COCHLEAR IMPLANTS FOR SEVERE TO PROFOUND DEAFNESS IN CHILDREN AND ADULTS

The comments in this submission from Auditory Verbal UK apply only to children with severe and profound deafness and their families.

What is it like to have the condition?

The most obvious symptom usually associated with severe or profound deafness is severely delayed communication using spoken language. A reduced competence in spoken language has consequences for literacy and therefore for access to information and education, and ultimately upon employability. A severe or profound hearing loss has an impact upon an individual’s education, social participation and therefore, on his or her economic and social independence.

Having one or more children with severe or profound hearing loss causes considerable stress on family life with a higher than usual level of need for care and supervision. For a child with severe or profound hearing loss, his or her social interaction is compromised both within and outside the family unit: Siblings have to learn to be tolerant of their brother(s) or sister(s) higher level of need and may, in turn, demonstrate behaviours requiring greater parent support and intervention. Peer-to-peer interaction needs greater adult guidance when one or more of the children has severely delayed communication skills. A child with severe or profound hearing loss will need greater support to access age appropriate educational and social activities.

What are the outcomes that matter most to patients?

The outcomes that matter most to hearing parents of severely and profoundly hearing impaired children are those associated with spoken language and communication skills, and the consequent benefit to social interaction and education. While the technology of the implants is of extreme importance, advances in the technology most go hand in hand with the development of excellent habilitation programmes.

Parents are most interested in evidence-based outcomes: AV UK has recently submitted a paper to a peer-reviewed journal detailing language outcomes in 37 children. Nearly 50% of the children on the study started their AV habilitation programme with hearing aids but went on to receive cochlear implants. Having embarked on a programme of Auditory Verbal therapy, the rate of language development in this sub-group of children more than doubled prior to implantation. The rate of language development increased again by a factor of more than 2 after cochlear implantation, giving an overall five-fold difference in the rate of language development. The results from our study illustrate that a child can make outstanding progress in spoken language development once they have appropriate access to the sounds of language in combination with excellent habilitation.
Brief details of one case study are given below: In the illustration the ‘Predicted Total Language Development Score’ is a measure derived from the ratio of the child’s equivalent total language age to the child’s chronological age at entry to the habilitation programme. The predicted total language development score is calculated by multiplying this ratio by the child’s chronological age.

**Case study:**

This child joined the AV habilitation programme shortly after her first birthday. She was diagnosed with a profound hearing loss at 9 months of age and used hearing aids until she was fitted with a cochlear implant at 23 months of age. There was a steady but low rate of progress for language acquisition prior to implantation. After receiving a cochlear implant, this child made an accelerated rate of progress of spoken language acquisition. It should be stressed that for severely and profoundly hearing impaired children yet to acquire spoken language, hearing technologies alone will not address their language and communication deficit. Severely and profoundly hearing impaired children need excellent habilitation for each child to achieve his or her full potential.

What difference does the technology make?

The group of children for whom an implant is available does not have an alternative treatment. Prior to the advent of cochlear implantation, a much smaller proportion of severely hearing impaired children and no profoundly hearing impaired children would have developed spoken language through listening alone.
The most obvious positive impact of cochlear implantation for children is that it gives the children the potential to access sounds across the frequency range of speech and thereby the potential to develop intelligible spoken language. The child with severe or profound hearing impairment becomes dependant upon the technology and there is a lifelong need to manage the technology. This negative aspect of cochlear implantation is far outweighed by the benefits of implantation.

Cochlear implants are appropriate for treating severe to profound sensory hearing loss. There are medical contra-indications for implantation such as auditory nerve aplasia and/or cochlear aplasia. Implantation does require invasive surgery and there is associated risk of infection in areas close to the brain as well as a small risk of facial nerve damage. Families of children who opt for cochlear implantation acknowledge that at some point there is likely to be a further need for surgery due to mechanical breakdown, infection or the desire to upgrade to new technologies. The child with a cochlear implant needs to be in contact for life with a clinical facility that can support, troubleshoot and manage the technology. Families and schools have to take on the responsibility of managing the cochlear implant, speech processor, radio aid, sound field systems etc.. At an appropriate age, the child needs to be responsible for maintaining the batteries, leads and ancillary equipment of their speech processor and implant for life.

The groups of children who do well with implants are those children who have either begun to develop auditory brain capacity through conventional amplification or who, by virtue of their age, have the potential to develop it. Specifically, young children with profound hearing impairment and children who have some prior benefit from hearing aids do well with cochlear implants. The group that benefits less are long term profoundly hearing impaired children with no auditory/oral communication.

**Using the technology**

In comparison to other technologies such as kidney dialysis, haemodialysis or chemotherapy, cochlear implantation fits in to family life with minimal disruption. However, for a family new to cochlear implantation, familiarisation will take considerable time and there is a need for high parental/adult supervision and awareness. The cochlear implant and speech processor are initially fitted and monitored at hospital but otherwise the technology is managed at the child’s home. From the user’s viewpoint, the level of sophistication of the technology is on a par with other electronic equipment used in everyday life.

There are financial costs associated with hospital appointments such as travelling costs and loss of earnings but these are generally equivalent to the costs incurred by families of children with lesser degrees of hearing loss for whom conventional hearing aids are the appropriate management.

1  No time for us. Relationships between Parents who have a Disabled Child  
   Published by Contact a Family  December 2003

2  An evaluation of Auditory Verbal therapy using the rate of early language development as an outcome measure.  
   S Hogan, C Labate, J Stokes, E Tyszkiewicz and A Woolgar  Submitted 2006