Chapter 12
Speech and Hearing after Cochlear Implantation in Children with Inner Ear Malformation and Cochlear Nerve Deficiency

Yasushi Naito, Saburo Moroto, Hiroshi Yamazaki, and Ippei Kishimoto

Abstract Despite wide possibilities of morphological deformities, our series have shown prevalence of several malformation types, IP-II (incomplete partition type II) being most frequent followed by IP-I (incomplete partition type I) and CC (common cavity). The speech perception and production outcomes after cochlear implantation were best in IP-II, which were comparable to those in controls without malformation, followed by IP-I and CC. It is important to note, however, that significant improvement in speech perception was observed even in CC anomaly, which is the severest malformation included in the present study. The number of functioning electrodes was less than default in some ears with CC and IP-I deformities, and adjustments of the current level and pulse width were necessary in some electrodes in these groups. The electrophysiological and audiometric data in CC deformity indicated that auditory neuronal elements are mainly distributed in the anteroinferior part of the cavity. Both the relative diameter of the vestibulocochlear nerve and the presence or absence of reproducible electrically evoked brainstem responses were significantly associated with cochlear implant outcomes in patients with cochlear nerve deficiency.

Keywords Common cavity • Incomplete partition • Map • Speech perception • Electrode • Spiral ganglion neurons • Cochlear nerve deficiency • EABR

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Table 12.1 Inner ear anomalies that underwent cochlear implantation in Kobe City Medical Center General Hospital

Anomalies		Number of ears	Number of patients
Inner ear	Common cavity[a]	10	10
	IP-I	13	11
	IP-II	18	14
	EVA	9	7
	CH-III[b]	6	3
	Lateral canal hypoplasia	1	1
	unclassified[c]	3	3
Internal auditory canal	IAC stenosis	3	2
	CNC stenosis	6	6
Total		69	57

CC common cavity, *IP* incomplete partition, *EVA* enlarged vestibular aqueduct, *CH* cochlear hypoplasia, *IAC* internal auditory canal, *CNC* cochlear nerve canal

[a]Five ears were with cochlear nerve deficiency

[b]Two patients had CHARGE syndrome, three ears with IAC stenosis, one ear with CNC stenosis, and two ears with duplicate IACs

[c]Waardenburg syndrome, CHARGE syndrome, and Down syndrome with inner ear anomaly

12.1 Introduction

Morphological abnormalities of the inner ear vary widely since there are multiple sites that can be malformed: the cochlea, the vestibule, and the internal auditory canal. The anomalies encountered in clinical practice, however, are not equally distributed, but several types prevail and others are rare. The number of the ears and patients who had inner ear and/or internal auditory canal anomalies and underwent cochlear implantation (CI) in our clinic is shown in Table 12.1. The IP-II anomaly was most frequent (26 % of all ears with anomalies), followed by IP-I (19 %) and common cavity (CC) (14 %). As for the anomaly of the internal auditory canal (IAC), stenosis of cochlear nerve canal (CNC) was more frequent than IAC stenosis. Not only inner ear and IAC anomalies but also hypoplasty of the cochlear nerve (cochlear nerve deficiency or CND) influence CI outcomes. In this chapter, we report CI outcomes of patients with inner ear anomalies, focusing on CC, IP-I, IP-II, and CND, and discuss on their pathophysiologies.

12.2 Speech Perception in CC, IP-I, and IP-II

12.2.1 Introduction

Common cavity anomaly lacks separation between the cochlear and vestibular part of the inner ear. In contrast, the cochlear and vestibular regions are individually identified in the inner ear of IP-I and IP-II, but both lack bony partitions within the cochlea,

Table 12.2 The subjects included in the present investigation

	Age at surgery (months)	Concomitant CND	Electrode array	Follow-up period (months)	CI-aided threshold (dB)
CC: 7 ears	30.4 ± 6.1	2 ears	CI24M: 1 ear, CI24RE(ST): 4 ears, CI422: 2 ears	35.8 ± 9.8	41.1 ± 3.9
IP-I: 9 ears	32.5 ± 20.4	None	CI24RE(ST): 8 ears, CI24R(CA): 1 ear	35.4 ± 9.1	34.9 ± 4.1
IP-II: 11 ears	71.1 ± 55.8	None	CI24M: 2 ears, CI24R(CS): 3 ears, CI24RE(CA): 5 ears, 90 K: 1 ear	35.8 ± 9.1	30.3 ± 4.1
Controls: 22 ears	32.8 ± 18.3	None	CI24R(CS): 1 ear, CI24RE(CA): 20 ears, 90 K: 1 ear	37.7 ± 13.9	28.4 ± 1.7

Aided thresholds = (500 Hz + 1,000 Hz + 2,000 Hz + 4,000 Hz)/4, controls: GJB2 gene mutation without anomaly

CND cochlear nerve deficiency, *CI* cochlear implant, *CC* common cavity, *IP-I* incomplete partition type I, *IP-II* incomplete partition type II

partly or completely. While the osseous structure of the basal turn including the modiolus is formed in IP-II, the bony modiolus is missing in IP-I. Thus, the primary auditory neurons exist at the center of the cochlea in IP-II, while the distribution of auditory neurons of IP-I varies and is not always located at the central region in the cochlea. Since patients with common cavity and IP-I anomalies have profound deafness at birth, cochlear implantation is the only strategy for them to obtain auditory perception. In contrast, patients with IP-II anomaly often have residual hearing, primarily in low frequencies, at birth, and there are children who acquire spoken language with hearing aids. Their hearings, however, usually deteriorate with age, and cochlear implants take over the role of hearing aids. Anatomical differences among these anomalies influence postoperative hearing and spoken language development.

