B-ENT, 2014, **10**, 265-269

Evaluation of cochlear implantation in children with inner ear malformation

H. Zhou[1], *X. Sun*[2], *Z. Chen*[1], *H. Shi*[1], *Y. Wu*[1], *W. Zhang*[1] *and S. Yin*[1]

[1]Department of Otolaryngology, Affiliated Sixth People's Hospital to Shanghai Jiaotong University, Shanghai, China; Otolaryngology Institute, Shanghai Jiao Tong University, Shanghai, China; [2]Department of Otolaryngology, Affiliated hospital of Luzhou medical college, Sichuan 646000, China

Key-words. Cochlear implantation; inner ear malformation; Categories of Auditory Performance; Speech Intelligibility Rating

Abstract. *Evaluation of cochlear implantation in children with inner ear malformation*. *Objective*: This study aimed to compare the outcomes of cochlear implantation (CI) in children with malformed versus normal inner ear anatomy.
Methods: We assessed 63 children with prelingual deafness, including 12 with inner ear malformations. All had undergone CI before the age of 5 y. We evaluated Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores before surgery and at 6, 12, and 24 months after surgery.
Results: In both groups, the CAP and SIR scores increased with time after implantation in follow-up assessments. No significant differences were found in the CAP or SIR scores between the two groups at any of the four follow-up assessments.
Conclusion: Children with inner ear malformation can benefit from CI. Although additional factors may influence the outcome of CI in children with inner ear malformations compared to children with deafness from other causes, early implantation may provide similar results.

Introduction

Cochlear implantation (CI) has an established role in the treatment of profound hearing impairments. Due to changes in candidacy for CI, there has been a significant increase in CI procedures since the first multichannel devices were implanted in the late 1970s. Accordingly, more children with anomalous inner ear anatomy are being considered as candidates for CI. Approximately 20-30% of patients with profound sensorineural hearing loss show inner ear anomalies on temporal bone computed tomography.[1,2] Furthermore, this population is increasing, based on the recent upward trend in our series and in cases reported in the literature.[3] Thus, the success of CI in this group of patients is of special interest.

Although several clinics have reported that CI provided a worthwhile benefit in children with inner ear malformations,[4,5] the results have been highly variable and complicated. Many factors that might affect the results of CI have been proposed.[6] However, it remains a challenge to define these factors and determine how to modify them to increase the efficacy of CI in patients with malformed inner ears. In this study, we monitored the development of auditory performance and speech intelligibility after CI in children under 5 years old. We compared findings in children with normal anatomy to those in children with inner ear malformation.

Methods

This study included 63 children with congenital deafness that had undergone CI before the age of 5 years. The CI had been performed at the Sixth Hospital Affiliated to Shanghai Jiaotong University between November 2006 and December 2007. Children were excluded when they had severe mental retardation or an absent or hypoplastic cochlear nerve, as demonstrated by MRI. Of the 63 children studied, 51 had normal inner ear

Zhou and Sun contribute equally to this paper.

anatomy, and 12 had malformations of the inner ear. The malformations included a common cavity (n = 2), a common cavity with an enlarged vestibular aqueduct (n = 2), an incomplete partition (type II; n = 3), cochlear hypoplasia (n = 4), and hypoplasia with an enlarged vestibular aqueduct (n = 1). During surgery, among the children with malformations, a full complement of active electrodes (24 electrode) was inserted in eight patients, and the remaining four patients received partial insertions (one received 22 electrodes and three received 20 electrodes). None of the patients enrolled in this study had worn a hearing aid before or after surgery. The communication mode for these patients was solely or mainly oral communication. All children were assessed before surgery, and at 6, 12, and 24 months after surgery with the Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores, administered by a speech and language therapist. All children received the MED-EL COMBI 40+ cochlear implant, and the speech processing strategy was continuous interleaved sampling.

At the time of the CI, the children's ages ranged from 10 to 58 months (mean ± SD, 25.1 ± 14.7 months). The children with normal anatomy (controls) were aged 10.5 to 58 months (26.4 ± 15.2 months), and the children with inner ear malformations were aged 10 to 58 months (24.6 ± 13.9 months). The difference in age between the two groups was not significant (Student's *t*-test, $P > 0.05$).

The CAP scale was used to evaluate speech perception in the children after implantation. CAP is an outcome assessment of auditory receptive capabilities. It comprises a hierarchical scale of auditory perception, ranging from 0 (displays no awareness of environmental sounds) to 7 (can communicate by telephone with a familiar person). The SIR scale was used to evaluate speech intelligibility in the children after implantation, by quantifying everyday spontaneous speech. The SIR rating scale consists of five speech performance categories, ranging from 1 (connected speech is unintelligible; pre-recognizable words in spoken language) to 5 (connected speech is intelligible to all listeners).

The CAP and SIR scores for the two groups were determined before surgery, and at 6, 12, and 24 months after CI. Each assessment was administered by the child's speech therapist in a familiar environment; either the child's home or school.

