Cochlear implantation in a child with auditory neuropathy spectrum disorder

JI Fei CHEN Ai-ting HONG Meng-di SHI Wei LI Jia-nan YANG Shi-ming
Department of Otolaryngology/ Head and Neck Surgery, Chinese PLA Institute of Otolaryngology, Chinese PLA General Hospital, 28 Fuxing Road, Beijing 100853, China

Abstract Objective To report outcomes of cochlear implantation (CI) in a child with auditory neuropathy spectrum disorder (ANSD) and to provide preliminary clinical evidence of the efficacy of CI in ANSD patients. Methods A 4-year-old boy with diagnosed auditory neuropathy spectrum disorder (ANSD) received implantation of a Nucleus CI24R after an unsatisfactory trial of amplification. Post-implantation performance in both hearing sensitivity and speech recognition was assessed in different sessions. Aided hearing thresholds were tested by behavioral audiometry. Mandarin Early Speech Perception Test (MESP), Meaningful Auditory Integration Scale (MAIS), category of auditory performance (CAP) and Speech Intelligibility Rating (SIR) were used to assess the benefits in auditory skills or speech recognition the boy obtained from CI. The tests were administered before surgery and at 3 months and 7 months after opening. Results The boy demonstrated improved auditory sensitivity by using CI. Concerning speech recognition and communication, both speech audiometry and questionnaires showed an obvious benefit from CI. Conclusions CI has worked efficiently in this ANSD boy. But because of limited understanding of ANSD and rehabilitation effect by cochlear implantation in this condition, the clinical decision to implant should be cautious and only after a thorough evaluation. Meanwhile, well controlled and long term studies are needed to confirm the efficacy of cochlear implantation in patients with ANSD.

Key Words Auditory neuropathy spectrum disorder, Cochlear implantation

Introduction

The term auditory neuropathy spectrum disorder (ANSD) is recognized as an expansion from auditory neuropathy (AN), which was first proposed by Starr et al in 1996 and refers to a group of auditory diseases demonstrating intact outer hair cells and dys-synchronization of auditory neural activities. The incidence of ANSD is about 8% in newly diagnosed hearing-impaired children. Children with ANSD will be affected in their hearing and speech development, which holdback their future learning and social communication. So far, there is no specific medicine for this group of diseases. Clinical evidence in audiological management has showed very limited benefits from acoustic amplification. Cochlear implantation (CI) is now recognized as the most possible approach to help ANSD patients in improving their capability in hearing and speech communication. Because there are no reliable methods in clinical practice to identify the exact lesion site and mechanisms for a certain ANSD patient, the efficacy of CI is difficulty to predict before surgery. There are divaricating opinions on the efficacy of CI on ANSD patients, especially in children. Here we will introduce one implanted ANSD case, including pre-operative assessment and a series of post-operative efficacy evaluations, in order to provide preliminary clinical evidence on the efficacy of CI in ANSD patients.

Materials and Methods

The patient

The patient was a 4 years old boy with a history of neonatorum asphyxia. The parents recognized poor
response to environment sounds when the child was 1 year and 2 months old. The initial diagnosis was bilateral profound sensorineural hearing loss and a pair of high power Phonak hearing aids were fitted. However, the boy made little progress in hearing and language with hearing aids despite speech training.

**Audiological assessment and diagnosis of ANSD**

The 4-year-old boy was able to cooperate on behavioral audiometry well and reliable pure tone thresholds as well as aided thresholds from both ears were obtained (Figure 1–A). His audiogram showed profound sensorineural hearing loss with 3FA (average threshold at 0.5k, 1.0k, 2.0k Hz) of more than 100 dB HL in both ears. Aided 3FA was around 40 dB HL with insufficient gains at frequencies over 2 kHz. Bilaterally aided close-set disyllable recognition score in sound field was 64%. Despite the fairly good aided performance in disyllable recognition, the parents complained of abnormal speech development and difficult speech communication with their son.

