Paediatric Programme
Annual report 2016 - 2017

NHS
Manchester University
NHS Foundation Trust

THE RICHARD RAMSDEN CENTRE FOR HEARING IMPLANTS

INVESTORS IN PEOPLE

POSITIVE ABOUT DISABLED PEOPLE

PAEDIATRIC PROGRAMME

The team

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Background

The Paediatric Programme at the RRCHI was established in October 1991 and to March 2016, 814 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. 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

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:

- 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

- 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)

- 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

- 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 that children aged 10 years and over should be referred to the Adolescent Programme.

Clinical activity between April 2016 and March 2017

Surgeries

A total of 52 children (a total of 69 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.

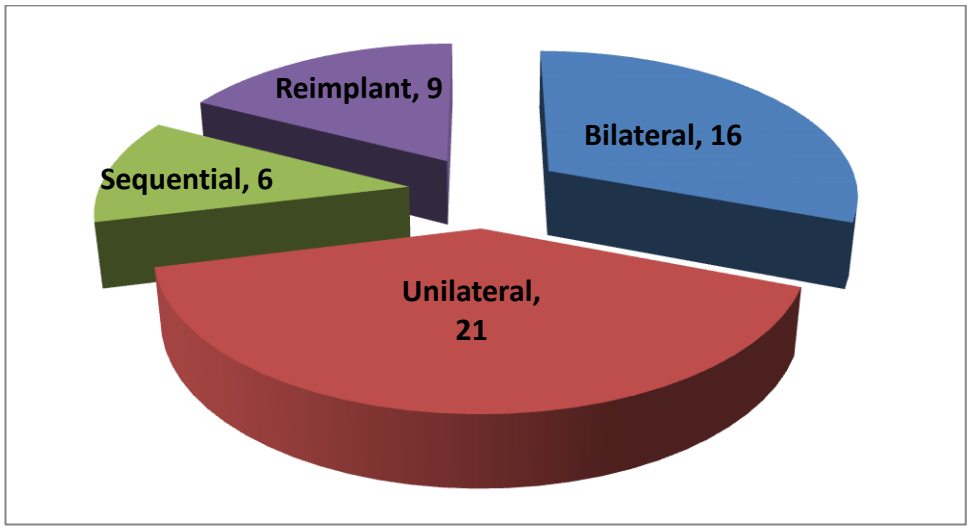

***Figure 1**: Breakdown of implanted ears for children implanted in 2016-2017*

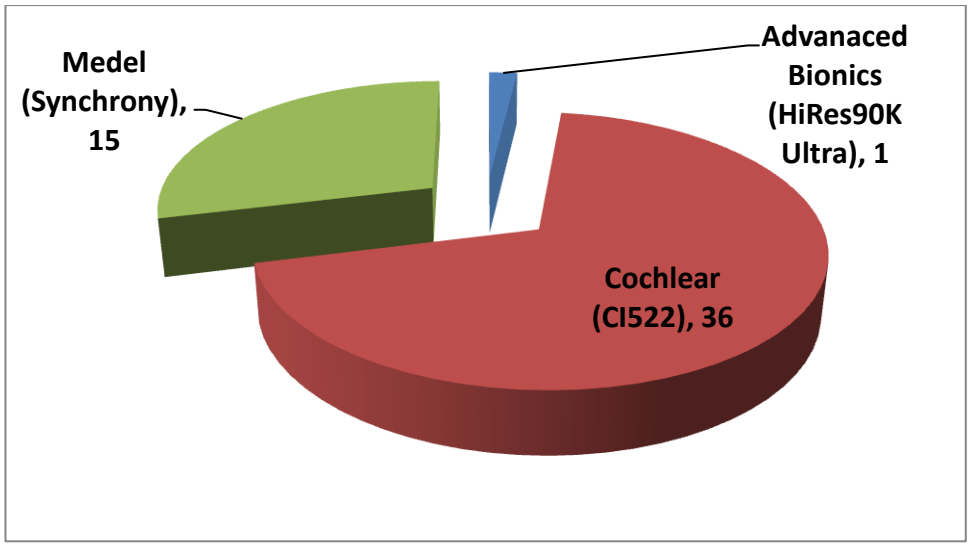

***Figure 2**: Breakdown of internal implants for children implanted in 2016-2017*

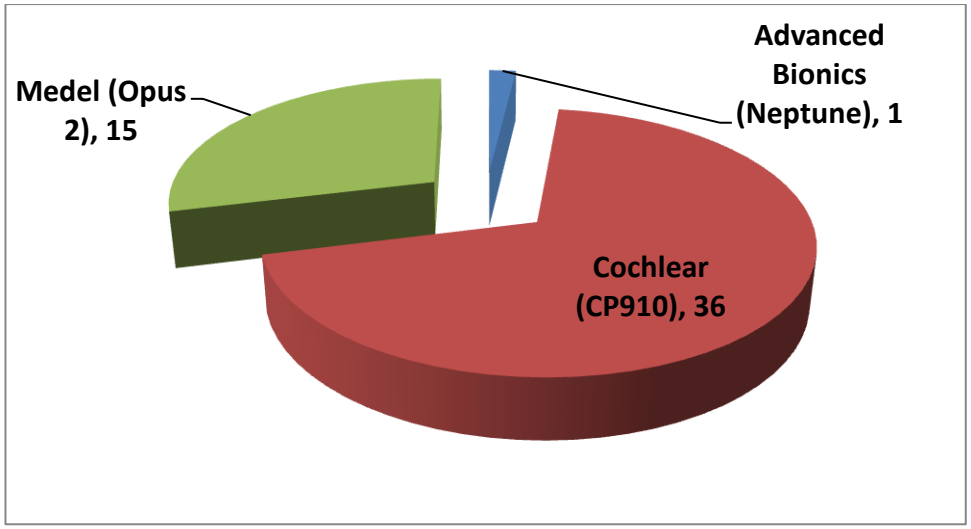

***Figure 3**: External devices for children implanted in 2016-2017*

Demographics

The average age of paediatric cochlear implant patients was 3 years (range = 13 months - 8 years). Figures 4 and 5 show the age distribution and hearing loss aetiology of the implanted children population at the RRCHI during the 2016-2017 financial year period.

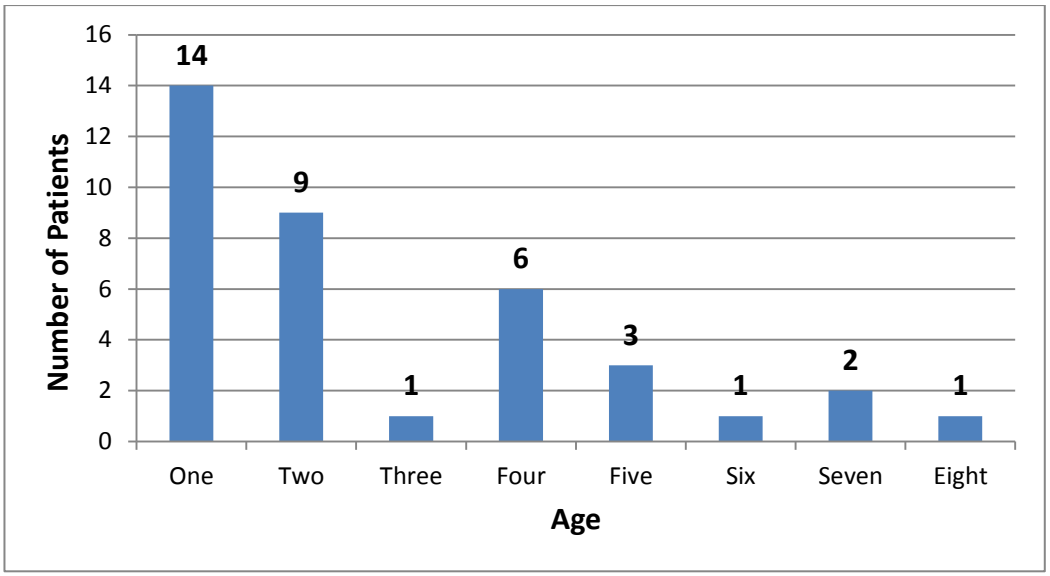

***Figure 4**: Age distribution of children receiving cochlear implants for the first time (reimplant and sequential patients not included)*

