COCHLEAR IMPLANT TEAM

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Introduction

Cochlear implantation has been established in the UK for over 3 decades and there are currently just over 17000 patients in the UK using cochlear implants (see Figure 1). In the UK, cochlear implants are funded by NHS England (Ref: D09/S/A) if patients meet the NICE guidance (Ref: TA166. This guidance is currently under review.

Figure 1: Total number of maintained patients in the UK (from BCIG’s annual data collection 2017-18)

The auditory implant programme in Manchester was established in 1988 by ENT Consultant Richard Ramsden, using funding to provide cochlear implants obtained from the HEAR (Help Ear & Allied Research) Charity. In the mid 1990’s government resources became available to fund cochlear implants for patients with severe-profound sensori-neural hearing loss. By the end of March 2018, 1274 adults have received a cochlear or an auditory brainstem implant in Manchester manufactured by Advanced Bionics, Cochlear and MED-EL.
Criteria for referral to the adult programme for cochlear implants

There is no maximum age for referral and patients with additional needs are not excluded (Ref: NICE TA166):

- Bilateral, severe to profound sensori-neural hearing loss (≥ 90 dB HL at 2 and 4 kHz).

- Limited or no benefit from hearing aids (a score of 50% or less on BKB sentence testing at a sound intensity of 70 dB SPL in the ear to be implanted).

- Simultaneous bilateral cochlear implantation is recommended as an option for adults who are blind or who have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness.

- Cochlear implantation is considered for adults only after an assessment by a multidisciplinary team. As part of the assessment adults should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate).

Clinical activity between April 2017 and March 2018

Procedures

During the 2017-18 financial year, a total of 115 cochlear implant procedures were performed in adults. Figure 2 shows a breakdown of the types of surgeries performed.
Three explantations were carried out, all due to medical problems (D) based on the International Classification of Reliability for Implanted Cochlear Implant Receiver Stimulators (see figure 3). Two of the explanted patients were successfully re-implanted. One patient continues to suffer from recurrent ear infections in the implanted ear which may be related to a pre-existing medical condition and their management is on-going.

Figure 3: International Classification of Reliability for Implanted Cochlear Implant Receiver Stimulators (Battmer et al., 2010)
Excluding explantation and repositioning procedures, a total of 110 devices were fitted in this financial year. Detailed charts on the types of internal devices, manufacturers and type of speech processor are shown in figures 4 to 6.
Figure 6: Speech processors by configuration

✦ Demographics

The average and median age of the patients implanted was 56 years (range = 19 - 94 years). Figure 7 shows the age distribution of the implanted adult population during the 2017-2018 financial year period.

Figure 7: Age of patient at implant

✦ Post-implant support

Patients typically attend 5 appointments with the adult team within their first six weeks of implant use. During these appointments the speech processor is
programmed and patients receive rehabilitation through an individualised auditory and communication skills training programme with a therapist. Where appropriate, the training programme includes tactics for using the telephone, music therapy and advice about using assistive listening devices. Patients are then followed up at three, nine and twenty-one months after their initial activation. Additional rehabilitation sessions are offered to patients as required. Following this, a patient led appointment is sent on an annual basis. Every 6 years the patient will be offered a speech processor upgrade.

**Outcomes**

Speech perception and lip-reading function is measured using standardized recorded test materials for each patient at the pre-implant stage. Following implantation, the tests are repeated at one week, three months, nine months and twenty-one months, and then annually as required. Patients who use English as a second language, or who cannot communicate in English are evaluated in their own language using language interpreters to translate the test materials to the appropriate language.

Figure 8 shows the average scores obtained using the Bamford-Kowal-Bench (BKB) sentence test in quiet at pre- and post-operative stages in this financial period.

![Figure 8: Speech perception scores](image-url)
Sound-field aided thresholds are also measured post operatively. Figure 9 shows the average aided thresholds at the 1 week, 3 month and 9 month stage, as well as their average pre-op unaided thresholds. Thresholds ≤ 40 dB HL at frequencies between 250 and 8000 Hz allow good access to normal conversational speech levels and everyday sounds and are considered optimal for cochlear implant users.

![Graph showing average aided thresholds at different stages](image)

**Figure 9: Threshold levels at pre op and post stages**

- **Service evaluation**

A satisfaction questionnaire is sent to all newly implanted users at their 9 month post implant stage to gauge their views on their cochlear implant progress and our service. All responses are anonymous. Below are a few responses collected during the last financial year:

- 91% of respondents report that their cochlear implant exceeded their expectations or that it was too soon to comment.
- 100% of respondents report using their cochlear implant every day (all day long or part of the day)
- 91% of respondents report obtaining great benefit from their cochlear implant
- 100% of respondents feel it was worthwhile having a cochlear implant
- 100% of respondents would recommend a cochlear implant to a friend or relative if they had a similar hearing problem
- 100% of respondents were satisfied with the treatment/support that they received from the staff on the cochlear implant programme
- 100% of respondents rated the treatment/support that the staff at the cochlear implant programme provided as very good

Below are statements from respondents with regards to why they felt this treatment was worthwhile:

“For so much of my life I have felt foolish and odd because I could not hear so I would withdraw into myself.”

