Cochlear implantation in asymmetrical hearing loss for children: Our experience

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Cochlear implantation in children with an asymmetrical hearing loss is now becoming more recognized as an appropriate and beneficial treatment option. In our programme, we have been implanting children with asymmetrical hearing loss since 2008.

Asymmetrical hearing loss is defined as hearing loss when one ear meets criteria for cochlear implantation and the other ear has better hearing and receives benefit from a hearing aid. There is a wide range of asymmetrical hearing loss: single-sided deafness is the end of the spectrum.

Amplification in asymmetrical hearing loss is usually difficult, it is often not possible to adequately aid the poorer ear; therefore patients often experience compromised hearing with adequate aiding of their better ear only. The impact of hearing with one ear on speech development, education, and employment is significant.

Keywords: Paediatric cochlear implantation, Asymmetrical hearing loss

Methods

We assessed 10 patients of whom 2 were excluded because their hearing loss progressed and they are now considered for sequential cochlear implantation. We discuss about eight patients with asymmetrical hearing loss who were unilaterally implanted at chronological ages of 14 months to 10 years (mean age 4.6 years). The aetiology of deafness was genetic, meningitic, progressive with unknown cause, and auditory neuropathy spectrum disorder (ANSD). The implants used were all cochlear devices and included Freedom, CI422 and System 5.

We used a standard assessment battery and a lengthy evaluation of functional benefit from conventional amplification through diagnostic therapy, in conjunction with the Auditory Speech Sounds Evaluation (ASSE) and other objective tests such as pure tone audiograms, automated toy test and localisation test. Outcomes were measured through a series of speech perception and speech and language assessments.

Results

Interdisciplinary team working is essential to ensure a robust clinical decision making process in these children. Ear specific functional and objective evaluation of hearing aid benefit and assessment of bilaterally aided hearing is crucial to draw a conclusion about the potential benefit of an implant versus hearing aids. Table 1 shows the study sample. Figs. 1 and 2 shows the pre- and post-implantation Categories of Auditory Performance II (CAPII) and Speech Intelligibility Rating (SIR) scores, respectively.

Patient 1

This child's hearing loss was first picked up through a targeted hearing screening. He was born premature at 25 weeks and his early medical history is complex. Initially, had clear otoacoustic emissions, which later disappeared. He was fitted with a hearing aid in the left ear at around 12 months of age, and in the right ear in May 2009, as part of the assessment for a cochlear implant. Auditory brainstem responses (ABR) results from Birmingham Children's Hospital in February 2009 showed no clear response to click stimulus in either ear, with a cochlear microphonic response present on the right and absent on the left. Repeat ABR in April 2009 confirmed ANSD in both ears.

The right-sided hearing aid was not found to be helpful, so following a trial, it was discontinued. He continued to wear a hearing aid on his better hearing left side but demonstrated very little functional benefit and was not able to develop spoken language despite reasonable aided levels on his left side. We felt that the ANSD was distorting the sound given by his left aid.

His parents opted to go ahead with a cochlear implant for his worst hearing ear – his right side – but there was a significant possibility that, following

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Table 1 Details of patients' age at implantation, age at assessment, hearing aids, aetiology, and hearing device

Patient	Implantation age	Aetiology	Assessment age	Special needs	Device	HAs
1	3 years	Prem, ANSD	2 years	Mild develop delay	Right System 5	12 months
2	14 months	Congenital	1 years	No	Left System 5	3 months
3	10 years	CMV	9 years	No	Right System 5	5 years
4	7 years	Meningitis	6 years	No	Right System 5	Only left
5	2 years	Mondini, WVAS	18 months	No	Right Freedom	3 months
6	5 years	Congenital	3 years	No	Left Freedom	8 months
7	3 years	Meningitis	3 years	Dyspraxia	Left Freedom	3 years
8	7 years	Idiopathic progressive	10 years	No	Right CI422	6 years

WVAS, Widened Vestibular Aquaduct Syndrome; CMV, cytomegalovirus; ANSD, auditory neuropathy spectrum disorder; HAs, Hearing aids.