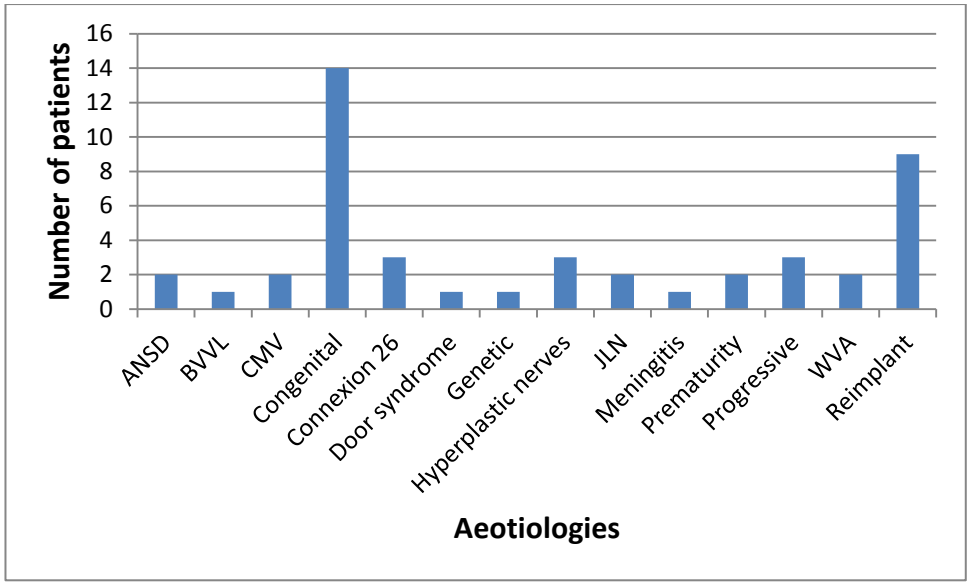

Figure 5*: Aetiology of children receiving unilateral or bilateral (not sequentials) cochlear implants*

Outcomes

Implant use

We carry out the Brief Assessment of Parental Perception (BAPP) after 1 and 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 1 and 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 one and 2 years of use can be seen in Figure 6 a and b. The majority are very keen to wear their processor especially by 2 years of implant use. One hundred percent of the parents who completed the questionnaire would recommend cochlear implantation to another family in a similar situation.

