Evidence-based practice for cochlear implant referrals for infants

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Summary:

This report provides an update on the importance of early implantation for spoken language development of children. The evidence calls for early referral of children with severe to profound hearing loss for cochlear implant candidacy evaluation in order to optimise the potential for spoken language development.

Background:

The implementation of Universal Newborn Hearing Screening has led to an increase of children identified with hearing loss soon after birth. For infants diagnosed with severe or profound hearing loss, a major habilitative consideration is referral for cochlear implant candidacy evaluation. It is generally agreed that early implantation leads to higher levels of spoken language or more rapid progress, but how early is “early”?

A sizable published literature on the effect of age of implantation on language development has appeared since 2000, with previous work showing the benefit of implantation before age 5 years (e.g. Kirk et al, 2002; Svirsky et al, 2004), before age 2 years (e.g. Manrique et al, 2004), and more recently before age 12 months (e.g. Tait et al, 2007; Dettman et al, 2007; Tomblin et al, 2005). Typically, results on small numbers of children were reported. For instance, Tomblin et al (2005) measured receptive and expressive language of 27 profoundly deaf children over 3 years, with only 1 child implanted under 12 months of age. The results showed that children who received an implant earlier are more likely to develop language at a rate commensurate with normal-hearing peers. Tait et al (2007) reported preverbal communication of 10 deaf children who received a cochlear implant before the age of 1 year showing that they developed preverbal communicative behaviours to an extent that was not significantly different from those of age-matched normal-hearing children. Dettman et al (2007) showed that 11 children who received an implant before 12 months of age developed language at a normal rate whereas children who received an implant between 12 and 24 months of age exhibited slower progress. The former group received hearing aid fitting at an age that was significantly earlier than that of the latter group. It is not clear whether the difference in rate of development was related to the differential age of hearing aid fitting for the two groups of children.

Child Outcomes study: findings to date

The Child Outcomes study is aimed to follow the development of 400 hearing-impaired children over a period of five years (for details: www.outcomes.nal.gov.au). Interim findings of 41 children with cochlear implants measured at 6 months after cochlear implantation are now available (Ching et al, accepted). The three-frequency average (3FA) hearing thresholds for all children were 90 dB HL or greater in the implanted ear.
On average, children who received a cochlear implant before 12 months of age developed auditory comprehension and expressive communication similar to their normal-hearing peers (standard scores between 85 and 115). On the other hand, children who received cochlear implants at a later age performed at 2 standard deviations (SD) below the normative mean of 100. The results revealed that age of implantation was significant ($p = 0.001$) after accounting for the effect of age of first hearing-aid fitting.

These results support the importance of early implantation. The evidence implies that when infants’ confirmed hearing loss exceeds 90 dB HL, implantation before 12 months of age would lead to rapid language gains. This is consistent with the report of Sharma and colleagues (2004) showing changes in neural response within central auditory pathways of two children shortly after cochlear implantation at ages 13 and 14 months and the changes appeared to be related to development of early communicative behaviours.

The Child Outcomes study has also found that early hearing aid fitting is associated with normal language development (Ching et al, 2008). Early use of hearing aids provides auditory stimulation that is vital for development of the auditory system and access to language.

**For babies with confirmed severe or profound hearing loss**

The cochlear implant should form a routine part of the discussion about devices for families of children who have a severe to profound hearing loss. In order to make an informed decision about their child’s communication development and audiological management, parents should be provided with factual information about the impact of hearing loss on the child’s development and the potential benefits of hearing aid and cochlear implant technology.

The evidence to date supports early fitting of hearing aids and early referral for cochlear implant candidacy evaluation.

**Guidelines:**

1. Fit hearing aids to all children with residual hearing as soon as possible after the family have agreed to proceed with the fitting.

2. For children with confirmed hearing level within the shaded area in the figure below (described here as $3FA \geq 80$ dB HL), encourage the family to consider cochlear implantation and refer for candidacy evaluation as soon as the family is ready.
Candidacy criteria: ‘moderate-to-profound’

- It is not necessary to wait for a hearing aid evaluation for these children, since a cochlear implant will almost always provide improved intelligibility over acoustic stimulation alone.
- There is no minimum age for referral.

3. For children with confirmed 3FA hearing levels between 60 and 80 dB HL, evaluate success with hearing aids by using functional performance measures such as the Parents’ Evaluation of Aural/oral function of Children (PEACH; Ching & Hill, 2007). Refer for cochlear implant candidacy evaluation if parent report or PEACH data suggests that auditory function is poor (see Appendix 1 and [http://www.outcomes.nal.gov.au/assessments-peach.html](http://www.outcomes.nal.gov.au/assessments-peach.html) for information about PEACH administration).