12.2.2 Speech Perception Test Results

We performed cochlear implantation in 69 ears of 57 pediatric patients with malformations in the inner ear and/or in the internal auditory canal. Among them, 27 patients reached the age range at which speech perception test was possible and had been followed up more than 1 year after surgery. The test results of 27 ears in these patients, 7 ears with CC, 9 with IP-I, and 11 with IP-II anomaly, were studied (Table 12.2). The results of 22 pediatric CI patients whose hearing loss had been confirmed to be due to GJB2 gene mutation and without inner ear malformation

were used as controls. Children with mental retardation and pervasive developmental disorders were excluded from the current study.

The mean age at implantation in IP-II was 71.1 months, which was much higher than the other groups (Table 12.2). The delay of CI surgery in IP-II children was due to their usable residual hearings that enabled them to, at least partly, acquire speech. But they lost hearing afterward and underwent cochlear implantation.

12.2.2.1 CI-Aided Thresholds

The CI-aided thresholds in CC, IP-I, IP-II, and control group are listed in Table 12.2. The thresholds of patients were highest in CC group, followed by IP-I. The aided thresholds of IP-II group were significantly lower than those of CC and IP-I, exhibiting no significant difference between controls.

12.2.2.2 Monosyllable Perception Scores

The monosyllable perception scores in each group are shown in Fig. 12.1. The scores were lowest in CC, followed by IP-I. The scores in IP-II and the control groups were about 80–90 % and did not differ from each other. The scores in CC and IP-I groups were significantly lower than those in IP-II and control groups.

12.2.2.3 Word Perception Scores

Figure 12.2 shows the word perception scores of CC, IP-I, IP-II, and control groups. The results are similar to monosyllable perception scores. The mean score of IP-II was 93.7 %, which was very close to 95.3 % in controls. The scores for IP-I and CC were 82.2 % and 54.3 %, respectively, which were lower than those of IP-II and controls, but the difference between each group is smaller compared to monosyllable tests.

12.2.2.4 CAP Score and SIR Scale

To assess the spoken language development in daily life situations, we examined Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) Scale.

Categories of Auditory Performance (CAP) is an index consisting of eight performance categories arranged in order of increasing difficulty [1]. The category 0 means no awareness of environmental sound, 1 awareness of environmental sounds, 2 response to speech sounds, 3 identification of environmental sounds, 4 discrimination of speech sounds, 5 understanding of phrases without lip reading, 6 understanding of

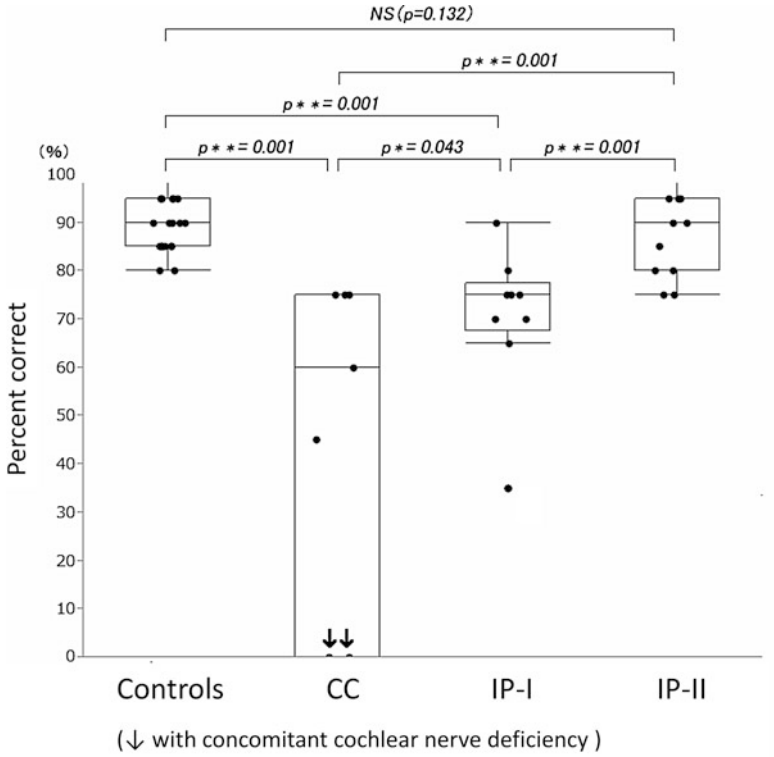

Fig. 12.1 Monosyllable perception scores

conversation without lip reading, and 7 use of the telephone. The mean CAP score in IP-II group was 6.4, which was the same as in control group, corresponding to the level of understanding conversation without lip reading, and sometimes telephone can be used. The mean scores of IP-I and CC children were 5.7 and 4.5, respectively, which were one and two levels below that of IP-II and controls (Fig. 12.3).

The Speech Intelligibility Rating (SIR) Scale is used as a framework to rank the child's spontaneous speech production into one of five hierarchic categories: (1) pre-recognizable words in spoken language, (2) connected speech is unintelligible but is developing for single words, (3) connected speech is intelligible to a listener who concentrates and lip-reads within a known context, (4) connected speech is intelligible to a listener who has little experience of a deaf person's speech (the listener does not need to concentrate unduly), and (5) connected speech is intelligible to all listeners (the child is easily understood in everyday contexts). SIR is not a performance test and was designed as a time-effective global outcome measure of speech production in real-life situations [2]. The mean SIR scores were as high as 4.8 and 4.6 in IP-II and control groups, and, again, the scores were 1 and 2 points lower in IP-I and CC groups, respectively (Fig. 12.4).

