West of England Cochlear Implant Programme

Paediatric Team

A report on activity and outcomes

1997 - 2012
Introduction

The West of England Cochlear Implant Programme (WECIP) was established in 1995 and the first child was implanted in 1997. By the end of 2012, a total of 219 children had received one or two cochlear implants (CIs). This document reports on the activity of the paediatric programme and outcomes of the children seen over this time period.

Our Activity

We implant children from the South West of England and beyond (Fig. 1.), Somerset, Bristol and South Gloucestershire being our top three referrers.

Fig. 1. Number of Children with Cochlear Implants by PCT

The causes of hearing loss in the children that we implant are very varied; in line with population data, for the majority of children the cause of their hearing loss is unknown (Fig. 2.).

Fig. 2. Aetiologies of Hearing Loss
Fig. 3. Number of Cochlear Implant Surgeries by Year

The number of implant operations has increased steadily, particularly following guidance from the National Institute for Health and Clinical Excellence (NICE TAG 166: NICE, 2009) recommending that children receive bilateral devices. Fig. 3. shows the number of CI operations performed (rather than the number of devices implanted). The number of CI devices implanted over the last 6 years (since the advent of bilateral simultaneous surgery) is given in Table 1; we implanted 50 CIs in 2012.

<table>
<thead>
<tr>
<th>Year</th>
<th>No. of Devices Implanted</th>
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<tbody>
<tr>
<td>2007</td>
<td>19</td>
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<tr>
<td>2008</td>
<td>24</td>
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<td>2009</td>
<td>28</td>
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<td>2010</td>
<td>28</td>
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<td>2011</td>
<td>30</td>
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<td>2012</td>
<td>50</td>
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Table 1. Transfers in and out of the Programme

Since 1997, a total of 19 children have transferred to WECIP from other implant programmes, usually because they have moved into our region. Conversely, 35 children have transferred out of the programme either to our adult team or because they have moved away.
Age at Implantation

Globally, the recommended minimum age for implantation is falling and at WECIP we would now consider implanting a child between 8 and 10 months of age. In some cases this might be later, depending on the child’s audiological profile and other developmental issues. Fig. 4 illustrates how the minimal age of implantation at WECIP has fallen over the years.

The age-range of the children that we have implanted, however, remains diverse as we implant children who develop severe-profound hearing loss later in life, as well as those referred early from the Newborn Hearing Screening Programme (Fig. 5.). We are also dependent on the age of referral to our programme and welcome babies and children being referred to us at the earliest stage possible. Initially, it might just be appropriate to offer advice about cochlear implantation.

Rapid referral is particularly important in children whose hearing loss has been caused by meningitis; cochlear ossification can occur very quickly in these children which will prevent full electrode insertion.

Fig. 4. Youngest Age at CI Surgery
Device Choice

At the WECIP, we offer devices made by Cochlear and MED-EL to children. Device choice has varied over the years and is mainly dependent on parental choice, although sometimes one particular device may be more appropriate for medical or lifestyle reasons. By the end of 2012, over the duration of the paediatric programme, 120 children (55%) had received one or two devices made by Cochlear, the remainder receiving MED-EL devices. The variation (Fig.6.) could be explained by the introduction of new products on the market, device problems, cosmetic aspects and other factors.

**Fig. 6. Percentage of CIs by Manufacturer**

The Rehabilitation Process

Surgery is the first stage of the child learning to listen and develop spoken language. This journey is monitored and supported by our Rehabilitation Team who will provide advice and recommendations over the coming months and years.

