An audit of referrals to the Southern Cochlear Implant Paediatric Programme

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Abstract

Aim To audit the age at referral and time to assessment and implantation in children presenting to the Southern Cochlear Implant Paediatric Programme and identify any delay in implantation, particularly in individuals with pre-lingual sensorineural hearing loss.

Methods All paediatric referrals to the Southern Cochlear Implant Programme from March 2003–March 2008 were evaluated retrospectively. The Student t-test was used to compare median time intervals between those with and without risk factors for Sensorineural hearing loss.

Results Seventy five children were referred, 42 with pre-lingual deafness and 33 with post-lingual deafness. The median age of referral was 17 months with a range of 1 to 203 months. Thirty-five children with pre-lingual deafness were accepted as candidates and implanted, 6 were declined as they were too old to receive benefit from cochlear implantation. Of these 6 children who were declined, 4 had not been adequately diagnosed despite having risk factors for sensorineural hearing loss. There was no significant difference in the age of referral in pre-lingually deafened children between those with risk factors and those without risk factors.

Conclusion The age at referral of pre-lingually deafened children to the Southern Cochlear Implant Programme is unacceptably high, particularly in those children who have known risk factors for sensorineural hearing loss.

Cochlear Implants (CI) are devices which replace the auditory function of the inner ear and directly electrically stimulate the cells of the spiral ganglion (which form the cochlear nerve within the cochlea). They are used in both adults and children with severe to profound sensorineural hearing loss (SNHL) where hearing aids provide insufficient amplification for the understanding of speech.

Children with SNHL may be categorised into having either prelingual or postlingual deafness. To optimise language outcomes it is very important to identify and manage prelingually deafened children early, preferably before the age of 6 months.1

For those children who require cochlear implantation there is clear evidence that implantation prior to age 2 leads to potentially better results in speech, language and reading skills.2 With the introduction of neonatal hearing screening in many parts of the world, and thus much earlier implantation, there is steadily accumulating evidence that implantation prior to age 1 year is associated with improved outcomes.3-8

At the other end of the spectrum, children over the age of 5 who have not received sufficient sound to develop speech and language are highly unlikely to ever do so because of reduced neural plasticity. In these instances a cochlear implant provides
awareness of environmental sound only, and unless the child has no hearing whatsoever does not provide any significant advantage over hearing aids.

New Zealand is in the process of introducing universal neonatal hearing screening. Prior to this New Zealand neonates only had their hearing tested if they had risk factors for SNHL or if there was parental concern. A risk factor registry can identify up to 66% of those with significant bilateral deafness.9

These risk factors include all of the factors predisposing to admission to a neonatal intensive care unit (such as prematurity, hypoxia, low birth weight), maternal infection, positive family history and a number of syndromes which are associated with hearing loss. The biggest single cause of deafness however is genetic, autosomal recessive, non-syndromic sensorineural hearing loss for which there is no “warning”. In New Zealand in 2005, 58% of children had no known risk factors for their hearing loss. It is likely that a significant proportion of these children had genetic, autosomal recessive non-syndromic sensorineural hearing loss.

The Southern Cochlear Implant Programme (SCIP) provides comprehensive CI services to the South Island and lower North Island. We are aware of significant delays in some referrals and hence have undertaken an audit to further categorise these. In addition, to try and improve our service to our patients, this study should serve as a baseline prior to the introduction of universal neonatal hearing screening in New Zealand.

Method

A retrospective chart analysis was made of the referrals to the Southern Cochlear Implant Programme (SCIP). The first 5 years of this programme, from March 2003 through to March 2008 were chosen. Data was collected in respect to the age of the child (expressed in months) and the following time points; diagnosis, referral to the program, assessment and then either implantation or observation. The children were divided into two categories, those with pre-lingual deafness and those with post-lingual deafness, based on individual chart review. Those with risk factors for sensorineural deafness were also identified.

Results

There were 75 children referred over this 5-year period, of which 53% percent were female and 48% male.

Following assessment 50 children (67%) were accepted as surgical candidates, 8 remain under review with progressive hearing loss, 5 were awaiting assessment and 12 have been declined a cochlear implant.

Thirty-three children were identified with post-lingual deafness (44%). Twelve were implanted, while 3 await surgery. Eight remain under review while 5 were awaiting assessment. Five children were declined a cochlear implant.

Forty-two were identified as having pre-lingual deafness (56%). The median age of referral was 17 months with a range of 1 to 203 months. With 6 children over the age of 5 years (5.5yrs, 5.9yrs, 8.5yrs, 8.9yrs, 10.9yrs, 17.4yrs), the average age was much higher, at 32 months. Thirty-five of these children underwent implantation. Seven children with pre-lingual deafness were declined an implant. One child moved away from the region and has had surgery elsewhere, while six children were declined surgery due to their age (Figure 1).
Figure 1. Flow chart of referrals to the Southern Cochlear Implant Paediatric Programme 2003–2008

The 35 children with pre-lingual deafness who received a cochlear implant were referred with a median age of 16 months (mean 18 months), assessed by 18 months (mean 21 months), approved by 20 months (mean 23 months) and implanted by 21 months (mean 24 months).

