MANCHESTER
AUDITORY IMPLANT CENTRE

ANNUAL REPORT 2010-2011
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Cover illustration: - topography of the cortical auditory evoked potential N1 from stimulation with a cochlear implant high (HF), mid (MF) and low (LF) frequency electrode.

From:- Cortical potentials evoked by electrical stimulation of the central auditory pathway In adults with an auditory brainstem implant. O’Driscoll M, Ramsden RT, El-Deredy W. in press.
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OVERVIEW AND SUMMARY

The Auditory Implant Centre in Manchester was established in 1988 and was the first centre in the UK to implant multi-channel devices. Manchester has since grown to be a world renowned centre of excellence in the field of cochlear implantation. The Manchester Auditory Implant Centre comprises three teams: Adult, Adolescent and Paediatric. The Manchester Programme was one of the first in the UK to recognise the need for a service specifically aimed at the adolescent population and the Adolescent Programme was established in 1997.

The Adult Programme includes a number of patients (adult, paediatric and adolescent) who have received Auditory Brainstem Implants (ABI). The ABI is suitable for those patients for whom a cochlear implant would be inappropriate, due to compromised auditory nerve function most commonly occurring as a result of vestibular schwannoma, or following the surgical removal of a tumour in cases of Neurofibromatosis Type II (NFII).

By the end of March 2011, 1199 patients have received an implant on the Manchester programme, the youngest of whom was 6 months old and the oldest, 91 years. The numbers of patients receiving implants on each of the four programmes, between 1988 and March 2011, are as follows:

- Adult Programme (established 1988) = 572
- Adolescent Programme (established 1997) = 92
- Paediatric Programme (established 1991) = 598
- ABI Programme (adult, adolescent and paediatric) = 56
- TOTAL: 1262 cochlear implants and 56 auditory brainstem implants

Clinical Activity 1988-2011

Since 1988 the annual activity on each of the three programmes has generally shown a steady increase (Figures 1-4). This increase in referral rates has resulted from a number of factors. There is now an increased awareness and wider acceptance, on the part of patients, parents and referrers, that cochlear implants are a safe and effective treatment; due in part to publication of the NICE guidelines. Well-publicised outcomes, in terms of speech perception and spoken language development, clearly indicate significant benefit for the majority of cochlear implant recipients. There have also been changes in referral criteria such that a cochlear implant is now considered to be an option for patients with useful residual hearing.

Patients are referred to the centre from a wide geographical area, covering three main strategic health authorities and over 50 Primary Health Care Trusts in the North West of England (Table 1). A small number of referrals are received from other areas and abroad.
Figures 1-4 – Annual numbers of adult, adolescent and paediatric patients implanted with cochlear and auditory brainstem implants

Figure 1 – Annual numbers of adult cochlear implant patients

Figure 2 – Annual numbers of adolescent cochlear implant patients

Figure 3 – Annual numbers of paediatric cochlear implant patients

Figure 4 – Annual numbers of auditory brainstem implants
ADULT PROGRAMME

The Adult Programme in Manchester was established in 1988, using funding obtained from the HEAR (Help Ear & Allied Research) Charity until government resources became available in the mid 1990s. By the end of March 2011, 572 adults have received cochlear implants on the programme.

Patients with good residual low frequency hearing may be suitable for an Electroacoustic stimulation (EAS) device which combines both electrical stimulation through a cochlear implant and acoustic hearing with a hearing aid. Currently, patients who fit this criterion are implanted with the MED-EL Sonata Flex cochlear implant and use the DUET processor.

Criteria for referral to the adult programme for cochlear implants (see Appendix 1):

- Bilateral, severe/profound sensorineural hearing loss (≥90 dBHL at 2 and 4kHz)
- Post lingual hearing loss
- Good oral communication skills
- Limited or no benefit from hearing aids
- There is no maximum age for referral and patients with additional needs are not excluded

Figure 5 – Audiometric referral criteria for cochlear and EAS implants (courtesy of MED-EL www.medel.com)
Clinical activity between April 2010 and March 2011

Number of patients implanted = 52

Devices used with adult patients:
- Cochlear CI512 = 26
- Cochlear SRA = 2
- Cochlear Split Array = 1
- MED-EL Sonata Flex Soft = 11
- MED-EL Sonata Flex EAS = 20
- AB HiRes 90K = 1

Average age of cochlear implant patients = 57 yrs (Range= 25-82 yrs)

Figures 6 and 7 show distribution of age and aetiology.