![Figure 1 Hearing thresholds at different sessions.](image)

Electrophysiological tests and other objective test results are shown in table 1. ABRs were absent in both ears at the highest click stimulation intensity of 100 dB nHL. To evaluate outer hair cell function, otoacoustic emissions (OAE) and cochlear microphonics (CMs) were measured. Although neither TEOAE or DPOAE were present, CMs were obtained from both ears on subtraction (Figure 2) on an ABR test. CMs were elicited using rarefaction and condensation clicks respectively. Rarefaction responses were subtracted from condensation responses to set off possible disturbing components, and averaged CMs of relatively large amplitudes were identified at intensities of over 75 dB nHL. An intensity–amplitude CM function was measured from the right ear over an intensity range of 75 to 100 dB nHL with a step size of 10 dB, which demonstrated an obvious non-linear curve (Figure 3), providing evidence of intact function of outer hair cells.

<table>
<thead>
<tr>
<th>Ear</th>
<th>ABR</th>
<th>OAE</th>
<th>CM</th>
<th>ASSR</th>
<th>500Hz</th>
<th>1000Hz</th>
<th>2000Hz</th>
<th>4000Hz</th>
<th>Acoustic Reflex</th>
<th>Image</th>
</tr>
</thead>
<tbody>
<tr>
<td>L</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>74</td>
<td>89</td>
<td>77</td>
<td>71</td>
<td>100</td>
<td>A</td>
<td>-</td>
</tr>
<tr>
<td>R</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>74</td>
<td>99</td>
<td>87</td>
<td>71</td>
<td>110</td>
<td>A</td>
<td>-</td>
</tr>
</tbody>
</table>

**Table 1** Electrophysiological tests, tympanometry, acoustic reflex test, and image test results
Figure 2  Waveforms of Cochlear Microphonics (CM) in the implanted ear. The waveform on the top (A) is an ABR waveform elicited by alternating clicks showing no neural activity. The second waveform (R) is elicited by rarefaction clicks and the third one (C) by condensation clicks. Phase-inversed CMs are seen in the initial 2 milliseconds in R and C. The bottom one (C–R)/2 is obtained by subtracting the R from C to set off the possible disturbing components. The red arrowhead marks CMs.

Figure 3  Intensity – Amplitude functions of CMs in right ear

Tympanogram was tested using a GSI Tympstar mid-ear analyzer with a 256 Hz probe tone. Acoustic reflex was tested at 500 Hz, 1 kHz, 2 kHz and 4 kHz. A-type tympanograms were shown in both ears. MRI and CT showed normal structures of temporal bone and auditory nerve development. These clinical data supported the diagnosis of ANSD in this 4-year-old boy.
Implantation surgery

The boy received a CI24RCA implant (Cochlear Inc.) in the right ear under general anesthesia. The electrodes array was placed in an appropriate position in the scala tympani (Figure 4).

Figure 4  X-ray of implanted electric array in right cochlea.

Hearing and speech ability evaluation

To assess the efficacy of CI, hearing and speech evaluations were done at opening session and at 3 and 7 months after opening. Evaluation included aided play audiometry, meaningful auditory integration scales (MAIS), Mandarin early speech perception test (MESP), categories of auditory performance (CAP) questionnaire and speech intelligibility rating (SIR).

MAIS was completed with the parents regarding their child’s hearing capabilities. Each item was scored on a 0 to 4 scale using information provided by the parents following instructions from the tester. Three MAIS scores in three categories (Total, Detection, and Perception) were analyzed using custom software with each score expressed as the percent of the total possible points. There’re a total of 10 items. The Detection score is for items 3–6; and Perception score is for items. 7-10

The MESP test, developed by Zheng Y et al. and based on the English Early Speech Perception test 9,10, is a closed–set assessment tool for evaluation of early speech perception abilities in children. The MESP is part of a Mandarin hierarchical test battery for assessment of speech perception in children as young as 2 yr of age. MESP includes 6 subtests, each of which measures a different category of early speech perception. The categories 1, 2 and 3 measure Speech Sound Detection, Speech Pattern Perception, and Spondee Recognition, while categories 4, 5, and 6 measure mandarin Vowel Perception, Consonant Perception, and Tone Perception.