The device(s) are usually switched-on 3 – 4 weeks after surgery. After this, children are seen regularly by Audiology for tuning and assessment. In addition, the specialist Speech and Language Therapists and Teachers of the Deaf of the Rehabilitation Team work closely with the child’s family and with local professionals supporting the child (e.g. Teachers of the Deaf, Speech and Language Therapists and mainstream teachers) in the child’s home and educational setting. Once a child has had his/her implant(s) for 3 years, follow-up in Audiology is usually on an annual basis (although of course we will see children as needed should any problems arise). Children will be routinely seen by the Rehabilitation Team for the first 10 years after switch-on. Additional visits are provided at any stage, for example if the child is experiencing difficulties at transition between educational placements, or is being fitted with an FM device.
Outcomes from Implantation

Assessment and monitoring progress forms the cornerstone of our rehabilitation programme. By regularly assessing a child’s listening and language abilities we are able to tailor rehabilitation recommendations to the child’s individual needs. The type of assessment performed depends on the child’s age and developmental stage. Assessment of a child’s hearing ability generally covers four main categories: sound detection, functional listening and communication development, speech production and speech recognition. A variety of different tools are used to measure outcomes, together with observation of the child’s functional listening and communication ability. Regular assessment of both a child’s hearing and speech and language ability enables us to document a child’s progress as well as informing our rehabilitation recommendations.

Fig. 7. Average Sound-Field Results at One Year Post Implantation

Sound Detection
Hearing for different sounds across the speech frequency range (500Hz – 4kHz) is usually assessed from about 3 months’ post implantation. Information gained from these audiometric sound-field results is used in tuning a child’s implant(s). Fig. 7. shows the average results obtained for 154 of our children at 1 year post-implantation.

N=154; (95% Confidence Limits shown)

Functional Listening and Communication Development
The Categories of Auditory Performance (CAP) scale enables hearing ability to be classified into eight mutually exclusive categories ranging from awareness of environmental sounds through to use of the telephone. The results from WECIP clearly demonstrate that with increasing CI experience, CAP scores improve with many children developing the ability to use the telephone (Fig. 8.). A total of 34.3% of our children can use the telephone 4 years after implantation (compared to 1.0% pre-implantation).

Fig. 8. Categories of Auditory Performance Scores

N.B. Shows the 3 highest categories of CAP score. Note how both the percentage of children in all three of these categories has increased as well as the percentage of children able to use the telephone.
The Development of Spoken Language

A child’s developing communication is monitored and assessed using both informal and formal assessments carried out by both WECIP and local professionals. We work closely with our local colleagues to ensure seamless care and to inform the development of local Family Support Plans and Targets.

The Speech Intelligibility Rating (SIR) Scale is used to assess the child’s development of spoken language. Speech production is classified on a scale ranging from pre-recognisable words (score of 1) to connected speech which is intelligible to all listeners and easily understood in everyday contexts (score of 5). Our results are given in Fig. 9, which shows how SIR scores improve with cochlear implant experience.

**Fig. 9. Speech Intelligibility Rating Scores**

The McCormick Toy Test is a closed-set test appropriate for the 3 – 7 year age group (or for older children who have limited language ability). At age 4 years, the average score of our children who were able to undertake testing was 44.4dBA (SD 10.3, n=50), with similar results at age 7 years (mean score = 45.2dBA, SD 8.8, n=59, Fig.10.). These scores are the sound level at which a child is able to correctly identify 71% of the toys. (45dBA is approximately the level of very quiet speech.)

**Fig. 10. McCormick Toy Test Results (age 7)**
Older children are usually able to participate in sentence testing and at WECIP we use the adaptive Bamford-Kowal-Bench (BKB) test which is an open-set test and linguistically more complex than the McCormick Toy Test. As with the McCormick Toy Test, our BKB scores represent the sound level at which a child is able to identify 71% of keywords correctly. The results for the 70 children (age 7 – 18 years) able to participate in BKB testing are given in Fig. 11a and Fig. 11b. The mean score is 46dBA (SD 8.3).

**Fig. 11a. Adaptive BKB Scores (in quiet)**

![Graph showing adaptive BKB scores in quiet](image)

**Fig. 11b. Adaptive BKB Scores (in quiet)**

![Bar chart showing adaptive BKB scores](image)

**Children with Complex Needs**

It is common for children with hearing loss to have learning difficulties and other special needs. A significant proportion of our children are therefore unable to undergo the formal testing outlined above. Rather, alternative measures of testing are used. Observation of the child in his/her setting is particularly useful and gives the best indication of how the child is progressing. The knowledge and experience of our Rehabilitation Team is often the most important resource in monitoring a child’s progress and enables individual targets to be set for each child.
Parents’ Views

A recent parental questionnaire has found satisfaction with the paediatric programme to be very good, with 98% of families reporting that they were either “greatly satisfied” or “satisfied” with the service provided by the Rehabilitation Team.