From the 75 referrals (both post-lingual and pre-lingual) there were 25 children with known risk factors for sensorineural deafness. Ten children were premature, 5 had a family history of deafness, 2 had Jervell and Lange-Nielsen syndrome and 2 had CHARGE syndrome. The remaining 6 children had a diagnosis of: Goldenhar syndrome, meningitis, Beckwith-Weidemann syndrome, Refsum’s syndrome, cytomegalovirus (CMV) infection and cerebral palsy. Despite having known risk factors only 13 of these children (52%) were screened for deafness.

Twenty children identified with pre-lingual deafness had known risk factors. Twenty of the 42 children (47.6%) identified with prelingual deafness had known risk factors. Eight (40%) were screened for deafness. Sixteen have been implanted. These children were referred (median age) by 14 months, assessed by 17 months, approved by 18 months and implanted by 19 months. Using the Student t-test there was no significant difference between the ages of referral (P=0.88), assessment (P=0.97), approval (P=0.86) and implantation (P=0.90) for those children with known risk factors compared to those without.

Twelve children have been declined an implant; 5 with post-lingual deafness and 7 with pre-lingual deafness. Of the 7 declined children with pre-lingual deafness 6 were declined surgery due to their advanced age, and 1 child moved out of the area. Four children with pre-lingual deafness and a known risk factor were denied surgery. The risk factors included prematurity (3) and CMV infection (1).

Discussion

This audit demonstrates a number of areas of concern relating to children with significant pre-lingual deafness. These children have insufficient hearing levels to access enough information to develop speech and language. As stated previously, it is highly desirable to implant these children at a young age, preferably by 12 months.
The mean age of referral for this group was 32 months, this figure being distorted somewhat by 6 children who were referred after the age of 5 years. All of these older children were declined cochlear implantation, not on audiologic grounds, but because they were too old to be able to utilise the technology to learn spoken language.

A major cause of late referrals to our Programme is the lack of universal neonatal hearing screening. This problem has been identified by the Ministry of Health and a Programme of Screening is currently being implemented. This should help significantly with early referrals, especially in those children with no risk factors for hearing loss. Of the children with prelingual deafness in our series, 22 of the 42 or 52.4% had no risk factors. This is comparable with the 58% of children with no risk factors in the New Zealand Deafness Notification Database for 2005.

The most concerning aspect of this audit is the lack of screening and late referral of children with known risk factors for SNHL. There was no difference in the referral ages between those children with risk factors and those without. A few children at risk had actually had their hearing tested with misinterpretation of results, only to be re-tested later following speech delay or parental concern. Failure to acquire language had been attributed to other causes in some children.

Many of these children with risk factors have chronic medical problems and profound deafness presents a huge extra burden to them and their family. It is particularly devastating for those children declined surgery due to being referred too late to benefit from cochlear implantation.

The late referrals of children with risk factors may be due to a number of factors. Firstly, lack of awareness may be an issue. Secondly poor resourcing and lack of experience in paediatric audiology is likely to be relevant. Testing young children electrophysiologically and behaviourally requires time, more than one tester, skill and experience. If there are insufficient resources to perform this vital testing, universal screening of hearing may not be the panacea we have hoped for.

There are minor “delays” associated with assessment of suitability for a cochlear implant. Some of the intervals between referral and assessment relate to waiting for consent from families and for relevant information from referral sources (principally Audiologists and Otolaryngologists). Some children were referred and then commenced on a 6–8 week trial of hearing aids locally, prior to assessment by the SCIP Team, which also added to the interval time between referral and assessment. Throughout the 5-year period, funding has been a huge issue for the combined Adult and Paediatric SCIP, but we have generally managed to implant children expeditiously.

It is important to stress that this group of children represent the most severe end of this spectrum of hearing loss. When mild and moderate deafness is included, the average age of detection in New Zealand is 33.9 months.

There will be a small number of children who have severe to profound SNHL who are not referred for consideration of cochlear implantation. Parents who are members of the Deaf (Signing) Community may not wish their own deaf children to undergo cochlear implantation for cultural reasons.
In summary, this audit of referral times to the Southern Cochlear Implant Programme demonstrates a lack of early diagnosis and referral in children who have known risk factors for significant sensorineural hearing loss. This factor plus lack of universal neonatal screening means that there are significant delays to cochlear implantation in children, which is likely to impact significantly upon results. Included amongst these delays are 6 children who were denied any benefits of cochlear implantation because they were referred after the age of 5 years, with no ability to utilise this technology.

Competing interests: None known.

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References: