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ORIGINAL ARTICLE

Vestibular evoked myogenic potentials of children with inner ear malformations before and after cochlear implantation

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Abstract

Conclusions: Our results show that among the patients with inner ear malformations, there were two patients with sensory cells of sacculus and inferior vestibular neurons and at least five patients with inferior vestibular neurons only, but no patients without sensory cells and vestibular neurons, as determined on the basis of vestibular evoked myogenic potentials (VEMPs). *Objective:* The aim of this study was to investigate whether sensory cells of sacculus or inferior vestibular neurons or both are present in cochlear implant patients with inner ear malformations, particularly common cavity (CC) deformity, using VEMPs. *Patients and methods:* Seven children with inner ear malformation who underwent cochlear implantation (CI) participated in this study. The patients had CC deformity ($n=2$), incomplete partition type I ($n=2$), incomplete partition type II ($n=1$), or a narrow internal auditory canal (IAC) ($n=2$). It was possible to record VEMPs before and after CI in three patients and not before but after CI in four patients. After surgery, VEMPs were recorded with the cochlear implant device switched both off and on. *Results:* Before the CI, two patients showed VEMPs and one patient showed no VEMPs, whereas in four patients no VEMPs could be recorded. All the patients showed VEMPs with the cochlear implant on.

Keywords: *Children, inner ear malformations, common cavity deformity, cochlear implantation (CI), vestibular evoked myogenic potentials (VEMPs), sensory cells of saccule, inferior vestibular neuron*

Introduction

Inner ear malformations represent a major inner ear disorder in approximately 20% of children with congenital sensorineural hearing loss [1]. They are usually characterized by profound hearing loss, and their development delays gross motor functions such as head control or independent walking, because such functions are related to abnormal inner ear structures [2]. However, it is not easy to unequivocally determine whether vestibular sensory cells of semicircular canals and otolith organs or primary vestibular afferent neurons are present in patients with inner ear malformations, particularly common cavity (CC) deformity. In an embryological study, it has been found that, in the human fetal developmental stage, the vestibular system develops earlier than the cochlear system [3]. Thus, it is speculated that sensory cells of vestibular end organs and vestibular afferent neurons may be present in patients with inner ear malformations, which is similar to early stage inner ear development.

Vestibular evoked myogenic potentials (VEMPs) are regarded as indicators of the neurophysiological function of the saccule–inferior vestibular nerve system [4–6]. If VEMPs are present, it denotes that sensory cells of both saccule and inferior vestibular neurons are present. In contrast, if VEMPs are absent, it suggests that a part of the sensory cells of saccule or inferior vestibular neurons that respond to auditory stimuli may be absent at least. In our previous study, we reported that VEMPs could be elicited with the cochlear implant switched on, and suggested that the electrical stimulation of a cochlear implant may directly stimulate the inferior vestibular nerve [7,8]. If VEMPs are evoked with the cochlear implant

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switched on, it suggests that some of the inferior vestibular neurons are present. In contrast, if VEMPs are absent with the cochlear implant switched on, it suggests that the inferior vestibular neurons may be absent.

In this study, we selected patients with cochlear implants who had inner ear malformation to compare VEMPs before and after cochlear implantation (CI). The aim of this study was to investigate whether sensory cells of saccule or inferior vestibular neurons, or both, are present in cochlear implant patients with inner ear malformations including common cavity (CC) deformity.

Patients and methods

Patients

The patients comprised seven children who underwent CI at the University of Tokyo Hospital. The mean age at implantation was 3.7 ± 1.1 years (range 2–5 years). All the patients underwent CT scan and audiological assessments including conditioned orientation reflex (COR) audiometry, play audiometry, standard pure tone audiometry, and auditory brainstem response (ABR) measurement. All the patients showed severe hearing loss bilaterally. Their hearing levels ranged from 101.3 dB to off the scale.

We adopted the classification of cochlear malformation of Sennaroglu and Saatci [9]. The children were divided into two groups, according to the VEMP performance both before and after CI (group A), or only after CI (group B). Although VEMPs were recorded in three patients before and after CI, unfortunately, in four patients, no VEMPs could be recorded before CI.

In group A, VEMPs were recorded before and after CI ($n=3$). The types of inner ear malformation were as follows: common cavity (CC) ($n=1$), incomplete partition type II (IP-II) ($n=1$), and narrow internal auditory canal (IAC) ($n=1$).

In group B, VEMPs were recorded only after CI. The types of inner ear malformation were as follows: CC ($n=1$), incomplete partition type I (IP-I) ($n=2$), and narrow IAC ($n=1$).

As controls, nine healthy volunteer children (five boys, four girls; mean age 5.2 ± 2.8 years, range 2–10 years) also participated in the study.