**Figure 6 – Age distribution of adults receiving cochlear implants in 2010-2011**

**Figure 7 – Aetiology of deafness for adults receiving cochlear implants in 2010-2011**
Post-implant support

Patients typically attend six appointments with the adult team within their first month of implant use. During these appointments the speech processor is programmed and patients receive rehabilitation through an individualised auditory training programme. The training programme includes tactics for using the telephone, music therapy and advice about using assistive listening devices and FM systems with the implant. After the first month, patients are assessed again at 3, 9 and 21 months after switch-on and offered additional rehabilitation sessions as required. Following this, each patient is offered a follow up appointment on a bi-annual basis.

Outcomes

Speech perception and quality of life outcomes are measured for each patient at one week, three months, nine months and 21 months post-implant and then bi-annually. Figures 8-10 show the average scores obtained on BKB sentences, CUNY sentences and AB monosyllabic words for patients tested in 2010-2011 (up to 9 months post-implant). N.B. In the BKB sentence test, the patient’s ability to discriminate speech in noise is only evaluated from 3 months of implant use.

Figure 8 – CUNY sentence scores

![Figure 8 – CUNY sentence scores](image)

Figure 9 – BKB sentence scores (auditory alone)

![Figure 9 – BKB sentence scores (auditory alone)](image)
Figure 10 – Monosyllabic (AB) word scores (auditory alone)
ADOLESCENT PROGRAMME

In the summer of 1997 a programme was developed to support adolescent cochlear implant users. This population is too old to be supported within a paediatric environment, but still needs regular one-to-one intervention and intensive school support. By providing a specialised service for this age group, the Manchester Programme is able to offer the support necessary for these cochlear implant users to gain maximum benefit from their devices. In addition to those patients referred directly to the adolescent programme, all children who have received an implant on the paediatric programme automatically transfer to the adolescent programme during the summer holidays prior to starting secondary school. At the point of transition to the adolescent programme, local services are contacted and offered support and training to suit their needs. This may include in-service training to the new educational placement. The Adolescent Programme currently supports a total of 239 young people.

Criteria for referral to the adolescent programme

From the age of 10 years until they leave full-time education, children can be referred directly to the adolescent programme, normally by a hospital Consultant or Audiologist, or via a local GP. Referral criteria are selected according to evidence based practice and experience. They are set to ensure that patients who receive a cochlear implant are those most likely to obtain benefit from the device. None of the criteria outlined below exclude patients with additional physical disabilities or learning difficulties. For adolescent patients, it is important that the patients themselves are involved at every stage of the assessment process and receive all of the information they need to reach their own conclusions about the cochlear implant as a potential option for them.

The Adolescent Cochlear Implant Programme accepts referrals for assessment for patients over the age of 10 years who are still receiving support from an educational department or school and fit into one of the following groups:

- Patients who were born with hearing and have recently acquired a profound hearing loss. A child with a suspected hearing loss following meningitis should, of course, be referred for assessment immediately so that cochlear implant surgery can be fast tracked in the event of any ossification of the cochlea.
- Patients who have had some benefit from appropriately fitted hearing aids in the past, but whose hearing has deteriorated to the point where powerful hearing aids are no longer helpful. Patients who were born with a profound hearing loss, have received some benefit for spoken language learning through consistent use of appropriately fitted amplification, but who might receive significantly more auditory information from a cochlear implant. NB. These patients should show evidence that they have learned spoken language through listening, thus demonstrating the integrity of the auditory pathway.
The Adolescent programme encourages patients to continue wearing a contralateral hearing aid if they have previously been good hearing aid users and have residual hearing in the non-implanted ear. Other factors need to be considered as part of the evaluation, including the patient’s communicative environment at home, the need for support services to be in place post implant and the need for an educational setting where the curriculum is delivered through speech.

**Clinical activity between April 2010 and March 2011**

Number of patients implanted = 16

Devices used with adolescent patients:
- Cochlear CI512 = 11
- MED-EL Sonata / Sonata Flex = 5
- AB HiRes 90K = 2

Number of unilateral implants = 7
Number of simultaneous bilateral implants = 1
Number of sequential second implants = 8

Average age of adolescent cochlear implant patients = 11 yrs (Range= 10-18 years)

Figures 11 and 12 show distribution of age and aetiology.