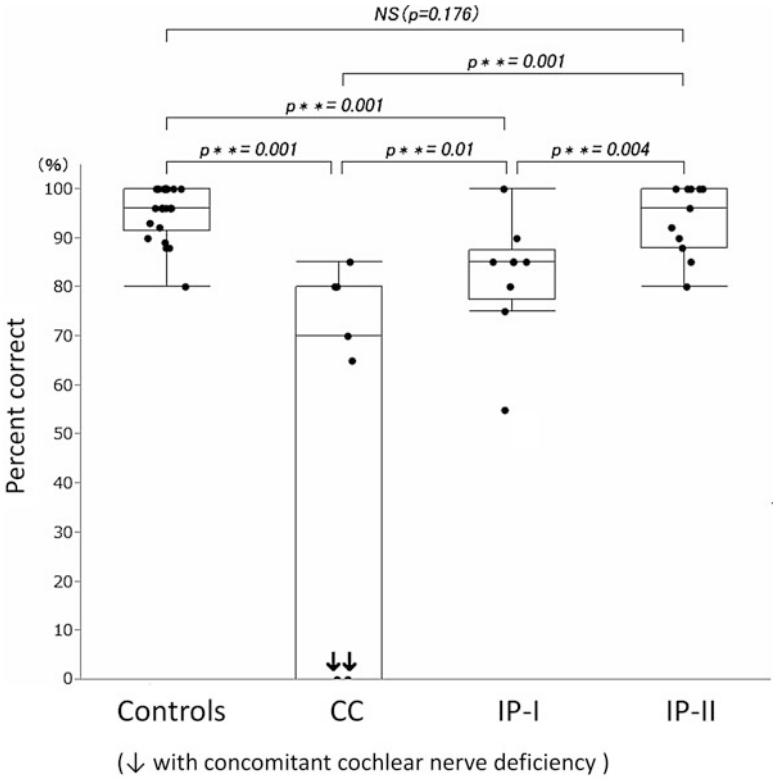

Fig. 12.2 Word perception scores

12.2.3 Mapping Characteristics in Children with an Inner Ear Anomaly

The CIs used in the current pediatric patients were all cochlear devices. In principle, electrode arrays with straight configuration (CI24M, CI24RE-ST, CI422) were selected in CC and IP-I patients, with an exception in which pre-curved electrode (CI24R-CS) was used in one IP-I patient. In contrast, pre-curved arrays (CI24R-CS, CI24RE-CA) were used more in IP-II and in control group with three exceptions in which straight-type electrode arrays (two CI24M and one CI422) were selected. The initial values of mapping parameters, pulse width, stimulation rate, and maxima (the number of electrodes for stimulation to extract sound features), are set at 25 μs, 900 Hz, and 8, respectively. The map for each patient is created by gradually raising the sound intensity from the T level (threshold level) until the charge reaches the C level (maximum comfort level) by observing the responses to the sound. If the charge amount corresponding to T level and C level is not attainable within default current range, a pulse width is widened to create a map at lower current levels. Such

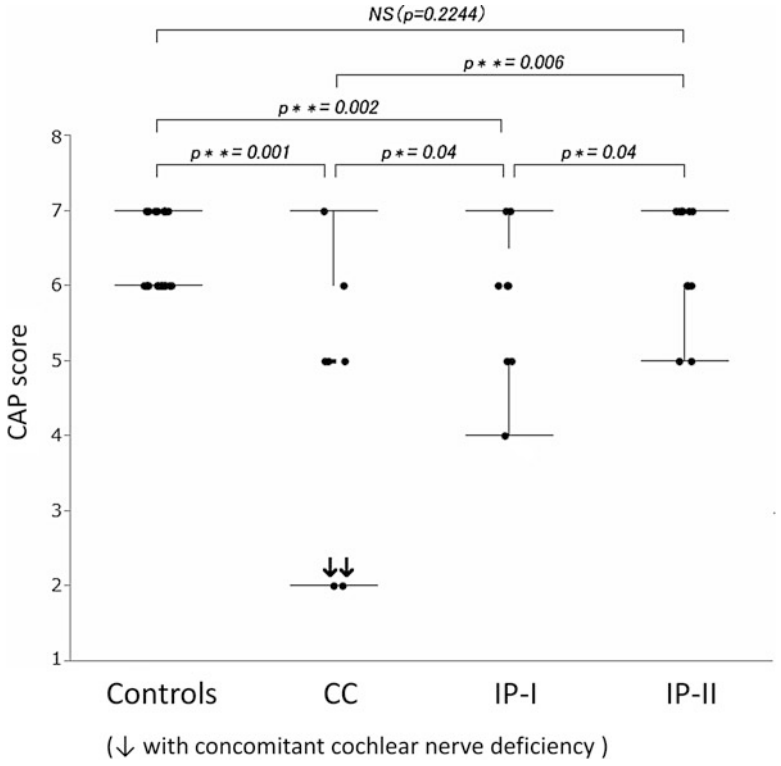

Fig. 12.3 CAP scores

adjustments are often necessary in anomalous inner ears, and there are even cases in which certain electrodes are determined to be unusable due to lack of auditory responses in spite of thorough adjustments.

12.2.3.1 Number of Usable Electrodes

The numbers of usable electrodes that elicited auditory responses ranged from 8 to 22 in CC group, 18–22 in IP-I group, and all 22 in IP-II group (Table 12.3). The numbers of usable electrodes were less in patients with smaller cavities in CC group.

12.2.3.2 The Amount of Charge Used in Electrodes

The amount of charge per phase for T levels (mean ± standard deviation) was 26.3 ± 13.4 nC for the CC group, 12.8 ± 3.3 nC for the IP-I group, 5.6 ± 1.8 nC for the IP-II group, and 4.7 ± 1.3 nC for the control group (Table 12.3). The amount of

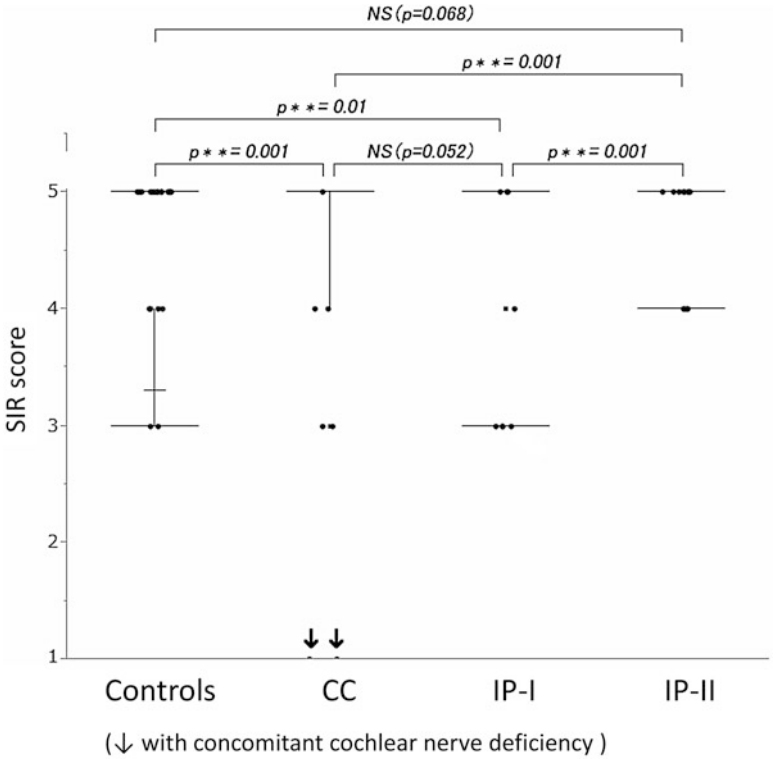

Fig. 12.4 SIR scale scores

charge used in the CC and IP-I groups was significantly greater than that of the control group ($p < 0.01$). There was no significant difference between the IP-II and control groups.

The amount of C level charge was 66.3 ± 35.1 nC for the CC group, 29.3 ± 5.3 nC for the IP-I group, and 15.4 ± 6.5 nC for the IP-II group, while it was 12.7 ± 3.4 nC for the control group (Table 12.3). Charge in CC and IP-I groups was significantly greater than that in the control group ($p < 0.01$). Again, there was no significant difference between the IP-II and control groups.

12.2.3.3 Modification of Routine Mapping Procedures

Our initial setting for pulse width was 25 μs, which was sufficient for one ear in the CC group (14.3 %), one ear in the IP-I group (11 %), and all ears in the IP-II and the control groups (Table 12.3). There was a need to set the pulse width wider than 25 μs in six ears in the CC group and eight ears in the IP-I group. Of these 14 ears, for two ears in the CC group and for six ears in the IP-I group, it was possible to ensure the appropriate amount of charge corresponding to C level by expanding the

Table 12.3 Mapping parameters

Groups	Number of functioning electrodes	Amount of charge per phase for T and C levels (nC)[a] (mean ± SD)		Pulse width and facial stimulation below C level		
		T level	C level	Pulse width =25 μs	Pulse width >25 μs without facial nerve stimulation	Facial stimulation below C level
CC (7 ears)	8 (1 ear)	26.3 ± 13.4[b]	66.3 ± 35.1[b]	1 ear (14.3 %)	2 ears (28.6 %)	4 ears (57.1)%
	17 (2 ears)					
	22 (4 ears)					
IP-I (9 ears)	18 (1 ear)	12.8 ± 3.3[b]	29.3 ± 5.3[b]	1 ear (11 %)	6 ears (67 %)	2 ears (22 %)
	22 (8 ears)					
IP-II (11 ears)	22 (all 11 ears)	5.6 ± 1.8	15.4 ± 6.5	11 ears (100 %)	None	None
Controls (22 ears)	22 (all 22 ears)	4.7 ± 1.3	12.7 ± 3.4	22 ears (100 %)	None	None

CC common cavity, *IP-I* incomplete partition type I, *IP-II* incomplete partition type II
[a]Amount of charge in nanocoulomb (nC) = amount of current (μ A) × pulse width (μs) × 1000
[b]$P < 0.01$ larger than controls (Kruskal-Wallis, Mann-Whitney *U* test, Bonferroni correction)

pulse width from 37 to 88 μs and without encountering facial nerve stimulation. Nevertheless, for four ears in the CC group and two ears in the IP-I group (Table 12.3), increasing the current level stimulated the facial nerve, and securing a charge amount corresponding to the C level was challenging. As a result of re-adjusting the map through further expansion of pulse width, for five out of the six ears, we were able to reach C level before encountering facial nerve stimulation. Nevertheless, for the one remaining ear, it was not possible to suppress the facial nerve stimulation, and maximum stimulation remained at a lower value than the charge amount corresponding to the C level.

12.3 Discussion on Speech Perception and Map Parameters

The results of cochlear implantation in patients with inner ear malformations have been reported by many authors. Despite wide possibilities of morphological deformities, our series have shown prevalence of several malformation types, IP-II being most frequent followed by IP-I and CC, which is similar to previous results including the one by Sennaroglu et al. [3]. These findings indicate general patterns for inner ear malformation occurrence and the importance of detailed analysis on CI outcomes in CC, IP-I, and IP-II.

It is important to check whether patients with mental retardation or developmental disorder are included in the study or not when interpreting the CI outcomes of patients with inner ear malformations. In overall, children suffering from developmental disorder [4] or mental retardation [4, 5] do not progress as well as the non-delayed children after cochlear implantation. In the current study, we excluded patients with developmental disorders and those with mental retardation. Thus, our results may reflect the difference in inner ear morphology and spiral ganglion cells between malformation and normal anatomy cases.

