An audit of paediatric referrals to the Southern Cochlear Implant Programme (2009–2014)

Jill Mustard, Megan Chinnery, Alice K Guidera, Neil Heslop, Philip A Bird

ABSTRACT

AIM: To determine the effect of the Universal Newborn Hearing Screening and Early Intervention Programme on the age of referral and implantation of prelingually deaf children in the Southern Cochlear Implant Programme.

METHOD: A retrospective review of data collected prospectively from March 2003 to August 2014.

RESULTS: 123 children were referred to the programme with prelingual deafness in the time period. There was a significant decrease in the age of referral (median 6.23 months vs. 21.50 months) and age at implantation (12.66 months vs. 24.0 months) in those that underwent newborn hearing screening. Reasons for delay in referral and implantation were identified.

CONCLUSION: The introduction of universal newborn hearing screening has significantly reduced the age at referral and implantation of prelingually deaf children. However, the screening programme must continue to undergo monitoring and regular audit. Efforts must also be made to reduce the time to referral, including reducing non-attendance rates, education for parents and service providers, and earlier referral of those with comorbidities so as to reduce the time to implantation.

The Southern Cochlear Implant Programme (SCIP) was established in 2003 in Christchurch, New Zealand, and expanded to include a clinic in the Wellington region in 2014. SCIP provides cochlear implant assessment, surgery, audiological and rehabilitation services for children and adults throughout the South Island and the lower North Island. A cochlear implant is a device that can replace the auditory function of the inner ear by electrically stimulating the cells of the spiral ganglion within the cochlea. It consists of a surgically implanted electrode array and an externally worn sound processor. Cochlear implants are a suitable treatment for people who get limited benefit from hearing aids. Cochlear implantation in prelingual infants has undergone marked advances in the past decade. Studies have proven that early detection of hearing loss and early intervention significantly improve long term language skills and cognitive ability.1 Children who receive cochlear implants by 12 months can demonstrate age appropriate speech perception and language skills at 3 years post implant.2 The US Food and Drug Administration (FDA) has approved cochlear implants for infants at 12 months of age since 2000, but there is now evidence that implantation prior to 12 months of age results in improved auditory outcomes, without an increased risk of complications.3,4

Earlier cochlear implantation is supported by findings that primary and secondary neonatal auditory cortices are disorganised and thought to be overlapped by visual projections. As the primary auditory cortex is stimulated, it makes sophisticated reciprocal connections with the secondary auditory cortex essential for language development and the visual projections become concentrated in the normal visual pathway. If there is no input, there may be atrophy of connections or compensatory abnormal projections, which can mean cochlear implantation is less successful.5 This auditory development begins at approximately 26 weeks.
of gestation, major developments occur within the first 12 months with significant ongoing cortical change until four years of age and refining up until 15 years of age. Children who receive implantation before 12 months of age do significantly better in terms of auditory and spoken language outcomes than those implanted between 12 and 24 months. There is now evidence that even earlier implantation yields even better results with no significant increase in risks to the child.

As a result of these findings, Universal Newborn Hearing Screening (UNBHS) has been introduced following World Health Organization (WHO) guidelines. Implementation of these guidelines was anticipated to reduce the age of referral and implantation. A recent dual centre retrospective study in the Netherlands and Germany confirmed a reduction in age in both centres following the introduction of newborn screening, though the scale of this reduction was markedly better in the Dutch population. The difference may be explained by a difference in follow-up systems. In the Netherlands, children are referred by the screening programme to an audiology clinic and are subject to a national tracking system. In Germany, parents are advised to contact an otolaryngologist for a diagnostic hearing test, but there is not a national tracking system.

A previous audit was undertaken by the Southern Cochlear Implant Programme (SCIP) prior to the introduction of UNBHS. At that time, infants hearing was only screened if risk factors for sensori-neural hearing loss were present, or if there were parental concerns. The audit found that the median age of referral was 21.5 months and median age at implant was 24 months, for children with prelingual deafness. At that time, analysis of national data found the mean age for confirmation of profound loss was 13 months, and the mean age of confirmation of severe hearing loss was 34 months.