**Figure 11 – Age distribution of adolescents implanted in 2010-2011**
Figure 7 – Aetiology of deafness for adults receiving cochlear implants in 2010-2011

Post implant support

Patients are offered an appointment for their initial programming 10-14 days following implant surgery. They typically attend four programming and habilitation appointments with the Adolescent team during their first two to three weeks of implant use. Habilitation sessions are then offered on a twice monthly basis for the first three months and on a monthly basis for the following three months, with further support as agreed with the patient. Programming appointments are offered at 3, 6, 9, 12, 18 and 24 months and annually thereafter. Speech perception tests are also carried out at these appointments. The habilitation programme is tailored to take into account the individual needs and preferences of each patient and specific activities are provided for practice and carryover at home. The aim is to encourage the individual to exploit the potential of the equipment, to advise on techniques for developing auditory processing skills and to enable the “mapping” of the new auditory signal onto known linguistic information. Speech and language assessments and diagnostic habilitation are used to identify areas that require remediation.

Adolescents are also offered the opportunity to meet and share experiences with other implant users on activity days and outings run by the programme. These have included tenpin bowling, a trip to the BBC Manchester news studios, an art workshop, a trip to an indoor climbing wall and a day of football skills/coaching with City in the Community linked with Manchester City Football Club.

Outcomes

Manchester Adolescent Outcome Scale (MAOS)
The Manchester Adolescent Outcome Scale (MAOS) was adapted from the Manchester Spoken Language Development Scale (MSLDS). It is a 10-point scale used to assess the overall functional benefit obtained from a cochlear implant by an adolescent patient.
PAEDIATRIC PROGRAMME

The Paediatric Programme in Manchester was established in October 1991 and to date, 598 children have received cochlear implants on the programme. Members of the team have experience in working with children of widely differing age, history of hearing loss, cultural and educational background. In addition, the team have developed particular expertise with children under the age of 24 months and children with additional disabilities.

All patients referred to the Paediatric Programme undergo a full audiological and speech and language assessment, involving a series of appointments with the team. All children with congenital hearing losses would normally be expected to be wearing appropriately-fitted high powered hearing aids throughout the assessment process. Diagnostic habilitation is used to assess the current benefit a child receives from hearing aids as well as to identify additional factors which may affect learning with a cochlear implant system. For children with a sudden, acquired hearing loss (particularly following meningitis, where there risk of cochlear ossification leading to surgical complications) a fast track programme is in place to enable surgical priority.

Criteria for referral to the paediatric programme

Criteria are selected according to evidence-based practice and experience. They are set to ensure that those children who receive a cochlear implant are those most likely to obtain benefit from the device. None of the criteria outlined below exclude children with additional physical disabilities or learning difficulties.

Referrals for assessment are accepted for:

- Children who were born with a profound hearing loss, receive no significant benefit from hearing aids, and are under the age of 4 years at the time of referral. NB. The younger a child is when he/she receives a cochlear implant, the more successful the outcome is likely to be. A child who receives a cochlear implant at the age of 4 years will be far less likely to successfully acquire spoken language than a child who receives a cochlear implant at the age of 18 months. We would therefore recommend that any child, however young, who is suspected of having a significant hearing impairment should be referred to the cochlear implant team as soon as possible (including cases where auditory neuropathy spectrum disorder may be indicated). This will allow the cochlear implant team to begin carrying out assessments, and informing parents about cochlear implants, in parallel with the ongoing audiological assessments and hearing aid fitting being carried out by the child’s local services.
As part of the assessment programme, the child will undergo an MRI (Magnetic Resonance Imaging) scan in order to confirm the presence of a cochlear nerve, and determine the suitability of the inner ear to receive an implant.

**There are circumstances under which children over the age of 4 years may also be referred to the paediatric programme. Referrals for assessment are accepted for:**

- Children under the age of 10 years who were born with normal hearing and have acquired a profound hearing loss, e.g. following meningitis. A child with a suspected hearing loss following meningitis should, of course, be referred for assessment immediately so that cochlear implant surgery can be fast-tracked in the event of any ossification of the cochlea.
- Children under the age of 10 years who have had some benefit from hearing aids in the past, but whose hearing has deteriorated to the point where powerful hearing aids are no longer helpful
- Children under the age of 10 years who were born with a profound hearing loss, have received some benefit for language learning through consistent use of powerful hearing aids but who might receive significantly more auditory information from a cochlear implant.

