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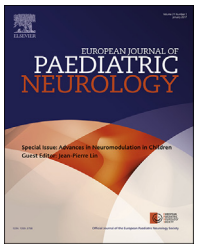

Original article

Paediatric cochlear implantation factors that affect outcomes

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ABSTRACT

Cochlear implantation is an established surgical intervention for individuals with bilateral severe to profound sensorineural hearing loss. The aim of the interevention is to provide the individual with a sensation of sound which they can learn to interpret with meaning. Outcomes vary considerably and the factors that impact on outcomes will be discussed.

Cochlear implantation is an established surgical intervention. A cochlear implant system consists of two components, an internal receiver stimulator surgically implanted and an external speech processor which powers the internal implant (see Fig. 1).

"The aim of cochlear implantation is to improve the hearing and quality of life of those with permanent functional severe to profound deafness who do not gain adequate benefit from optimally fitted hearing aids" (BCIG Quality Standards 2016).[19] The cochlear implant has the potential to give an individual access to the frequencies important to access spoken language (see Fig. 2).

To assess and support individuals to gain optimal benefit from the technology the surgery is embedded within a strong multidisciplinary team who, in the UK, adhere to the British Cochlear Implant Group Quality Standards ('Cochlear Implant Services for Children and Adults Quality Standards', April 2016).

In 2009 the National Institute for Health and Care Excellence carried out a review of the evidence base on cochlear implantation and concluded that bilateral simultaneous cochlear implantation should be offered to children who have a significant hearing loss of 90 dB or greater at 2 and 4 kHz.[18] For this degree of hearing loss hearing aids are typically unable to provide enough gain and clarity to provide access to spoken language. Without cochlear implantation these children would typically need access to a formal sign language system to develop a way to understand and communicate with others.

Almost four hundred children a year are born in England with a permanent bilateral severe to profound sensorineural hearing loss. As 90% of these children are born into hearing families with no experience or history of hearing loss or sign language the diagnosis can have a significant impact on the family. As an adult it can be a real challenge to learn and be able to use sign language fluently. The parents' ability to communicate effectively with their child through sign may then be compromised, as is the child's ability to learn sign language fluently from non-native sign language users. For deaf children of deaf parents, whose first language is sign language, they are often much more competent at developing fluent Sign Language themselves at an age appropriate rate. It is often the desire of hearing families, despite many additionally embracing sign language, for their deaf child to also have the potential to access spoken language and experience the world in a similar way to their own life experiences.

The comprehensive evidence base available clearly indicates a range of factors that impact on a child's ability to

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access the hearing world, develop receptive and expressive spoken language and intelligible speech.

Over the years advances with the implant technology, the speech coding strategies and the use of front end hearing aid processing techniques, made possible by cochlear implant companies joining forces with hearing aid manufactures, has contributed to significant improvements in outcomes.

Also of significant importance is age at implantation, if a child is congenitally profoundly deaf. The human brain's sensitive period for spoken language acquisition combined with the possibility of cross modal cortical reorganisation when areas are not stimulated by a certain sense (in this case hearing) is well documented in the literature[10] and also well explained by other articles in this specialist edition.

Because of this strong body of evidence a Newborn Hearing Screening Program was rolled out across the UK in 2008. This provides the best opportunity for early amplification and early implantation to stimulate the hearing pathway and auditory cortex during the brains sensitive period for language acquisition and to minimise the risk of cross modal reorganisation.

For profoundly deaf children to have the potential to develop spoken language and intelligible speech in line with their hearing peer group the optimal age of cochlear implant device activation is under or around 12 months of age. This not only allows the sensations delivered by the cochlear implants to support the organization of the neural pathways but also allows the child to integrate hearing to support the development of other age appropriate skills, for example the development of shared attention. Tait[15,16] compared the shared attention skills of normally hearing children and deaf children implanted before their first birthday and indicated that at 1 year post implant there was no significant difference between the two groups. Shared attention is an important foundation skill for spoken language.