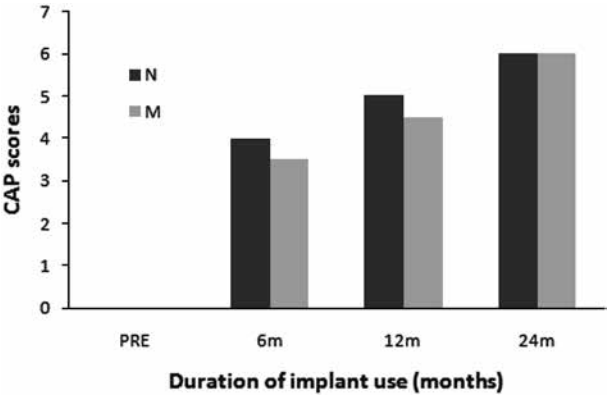

Figure 1
Median CAP scores increased after the time of cochlear implant. CAP scores did not differ significantly between groups at any time point (Mann-Whitney rank-sum test, $P > 0.05$). N: no inner ear malformation; M: with inner ear malformation; CAP: Category of Auditory Performance.

The CAP and SIR scores assessed at each time point were compared between groups with the Mann-Whitney rank-sum test. The two groups were also compared in terms of the percentage of children that achieved level 3 or higher scores (both CAP and SIR) at each of the four time points; these comparisons were performed with the Fisher Exact Test.

Results

Comparison of CAP scores between the two groups

The median CAP scores for the two groups are shown in Figure 1. CAP scores in both groups increased with time after implantation in follow-up assessments. The median CAP score for the control group was 0 before implantation, and it increased to 4, 5, and 6 at 6, 12, and 24 months after CI, respectively. Similarly, the median CAP score for the malformation group increased from 0 before implantation to 3.5, 4.5, and 6 at 6, 12, and 24 months after implantation, respectively. The median CAP scores did not differ significantly between the two groups at any time point (Mann-Whitney rank-sum test, $P > 0.05$).

The percentages of children with CAP scores at level 3 or higher before and at 6, 12, and 24 months after CI were not significantly different between the two groups at any time point (Chi-squared test, $P > 0.05$). In the control group, the percentage

Table 1
Distribution of patients in CAP categories before and after cochlear implantation

CAP	Before		6 months		12 months		24 months	
Category	N	M	N	M	N	M	N	M
0	44 (86.3)	11 (91.7)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
1	5 (9.8)	1 (8.3)	3 (5.9)	1 (8.3)	2 (3.9)	0 (0)	0 (0)	0 (0)
2	2 (3.9)	0 (0)	12 (23.5)	3 (25.0)	4 (7.8)	2 (16.7)	0 (0)	0 (0)
3	0 (0)	0 (0)	9 (17.6)	2 (16.7)	2 (3.9)	0 (0)	3 (5.9)	1 (8.3)
4	0 (0)	0 (0)	11 (21.6)	2 (16.7)	5 (9.8)	4 (33.3)	2 (3.9)	1 (8.3)
5	0 (0)	0 (0)	14 (27.5)	3 (25.0)	13 (25.5)	1 (8.3)	7 (13.7)	1 (8.3)
6	0 (0)	0 (0)	2 (3.9)	0 (0)	21 (41.2)	4 (33.3)	24 (47.1)	5 (41.7)
7	0 (0)	0 (0)	0 (0)	1 (8.3)	4 (7.8)	1 (8.3)	15 (29.4)	4 (33.3)
Total	51	12	51	12	51	12	51	12

Values represent the numbers of patients (%), unless indicated otherwise. CAP: Categories of Auditory Performance; N: no malformation (control group); M: with malformation (test group).

of children at level 3 or higher increased from 0 before surgery to 70.6, 88.2, and 100% at 6, 12, and 24 months after implantation, respectively. Similarly, in the malformation group, the percentage increased from 0 before surgery to 66.7, 83.3, and 100% at 6, 12, and 24 months after implantation, respectively (Table 1).

Comparison of SIR scores between the two groups

The increase in median SIR scores was similar to that in the CAP scores in both groups (Figure 2), and no significant difference was observed between groups at any time point (Mann-Whitney rank-sum test, $P>0.05$). In the control group, the median SIR score was 1 before CI, and it increased to 2, 4, and 4 at 6, 12, and 24 months after implantation, respectively. In the malformation group, the median SIR score increased from 1 before implantation to 2, 3.5, and 4 at 6, 12, and 24 months after implantation, respectively.

Based on a previous study, intelligible speech was defined as a SIR score of 3 or more.[7] The percentages of children with SIR scores of 3, 4, and 5 before and at 6, 12, and 24 months after CI were not significantly different between groups (Chi-squared test, $P>0.05$). In the control group, the percentage increased from 0 before surgery to 43.1, 72.5, and 94.1% at 6, 12, and 24 months after CI, respectively. In the malformation group, the percentages were 0 before surgery and 41.7, 66.7, and 91.7% at 6, 12, and 24 months after CI, respectively (Table 2).