“Their support has brought me out into the sunshine”

“It has given me my life back. I can go out now and be part of a conversation where before you are isolated”

“I am no longer excluded from conversation”

“It has given me confidence to go places alone. It has completely changed my and that of my family”

“I can now hold a conversation clearly and not afraid to be in company”

Among the disadvantages noted by cochlear implants users, the following were reported: one patient reported short battery life, another patient noted louder but non-troublesome tinnitus and one other patient reported discomfort when wearing their device.
Introduction

The Paediatric Programme at the RRCHI was established in October 1991 and to March 2016, 864 children (ages 0-11) 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. We also work with children with a wide range of additional disabilities.

All patients referred to the Paediatric Programme undergo a full audiological 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 may be 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. In addition, 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. Some children may also require a CT scan but this is not routinely undertaken. 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 for cochlear implants

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. 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 less likely to successfully acquire fluent 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. 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.

In cases where a child receives a unilateral implant, the Paediatric Programme encourages children to continue wearing a contralateral hearing aid if they have residual hearing in the non-implanted ear.

None of the criteria outlined below exclude children with additional physical disabilities or learning difficulties. Referrals for assessment are accepted for:

• Referral from first ABR where levels indicate cochlear implant may be appropriate.

• Children who were born with a profound hearing loss, receive no significant benefit from hearing aid and are under the age of 4 years at the time of referral

• 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 and / or have shown 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)

- Children diagnosed with ANSD and have consistent behavioural hearing thresholds bilaterally in the severe-profound SNHL range (≥90dBHL at 2 and 4 kHz) should always be referred to the cochlear implant programme. There is no minimum age of referral but it is still important to refer these children early. Children with ANSD and with additional needs should not be excluded. Children with ANSD can also be referred to the implant programme if they are making poor progress with their hearing aids despite having unaided thresholds outside implant criteria. We classify poor progress as:
  - Unable to consistently discriminate the ling sounds
  - Unable to imitate pattern perception (duration and number of syllables in a word)
  - Only able to understand simple instructions with visual cues e.g. pointing
  - Not making the appropriate progress with spoken language development with their hearing aids
  - Have not achieved behavioral hearing levels by 14 months of age.

- Children with one ear in the profound range and the other ear in the moderate / severe range. We accept referrals of all children who meet this criteria, but those over the age of 4 must be developing spoken language.

- Children with auditory nerves dysplasia. Children with suspected thin or absent nerves should also be referred to the paediatric team. We can assess these children using electronic auditory brainstem response to assess the ability of the auditory nerve to carry auditory responses. If good traces are obtained then these children may be still eligible for a cochlear implant. Those with absent nerves or have poor eABR traces can then be referred for an Auditory Brainstem Implant if the parents and professionals feel it is appropriate.
Please note:

• Early referral is encouraged (from ABR if possible / appropriate) and is extremely important to help children who need a cochlear implant to get one as quickly as possible.

• If you are unsure about a referral or unable to obtain behavioural levels please speak to us beforehand or send a referral. We are happy to see people for second opinions.

• That children aged 10 years and over should be referred to the Adolescent Programme.

✦ Clinical activity between April 2017 and March 2018

✦ Surgeries

A total of 55 children (a total of 89 cochlear implants) have been implanted during this financial year. A detailed info-graphic on the types of implants, processors and configuration of implantation is shown below.

Figure 10: Breakdown of implanted ears for children implanted in 2017-2018
The average age of paediatric cochlear implant patients was 3 years (range = 9 months - 8 years 10 months). Figures 13 and 14 show the age distribution and...
hearing loss aetiology of the implanted children population at the RRCHI during the 2017-2018 financial year period.

**Figure 13:** Age distribution of children receiving cochlear implants for the first time (reimplant and sequential patients not included)

**Figure 14:** Aetiology of children receiving unilateral or bilateral cochlear implants (reimplant and sequential patients not included)

BVVL = Brown Vialetto Van Laere Syndrome, ANSD = Auditory Neuropathy Spectrum Disorder, CMV = Cytomegalovirus, WVA = widen vestibular aqueducts
We carry out the Brief Assessment of Parental Perception (BAPP) after 2 years of implant use. This is a questionnaire gives the parents perception of their child’s implant use and willingness to wear implant and whether they would recommend a cochlear implant to other parents. It also asks for comparisons in their child’s in behaviour, contentment, communication learning and getting on with friends pre-implant compared to post. After 2 years of use, all patients wore their processor(s) at least some of the day, with the majority wearing it full time. The willingness for paediatric patients to wear their processor(s) at 2 years of use can be seen in Figure 15. The majority are very keen to wear. One hundred percent of the parents who completed the questionnaire would recommend cochlear implantation to another family in a similar situation.