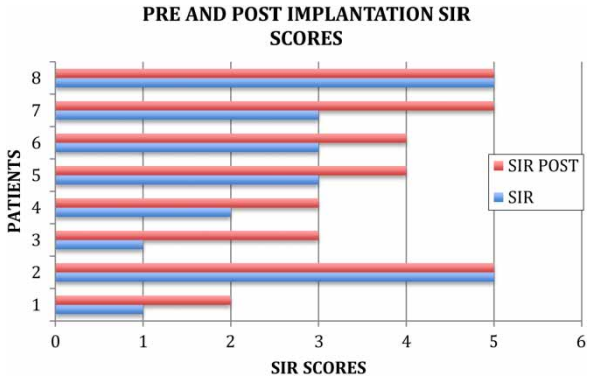


Figure 1 SIR scores pre- and post-implantation.

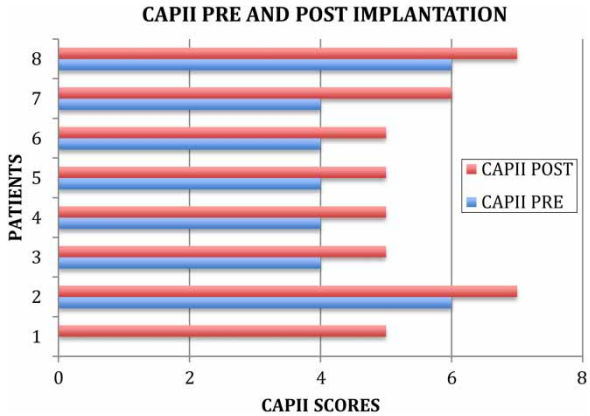


Figure 2 CAPII scores pre- and post-implantation.

a successful period of unilateral CI use, we may strongly recommend implanting the left ear too.

At 2 years post-implant, the child is wearing both devices consistently and reliably. He is able to follow routine instructions through audition alone and spoken language is slowly developing. Unfortunately, a change in family circumstances has resulted in high non-attendance rates at both our centre and at school, and this has hindered progress (audiogram in Table 2).

Patient 2

His hearing loss was first picked up through the neonatal hearing screening programme and he was fitted with hearing aids at 3 months. There is a family history of hearing loss. He has had some benefit from acoustic hearing aids so a prolonged assessment period was used to look at the likely increased benefit from a cochlear implant (audiogram in Table 2).

We explained that he has a progressive hearing loss, which is now severe to profound. He obtains some benefit from his hearing aids, but a prolonged hearing aid trial combined with therapy, has demonstrated that he is finding it increasingly difficult to learn language with his hearing aids.

We offered a left-sided cochlear implant. If his hearing loss continues to deteriorate, we will offer a right-sided implant at a later stage.

Patient 3

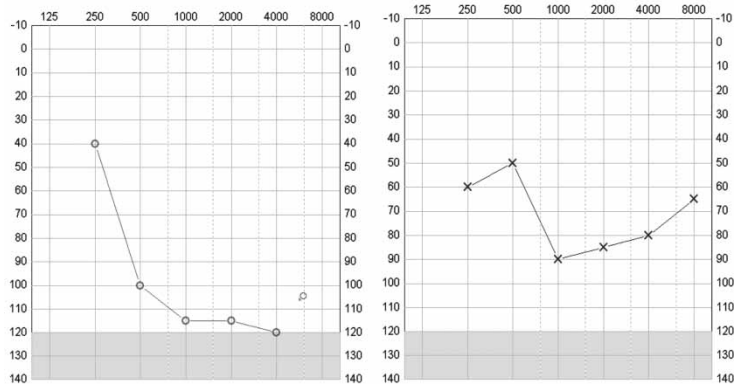
Cytomegalovirus was diagnosed at birth and he was treated with gancyclovir for 6 weeks. He has a progressive sensorineural hearing loss and as a result did not wear hearing aids until he was 5 years old. There has been a further deterioration in his hearing levels and he ceased wearing a hearing aid on his right side in July 2010. He has a moderate-to-severe sloping high-frequency sensorineural hearing loss on his left side for which he is appropriately aided (audiogram in Table 2).