On speech perception and production outcomes, the results in IP-II were comparable to those in controls and significantly better than those observed in CC and IP-I, which are in line with the findings in previous studies [6–8]. Although osseous modiolus and interscalar septa of cochlear upper turns are missing in IP-II anomaly, neurosensory elements and SG cells exist not only in the basal turn but in the upper region in approximately the same location as in the cochlea without anomaly [9], which may be the reason for IP-II's good CI outcomes. We may not have to expect significant disadvantage in CI-mediated speech perception in patients with IP-II anomaly when considering their indication for CI.

Although speech perception scores of CC patients using CI were lower than those in IP-II and control groups, it is important to note that significant improvement in speech perception was observed even in CC anomaly, which is the severest malformation included in the present study. Similar positive effects of CI on spoken language development in CC patients have been reported [7, 8].

The number of functioning electrodes was less than default in three ears in CC and one ear in IP-I groups, but all electrodes could be activated in all ears in IP-II and in control groups. Vera et al. [10] also reported that the number of functioning electrodes was significantly less in patients with malformed inner ear compared to those in patients without malformation. Significant differences were observed between the major and minor malformation groups in their study. As for mapping parameters, we found that the amount of charge per phase for T and C levels was significantly higher in CC and IP-I groups, which is also the same tendency observed in the previous investigation [10]. In most patients in CC and IP-I groups in the present study, pulse width had to be adjusted wider than the default value of 25 μs, suggesting that more charge was necessary to activate sufficient number of SG neurons and bring about sound sensation.

In the present study, four (60 %) of the seven CC patients and two (22 %) of the nine IP-I patients experienced CI-mediated facial nerve stimulation (FNS), which is consistent with the previous study reporting the high frequency of FNS among patients with inner ear malformations who had implants [11–13]. In cases with a severe inner ear malformation, high current level and/or increased pulse width are often required to achieve good auditory performance [13, 14], suggesting a necessity to adjust the current level to an appropriate value that is high enough to provide sufficient auditory input but lower than the threshold for FNS. The stimulus amplitude cannot be increased higher if it reaches the level of facial nerve stimulation, which practically limits the dynamic range of the CI map.

Lack of tonotopy in the cochlea and smaller number of SG neurons in inner ears with malformations [15–17] may underlie their relatively lower CI outcomes. It has been reported that at least 10,000 SG neurons may be necessary for speech discrimination by CI [18]. However, there is also a report discussing that benefit from CI can be obtained in patients with as few as 3,300 SG cells [19]. Kahn et al. [20] reported that significant correlation between psychophysical measures and SG neuron counts was found in only two of the five subjects they examined. Auditory perception by CI with fewer SG cells may be achieved by higher neural synchrony of SG cells activated by direct electrical stimulation. Possible redundancy in cochlear innervation [6] and plastic reorganization of cortical language networks [21] may also contribute to successful perception and production of speech through CI. The shape and placement of the electrode array in the inner ear cavity, especially in CC deformity, influence the outcomes of CI, which will be discussed in the following section.

12.4 Distribution of Auditory Neurons in Common Cavity Anomaly

12.4.1 Introduction

Effective stimulation of SG neurons by CI electrodes is necessary for better CI outcome, but the spatial distribution of SG cells and auditory nerve fibers is unclear in CC deformity because of no differentiation between the cochlea and vestibule in addition to the lack of a modiolus. Electrically evoked auditory brainstem responses (EABRs) using CI-mediated stimulus can be used for the objective evaluation of auditory neuronal responses in the brainstem [6, 14, 22]. In this section, we show the results of our previous EABR investigation [23] on the spatial distribution of auditory neurons in CC deformity.

12.4.2 CI-Mediated EABR Findings in CC Patients

We retrospectively examined five patients with CC deformity with congenital profound sensorineural hearing loss who underwent cochlear implantation at our hospital from 2005 to 2013. Mean age at implantation was 27.4 months, and the mean follow-up period was 26.0 months. Nucleus device with 22 active electrodes (Ch1–Ch22), including CI24RST, CI24REST, or CI422, was implanted in all cases. Intraoperative EABR testing was performed with Nucleus Custom Sound EP software using MP 1 + 2 mode. The EABR was recorded with a filter setting of 20 Hz to 3 kHz on the opposite side to minimize artifacts of the implanted device.

In Case 1 (Fig. 12.5), the radiograph obtained during the initial cochlear implantation demonstrated that most of the electrodes were located within the CC deformity, but the CI-aided performance was still poor even after 1 year of use of CI. EABR elicited a reproducible evoked wave V (eV) only at 2 of 11 tested electrodes. Thus,