NB. These patients should show evidence that they have learnt spoken language through listening, thus demonstrating the integrity of the auditory pathway. The Paediatric programme encourages children to continue wearing a contralateral hearing aid if they have previously been good hearing aids users and have residual hearing in the non-implanted ear.

**Clinical activity between April 2010 and March 2011**

Number of patients implanted = 59

Devices used with paediatric patients:
- Cochlear Freedom = 1
- Cochlear CI512 = 28
- MED-EL Sonata Flex Soft = 23
- AB HiRes 90K = 7

Number of simultaneous bilateral implants = 38
Number of sequential second implants = 17
Number of re-implants = 4

Average age of paediatric cochlear implant patients = 4 yrs (Range= 13 months - 11 years)
Figures 13 and 14 show distribution of age and aetiology.

**Figure 13 – Age distribution of children implanted in 2010-2011**

**Figure 14 – Aetiology of hearing loss for children receiving cochlear implants in 2010-2011**

- AUDITORY NEUROPATHY (1)
- BA-HT (1)
- CONGENITAL UNKNOWN (26)
- COINFECTION (5)
- CONGENITAL (1)
- Combined syndrome (1)
- HYPOPLASTIC NERVE (1)
- MI NINETY (7)
- PREMATURITY (6)
- PREOXACETATE UNKNOWN (8)
- RETINOPATHY OF PRETERM (2)
- RETIROPHTALMIC SYNDROME (1)
- NOT VERTEBRAL ACCESSIBLE (3)
- UNKNOWN (1)
Post-implant support

Children are generally offered fortnightly habilitation sessions during their first two years of cochlear implant use. These sessions are designed to ensure that the child obtains maximum benefit from the cochlear implant. Therapists work with parents or caregivers to help the child to develop spoken language through listening (Auditory Verbal Therapy). Children also have regular appointments for reprogramming of the speech processor.

After two years of implant use, primary responsibility for a child’s habilitation programme is handed back to the local support services. However, the implant team continues to provide advice, support and training to local professionals. Children continue to be seen annually by the cochlear implant team for equipment checks, reprogramming and speech and language assessments.

Outcomes

Manchester Spoken Language Development Scale (MSLDS)
The Manchester Spoken Language Development Scale (MSLDS) was first developed in 1999 and revised in 2004. Its purpose was to provide the team’s habilitationists with a simple and meaningful way of categorising the level of auditory spoken language learning achieved by the children in their care and of charting their progress over time. The high level of contact between the team and children in their first two years of cochlear implant use enables habilitationists to pinpoint a child’s current level of development. For longer term implant users, reports from parents and local support professionals, together with the child’s performance on standardised assessments administered at the annual review, are used to determine their level of attainment on the scale.
AUDITORY BRAINSTEM IMPLANT PROGRAMME

Manchester is one of two centres in the UK providing Auditory Brainstem Implants (ABI) for adults with Vestibular Schwannoma or Neurofibromatosis Type II (NFII). Like the Cochlear Implant, the ABI has both internal and external components. Following the removal of a tumour from the auditory nerve, the ABI can provide a patient with a sensation of sound by directly stimulating the cochlear nucleus in the brainstem. Patients undergo a programme of auditory rehabilitation similar to that of cochlear implant recipients.

Since 1993, 56 patients have been implanted with an ABI. 53 of these have been adults/adolescents, and 3 were children.

Clinical activity between April 2010 and March 2011

Number of patients implanted = 7

Average age of auditory brainstem implant patients = 35 yrs (Range= 2-59 yrs)
**RECENT PUBLICATIONS**

**Cochlear implantation in Donnai-Barrow syndrome.**

**Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement.**

**The outcome of cochlear implantation in adolescents.**
Bruce IA, Broomfield SJ, Melling CC, Green KM, Ramsden RT. Cochlear Implants Int. 2011 May;12 Suppl 1:S82-3.