Some parental and child testimonies are given below.

The Mother of a girl who received bilateral cochlear implants at the age of 8 months (in October 2012) says:

“When our daughter was diagnosed permanently and profoundly deaf, our world was shattered. In the early stages the West of England Implant programme provided us with hope - a light at the end of the tunnel. Pre- and post-implantation the entire team have given us constant but gentle advice, guidance and support, giving us clear and realistic goals - quite literally turning our journey from one of dread to one of excitement and expectation.

Any implant programme could have taken us through an implant journey, but it takes a special team of people, working so seamlessly together to give parents the reassurance and the confidence to undertake implantation without tremendous fear and panic. Without the programme our daughter’s world would have been one of silence and possible isolation. And whilst we are proud of her deafness, we are also blessed she now has a future of sound and speech...

One day she will hear us say ‘I Love You’ and just as importantly, say it back to us. Once that was just a distant dream. Now, thanks to the Implant Programme, it is our wonderful reality!”

A 16 year-old boy who has recently received a second implant says:

“Getting a Cochlear Implant has had a huge impact on my life. Since I have had the cochlear implant my clarity of sound is much better and it has also enabled me to communicate with people of different communities. I also feel the cochlear implant has enabled me to become more independent and so less reliant on others. I have recently had a second implant in the hope that it will improve my location of sound and I am looking forward to being switched on.”

His Mother says:

“He was born with a moderate to severe hearing loss and immediately took to the advantages his hearing aids gave him. In spite of being given every opportunity to learn to sign and to join the Deaf community, he always preferred to be oral and took every opportunity to speak! As his hearing decreased he was finding school more and more difficult. Although he was only 9 we discussed the possibility of an Implant with him and he was very enthusiastic about the possibilities.

Once he had his implant switched on he kept us all informed of his progress with comments such as “Oh that’s what a motorbike sounds like now.” He adapted well to the sound and has continued to grow in confidence in all situations. He has become a very confident young man who is expected to receive mostly A and B grades in this year’s GCSE results, he has carried out a variety of voluntary work as part of his Duke of Edinburgh Awards (he is currently working towards his Gold award) and has been involved in many sports and leisure activities. He is able to use both landline and mobile phones confidently and is currently looking forward to having his second implant switched on. As far as we as a family are concerned, the only downside to his implant is that we are so used to him getting on with everything that we sometimes forget we need an alternative means of communication when doing water-sports!”
Non-Users
As with any medical intervention, there will be occasions when circumstances arise when a child does not perceive benefit from implantation. This can be for a variety of medical or psychological reasons or distinct conditions such as autism, which is unlikely to have been diagnosed at the time of surgery. At WECIP we have 4 children (1.8%) who have become non-users of their cochlear implant. The decision to stop wearing a device is usually taken jointly by the family and WECIP.

Device Failures
It is rare for the internal part of a cochlear implant system to fail. At WECIP, 11 (5.0%) of our implants have failed, a rate which is similar to other UK centres. All of these children received a new device within a few weeks of their device failure without any long-term adverse effects.

Research and Development
Provision of a high quality clinical service has always been the priority of WECIP and will remain so. However, we are also a research-active programme and believe that evidence-based and reflective practice are both essential in maintaining and improving the quality of our service.

Several members of the team have published research and service evaluation papers in the national and international peer-reviewed literature and have presented their findings at national and international conferences. Research topics have been varied and include Auditory Neuropathy Spectrum Disorder, bilateral implantation, surgical technique, use of radiology, hearing preservation, outcomes in children with complex needs, parental experience of cochlear implantation, and multidisciplinary working.

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