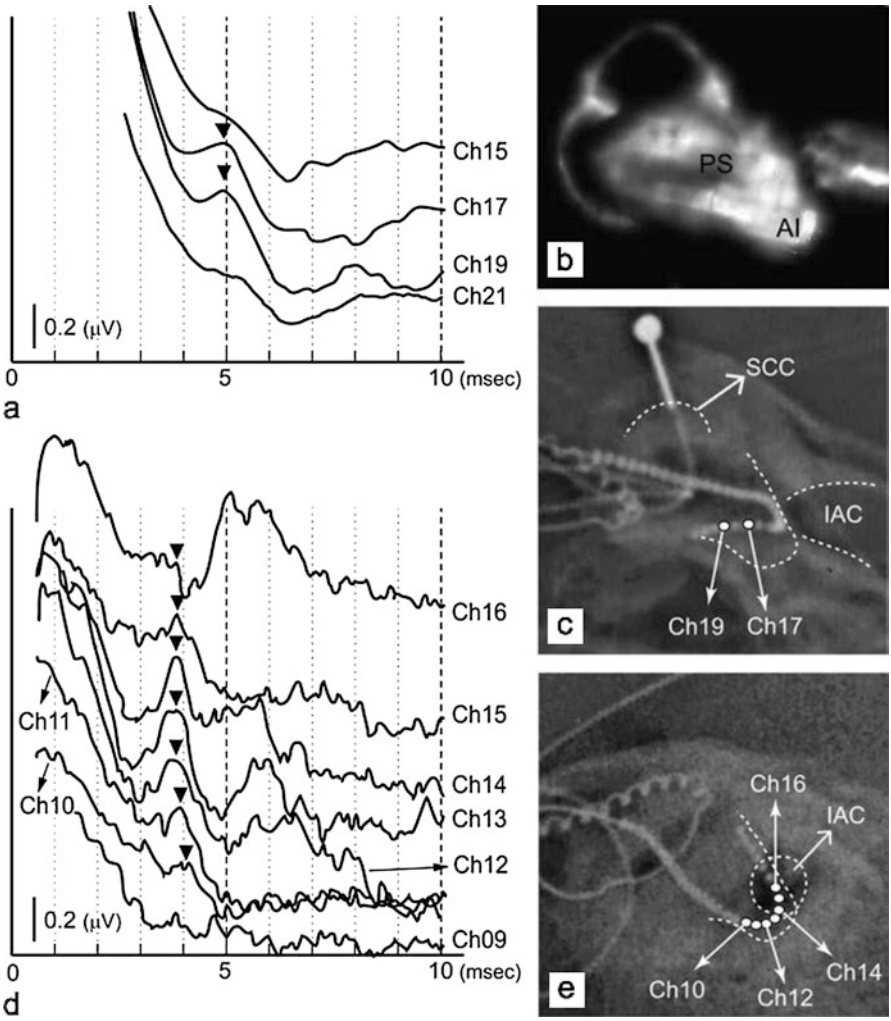

Fig. 12.5 Results of EABR testing in Case 1 before and after the reimplantation. (**a**) EABR testing after the initial implantation with the showing eVs in Ch17 and Ch19 among the 11 tested electrodes. The latency of these eVs is approximately 5 ms (*arrowheads*). (**b**) A maximum intensity projection of the T2-weighted magnetic resonance image of the CC deformity on the implanted side. The anteroinferior part of the CC deformity (AI) is smaller than the posterosuperior part (PS). (**c**) Radiograph of the initial implantation. (**d**) EABR testing after the reimplantation showing a distinct eV in 7 of 22 electrodes. The latency of these eVs ranges from 3.8 to 4.1 ms (*arrowheads*). (**e**) Radiograph after the reimplantation demonstrating that electrodes with a positive eV (*circles*) are located in the anteroinferior part of the CC deformity (*dotted line*) (Cited from Ref. [23] with permission)

we performed reimplantation surgery with wider labyrinthotomy, resulting in successful placement of the electrode array in the anteroinferior region of the inner ear cavity, obtaining appropriate eV at seven electrodes. In the other four patients, postoperative CT images showed the optimal position of the electrode array, requiring no revision surgery. Although the size and shape of each CC deformity differed among the cases (Fig. 12.6a–d), electrodes inserted in the anteroinferior cavity successfully elicited eVs in all four cases, similarly to Case 1 (Fig. 12.6e–h).

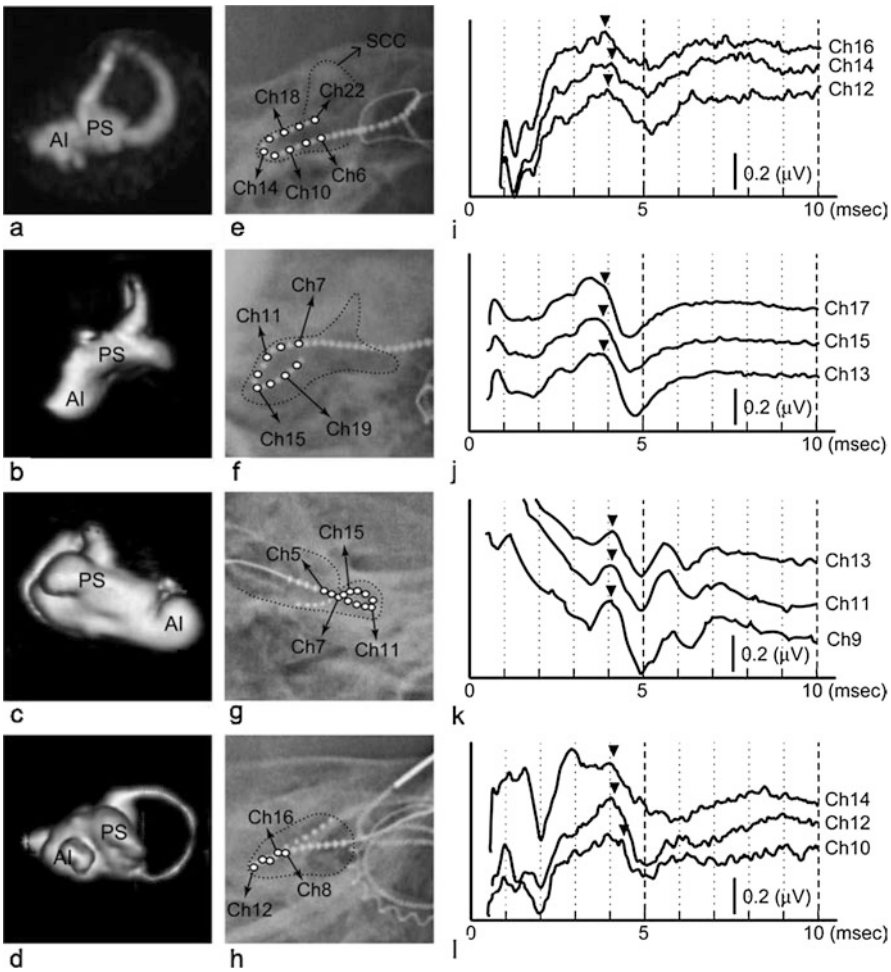

Fig. 12.6 Results of EABR testing in Cases 2–5. (**a–d**) Maximum intensity projection of T2-weighted magnetic resonance images of the CC deformity on the implanted side. *AI* and *PS* indicate the anteroinferior and posterosuperior parts of the CC. (**e–h**) Electrodes with a positive eV (*circles*) are located in the anteroinferior part of the CC. (**i–l**) EABRs for three representative electrodes. The latency of these eVs is approximately 4 ms in all cases (*arrowheads*) (Cited from Ref. [23] with permission)