**Brain stem responses evoked by stimulation with an auditory brain stem implant in children with cochlear nerve aplasia or hypoplasia.**

**Cochlear Implantation after Bacterial Meningitis in Children: Outcomes in Ossified and Nonossified Cochleas**

**Outcome of cochlear implantation in asymptomatic congenital cytomegalovirus deafened children.**

**Outcome of cochlear implantation in asymptomatic congenital cytomegalovirus deafened children.**

**Cochlear implantation following cerebellar surgery**
Saeed S, Mawman D, Green KM. Cochlear Implants International August 2011 12 (3): 185-188

**Bilateral cochlear implantation after ethylene glycol intoxication: a case report.**
Cochlear implantation in far advanced otosclerosis. A surgical, audiological and quality of life review of 35 cases in a single unit
Flook EP, Broomfield SJ, Saeed S, Bruce IA, Green KM
Int. Adv. Otol. 2011; 7:(1) 35-40

Cochlear implantation in children with cerebral palsy.
Steven RA, Green KM, Broomfield SJ, Henderson LA, Ramsden RT, Bruce IA.

Hearing preservation via a cochleostomy approach and deep insertion of a standard length cochlear implant electrode.
Bruce IA, Bates JE, Melling C, Mawman D, Green KM.
Otol Neurotol. 2011 Dec;32(9):1444-7

Goswamy J, Bruce IA, Green KM, O'Driscoll MP.

Broomfield SJ, Bruce IA, Henderson L, Ramsden RT, Green KM.

Cochlear implantation in a child with CINCA syndrome who also has wide vestibular aqueducts.
Bates JE, Bruce IA, Henderson L, Melling C, Green KM.
APPENDIX 1 - REFERRAL CRITERIA

NICE technology appraisal guidance 166

1.1 Unilateral cochlear implantation is recommended as an option for people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids, as defined in 1.5. If different cochlear implant systems are considered to be equally appropriate, the least costly should be used. Assessment of cost should take into account acquisition costs, long-term reliability and the support package offered.

1.2 Simultaneous bilateral cochlear implantation is recommended as an option for the following groups of people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids, as defined in 1.5:
   • Children
   • Adults who are blind or who have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness.

1.3 Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.

1.4 People who had a unilateral implant before publication of this guidance, and who fall into one of the categories described in 1.2, should have the option of an additional contralateral implant only if this is considered to provide sufficient benefit by the responsible clinician after an informed discussion with the individual person and their carers.

1.5 For the purposes of this guidance, severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz without acoustic hearing aids. Adequate benefit from acoustic hearing aids is defined for this guidance as:
   • For adults, a score of 50% or greater on Bamford–Kowal–Bench (BKB) sentence testing at a sound intensity of 70 dB SPL
   • For children, speech, language and listening skills appropriate to age, developmental stage and cognitive ability.

1.6 Cochlear implantation should be considered for children and adults only after an assessment by a multidisciplinary team. As part of the assessment children and adults should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate).
1.7 When considering the assessment of adequacy of acoustic hearing aids, the multidisciplinary team should be mindful of the need to ensure equality of access. Tests should take into account a person’s disabilities (such as physical and cognitive impairments), or linguistic or other communication difficulties, and may need to be adapted. If it is not possible to administer tests in a language in which a person is sufficiently fluent for the tests to be appropriate, other methods of assessment should be considered.
APPENDIX 2 – LOCAL CLINICAL CRITERIA FOR TREATMENT

Adults

Candidates should have a severe, profound hearing loss in both ears (average hearing threshold level >70 dB in the better hearing ear).

Irrespective of the degree of hearing loss at birth, candidates should have acquired the ability to use spoken language and should be oral communicators. In general, the onset of profound total deafness will have occurred after the acquisition of spoken language.

Candidates should receive no material benefit from acoustic hearing aids (e.g. aided thresholds >55 dB(A) at 2-4 kHz after 4-12 weeks of acclimatisation).

Candidates should display no ability to understand speech using acoustic hearing aids (≤70% of keywords identified in the best aided condition and 50% of the key words correctly identified in the ear to be implanted when the IHR/UCL recording of the BKB Sentence Test is presented at a level of 70 dB(A), without lip reading).

Candidates should display no morphological abnormality of the inner ear that would prevent the placement of electrodes close to surviving fibres of the auditory nerve. 2,3,4,5

Candidates should be physically fit for surgery and able and willing to participate in device tuning and rehabilitation.

Candidates should be committed to their own health care, to the proposed treatment and to oral communication.

The candidate should have realistic expectations regarding the outcomes of implantation.

Adults should be considered psychologically sound and able to participate in the programme.