![Percentage of paediatric patients willingness to wear their processor(s) after their second year of use](image)

Figure 15: Paediatric patients’ willingness to wear their after 2 years of use

The paediatric patients are seen regularly in their first year of implant use to establish good listening levels. We categorize good listening levels to be between 20 and 40 dB HL using sound field testing (warble tone). With some children who have
developmental delay it is often not possible to test aided levels. We therefore rely on objective and behavioral testing to establish that the processor is set optimally for the patient. Figure 16a shows the average aided levels achieved 1 year post implant. These levels are also checked at two years of use and remain stable (Figure 16b).

**Figure 16**: a) Average aided levels for the first year of implant use

**Figure 16**: b) Average aided levels for the second year of implant use

Within the first year of implant use, 100% of paediatric patient that were able to do behavioural testing achieved average aided thresholds of 40 dB HL or better.
**Post-Implant Support**

Children are generally offered regular habilitation sessions during their first three 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. Our habilitation programme is based on Auditory Verbal Therapy. Children also have regular appointments for reprogramming of the speech processor.

Over time primary responsibility for a child’s habilitation programme is handed back to the local support services. However, the implant team is always available to provide advice, support and training to local professionals if required/requested. Children continue to be seen annually by the cochlear implant team for equipment checks, reprogramming and speech perception assessments.

**Outcomes – PLS**

The Preschool Language Scales is standardized on normally hearing children aged from infancy to 6 years 11 months. The purpose of this assessment is to assess children’s receptive and expressive language capabilities. Responses range from parental report to picture selection and completion of open-ended sentences. 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 standardized assessments administered at the annual review, are used to determine their level of attainment on the scale. Figure 17 shows the outcomes of the PLS preimplant, and 1 and 2 years of implant use. The data show that the children on average of their age equivalent scores are catching up to their chronological age.
Figure 17: Average difference between chronological age and language age as a percentage for paediatric patients at preimplant and 1 and 2 years post implant use.
Introduction

The implant centre in Manchester has the largest population of auditory brainstem implant (ABI) users in the UK. The majority of these patients have Neurofibromatosis Type 2.

The auditory brainstem implant (ABI) depicted in figure 18 evolved from cochlear implant technology to address the problems of rehabilitating patients with total deafness with a damaged or absent cochlear nerve, and are therefore unsuitable for cochlear implantation. The great majority of these patients suffer from the genetic disorder called neurofibromatosis type 2 (NF2). NF2 affects one in 33,000 people and is characterised by the development of bilateral vestibular schwannomas (tumours on the hearing and balance nerves). More recently other indications for ABI have been approved including cochlear nerve aplasia or severe dysplasia in infants, extreme degrees of inner ear dysplasia even in the presence of normal looking auditory nerves, severe cochlear obliteration from otosclerosis or meningitis, and head injury with cochlear nerve avulsion. These conditions prevent effective cochlear implantation.

Figure 18: The auditory brainstem implant and external speech processor

The ABI stimulates the cochlear nucleus complex on the brainstem directly and bypasses any damaged or absent cochlear nerve. The ABI electrode array is placed on the surface of the cochlear nucleus complex on the floor of the
foramen of Luschka in the lateral recess of the fourth ventricle of the brainstem (see figure 19).

Figure 19: Placement position of the auditory brainstem implant

The external speech processor for an ABI is activated about 6 weeks post-surgery and patients undertake a similar programme of tuning and rehabilitation to cochlear implant patients. The ABI provides limited speech understanding for patients although it can help patients to understand speech better with lipreading (see figure 20). It also helps patients to hear and identify everyday sounds.

Figure 20: Comparison of CUNY scores (N = 49) using the ABI only, lipreading (LR) only and the ABI with lip-reading (Ramsden et al., 2016)

Manchester is one of only two centres in the UK commissioned to provide ABI’s. For adults, the ABI surgery takes place at Salford Royal NHS Foundation Trust and for children the surgery takes place and the Royal Manchester Children’s Hospital. The ABI programming and rehabilitation takes place within the Richard Ramsden Centre for Hearing Implants.

To date, the team has performed 82 ABI surgeries in 73 adults and 9 children.
In November 2016, Channel 4 broadcasted an observational documentary LIVE from the Richard Ramsden Hearing Implant Centre titled “Breaking the Silence”. The programme allowed viewers to share the moment eight patients heard for the first time when their cochlear implant was switched on. Staff and patients from the Manchester implant centre as well as other centers from around the UK (see picture 21), were involved in the ground breaking documentary.

This documentary can be viewed online in the following link: http://www.channel4.com/programmes/breaking-the-silence-live/on-demand/63119-002

Following the live broadcast, our Head of Department, Martin O’Driscoll, appeared on the BBC Breakfast TV programme to talk about the benefits of cochlear implantation.
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