The ASSE discrimination test showed that he was able to discriminate six of the eight speech sound pairs presented (could not discriminate m–z and v–z). The Automated Sentence Test (recorded voice) was completed to assess his speech reception threshold (SRT). This test is an automated version of the Bamford- Kowal-Bench (BKB) speech test which has an adaptive speech level and a fixed noise level of 55 dBA. The child has to repeat unfamiliar sentences through listening alone with and without the presence of background noise. In quiet, he was able to discriminate the sentences at a level of 45.3 dB.

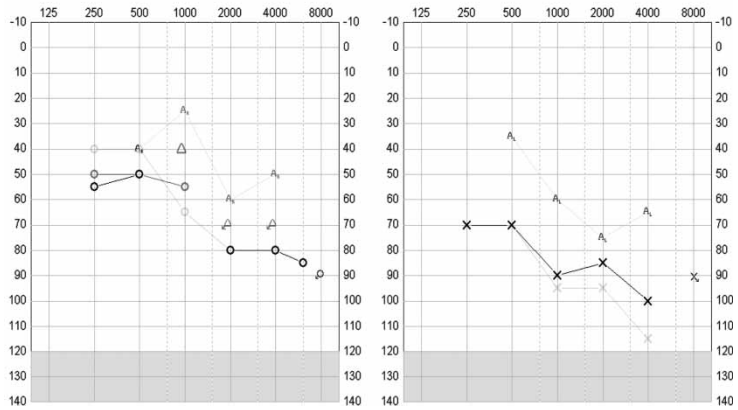
The SRT represents the signal-to-noise ratio at which the child could understand speech in noise. A lower score represents better performance. For example, the above child could repeat unfamiliar sentences when the background noise was only 0.3 dB

