The gift of hearing:

Advances in Paediatric Cochlear Implantation

Dr Annette Ang
Consultant, Department of Otolaryngology, KK Women's and Children's Hospital
Associate Program Director, SingHealth Otolaryngology Residency Program

Permanent deafness in early childhood is often related to the loss of function of the cochlea - the inner ear.

The incidence of reported loss of cochlear function ranges from 1.2 to 1.7 per 1,000 live births. Of these children, 20 to 30 percent experience profound hearing loss. The prevalence increases up to age six, due to late-onset deafness which may be the result of meningitis, delayed onset of genetic hearing loss, late diagnosis or exposure to ototoxic agents.

Early diagnosis and intervention for congenitally deaf children is important for their cortical development, as auditory input impacts the development of the immature brain. Proper complex auditory function and speech perception cannot be well established when hearing is restored late in life to congenitally deaf people.

Diagnosis

In April 2002, KK Women's and Children's Hospital (KKH) became the first hospital in Singapore to implement a Universal Newborn Hearing Screening Programme (UNHS) using Automated Auditory Brainstem Response technology to identify hearing loss early in life and facilitate early intervention. Achieving diagnosis by the age of three months and successful intervention, such as hearing aids, by six months has been shown to help children with deafness perform at the same academic level as their hearing peers.

Trends for Paediatric Cochlear Implantation

Since its establishment in 2005, the KKH Cochlear Implant Programme has brought about a sea change in trends for paediatric cochlear implantation, resulting in greatly improved results for children in Singapore and Southeast Asia.

1. Cochlear implantation at a younger age

Early cochlear implantation in children has been shown to result in superior speech and language outcomes. Children who receive a cochlear implant between the ages of 12 and 36 months are found to outperform those who receive a cochlear implant between the ages of 37 and 60 months.

In the eight years since the KKH Cochlear Implant Programme was set up, the mean age for cochlear implantation has dropped from nine years to three years.

2. Increased popularity of bilateral cochlear implantation

Binaural hearing, or hearing from both ears, is proven to have greater benefit for speech perception in a noisy environment, in addition to better sound localisation. Hence KKH routinely educates parents on the option of bilateral cochlear implants for children, and its superior results in comparison to unilateral implantation.

For children in whom a second cochlear implant is not possible, the use of a hearing aid in the other ear is strongly advocated. This allows the child to have bimodal hearing, which despite the asymmetry in hearing, has shown greater benefit in speech and language development.

3. Hearing-preservation cochlear implants

Early laboratory studies have shown that the inner and outer hair cells of the cochlea can survive after implantation with an electrode. This allows children with high frequency hearing loss and normal residual low frequency hearing to be implanted with a cochlear implant that preserves their native low frequency hearing.

The hearing-preserving cochlear implant allows for better word recognition in the presence of background noise and better appreciation of music. To date, an estimated 85 percent of patients in Australia and North America have achieved preservation of residual hearing.

Conclusion

Cochlear implantation is now an established strategy for the management of permanent hearing loss in childhood. With the aid of a cochlear implant, deaf children are now able to enter mainstream schools and achieve equivalent academic standards to their hearing peers.

The field of cochlear implantation and otolaryngology is expanding at a rapid pace as new discoveries are translated into clinical applications. At KKH, studies are underway to improve management and clinical outcomes for conditions such as laryngomalacia, mastoiditis and abscesses in the head and neck region.

Dr Annette Ang's main areas of interest are paediatric otolaryngology and the management of complex paediatric airway diseases. Dr Ang graduated in Singapore, and pursued a clinical fellowship at the Weill Medical College of Cornell University, New York City.

References:

1. A Kral, G O'Donoghe. Profound deafness in childhood. N Eng J Med 2010: 363:15
2. Niparko JK, Tobey EA, Thal DJ et. Al. Spoken language development in children following cochlear implantation. JAMA 2010; 303: 1498 -1506
3.Houston DM, Miyamoto RT. Effects of early auditory experience on word learning and speech perception in deaf children with cochlear implants: implications for sensitive periods of language development. Otol Neurotol. 2010: 31: 1248-1253
4.Nikolopoulos TP, O'Donoghue GM, Archbold S. Age at implantation; its importance in pediatric cochlear implantation. Laryngoscope 1999: 109: 595-599
5.P Kileny, T Zwolan, c Ashbaugh. The influence of age at implantation on performance with a cochlear implant in children. Otol Neurotol. 2001. 22 (1): 42-46
6.Ching TY et. Al. Should children who use cochlear implants wear hearing aids in the opposite ear? Ear Hear 2001; 22: 365-380
7. Shepherd RK, Clark GM, Xu SA et. al. Cochlear pathology following reimplantation of a multichannel scala tympani electrode array in the macaque. Am J Otol. 1995 mar; 16 (2): 186-99
8.Hodges AV, Schloffman J, Balkany T. Conservation of residual hearing with cochlear implantation. Am J Otol. 1997 Mar: 18 (2): 179-83
9.Von Ilberg V, Kiefer J, Tillein J et. Al. Electric-acoustic stimulation of the auditory system. New technology for severe hearing loss. ORL J Otorhinolaryngol relat. Spec. 1999 Nov-Dec; 61 (6): 334-40
10. Gstoettner W, Kiefer J, Baumgartner WD et. al. Hearing preservation in cochlear implantation for electric acoustic stimulation. Acta Otolaryngol. 2004 May; 124 (4): 348-352f