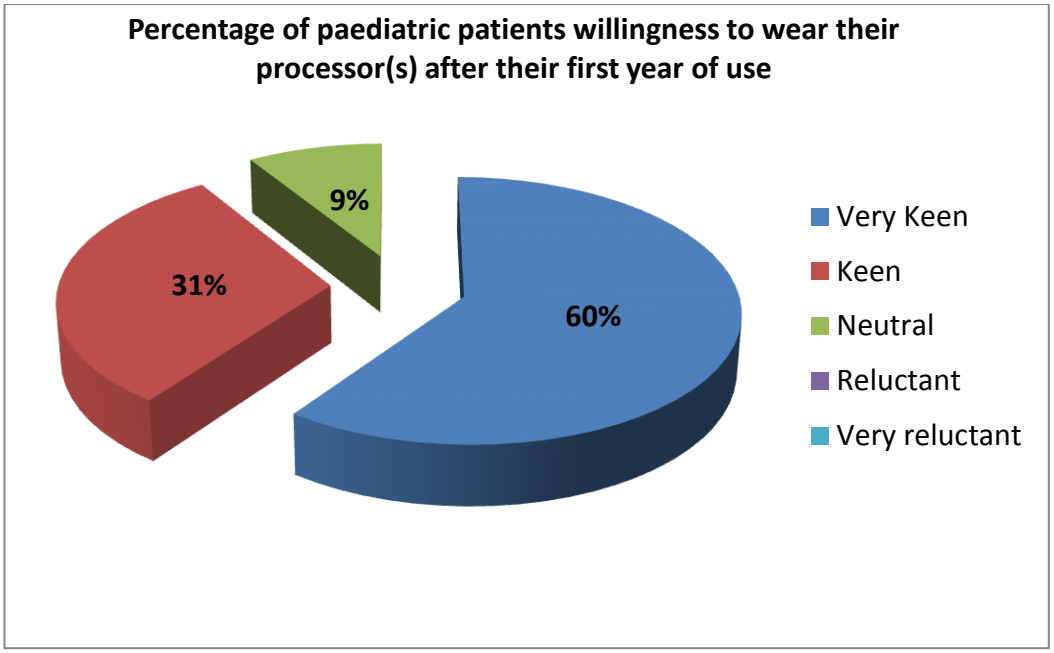

***Figure 6**: a) Paediatric patients' willingness to wear their after 1 year of use*

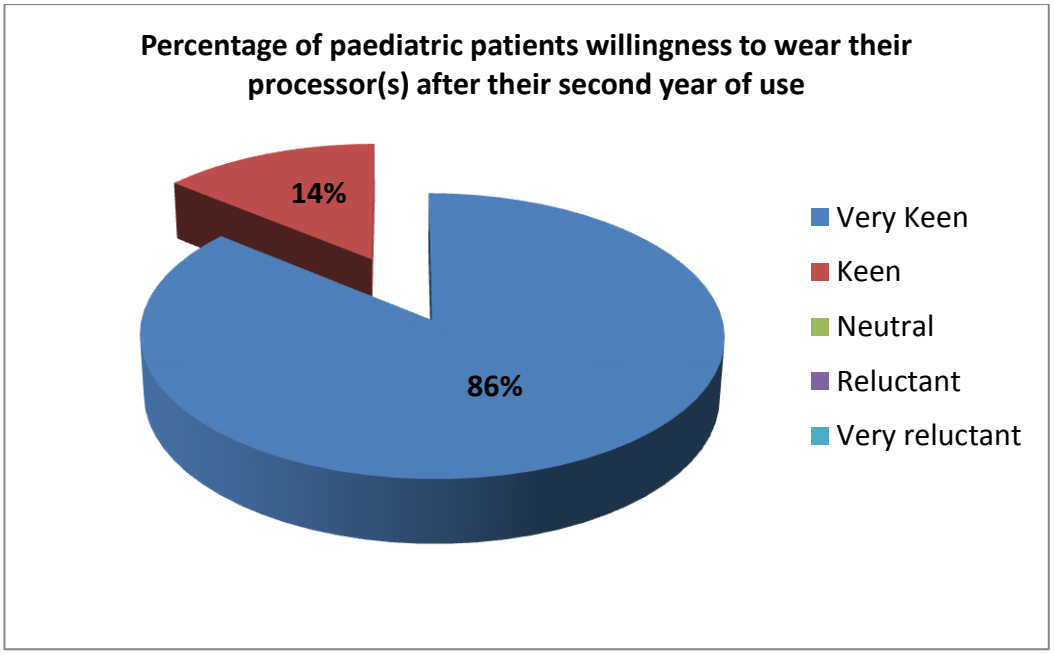

***Figure 6**: b) Paediatric patients' willingness to wear their after 2 years of use*

Aided levels

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 7a shows the average aided levels achieved 1 year post implant. These levels are also checked at two years of use and remain stable (Figure 7b)