Children implanted under 12 months are more likely to fulfill their full potential and reduce or eliminate the need for them to 'catch up' or learn spoken language at faster rates than normal, to achieve age appropriate norms. A study by Dettman[6] concluded children implanted under one achieved rates of receptive and expressive language growth at a rate comparable to normal hearing peers and significantly better

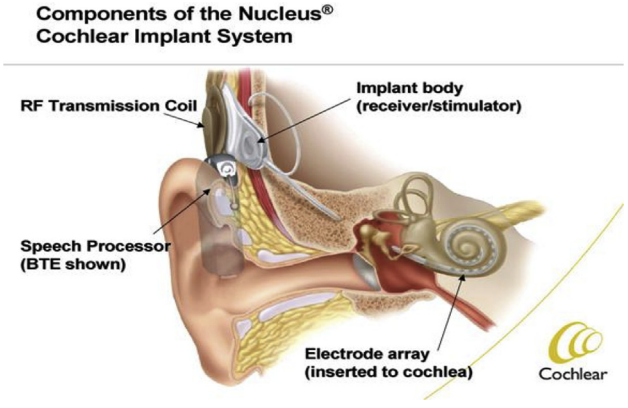

Fig. 1 – An illustration of a cochlear implant in situ.

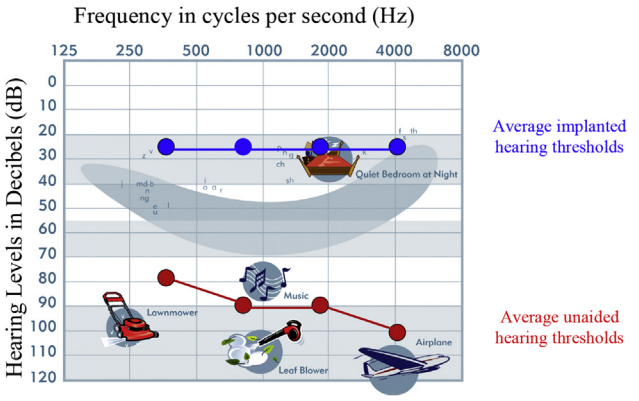

Fig. 2 – An graph indicating typical unaided and aided hearing levels with a cochlear implant.

than those children implanted between 12 and 24 months. Nott[12] found children implanted under one were closer to hearing peers in the time they acquired their 1st and 100th word. Nicholas and Geers[11] used a test standardised on hearing children and found that those implanted between 12 and 16 months were more likely to have age appropriate spoken language by four and a half than those implanted between 17 and 36 months.

For speech intelligibility the outcomes of early implantation are also very positive. Babbling, which is consonant vowel sequences produced in one breath unit combined with continuous phonation, represents the point at which infants produce mature phonetic syllables that are the building blocks of words and intelligible speech. Schauwers et al.[13] reported that onset of babbling in younger implanted infants was comparable to that of normal hearing peers. This is important as phonetic complexity of babbling correlates positively with speech and language outcomes at 4 years old.[17]

Early implantation also allows parents to do what comes naturally and interact with their child at a more age appropriate language level and with age appropriate activities rather than for example having an older child with whom the parents are being asked to work on awareness of symbolic animal and transport sounds which seems counter intuitive to what feels natural to do with an older child.

It is important however to be aware that longitudinal cochlear implant outcomes are still to be reported on. There are longitudinal population studies in Australia (the LOCHI study) and in America (the NAL study). As language becomes increasingly complex and social communication skills develop into teenage years and beyond it will be important to keep monitoring the outcomes of these very young implanted children to see if they continue to develop skills in line with their hearing peer group. The indications from the early years outcomes to date are positive however more recent publications by Convertino et al.[2] indicate that as children move through secondary school the advantage of early implantation seen at primary school does not always continue through secondary school. Aspects of word and world knowledge that requires an ability to 'overhear' language by picking it up incidentally is not always accessible to cochlear implant users due to the challenges of hearing speech in background noise.