Before implantation, no patient could detect sounds, that is, their preoperative CAP score was zero, but auditory perception improved after activation of the CI in all patients. The postoperative CAP score reached to 6 in Cases 1 and 2 who had used their CI for more than 2 years, and their speech discrimination scores of closed-set infant words were 76 % and 80 %, respectively. The other three patients, Cases 3, 4, and 5, who had used their CI for less than 2 years, showed CAP scores of 4, 3, and 3, respectively, and Case 3 showed 40 % of the infant word discrimination score.

12.4.3 Discussion on CI-Mediated EABR in Common Cavity Anomaly

The present results demonstrated that reproducible eVs were elicited by activating electrodes that were located at the anteroinferior part of the CC deformity in all patients. The electrophysiological and audiometric data indicate that auditory neuronal elements are mainly distributed in the anteroinferior part of the CC deformity. In the normal development of an inner ear, the ventral portion of the otic vesicle elongates in the ventral direction, initiating cochlear development [24]; therefore, the anteroinferior part of CC deformity might be programmed to differentiate to a cochlea. These findings support our conclusion regarding the anteroinferior distribution of auditory neuronal tissue in CC deformity.

Case 1 who showed eV only at 7 (31.8 %) of 22 electrodes exhibited 6 in CAP score and 76 % in the infant word discrimination test at 4 years after the initial implantation, which are similar to those observed in the 2-year postoperative Case 2 who showed eV at almost all electrodes (81.8 %). These data suggest that even if the only limited number of electrodes shows eV in EABR testing, the patient might achieve sufficient CI-aided auditory performance after long-term use of the CI with an appropriate program.

12.5 Cochlear Nerve Deficiency

12.5.1 Introduction

Hypoplasia and aplasia of the cochlear branch of the vestibulocochlear nerve, called cochlear nerve deficiency (CND), are defined by an absent or a small cochlear branch of the vestibulocochlear nerve (cochlear nerve) on MRI [25–27]. Several studies have reported that congenitally deaf children with CND show significantly poorer auditory performance using CI than children without CND [26, 28]. However, many patients with CND understood some words in a closed-set word

discrimination test using CI [26, 28]. Previous studies investigating CI children with CND demonstrated that the CI outcomes were correlated to the type of malformation on CT and MRI and the result of intracochlear EABR [26, 28]. In this section, we show the results of our previous collaborative research by the University of Melbourne and Kobe City Medical Center General Hospital [29], aiming to establish a strategy of preoperative and intraoperative objective examinations to discriminate CND patients with poor CI outcomes from those with satisfactory CI outcomes.

12.5.2 Patients, Methods, and Results

A retrospective examination of 19 congenital deaf children with CND who underwent cochlear implantation at Kobe City Medical Center General Hospital or Melbourne Cochlear Implant Clinic from 2003 to 2013 was conducted. The mean age at implantation was 26.7 ± 11.5 months, and the median follow-up period was 34 months. Nucleus devices were implanted. Simultaneous and sequential bilateral cochlear implantations were performed in one and four children, respectively.

Narrow internal auditory canal (NIAC) was defined by the width of midpoint of the IAC being narrower than 2 mm [6]. Hypoplasia of bony cochlear nerve canal (HBCNC) was defined by less than 1.4 mm in the diameter of the bony cochlear nerve canal as well as a normal width of the IAC on CT images [25]. NIAC was identified on the implanted side of six patients, HBCNC in six patients, cochlear aplasia (CA) in one patient, CC in five patients, and CH-III in two patients. MRI was acquired using a 1.5-T or 3.0-T system, which failed to visualize a definitive bundle of a cochlear nerve at the fundus of the IAC in all patients, on the basis of which CND was diagnosed. For each case, the relative diameter of the vestibulocochlear nerve compared to the facial nerve was evaluated at the cerebellopontine angle (CPA). The vestibulocochlear nerve was smaller than the facial nerve at the CPA in seven children, whereas it was equal to or larger than the facial nerve in the remaining 12 (Fig. 12.7). Intracochlear EABR testing was performed in the operation room using Nucleus Custom Sound EP software, of which details are described in our previous report [23].

Auditory performance with the CI was evaluated using CAP scores [1]. Preoperative and postoperative CAP scores in this population with CND were 0.2 ± 0.4 and 3.0 ± 2.1, respectively, and significant improvement in the auditory performance was observed after cochlear implantation. Children with relatively thin vestibulocochlear nerves "CN7 > CN8" had significantly poorer performance: postoperative CAP scores 1.1 ± 1.5, compared to those with more normal sized nerves "CN7 <= CN8," CAP score 4.1 ± 1.5 (Fig. 12.8a). With respect to the EABR testing, the postoperative CAP score was 4.3 ± 1.2 in those with "positive eV," which is significantly higher than 1.8 ± 1.9 in those with "negative eV" (Fig. 12.8b).