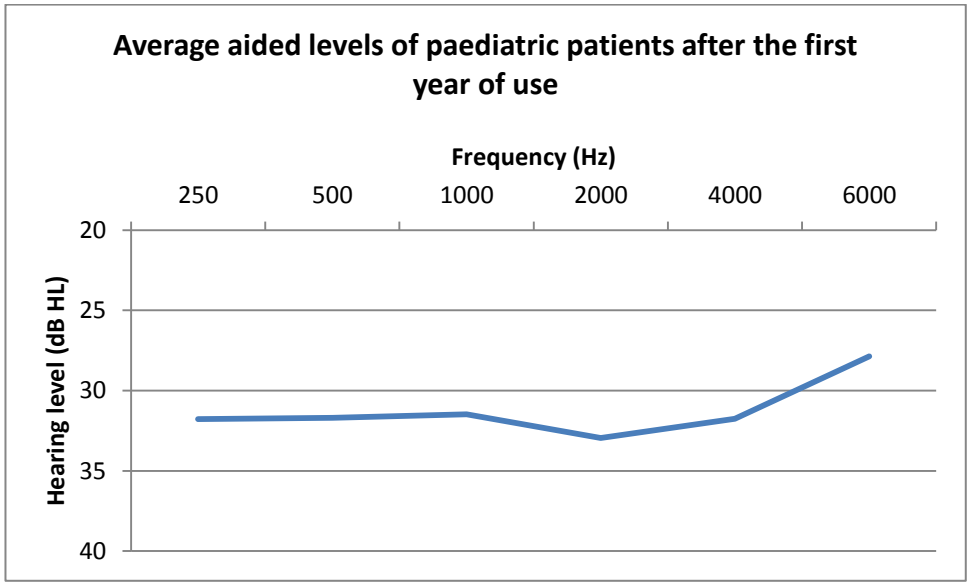

***Figure 7**: a) Average aided levels for the first year of implant use*

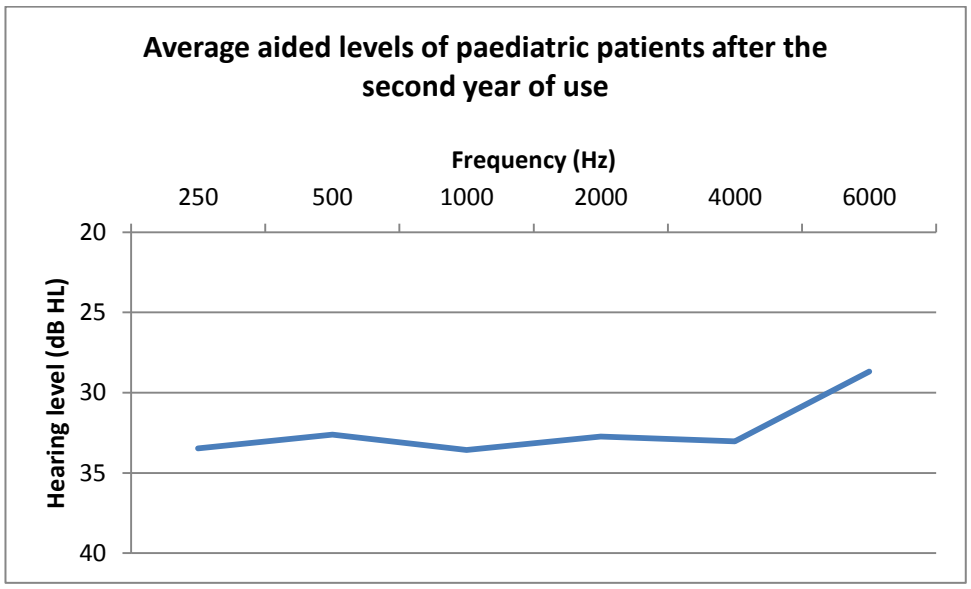

***Figure 7**: b) Average aided levels for the second year of implant use*

Within the first year of implant use, 93% 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.

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 8 shows the outcomes of the PLS at 1 and 2 years of implant use. At 1 year of use children are normally performing, on average, 3 months below their chronological age. By 2

years of implant use, children have caught up further and are on average performing at an age appropriate level.