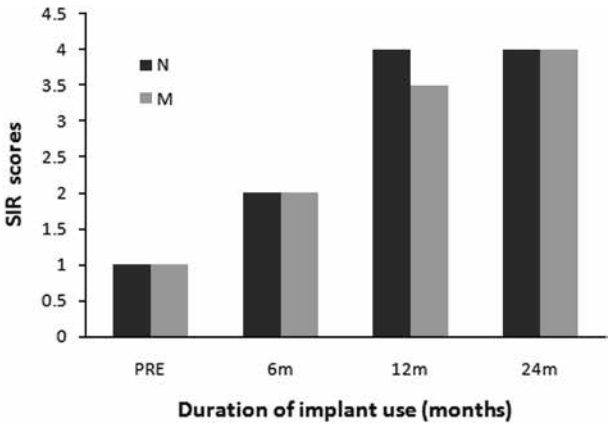

Figure 2
Median SIR scores increased after the time of cochlear implant. SIR scores did not differ significantly between the two groups at any time point (Mann-Whitney rank-sum test, $P>0.05$). N: no inner ear malformation; M: with inner ear malformation; SIR: Speech Intelligibility Rating.

Discussion

In the present study, children under 5 years old with inner ear malformations had CI outcomes similar to those for children with normal inner ear anatomy, based on scores of speech perception and speech intelligibility. The CAP and SIR scores increased after surgery in a similar manner in both groups of children.

Previous studies of CI in children with inner ear malformation have reported inconsistent results. Some studies demonstrated good results, comparable to those in patients with normal inner ear

Table 2
Distribution of patients with SIR scores before and after cochlear implantation

SIR	Before		6 months		12 months		24 months	
Category	N	M	N	M	N	M	N	M
1	47 (92.2)	11 (91.7)	11 (21.6)	3 (25.0)	3 (5.9)	1 (8.3)	0 (0)	0 (0)
2	4 (7.8)	1 (8.3)	18 (35.3)	4 (33.3)	11 (21.6)	3 (25.0)	3 (5.9)	1 (8.3)
3	0 (0)	0 (0)	10 (19.6)	3 (25.0)	6 (11.8)	2 (16.7)	2 (3.9)	1 (8.3)
4	0 (0)	0 (0)	10 (19.6)	1 (8.3)	24 (47.1)	5 (41.7)	21 (41.2)	5 (41.7)
5	0 (0)	0 (0)	2 (3.9)	1 (8.3)	7 (13.7)	1 (8.3)	25 (49.0)	5 (41.7)
Total	51	12	51	12	51	12	51	12

Values represent the numbers of patients (%), unless indicated otherwise. SIR: Speech Intelligibility Rating; N: no malformation (control group); M: with malformation (test group).

anatomy.[8,9] However, in other studies, the results were variable and less certain.[10,11]

One factor thought to affect the results of CI in children with inner ear malformation is the number of spiral ganglion cells in the brain. Spiral ganglion cells are the primary neural elements in auditory stimulation.[12] Thus, many authors believe that cochlear implants may have limited benefits in patients with reduced spiral ganglion cell numbers. However, a case study on temporal bone histopathology demonstrated that, despite significant degeneration of spiral ganglion cells (1,469 residual cells), the patient achieved a word recognition score of 28% by 6 months after CI.[13] Furthermore, a retrospective review concluded that the number of residual spiral ganglion cells is unlikely to determine the level of word recognition achieved after CI.[14]

Inner ear malformation often involves concomitant hypoplastic cochlear nerves. For children with a compromised neural connection between the cochlea and the brain stem, there may be no response to input from the implant. It has been suggested that patients with internal auditory canal narrowing, suggestive of eighth nerve aplasia or hypoplasia, should be considered poor candidates for CI. Several studies have demonstrated poor CI results in patients with hypoplastic cochlear nerves.[15,16]

The factors that determine the results of CI in children with inner ear malformations are not clear. Many variables, including age at implantation, duration of deafness, mode of communication, and preoperative speech perception, might influence the results of CI in children with either normal or abnormal inner ear formations. However, all studies agreed that earlier implantations produced better outcomes.[17]

The good outcomes in this study may have been due to the fact that implantations were performed earlier in development compared to other studies. Although children younger than 5 years were evaluated in previous studies, the mean age was typically older than 4 to 5 years, and patients with prelingual and postlingual deafness were typically combined for analyses.[18,19] Because the cortical adaptations to a CI decrease with increasing implantation age, early implantation has been advocated to minimize secondary degeneration of ascending auditory pathways and to promote central auditory maturation during potentially critical periods in development.[20,21] The post-implantation outcome was also shown to be directly dependent on the severity of developmental delay, due to the low cortical processing ability of children with developmental delays.[22]

Our results showed that children with inner ear malformations can benefit from CI. Although additional factors may influence the outcome of CI in children with inner ear malformations, we found that early implantation provided results similar to those achieved in children with deafness due to other causes.

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Zhengnong Chen, M.D., Ph.D.
Otolaryngology Institute, Shanghai Jiao Tong University
Shanghai 200233, China
Tel.: +86 2164834143
Fax: +86 2164834143
E-mail: jassey@126.com

Weitian Zhang, M.D., Ph.D.
Sixth People's Hospital
Shanghai Jiao Tong University
Shanghai 200233, China
Tel.: +86 2164834143
Fax: +86 2164834143
E-mail: walker.zhang@yahoo.com.cn