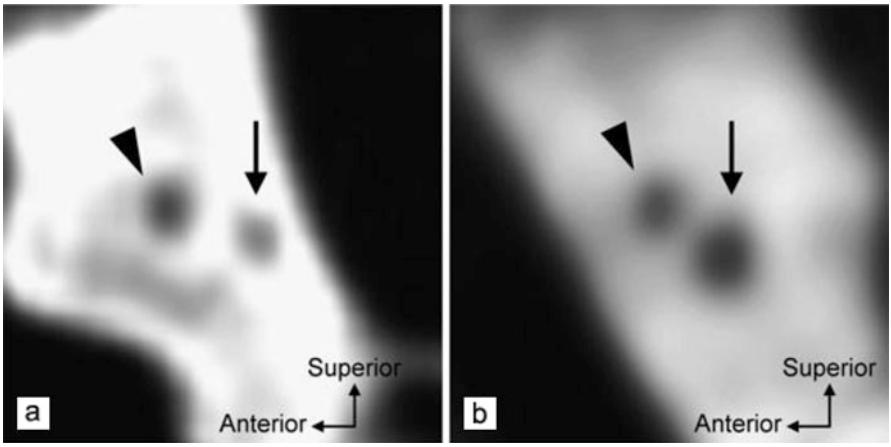

Fig. 12.7 Evaluation of relative diameter of the vestibulocochlear nerve compared to the facial nerve at CPA using MRI. MRI shows the vestibulocochlear nerve (*arrows*) and the facial nerve (*arrowheads*) at the CPA. (**a**) "CN7 > CN8," (**b**) "CN7 <= CN8" (Cited from Ref. [29] with permission)

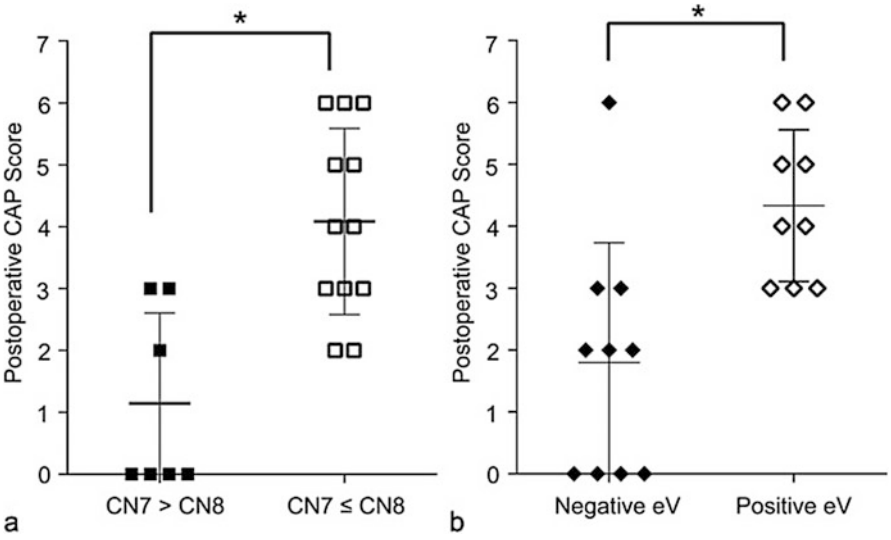

Fig. 12.8 A relationship between objective examinations and postoperative CAP scores. (**a**) A relationship between the MRI findings and postoperative CAP scores. (**b**) A relationship between the EABR results and postoperative CAP scores (Cited from Ref. [29] with permission)

Although the results of MRI and EABR testing were significantly associated with postoperative CI outcomes, each examination failed to clearly discriminate patients with poor CI outcomes from those with satisfactory CI outcomes. Combination of the results of MRI and EABR testing allowed better discrimination between children with limited or no benefit from a CI and those with moderate or good CI-aided auditory performance. All of the six patients who were categorized into both "CN7 > CN8" and "negative eV" exhibited less than or equal to 3 in the postoperative CAP scores, and four of them (66.7 %) showed no response to sound (CAP score of 0) even after 2 years of CI use. On the contrary, all of the eight children who showed "CN7 <= CN8" on MRI and "positive eV" on EABR testing reached greater than or equal to 3 in the CAP scores within 2 years after implantation, and six of them (75.0 %) discriminated at least some speech sounds without visual support (CAP score of 4).

12.5.3 Discussion on CI in Cochlear Nerve Deficiency

In this investigation, we found that both the relative diameter of the vestibulocochlear nerve in the CPA as seen on the preoperative MRI and the presence or absence of reproducible eVs with typical latency in the intraoperative EABR testing were significantly associated with postoperative auditory performance with CI. It was also demonstrated that the combination of MRI and EABR testing achieved more precise discrimination immediately after cochlear implantation between patients with no or limited benefit from CI and those with moderate to good CI outcomes than independent use of either.

CND is thought to diminish development of auditory perception with CI because of a small number of SG neurons [30]. A previous histological study showed that the count of the SG neurons was predicted by the maximum diameter of the main trunk of the vestibulocochlear nerve [31]. Theoretically, the counts of SG neurons relate to the size of the cochlear nerve more strongly than the main trunk of the vestibulocochlear nerve; however, accurate measurement of the diameter of the cochlear nerve is often difficult. Therefore, evaluation of the vestibulocochlear nerve at the CPA is reasonable to prevent underestimation in specific types of malformations.

Regarding the other groups, "CN7 > CN8/positive eV" and "CN7 <=CN8/negative eV," interpretation is not straightforward because the results of MRI and the EABR testing are contradictory. In the patient categorized in "CN7 > CN8/positive eV," the detection of eV suggests the auditory brainstem was activated by CI, but the number of SGNs was not enough to discriminate speech sounds. Among the four subjects with "CN7 <=CN8/negative eV," three children showed 2 or 3 in the postoperative CAP score, suggesting hypoplasia of the cochlear nerve component.

The current data may be informative to decide the treatment strategy in congenitally deaf children with CND.

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