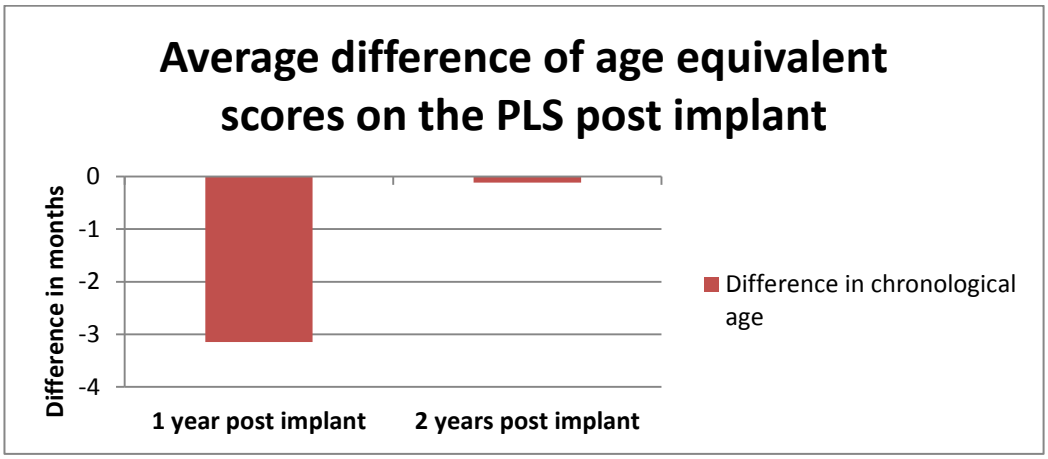

***Figure 8**: Average difference in age equivalent scores for both hearing age and chronological age of individual paediatric patients at 1 and 2 years post implant use*

PAEDIATRIC AUDITORY BRAINSTEM IMPLANT (ABI) PROGRAMME

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Surgeons

Simon Freeman (ENT Surgeon)

Simon Lloyd (ENT Surgeon)

Scott Rutherford (Neurosurgeon)

Charlotte Hammerbeck-Ward (Neurosurgeon)

Background

Manchester is the main UK centre for providing paediatric ABIs. This is for children who have cochlear nerve aplasia or hypoplasia, or have obliterated cochleas and cochlear implantation is not an option. Like cochlear implants, an ABI has both internal and external components. An ABI can provide children with a sensation of sound by directly stimulating the cochlear nucleus in the brainstem. Children undergo a programme of auditory rehabilitation similar to that of cochlear implant recipients. However, the benefit and sound perceived with an ABI is not equivalent to that of a cochlear implant and outcomes can be variable.

Currently the Manchester Programme looks after 15 children with ABIs. Two children have received bilateral devices which makes a total of 17 ABIs. 9 devices were implanted in Manchester and 8 devices were implanted in Verona, Italy. Two children have become non-users of their ABI.

Criteria for referral to the Paediatric ABI Programme

ABI are considered for children with hypoplastic or absent cochlea nerves or severe cochlea ossification. Due to the complexity of programming the ABI, we usually only recommend an ABI to Children who are developmentally able to perform behavioural audiological testing. Generally referrals are from paediatric cochlear implant centres who have diagnosed cochlear nerve aplasia or hypoplasia. As with cochlear implants, we only accept referral for children who are aged 4 years or younger.

Clinical activity between April 2016 and March 2017

During the financial year 2016 to 2017 we have implanted 2 MED-EL ABI devices. One child had cochlear nerve aplasia and the other had cochlear nerve hypoplasia. These children were aged 1 year 9 months and 2 years 10 months at the time of surgery.

Outcomes

Aided levels

As with cochlear implantation, aided levels are measured during programming appointments. We are aiming for levels between 20 and 40 dB HL at frequencies 500 to 4000 Hz. The average aided levels measured at the last review for our paediatric patients are shown below.