The national implementation of Universal Newborn Hearing Screening and Early Intervention Programme (UNHSEIP) in New Zealand was a phased process spanning 3 years, from 2007–2010. Since August, 2010, all 20 District Health Boards (DHBs) have been offering screening to families of all newborn babies. The SCIP provides services to 15 of these DHBs.

The purpose of this audit is to determine the effect of the UNBHS on age of referral and implantation, with a focus on those who were considered to have delayed referral, so that preventable delays can be avoided in the future.

**Method**

The SCIP database prospectively collected data from all referrals to the service from March, 2003, to August, 2014. At referral, an assessment is made as to whether the child is pre/peri lingual or post lingual. After initial count, postlingual children were excluded from further analysis. Children referred peri/prelingually were identified; fields used in calculations were: date of birth, date of referral, and date of implantation. It was also recorded if these children had undergone UNBHS. Individual case notes of those implanted following UNBHS who were implanted after 14 months of age were reviewed to identify contributing factors for the delay in implantation.

**Statistical Analysis**

Data were analysed using Microsoft Excel for Mac (2011) version 14.2.5 and GraphPad software (2014). The data were mostly normally distributed. However, the Mann-Whitney U test was used to compare the median data, as there were several major outliers from the mean in all datasets. Data pertaining to the reasons for delay are presented descriptively.

**Results**

In total, 219 children were referred to the programme, of which 123 were prelingual or perilingual at time of referral; the remaining 96 were referred post lingually. The first infant to be referred to the SCIP from UNBHS was in April of 2010, 44 were subsequently referred following UNBHS. The remaining 79 were referred prior to UNBHS.

The infants referred to SCIP who underwent UNBHS were significantly younger (median 6.23 months vs. 21.50 months, Mann-Whitney p<0.001), and underwent implantation at a significantly younger age (median 12.66 months vs 24.00 months).
Table 1: Age at referral and implantation.

<table>
<thead>
<tr>
<th></th>
<th>Prior to UNBHS</th>
<th>Following UNBHS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at referral (months)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>n</td>
<td>79</td>
<td>44</td>
</tr>
<tr>
<td>Median (Range)</td>
<td>21.50 (1.38–80.88)*</td>
<td>6.23 (1.41–50.66)</td>
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<tr>
<td>Age at implantation (months)</td>
<td></td>
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<tr>
<td>n</td>
<td>77</td>
<td>36</td>
</tr>
<tr>
<td>Median (Range)</td>
<td>24.00 (4.08–83.64)*</td>
<td>12.66 (7.20–55.79)</td>
</tr>
<tr>
<td>Time (months) between referral and implantation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>n</td>
<td>4.08 (0.36–14.96)</td>
<td>5.65 (1.68–18.71)*</td>
</tr>
</tbody>
</table>

* p<0.001

Figure 1: Age of individual infants at referral and implantation (Prior to UNBHS vs UNBHS)
months, Mann-Whitney p<0.001) than those who did not undergo UNBHS, but there was a longer mean time between referral and implantation in those that underwent UNBHS (median 5.65 months vs. 4.08 months, Mann-Whitney p<0.001) (Table 1). Figure 1 demonstrates the range of ages at referral and implantation, and compares those that underwent UNBHS and those that did not (the median is depicted by a black line).

Thirteen children who underwent UNBHS were implanted after 14 months of age, which we consider clinically unacceptably late. The following causes for delay were identified, with some children presenting with more than one contributing cause for delay:

- Additional disabilities (3)
- Auditory Neuropathy Spectrum Disorder (3)
- Did not attend appointments (3)
- Late referral from audiology (2)
- Progressive hearing loss (2)
- Health issues requiring other interventions (1)
- Screener error (1)
- Passed screening test (1)
- Passed screening in another country (1)

**Discussion**

This audit has confirmed that the introduction of UNBHS has significantly reduced the time to referral and surgery for cochlear implants in children with prelingual deafness. The median age of implantation of screened infants was 13 months, compared to 24 months for unscreened infants.

