEARING LOSSES FOR RIGHT AND LEFT EARS

We are continuing to trial inclusion of this special group within the database during 2014.

## Appendix C: Family History

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

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

Of the 23% of 2013 cases where a family history was specified, and where the nature of the relationship was specified, two thirds (68%) were parent(s), sibling(s) or grandparent(s), while the remainder were reported as more distant relatives.

Of the 2013 cases where the family member or members with hearing loss include the child/young person's sibling(s) and/or parent(s) and/or grandparent(s):

- Families were asked to tell the audiologist whether the relative still had the hearing loss to get some kind of indication as to whether the hearing loss may be/have been permanent. In 96% of cases the family of the notified child or young person confirmed that the hearing loss was still present, while the family were unsure whether this loss was still present in 4% of cases.
- The majority of these family members use/used one or more hearing aids or cochlear implants (49%), while 44% did not, and in the remainder of cases there was uncertainty regarding whether the relative used such a device.
- The majority of these close relatives (89%) had their hearing loss from childhood, while 9% did not, and the families were unsure in the remainder of cases.

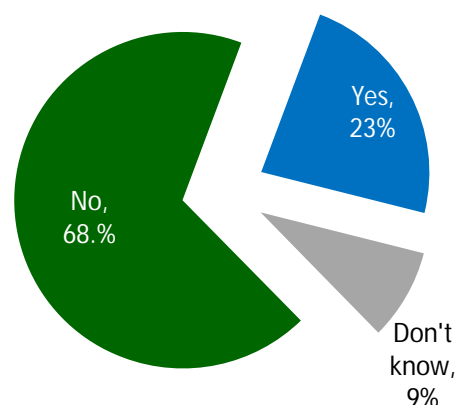


FIGURE 16: IS THERE A FAMILY HISTORY OF HEARING LOSS (2013, ALL CASES)

## Appendix D: Notifications and ethnicity

The method used in this report to classify ethnicity is the total response method, where every person identifying with a particular ethnicity is included in that specific grouping<sup>54</sup>. For example, if someone considers their child to be of Samoan and Māori ethnicities they are recorded under both these groups. This means the total number of ethnic groups selected by respondents is usually greater than the number of respondents.

Using this method provides a more detailed and realistic measure of the relative size of the groups identifying with a particular ethnicity when compared with older survey methods which required respondents to select only one ethnicity, the one with which they mostly identified. Using this method also aligns the database with The New Zealand Census, which began explicitly instructing respondents that they could select more than one category for their ethnicity in 1996.

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

The New Zealand Census (2006) categorises respondents into five major groupings and these groupings will continue to be used for the next Census. These groups are: Māori, Pacific Peoples, Middle Eastern/Latin American/African (MELAA), New Zealand European and Asian.

## Appendix E: Estimating the total number of new diagnoses per year

As no prevalence data exists for permanent hearing loss among New Zealand children and/or young people, it is not possible to accurately estimate how close the database is to collecting data on all new cases of permanent hearing loss which meet the inclusion criteria.

However, we can use a number of methods to provide some indication of the number of new diagnoses of hearing loss annually among children and young people. These methods are listed below. It is likely that the database has been receiving notifications for between 50% and 70% of all cases diagnosed each year, since 2010.

1. **Previous notification counts:** By reviewing the number of notifications to the previous DND in recent years<sup>i</sup>, it would seem reasonable to assume that between 50% and 80% of all new diagnoses may have been notified in 2013, the same proportion estimated since the re-launch of the database in 2010. However, this approach is somewhat flawed, as the criteria for inclusion in the database have changed since the database was re-launched in 2010, and so comparisons are problematic.
2. **Overseas prevalence estimates:** Although there are a number of difficulties using prevalence from overseas jurisdictions, estimates of overall prevalence in children and young people from international data<sup>55,56</sup> would suggest that given there are approximately 1.167 million children and young people in New Zealand to the age of 19 years old<sup>57</sup> there may be approximately 245 new hearing loss diagnoses made annually which fit the new criteria. Using this method the notifications collected in 2011/12 may comprise approximately 76% of the number of 2011 diagnoses.
3. **Newborn hearing screening figures:** The number of cases diagnosed each year from newborn hearing screening can be used to estimate the number of diagnoses missing from the database. The most recent data show that 46 children were diagnosed with hearing losses which were likely to meet the criteria for the database in the nine month period to the end of December 2012.

During the 2012 notification year, 45 cases were diagnosed as a result of referral from the UNHSEIP. By increasing the number of notifications to take into account the fact that the screening programme figure only covers a nine month period, this suggests that the DND may be receiving up to 73% of notifications which were identified directly through newborn hearing screening.

This process is also flawed however, firstly as only 60% of referred cases during this period had audiological results reported to the National Screening Unit (NSU). We do not know whether the prevalence of hearing losses among those referrals reported to the NSU is likely to be the same as the proportion of cases referred through screening but not reported to the NSU.

Secondly, while the types of hearing loss are included in the UNHSEIP monitoring reports, no audiometric data are reported. As a result, we are not able to be definitive about the number of children who might meet the threshold average for inclusion in the database.

4. **Audiologist survey:** In April 2014 audiologists around the country were asked to complete a very short survey to get an indication of the number and proportion of newly diagnosed cases which meet the criteria and which were not notified to the database for the 2013 year. This survey shows we are missing at least 26 notifications for the 2013 year. Please see the following section for additional detail on this survey.

<sup>i</sup> i.e. 2000-2005 We are unsure of the level of over-reporting of cases being notified in more than one reporting period as no record exists of which records were included in the analysis for specific reports before 2006.