4. If a child has been diagnosed with a condition that is likely to lead to a progressive loss (e.g., Large Vestibular Aqueduct Syndrome) then it may be appropriate to mention implantation as an option for the future.

Notes:
- Some families may feel more comfortable about an early referral to the implant clinic if you reinforce that a referral places them under no obligation to proceed with an implant for their child; they can attend for information only.
- If a parent wishes to obtain more information about implantation for a child with a lesser degree of hearing loss, the family should also be assisted even though the child is unlikely to be a candidate. The process may be an important part of helping the parents to understand and accept their child’s hearing loss.
- If a child’s hearing loss is potentially suitable for a cochlear implant, then appropriate information should be provided to the family as part of the initial discussion process. Avoid making judgements that someone is “not ready” or won’t want to receive the information (e.g., Deaf families).
- Some families of children with severe to profound loss may choose to decline the offer of referral to a cochlear implant clinic for a range of cultural, medical or emotional reasons. Their choice should be respected. It may be appropriate to revisit the issue at a later stage.
Babies with Auditory Neuropathy/Auditory Dys-synchrony

Extra caution must be exercised for babies diagnosed with AN/AD when considering referral for cochlear implant candidacy for two reasons. Firstly, it may not be possible to establish hearing thresholds reliably and hearing loss may change in either direction over the first 12 months of life. Secondly, not all forms of AN/AD benefit from cochlear implantation, as the success of implantation is expected to be greater the closer the lesion is to the inner hair cells. However, if there is strong evidence to support the presence of a bilateral profound hearing loss in a child diagnosed with AN/AD, refer the family for information on cochlear implantation (see Rance, 2005; King et al, 2005 for guidelines on management).
Provide bimodal fitting to recipients of unilateral cochlear implants

There is now compelling evidence to support the provision of binaural hearing to children who receive a cochlear implant in one ear by fitting a hearing aid in the contralateral ear (Ching et al, 2005; see Ching et al 2007 for a review). The use of a cochlear implant and a hearing aid in opposite ears is commonly referred to as “bimodal fitting”. Once the cochlear implant map has stabilised, the hearing aid should be fine tuned for optimal performance with the cochlear implant (see http://www.outcomes.nal.gov.au/Resources-clinical%20tools.html)

Guidelines:
1. Assess hearing of non-implanted ear
2. Fit a hearing aid according to standard procedures
3. Liaise with the child’s cochlear implant centre to ascertain that a stable map has been established
4. Adjust loudness and/or frequency response of hearing aid to achieve optimal fitting
5. Consider FM fitting

Bilateral cochlear implantation

Bilateral cochlear implantation is increasingly being accepted as the method of choice for managing infants and children with bilateral profound hearing loss (Offeciers E et al, 2005).

There is some evidence on cortical reorganisation showing that reduced plasticity is linked to absence of auditory stimulation for more than 7 years, and that central auditory development is affected by whether stimulation is provided unilaterally or bilaterally (Sharma et al, 2007). A related aspect is that of cross-modal reorganisation of higher order cortex by other sensory modalities as a result of functional decoupling of the primary auditory cortex. These reinforce the importance of providing bilateral stimulation as early as possible.

For children who receive a cochlear implant in one ear and who do not have residual hearing in the non-implanted ear, bilateral cochlear implantation provides bilateral auditory stimulation. For children with residual hearing in the non-implanted ear, current evidence does not support the superiority of two implants over the combined use of a hearing aid and a cochlear implant for providing binaural hearing (see Ching et al, 2007 for a review).

Consult your local cochlear implant clinics about bilateral implantation protocols and whether they undertake sequential or simultaneous implantation of the two devices.

Source: The PDF version of this document may be downloaded from the NAL website (www.nal.gov.au) or the Child Outcomes Study website (http://www.outcomes.nal.gov.au/Resources-Papers.html).
Selected References


Appendix 1.

PEACH scores as a function of age (Ching & Hill, 2007). The solid curve shows the relationship between PEACH scores and age in months and the broken lines denote ± 2 standard deviations, all for children with normal hearing. The dashed curve represents an estimated minimum acceptable performance for children with 3FA > 60 dB HL. A score below this line would be an important consideration for referring a child for cochlear implant candidacy evaluation.
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