sense
for deafblind people

Factsheet 8

Cochlear implants and the child with CHARGE syndrome

ALEXANDRA WHEELER, MSC Audiology (Paediatric Habilitation), **CERT ED** Hearing Impairment, **BA HONS** Specialist Teacher of the Deaf/Educational Audiologist, Paediatric Cochlear Implant Programme, University College Hospitals NHS Foundation Trust

Cochlear implantation (CI) is only indicated for children who have a hearing loss of greater than 90dBHL at the high frequencies of speech – which are 2 and 4 KHz. (NICE Guidelines). Their hearing loss may be less severe at lower frequencies, but this need not affect the decision to implant. However, the decision to implant children with CHARGE syndrome has to be based on very careful assessment of the evidence available.

Cochlear implants

Cochlear implantation gives a sensation of hearing by stimulating nerve endings in the cochlea directly, which in turn send auditory information to the brain along the auditory nerve. Stimulation is carried by electrodes which are surgically implanted into the cochlea and respond to information transmitted by a speech processor worn on the head. Implantation may be unilateral but is usually bilateral unless there are contra-indications (see below).

The CHARGE ear

Children with CHARGE syndrome who have been diagnosed with a severe to profound hearing loss are likely to have malformed cochleae (cochlear dysplasia) and may have thin ('hypoplastic') or absent auditory (cochlear) nerves (Collins and Buchman, 2002). A malformed cochlea may have only a single cavity, unlike the usual snail shape with several turns (Bamiou *et al.* 2001). In this case, a full insertion of the electrode array may not be possible.

Even if the cochleae are normal, if the auditory nerve is absent then cochlear implantation will not be possible as there is no means of transmitting the auditory information from the cochlea to the brain. However there may be a difference between the two ears, in which case unilateral (rather than bilateral) cochlear implantation might be possible (Bamiou *et al.* 2001). Although Auditory Brainstem Implantation (ABI) is possible in cases of absent auditory nerves, it is rarely available in the UK at present and long term outcomes have yet to be determined.

Pre-assessment

It is most important that children who are undergoing CI assessment should have Computed Tomography (CT) scanning and where possible Magnetic Resonance Imaging (MRI) at an early stage to assess the condition of the inner ears and nerve pathways (Bamiou *et al.* 2001; Holcomb *et al.* 2012).

Auditory Brainstem Response (ABR) testing will be carried out to provide objective evidence of auditory responses. Where there are doubts about the presence of an auditory nerve, electrical ABR (eABR) may be used to provide additional information and may indicate that a child is a candidate for cochlear implantation, even when it has been difficult to identify a nerve on the MRI scan (Warren *et al.* 2010).

The information that cochlear implantation is not possible may be devastating news for parents. They should be made aware of this possibility at the start of the assessment – particularly in cases where no hearing aid benefit has been demonstrated and the child presents with delayed motor milestones (Bamiou, 2001).

Follow up after implantation

Follow-up may also be more intensive and time-consuming. In children with cranio-facial abnormalities, the close proximity and atypical course of the facial and auditory nerves may result in non auditory stimulation (where the muscles of the face respond) as the cochlear implant is stimulated (Bajaj *et al.* 2012; MacArdle *et al.* 2002) and requires careful mapping by an experienced audiologist.

There is also a risk that the facial nerve could be damaged during surgery, perhaps permanently, and the surgeon should discuss this with the family (Bajaj *et al.* 2012). Where only partial insertion of the electrode array is possible (in cases of malformed cochleae), the audiologist will re-map the available electrodes to provide the best possible response.

Outcomes

Where a decision is made to offer cochlear implantation to a child with malformed cochleae and/or thin nerves, parents should be counselled that the outcomes will be more limited than might otherwise be expected. This is due to the reduced level of auditory information reaching the brain due to the much smaller number of nerve impulses.

Research evidence regarding outcomes from cochlear implantation is based on small numbers. However, a range of papers demonstrate that whilst results vary, children often develop auditory awareness, including awareness of speech, but not to the level of children with normal inner ears. This applies to children who have nerve deficiency, regardless of a diagnosis of CHARGE syndrome (Arndt *et al.* 2009; Kutz *et al.* 2011; MacArdle *et al.* 2002; Lanson *et al.* 2007; Southwell *et al.* 2010).

Where a child has additional difficulties as well as being deaf, this will also have an impact on outcomes. The more complex the child, the more likely it is that outcomes will be affected (Birman *et al.* 2012; Edwards, 2007) but parents have reported improved connectedness with the world, greater attentiveness and willingness to communicate (Berrettini *et al.* 2008).

Created: November 2013
Review due: November 2015
www.sense.org.uk

REFERENCES

Arndt, S., Laszig, R., Beck, R., Schild, C., Maier, W., Birkenhager, R., Kroeger, S., Wesarg, T., Aschendorf, A. (2009) Spectrum of Hearing Disorders and Their Management in Children with CHARGE Syndrome. *Otology and Neurotology*. 31, pp. 67–73.

Bajaj, Y., Gibbins, N., Fawkes, K., Hartely, B., Jephson, C., Jonas, N., Albert, D., Bailey, M., Wyatt, M., Cochrane, L. (2012) Surgical aspects of cochlear implantation in syndromic children. *Cochlear Implants International*. 13(3), pp. 163–7.

Bamiou, D.E., Worth, S., Phelps, P., Sirimana, T., Rajput, K. (2001) Eight Nerve Aplasia and Hypoplasia in Cochlear Implant Candidates: The Clinical Perspective. *Otology and Neurotology*. 22, pp. 492–496.

Berrettini, S., Forli, F., Genovese, E., Santarelli, R., Arslan, E., Chilosi, A., Cipriani, P. (2008) Cochlear implantation in deaf children with associated disabilities: Challenges and outcomes. *International Journal of Audiology*. 47, pp. 199–208.

Edwards, L.C. (2007) Children with Cochlear Implants and Complex Needs: Review of Outcome Research and Psychological Practice. *Journal of Deaf Studies and Deaf Education*. 12(7), pp. 258–269.

Kutz, J.W., Lee, K.H., Isaacson, B., Booth, T.N., Sweeney, M.H., Roland, P.S. (2011) Cochlear Implantation in Children with Cochlear Nerve Absence or Deficiency. *Otology and Neurotology*. 32, pp. 956–961.

Lanson, B.G., Green, J.E., Roland, T.J., Lalwanin, A.K., Waltzman, S. (2007) Cochlear Implantation in Children with CHARGE syndrome: Therapeutic Decisions and Outcomes. *The Laryngoscope*. 117, pp. 1260–1266.

MacArdle, B., Bailey, C., Phelps, P.D., Bradley, J., Brown, T., Wheeler, A. (2002) Cochlear implants in children with craniofacial syndromes: assessment and outcomes. *International Journal of Audiology*. 42(6), pp. 347–56.

Warren, F.M., Wiggins, R.H., Pitt, C., Harnsberger, R., Shelton, C. (2010) Apparent Cochlear Nerve Aplasia: To Implant or Not to Implant? *Otology and Neurotology*. 31, pp. 1088–1094.