CAP is an eight–point hierarchical scale to assess children’s hearing performance in their everyday lives with high reliability even when administered by different observers. Table 2 shows the scaling criteria, with categories from 1 to 8 modified from the original scaling.

<table>
<thead>
<tr>
<th>Category</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>Use of telephone with known listener</td>
</tr>
<tr>
<td>7</td>
<td>Understanding of conversation without lip-reading</td>
</tr>
<tr>
<td>6</td>
<td>Understanding of common phrases without lip-reading</td>
</tr>
<tr>
<td>5</td>
<td>Discrimination of some speech sounds without lip-reading</td>
</tr>
<tr>
<td>4</td>
<td>Identification of environmental sounds</td>
</tr>
<tr>
<td>3</td>
<td>Response to speech sounds</td>
</tr>
<tr>
<td>2</td>
<td>Awareness of environmental sounds</td>
</tr>
<tr>
<td>1</td>
<td>No awareness of environmental sounds</td>
</tr>
</tbody>
</table>

Table 2  Categories of Auditory Performance (CAP)
SIR is used to assess speech production in pediatric cochlear implantation. It scales speech intelligibility of children in 5 categories according to the criteria in table 3. Both CAP and SIR were scaled by the parents under the instruction of an audiologist in this case.

<table>
<thead>
<tr>
<th>Category</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>Connected speech is intelligible to all listeners. Child is understood easily in everyday contexts</td>
</tr>
<tr>
<td>4</td>
<td>Connected speech is intelligible to a listener who has little experience of a deaf person’s speech.</td>
</tr>
<tr>
<td>3</td>
<td>Connected speech is intelligible to a listener who concentrates and lip-readings</td>
</tr>
<tr>
<td>2</td>
<td>Connected speech is intelligible. Intelligible speech is developing in single words when context and lip-reading cues are available.</td>
</tr>
<tr>
<td>1</td>
<td>Connected speech is unintelligible. Pre-recognizable words in spoken language, primary mode of communication may be manual.</td>
</tr>
</tbody>
</table>

Results

Improvement in audibility and speech with CI

The boy had used CI regularly for 7 months by the time these data were reported. He had been sent to a speech rehabilitating center for listening and speech training for hearing loss children, while his parents took an active part in his rehabilitation. By now, the boy has made obvious progress in audibility and speech with the using of CI.

Aided hearing thresholds at each frequency before implantation, 3 months after opening, and 7 months after opening are shown in Figure 1–A. Profound sensorineural hearing loss of more than 100 dB HL was found with 3FA in both ears before implantation while aided 3FA was around 40 dB HL with insufficient gains at frequencies over 2 kHz. CI aided thresholds in right ear reached an average around 30 dB HL after surgery, and those with hearing aid in left ear remained the same as before operation (Figure 1–B). Figure 1–C shows a stable hearing thresholds pattern of both naked ears and CI aided thresholds in right ear at 7 months post opening. It demonstrates sufficient hearing sensitivity provided by CI to this boy.

Figure 5 demonstrates a detail change in the boy’s MAIS scores on different test sessions. A progressive tendency can be seen in scores of three MAIS categories, i.e. Detection, Perception, and Total. The sound detection ability score increased from 38% with hearing aids before implantation to 81% at 7 months post opening the CI device. The perception ability increased from 44% to 81%, and the total score also showed a stepwise progress from 48% to 83% during the period of using CI.
Concerning the child’s speech recognition and speech production, MESP, CAP and SIR were administered at 3 months and 7 months after opening of the CI, whose results are shown in Figure 6. MESP test demonstrated a large increment in speech recognition. At 3 months post opening, the boy reached a level of recognizing spondees, and at 7 months he passed the top category of tone perception, which allowed him to recognize some simple sentences in noise condition. Similarly, CAP scaling improved from 5 at 3 months to 6 at 7 months. SIR levels was relatively low, but also improved from 2 to 3, indicating that connected speech the boy produced was intelligible to a listener who was focused and used lip–reading.
Parents’ narration about the progress of the child

Detail performance of hearing and speech of the child in everyday life was reported by the parents during the interview, which was a complement to standard tests.