It is noted that there was a small, but significant, increase in the time from referral to implantation in the infants that underwent UNBHS. This can be explained, as the screened infants were referred at a significantly younger age than their unscreened counterparts, sometimes as young as 2 to 3 months of age. This necessitated a longer wait time to implantation—until they were of an appropriate age for surgery. Also, their unscreened counterparts, who were referred at an older age, were processed with some urgency so as to achieve the best possible results.

The Ministry of Health UNHSEIP has stated a core goal of “initiation of appropriate medical and audiological services ... by 6 months of age”, which was largely achieved (median referral age was 6 months). The SCIP aims to implant these children by 12 months of age and was still able to achieve this in half the population, with only a slight increase in delay from referral to implantation. However, there is emerging evidence that even earlier implantation is beneficial and the SCIP is moving towards implantation as close to 6 months of age, as is possible.

Two infants who experienced significant delays in referral and implantation were reported to have progressive loss—initially, they did not meet the severity criteria for cochlear implantation, but their hearing went on to deteriorate. These are examples of unavoidable delay, as it would be inappropriate to implant mild-moderate hearing loss. These children had been fitted with hearing aids and underwent appropriate ongoing monitoring, which is essential to prevent subsequent delay in referral should there be progression of the hearing loss.

Two infants who had undergone newborn hearing screening were reported to have passed. It is not known whether they were true passes and the children’s hearing rapidly deteriorated, or whether these were false passes. At the time of the study, in New Zealand the initial method of screening for well babies included otoacoustic emissions (OAEs), those that fail are referred for diagnostic Auditory Brainstem Response (ABR) testing. Those that are deemed to be high risk (eg, family history, intrauterine infection or NICU admission) undergo Automated ABR automatically (AABR) by screeners.

The sensitivity of OAEs is reported to be between 67–100% and AABR 99–100%. These methods of screening are adequate for population-based screening, but they are not 100%. There must be an awareness that the test can be wrong and considered when there are parental concerns or failure to respond. Similarly, a test that requires human administration is susceptible to error, as was evidenced in one case. The equipment used is mostly automated, but adequate provider training must be given to reduce the chance of error. Following a review of newborn hearing screening in March 2014, a revised screening regime is currently being rolled-out nationally,
whereby all babies will be screened using only one test, automated Auditory Brainstem Response (aABR) testing, with one standard screening device. The new regime is expected to reduce risk of protocol error, provide more concise screener scripts and explanations to parents, potentially reduce workload stress, and potentially earlier referral to audiology.18

A screening incident in 2012, whereby some screeners did not screen babies according to the screening protocol, resulted in “delayed referral from audiology” for one of the two children in this category. It is estimated that non-protocol screening occurred in 1.4% of all babies who have completed screening in New Zealand since the programme began full implementation across all DHBs in 2010.19 All babies identified as incorrectly screened were invited for rescreening. Only one baby was identified with profound congenital hearing loss and was referred for bilateral cochlear implants at 10 months and implanted at 14 months. This incident highlights the importance of vigilance in monitoring any screening programme, regular quality control checks and adequate education of the screeners. The revised screening regime eliminates the possibility for this screener error to recur.18

Delay in referral and implantation due to lack of attendance is preventable. Children ‘lost to follow-up’ have been recognised as problematic and an area for improvement in the UNBHS Programme in New Zealand, and also in UNBHS programmes in other countries.2,20 The significant reduction in age at implantation in the Netherlands, compared to the more modest reduction in Germany, reflects the national programmes of those countries; the first providing targeted follow-up and automatic referral, and the second relying on parent self-referral and follow-up following a failed screening result.10 The first few months of an infant’s life are a challenge for parents however this is, as has been explained, a critical period for auditory and language development. Enhanced education of parents, audiologists and the primary care physicians will emphasise the importance of timely treatment and potential consequences of treatment delay. The introduction of telephone and SMS reminders have been shown to decrease the rate of non-attendance across many disciplines internationally21 and should be considered in areas where this is a problem.