Table 2 Audiograms

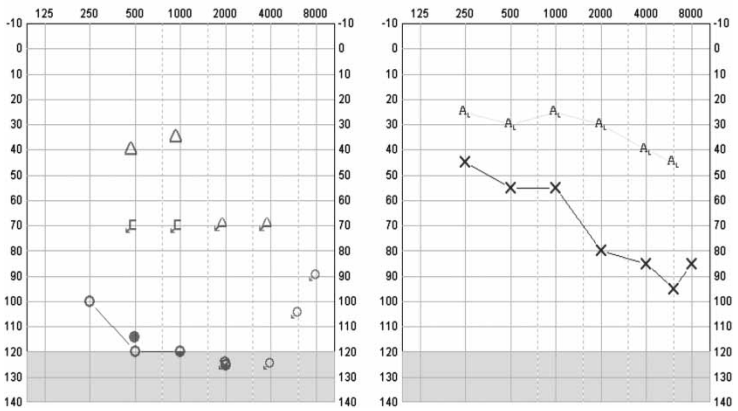
PATIENT 1



PATIENT 2



PATIENT 3



PATIENT 4

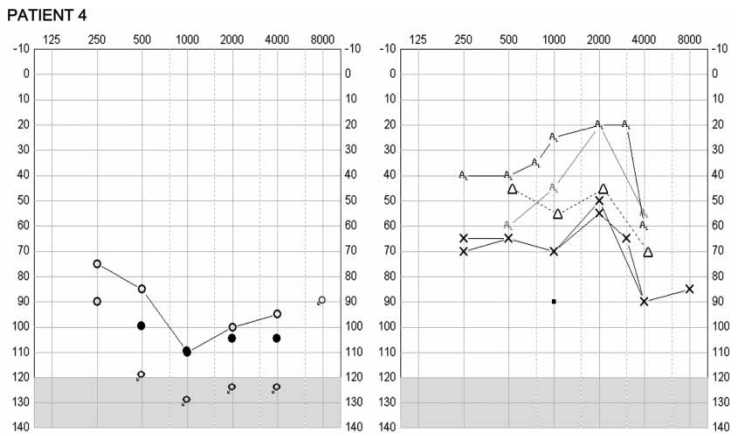


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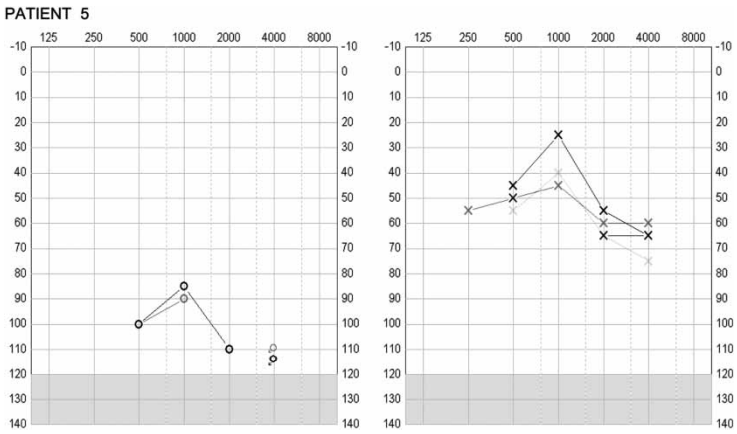
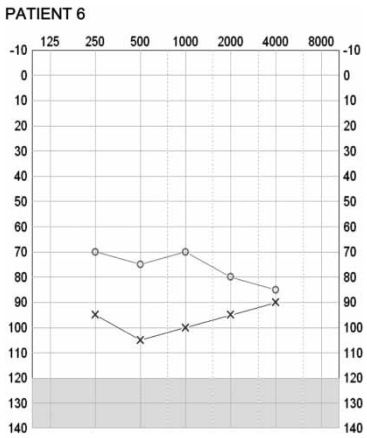
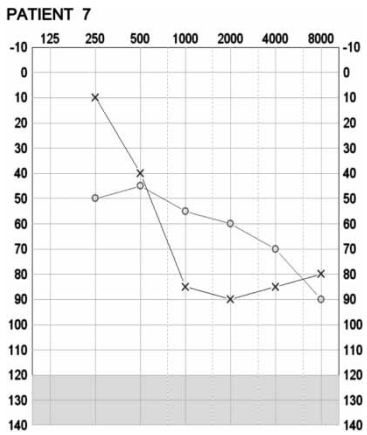
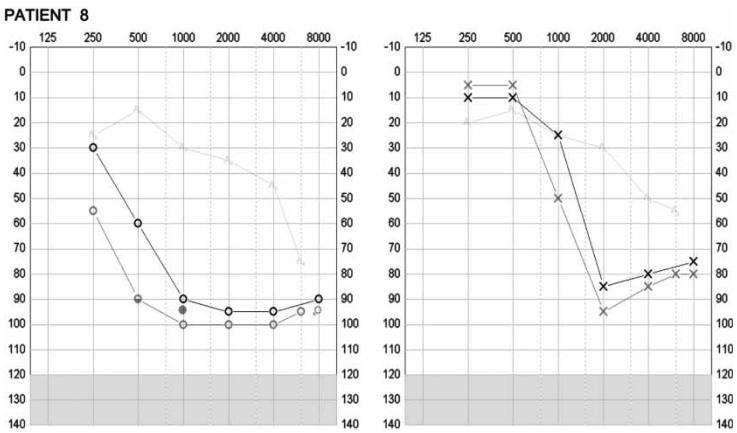
Table 2 ***Continued***









quieter than the speech, when presented from the centre speaker.