## 2014 Survey of Audiologists

A survey was distributed as widely as possible to audiologists who would be likely to diagnose children and young people with hearing loss, including through clinics and chains, in an attempt to reach non-NZAS members. NZAS members receive communications about the database through their administrators and a request to participate was also sent to each audiologist who had provided a notification since the database was re-launched in 2010.

Respondents completed the survey anonymously, although they were asked whether they were in the public or private sectors – 74% of respondents were employed in the public sector, while the remainder came from the private sector.

Of the 82 audiologists responding to the survey, 59% (n=48) reported that they were the first to diagnose one or more children and young people whose hearing loss met the criteria during 2013. Other audiologists who did not diagnose children which met these criteria may not have completed and submitted the survey on reading its purpose. In comparison, 49 audiologists provided notifications to the database for the 2013 year.

We know from the responses to the surveys, that 17% of the responding audiologists did not send in any notifications during the 2013 calendar year, although there is no way of knowing whether the sample which responded to the survey was representative of those who have not been notifying cases to the database.

A further 42% reported providing a notification for all cases, and the same number reported providing notifications for some of their cases. A total of 26 cases were reported as not being notified to the database from these audiologists.

Form fatigue was the single most mentioned reason for not sending in notifications (50%), followed by the issue of difficulties gaining consent (35%). Difficulties providing information by the 31<sup>st</sup> January cut-off date was also mentioned by 22% of respondents. One audiologist surveyed reported being unaware of the database.

Selected comments are below:

*"The database should be linked to Accessable. Accessable should have a tick box in order to inform the deafness notification database that the child/baby is newly diagnosed and needing amplification device. In my opinion, the consent is not needed because we don't get their full information anyway. In your reports, you don't publish their personal details. You just report the numbers (and others). The difficulty of getting their immediate consent for this plus the additional work of notifying online is burdensome."*

*"I appreciate that the Deafness Notification Database is very important. Regular polite reminders by e-mail from you are helpful to keep the notification process current in our minds amidst all the other paperwork we are asked to do. Completion of the form is not difficult...thanks for your efforts."*

*"In a hospital setting when you diagnose a child's hearing impairment, you need to send a report to the referrer, write a referral to ORL, write a referral to AODC, fill in Accessable application form for hearing aid(s), fill in manufacturer's order form for hearing aids and fill in manufacturer's order form for earmoulds, get parents to sign deafness database consent form, and fill in online deafness database form. Sometimes, I forget about the deafness database form in the midst of doing all the other things."*

*"Hmm, could this be made easier to feed into thru an App or programme linked to NOAH that might prompt clinicians to refer to the DN database?"*

*"Some children were diagnosed. Consent was not obtained at the initial diagnosis appointment, and then subsequent appointments were DNAd [did not attend] By the time children were seen again it was past the cut-off date."*

*"I find the consent a real issue specifically for children who have a hearing loss that you are just going to monitoring and see again in 1 year. I cannot ethically and morally make getting the families consent a priority when I am telling them their child has a hearing loss when they are upset and have lots and lots of questions regarding the loss. Getting them to sign a form to me isn't sensitive to the family. If they are coming back for hearing aids we will see them regularly so it's easy to find a time to address this. A suggestion would be to add the Deafness database notification to the consenting the procedure for NBHS then you obtain consent then..."*



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# Glossary

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

**Aetiology:** The cause or set of causes, in this case, of a child or young person's hearing loss.

**Audiometric data:** Audiometric data is about a person's hearing acuity given variations in sound intensity and pitch (frequency), involving thresholds and differing frequencies. The database collects information at 0.5, 1.0, 2.0 and 4.0 kHz wherever possible, and at higher frequencies for children and young people whose hearing loss meets the criteria for inclusion as a 'high frequency hearing loss'.

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

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

**Bilateral hearing loss:** Hearing loss affecting both ears.

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

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

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

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

**False negatives:** False negative is a term used to describe screened children who are incorrectly categorised as having a low risk of the target condition. In this report, this term relates to potential false negatives resulting from the newborn hearing screening programme (UNHSEIP) (i.e. a child who passed the screening test where it is possible that they had a hearing loss at the time the screening was conducted).

**Full Time Equivalents or FTE:** These are used to measure the number of full time equivalent positions for audiologists and generally equate to approximately one full time equivalent for every 38 hours worked per week.

**Inclusion criteria:** The current Deafness Notification Database contains information about children and young people 18 years or younger, born in NZ or overseas, with:

- a permanent hearing loss in one or both ears

- an average loss of 26dBHL or greater over four audiometric frequencies (0.5, 1.0, 2.0 and 4.0 kHz)<sup>i</sup>

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

**Notifications:** Notifications contain data about an individual case of hearing loss, including demographic information, information on the hearing loss and its diagnosis. Information is provided to the DND database with the consent of the young person who has been diagnosed with a hearing impairment, or their parent in the case of babies and children. This information has been provided to the database manager via an online form since 2010.

**Suspicion age:** For the purposes of this database, this is the age at which the hearing loss was first suspected. This may relate to the age the child was referred from the newborn hearing screening programme.

**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 26dBHL four frequency average criterion.

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

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

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

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<sup>i</sup>Based on feedback from the audiological community, high frequency hearing losses which would not meet the original criteria have been collected as a trial from July 2011. We will continue to trial inclusion of this special group within the database. A limited analysis of data from high frequency hearing losses notified in 2013 can be found in Appendix B: High frequency hearing losses, on page 40.

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