A diagnosis of Auditory Neuropathy Spectrum Disorder (ANSD) contributed to later referral for three children. Two of these children also presented with additional disabilities, one received poor follow-up from Audiology. Management of ANSD can be a challenge. Outer hair cell function is preserved and therefore these children are not always identified by otoacoustic emissions, so detection and diagnosis may be delayed. Sound may be detected, but the signal may be distorted and lack clarity, causing delays in speech and language development. It is only recently that cochlear implants have been confirmed as a successful treatment in cases of ANSD.22,23 Children with a diagnosis of ANSD, without associated cognitive or developmental disorders, can acquire speech and language outcomes comparable to other children who receive a cochlear implant.24 Some children with ANSD benefit from hearing aids. However, ANSD patients who do not benefit from conventional amplification do well when implanted at a young age.25 Careful monitoring of the functional benefit derived from hearing aids should facilitate prompt referral for a cochlear implant in cases of ANSD.

Prelingually deaf children are an extremely heterogeneous group, and include children with congenital malformations or co-morbidities that may delay suitability for general anaesthesia. Additional disabilities are no longer a contraindication for cochlear implantation, although outcomes may be significantly different than for children with no additional disabilities.26,27 Speech and language outcomes may be poorer than for children without additional disabilities, however benefits have been noted in general communication, and it is appropriate to provide children with additional disabilities with the same opportunity to access audibility as any other child with a hearing impairment.28 It is critical that referral for cochlear implantation is not delayed until other issues are ‘sorted out’, as this leads to further delays to implantation and language development.
Finally, age of implantation is only one of the variables known to affect outcomes post implant, together with duration of deafness, additional disabilities, number of active electrodes, neural survival and mode of communication.29,30 However, the age of implantation is one variable that we can aim to improve.

Conclusions

- The introduction of newborn hearing screening has significantly reduced the age at which children are referred for cochlear implantation.
- The Southern Cochlear Implant Programme can continue to improve its practice, as international trends move towards implantation even earlier than 12 months.
- Children with moderate to severe loss must be regularly monitored so as not to delay referral, should the hearing loss progress.
- Parents of children with hearing loss must be supported and educated so they understand the importance of early intervention. Efforts must be made to reduce non-attendance of appointments.
- Screening programmes and the screeners must be audited and regularly monitored. Adequate training must be given to screeners and they must be educated as to the importance of early intervention.
- Children with other co-morbidities should not have their referral delayed.

REFERENCES:

1. Yoshinago-Itano C. From screening to early identification and intervention: Discovering predictors to successful outcomes for children with significant hearing loss J Deaf Stud Deaf Educ 2003;8:11-30
6. Ruben RJ. A time frame of

Competing interests:
Philip Bird reports he is a trustee of the Southern Charitable Hearing Trust which administers the Southern Cochlear Implant Programme.

Author information:
Jill Mustard, Clinical Manager, Southern Cochlear Implant Programme, Christchurch; Megan Chinnery, Habilitationist, Southern Cochlear Implant Programme, Christchurch; Alice K Guidera, Otolaryngologist, Department of Otolaryngology, Head and Neck Surgery, Christchurch Hospital, Christchurch; Neil Heslop, General Manager, Southern Cochlear Implant Programme, Christchurch; Philip Bird, Otolaryngologist, Department of Otolaryngology, Head and Neck Surgery, Christchurch Hospital, Christchurch, and Southern Cochlear Implant Programme, Associate Professor, Department of Surgery, University of Otago, Christchurch.

Corresponding author:
Jill Mustard, Southern Cochlear Implant Programme, Milford Chambers, St George's Hospital, 249 Papanui Rd, Christchurch.

jill.mustard@scip.co.nz

URL:


27. Edwards L. Children with cochlear Implants and complex Needs:
A Review of Outcome Research with Psychological Practise Journal of Deaf Studies and Deaf Education 2007 12:3