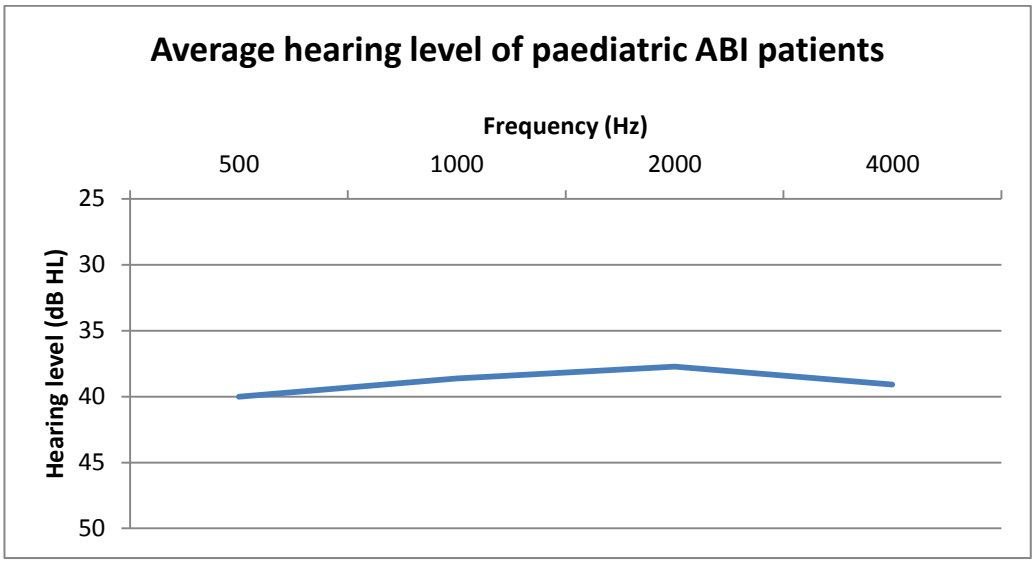


Figure 9: Mean audiological thresholds of the paediatric ABI patients at their last review

Post-implant support

Children are offered regular habilitation sessions during their first two years of implant use. These sessions are designed to ensure that the child obtains maximum auditory input as possible. Children also have regular appointments for reprogramming of their processor. Outcomes for auditory brainstem implants can be variable. We measure Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) questionnaires with each of our patients who use their ABIs. CAP is an 11 point scale outcome measure used to assess auditory receptive abilities by a paediatric patient. The 11 categories are outlined in Figure 10 below:

0: No awareness of environmental sounds or voice
1: Awareness of environmental sounds
2: Responds to speech sounds
3: Identification of environmental sounds
4: Discrimination of speech sounds without lip reading
5: Understanding of common phrases without lip reading
6: Understanding of conversation without lip reading
7: Use of telephone with known speaker
8: Follows group conversation in a reverberant room or where there is some interfering noise, such a classroom or restaurant
9: Use of telephone with an unknown speaker in unpredictable context
10: Use of telephone with an unknown speaker in unpredictable context

Figure 10: The 11 point scale use in the CAP

The SIR has 5 categories which grow in complexity and the clinician applies the category which best fits the patients spoken language abilities. The categories can be seen in the table below.

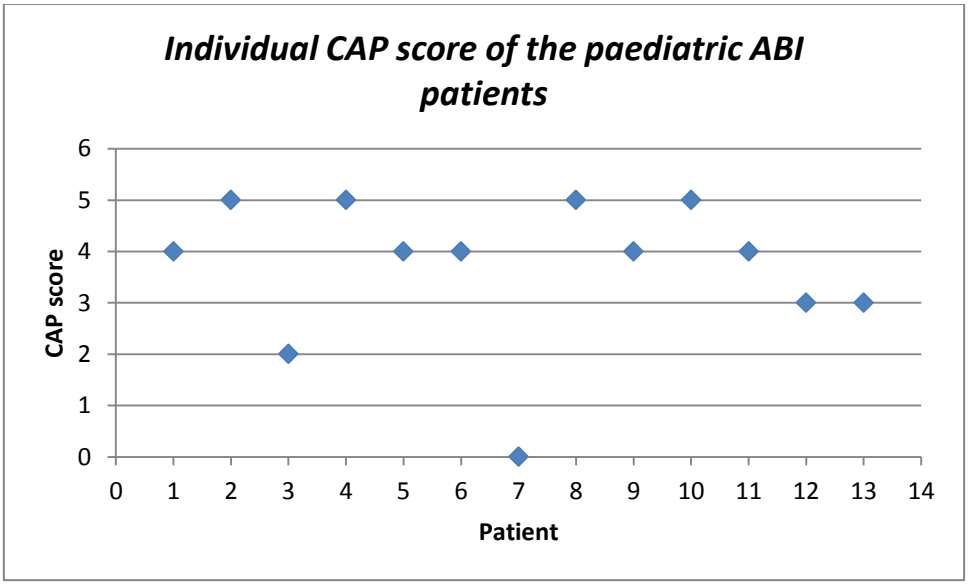
Category	Category notes
1	Speech like sounds, pre-linguistic babble. Open vowels- canonical babble CVCVC. Some children may be at the stage of pre- speech and use a range of blows raspberries, lip smacking, blowing laughing, sighing etc. May be very difficult to transcribe using English phonemes. The child's primary mode of everyday communication may be manual.
2	Where keyword is known, speech production errors are phonologically based e.g. 'shoe' would be pronounced as 'do' as opposed to a lateralised /Σ/ -> /K/ or theinitial consonant might be omitted. Analysis would include phoneme repertoire andphonological analysis (if appropriate). Useful descriptors would include the child's syllable structure and any use of syllable stress. Connected speech is either unintelligible or has not emerged.
3	Phonetic errors may include features of deaf speech e.g. ingressive, implosives or silent articulation. Speech production errors continue to be primarily phonological e.g. manner, place, voice. A more detailed