After using CI for 3 months, the boy could respond to simple sentences within a few words or some nominal phrases, and could act correctly to short imperative sentences such as Come here, Sit down, as so on. The boy could also respond to simple selective questions such as Do you like it? by nodding or shaking his head. He could not understand longer sentences at 3 months session. At 7 months after opening the device, the boy could respond to imperative sentences with more complex structures, such as go and fetch something, or put something somewhere, and then take it to some place. The child could make correct selection according to the demand from up to 6 nominal options.

With regard to speaking, the boy’s reading ability improved on CI at 3 months, with enlarged glossary compared with pre-operation. But his pronunciation was relatively poor especially for consonants of g, k, h, sh, s. Consecutive sentences were mostly less than three words, and signs were needed frequently at 3 months session. At 7 months after opening the device, the intelligibility of the child’s speech had improved so as to parents and teachers could understand his meaning by longer sentences of five to six words without signs. People who were not familiar with his pronunciations could feel the progress in his speech intelligibility.

Discussion

The diagnosis of ansd

Before audiological management is administered in a patient, an accurate and reliable diagnosis is required, especially for children who may have ANSD. The term ANSD is now recognized in a broad sense as referring to a group of auditory diseases demonstrating intact outer hair cells and dys–synchronization of auditory neural activities. In clinical practice, two necessary evidences must be confirmed for a definite diagnosis of ANSD. One is the abnormality or absence of ABR which demonstrates dys–synchronization of neural activities, and the other is presence of OAEs or CMs which demonstrates normal function of outer hair cells.

There was an uncertainty in diagnosis of this case in that the child’s OAE was absent while the absence of ABR at high intensity was consistent of profound hearing loss, which led to a diagnosis of sensorineural hearing loss. On further investigation, CMs were recorded on ABR testing using rarefaction and condensation clicks, as well as intensity–amplitude function. The presence of CMs with large amplitudes recorded from tympanic membrane electrodes indicated presence of hair cell function. Meanwhile, CMs intensity–amplitude function expressed a non–linearity. Brownell has proved that the nonlinearity of CMs is decided by OHC. The above-mentioned evidence indicates that OHC function was normal, and the site–of–lesion might be inner hair cells, the synapses between IHCs and the VIIth nerve dendrite, or the VIIth nerve. Another piece of clinical evidence that may help support the diagnosis is the ASSR threshold (Table 1). Previous literature regarding ASSR in assessing hearing thresholds indicated that ASSR thresholds show good consistency with pure tone thresholds on behavioral audiometry in children with severe to profound sensorineural hearing loss, while those obtained in ANSD children fit poorly with pure tone thresholds. As far as this case is concerned, ASSR thresholds at frequencies from 0.5 kHz to 4.0 kHz were better than those obtained by play audiometry by around 20 dB, which is not typical in SNHL children. These analyses facilitate the diagnosis of ANSD.