The localization test was completed to assess whether he was able to determine where a sound comes from in the best-aided condition. A person with hearing within normal limits can localize sounds to a radius of 2 degrees. Hence, the wider the angle reported the poorer the localization. He was able to localize sounds to 54 degrees (25 trials completed). CAPII rating scale = 6, SIR rating scale = 5.

In the funding request it was pointed out that as a committed hearing aid user with strong spoken language skills, he is in a position to gain considerable benefit from the use of a cochlear implant system. With appropriate support, there is every reason to hope that he will continue to fulfil his considerable potential. He has a progressive hearing loss and gains no functional benefit from his hearing aid in his right ear, and limited benefit in his left. He would benefit from a cochlear implant for his right ear.

Patient 4

He had meningitis at the age of 2 years, which resulted in a profound hearing loss in his right ear and a moderate–severe hearing loss in his left ear. He gained no benefit from a hearing aid in the right ear, and only wears a hearing aid in the left ear (audiogram in Table 2).

In a quiet, stable situation, he was able to process a range of spoken utterances within his language repertoire, but relied on lipreading to supplement information from his hearing aid. He has limited strategies for dealing with communication breakdown and in a noisy, busy setting such as a classroom, it is likely that his access to spoken information is severely compromised. His speech was intelligible in the controlled environment of the clinic, for short utterances but his family report that for longer and more complex utterances, it is often difficult for others to understand him. As the assessment result showed, his language attainment is severely delayed for his chronological age.

The overall picture that emerges is of a young boy struggling to learn and communicate with very limited access to sound. He will require consistent and sensitive support to enable him to adapt to new hearing technology, but it is likely that the enhanced access to sound provided by a cochlear implant system would be of benefit to him.

Patient 5

This was the first patient with asymmetrical sensorineural hearing loss that was implanted in our team, a 2-year old with progressive hearing loss. His elder brother had a cochlear implant and the team already knew the family. Both children had Mondini dysplasia and patent vestibular aqueducts. His audiology results are shown in Table 2.

His case triggered huge discussions within our team. His pre-implant assessment showed that he had severely delayed language abilities at this stage, and required urgent intervention to ensure that he fulfils his potential as a listening and speaking child who can access mainstream education (CAP2 = 4, SIR = 2).

Reporting from the mother suggested that some previously acquired spoken language knowledge has become lost or degraded in the past few months, which was consistent with a possible history of deterioration in his hearing levels. His receptive understanding was heavily dependent on additional contextual information, and his expressive vocabulary consisted of a small repertoire of single words and short phrases. He had made very little progress and was mainly at the babbling stage you would expect to see in a child of 6–9 months old (he was 2 and 1/2 years old at the time of the report). He was understandably frustrated and communication was becoming extremely challenging. The hearing aid on his left side alone was either not providing enough access to speech, or he was not able to interpret the information clearly enough. In our application for funding, we concluded that in our experience, an implant provides a clear and consistent signal and children make progress in cases such as these (audiogram in Table 2).

Patient 6

Her hearing loss was first identified through the neonatal hearing screening programme at 2 weeks of age. She was confirmed with a bilateral severe sensorineural hearing loss in April 2005 and fitted with hearing aids at 8 weeks of age. She has better hearing in her right ear. There is no family history of hearing loss (hearing results in Table 2).

Her assessment showed that her speech was intelligible most of the time but she could revert to babble when under pressure. Lack of use of the high-frequency sounds such as s/z in plural forms, possessives and tenses was evident as was the use of some simplifying processes still evident in her speech (SIR = $\frac{3}{4}$).

It is important to note that these results were obtained in quiet listening conditions. She would find group teaching and discussion far more challenging and it is reported she is very reliant on pre- and post-teaching by her support assistants. It was reported that in large group teaching situations she found it difficult to process the information and that she appeared to 'switch off' in these challenging situations.