	phonological analysis is now achievable now that the child's phoneme repertoire has expanded. The burden of interpretation is placed on the adult who takes the lead in soliciting contextual information. Developmentally the child may not have reached a language level to conduct a conversation.
4	Speech production is more consistent across contexts/use of the same word. The listener can readily tap into the child's phonological system. Any phonetic errors present do not impact on intelligibility. Syllable structure should be relatively consistent. The child may use suprasegmental cues to aide conversation. Speech errors more in line with typical development than with deaf speech. Child is taking more of a lead in communication. The topic can now be changed without the adult losing the thread of the narrative. Fewer cues are required to support intelligibility.
5	Speech errors would be phonetic imprecisions if present. Some suprasegmental features could still be atypical but this does not affect intelligibility. Conversation is balanced between speakers. Topics readily changed

Figure 11: The 5 categories of the SIR

The average scores for our paediatric patients on both the CAP and the SIR can be seen below. Generally patients are able to discriminate some speech sounds without lip reading with their ABIs and have some speech although intelligibility may be poor.

a)



b)

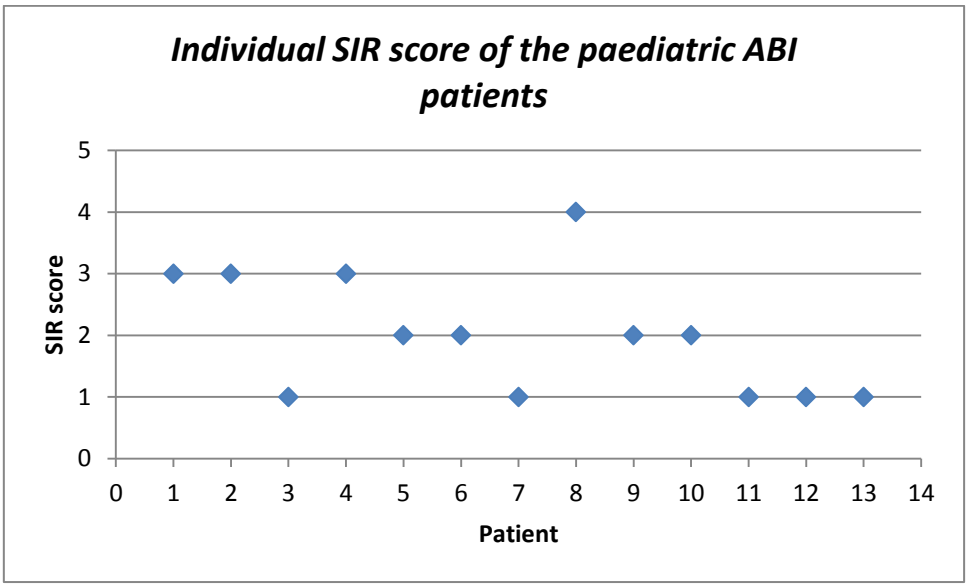


Figure 12 a and b: Individual CAP and SIR scores of the paediatric ABI patients at their last review