The efficacy of implantation in ANSD children

The toughest challenge in cochlear implantation in ANSD patients is to predict the efficacy. Several studies have reported the degree to which the device works in such patients, but various or even contrary conclusions have been drawn with different research protocols or methods. Several recent studies have declared that CI may work in most ANSD children. But a question remains: by what criteria can we declare CI is working in an ANSD patient? In a review by Jeffrey Simmons in 2009, the cited data demonstrated that about 88% of ANSD patients who received CI implantation benefited from the device while 12% of the patients were implanted in vain. Four criteria have been used to determine the device’s efficacy in these reports, including: 1) the difference in speech recognition performance after
Implantation compared with patients with SNHL (accounted for about 47% in the so-called effective group); 2) speech recognition performance improvement after implantation compared to before implantation (accounted for about 39%); 3) hearing threshold improvement upon using CI (about 6%); and 4) presence of EABR waveforms after implantation. In fact, speech recognition is the main complaint in ANSD patients and the most challenging disorder for language development in children. EABR and aided threshold thus are probably not appropriate standards to judge the efficacy of CI in ANSD children. Excluding the last two groups of patients, the proportion of improved cases accounts for about 75.7%, which is close to the figure from a large-sample research reported by Gibson et al. 17

In clinical practice, how can we decide if CI is beneficial for ANSD patients, especially pre-lingual children? In this case, several assessment methods were used on the 4-year-old ANSD child, including questionnaires and speech test for children. The results proved that sufficient and stable hearing sensitivity was provided by CI to this boy. MAIS scores in categories of Detection, Perception, and Total all showed significant improvement 7 months after opening the device. MESP test demonstrated a large increment in speech recognition from a level of recognizing spondees at the session of 3 months post opening to the top category of tone perception at 7 months. CAP scaling was improved from 5 to 6, and SIR levels improved from 2 to 3, which meant that both audibility and speech intelligibility were improved obviously by using CI for about 7 months. Besides standard test procedures and questionnaires, parent observation was considered to play an important role in the assessment of the young child. In the detail narration and contentment from the child’s parents, a definite progress was confirmed, which provided an important qualitative assessment of the efficacy of CI in this pre-lingual child.

**The decision of implantation**

According to previous reports of CI implantation in ANSD children, the device may work in some of the children (about 75%), but what is hidden behind the figure is the uncertain site of lesion and disorder mechanism in a certain individual. To date, no effective clinical measurement is able to provide the exact site of deterioration of ANSD from the possible sites of inner hair cells, synapse, and auditory nerve, 3 preventing definite prediction of benefit of implantation for a particular individual. This fact has led to a practice question: how to make a clinical decision for an ANSD patient (especially a pre-lingual child) regarding a CI implantation.

As far as this child was concerned, the decision to implant was made by both the audiologist and parents in view of three basic facts: 1) Before implantation, the child had an experience of using hearing aids on both ears and accumulated some listening skills which helped him reach to a relatively high level of aided hearing sensitivity and speech performance, indicating that audiological interference would likely help this child in his hearing and speaking; 2) The child benefited from hearing aids in speech recognition, which is the main disorder in ANSD patients, indicating a relatively better basis in his speech recognition performance, providing an optimistic signal to post operation efficacy with CI; 3) The parents of this boy showed a reasonable expectation to implantation while taking active part in his rehabilitation both before and after implantation surgery. The participation of parents has been proved to be a very important factor in the rehabilitation of children using CI. 22

In conclusion, this 4-year-old boy diagnosed with ANSD has benefit from cochlear implantation in both hearing sensitivity and speech recognition performance after a period of 7 months of using the CI. This case provides preliminary clinical evidence to the efficacy of CI in ANSD patients. This relatively successful case provides us with some key points in making a clinical decision regarding implanting children with ANSD, including hearing aids experience, relative better speech recognition basis before implantation, and active participation of parents in the whole rehabilitation process. However, it should be recognized that this conclusion is neither the aim nor the end of the work. For cochlear implantation in ANSD children, much more work especially well controlled and long term research is needed to identify site of lesion, predict and assess treatment efficacy, and help make clinical decision.
Financial funds

This work was supported by grants from the National Basic Research Program of China (973 Program) (#2012CB967900; 2011CBA01000), National Hi-Tech Research and Development Program of China (863 Program) (#2007AA02Z150) to SMY, the National Natural Science Foundation of China (NSFC) (#30730040) to Yang Shi-ming.

References

13 Brownell WE. Outer hair cell electromotility and otoacoustic emissions. Ear Hear 1990; 11:82.

(Received November 21, 2011)