Patient 7

His hearing loss was found in April 2006 following admission to hospital with meningo-enchephalitis and endophthalmitis. Auditory brainstem testing with an AC click demonstrated responses of 70 dB(nHL) on the left and 65 dB(nHL) on the

right. Testing afterwards confirmed a moderate sensorineural hearing loss. However, subsequent testing has shown some deterioration to a severe hearing loss. He also has dyspraxia (audiogram in Table 2).

His severe hearing loss was 'borderline' for our audiological criteria for a cochlear implant. However, he has had a trial of the most up to date hearing aids available and he was still unable to detect and make use of the higher-frequency speech sounds.

He became deaf through meningitis. Since this time his hearing loss had slowly deteriorated and at the time of funding application had a severe bilateral sensorineural progressive hearing loss. He was not making the progress we would normally expect with speech and language development and was not able to access the range of sounds that one would predict from his unaided audiogram. He has undergone a full hearing aid trial but any improvements have been negligible. Due to the progressive nature of his hearing loss, combined with the fact he is falling further and further behind with his communication and academic performance, we proposed to offer a unilateral cochlear implant, to be worn with a hearing aid in his better hearing ear.

Patient 8

A 10-year-old girl with a bilateral sensorineural hearing loss that was progressive in nature was also assessed. Our detailed pre-implant assessment concluded that her hearing was likely to continue to deteriorate. Her speech had recently lost clarity and she was now strongly reliant on lipreading to keep up with conversation. She was missing a significant amount of conversation and was visibly tired at the end of a session. As a result, she was struggling to keep up in school.

Her right ear was within the criteria for a cochlear implant and her left ear was outside of criteria. She would continue to lose speech clarity and fall behind with her language and academic development if we waited for her left ear to deteriorate further. Her audiology assessment is shown in Table 2.

Due to the progressive nature of her hearing loss, there is a strong possibility that she will need to have a second cochlear implant for her left ear in the future.

Discussion

Although this is a small series all our patients found cochlear implantation beneficial as shown in Figs. 1 and 2. In all funding requests for the above eight patients we pointed out that one ear is within criteria and that the other ear is out of criteria for cochlear implantation. The issue of implantation of patients with one ear that is within implant criteria and poor hearing in the contralateral ear that is not within implant criteria will be a topic under debate in future.

The far end of the spectrum of asymmetrical hearing loss is single-sided deafness. Historically, a Bone anchored hearing aid (BAHA) has been the implantable device recommended for single-sided deafness. However, it does not treat the deaf ear. There have been many recent studies (Steizig *et al*., 2011; Hassepass *et al*., 2013) that demonstrate the efficacy of using a cochlear implant to stimulate the deaf ear. A cochlear implant has a role in single-sided deafness but as with bilateral profound hearing loss in children, improved outcomes will be linked with early implantation. Such decisions will make cochlear implantation for single-sided deafness in children a challenge.

In partial deafness, decisions need to be made with focus on functional hearing as we have shown in our series. The situation needs to be explained in the funding application and clearly state that the one ear is out of criteria; however, the functional hearing is impaired and the child will not be able to progress with hearing aids. The direct link of deafness with brain neuroplasticity and speech development in the paediatric population should be emphasized in every funding application.

Conclusion

Asymmetrical hearing loss in children should not be overlooked. There are gradually more patients with asymmetrical hearing loss receiving cochlear implants worldwide (Steizig *et al.,* 2011; Hassepass *et al*., 2013). There are clear benefits of binaural and bimodal hearing and if this is combined with a hearing preservation technique the outcomes for children are improved.

Children with asymmetrical hearing loss should be assessed early. The aim is to see whether hearing aids are sufficient for speech and language development. If there is no improvement and hearing becomes increasingly difficult, it is clear that an implant will be extremely beneficial for the child. Improved outcomes are associated with early implantation in children as directly linked with brain neuroplasticity. Early assessment and early application for funding is recommended.

Acknowledgements

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