Characteristics of CI device

The types of CI device are summarized in Table I. All the patients received multichannel cochlear implants: one child received Nucleus 22; 5 children, Nucleus 24; and one child, 24R (Table I). The patients with Nucleus 24 were coded with ACE, and the patients with Nucleus 22 and 24R were coded with SPEAK (Table I). For speech processors, five patients used Sprint and two patients used ESPrit 3G (Table I).

After mapping studies, it was demonstrated that the C levels were higher in patients with inner ear malformation than in patients with a normal inner ear (Figure 1). One child with CC had a large pulse width. Both patients with narrow IAC had a large pulse width (Table I).

Tests of vestibular functions

The vestibular functions of the patients were evaluated by ice-water caloric test, damped-rotational chair test, and VEMP measurement.

Caloric test. The child is placed in dorsal decubitus at 30°. The ice-water (4°C and 2 ml) caloric test was performed to irrigate the external auditory meatus to induce a thermal gradient across the horizontal semicircular canal of one ear. Horizontal and vertical eye movements were recorded by electronystagmography (ENG) using standard ENG electrodes.

Rotational chair test. For the rotational chair test, a rotational chair (Nagashima Co. Ltd, S-II) was accelerated to a maximum rotational velocity of 160°/s, then reduced by $4°/s^2$. The mother sat in the chair and rotated together with the child in her arms. The test was performed in both clockwise and counterclockwise directions. Eye movements were recorded by ENG, and the duration and numbers of beats of per rotatory nystagmus were calculated.

VEMP. The children were placed in a supine position. The active electrode was placed on the upper half of the sternocleidomastoid muscle (SCM), the reference electrode on the lateral end of the upper sternum, and the ground electrode on the midline of the forehead. During VEMP recording, the mother pulled the child's arms, and one member of staff encouraged the child to lift his/her head up or to turn his/her head to the contralateral side to induce hypertonicity in the SCM. Another member of staff monitored the electromyography (EMG) activity during the recording to maintain muscle activity at a constant level.

The EMG signal from the stimulated side was amplified and averaged using a Neuropack evoked-potential recorder (Nihon Kohden Co. Ltd, Tokyo, Japan). A headphone (type DR-531; Elega Acous Co. Ltd, Tokyo, Japan) was used to evoke VEMPs. The

Table I. Profiles of patients.

Patient no.	Sex	Ear	Age at surgery (years)	Type of inner ear malformation	CI type	Speech processor	Strategy	Pulse width
1	M	L	5	Commn cavity	24M	ESPrit 3G	ACE	25
2	M	R	5	IP-II LVAS	24M	Sprint	ACE	25
3	M	R	4	IP-I, narrow IAC	24R	Sprint	SPEAK	200
4	M	L	3	Common cavity	24M	Sprint	SPEAK	50
5	M	R	3	IP-I	22M	ESPrit 3G	SPEAK	0
6	M	R	4	IP-I	24M	Sprint	ACE	25
7	M	L	2	Narrow IAC	24M	Sprint	ACE	100

IP-I, incomplete partition type I; IP-II, incomplete partition type II; IAC, internal auditory canal; LVAS, large vestibular aqueduct syndrome; CI, cochlear implantation.

stimulation rate was 5 Hz, the bandpass was filtered at 20–2000 Hz, and the analysis time was 50 ms. VEMPs in response to 100 stimuli were averaged twice. We analyzed the amplitude of the first positive-negative peak, p13-n23, ipsilateral to the stimulated ear, and the latencies of p13 and n23. VEMPs were recorded before and after CI whenever possible. After CI, in all the seven children, VEMPs were recorded from the operated ear with the cochlear implant device switched both off and on.

Results

Controls

Clear VEMPs were obtained from all the healthy volunteer children. The means $\pm$SD latencies of p13 and n23 were 10.5 ± 0.5 and 16.0 ± 1.4 ms, respectively.

Group A patients

Patient 1: 6-year-old boy with left CI (Figure 2). CT scan demonstrated deformity of the CC communicating with the IAC. The lateral semicircular canal (LSCC) was dilated, but the superior semicircular canal (SSCC), and posterior semicircular canal (PSCC) were normal. The hearing level was 108.8 dB HL; after CI, the hearing level was 40 dB HL with the CI on.

Before CI, the caloric tests showed no responses in both ears, but the rotational chair test showed normal responses to both rotations.

Before CI, VEMPs were present in both ears. After CI, the VEMPs were present with the CI off and on.

Patient 2: 5-year-old boy with right CI. CT scan demonstrated IP-II deformity, dilated vestibules, enlarged VA, and normal LSCC, SSCC, and PSCC. The hearing was 118.8 dB HL. After CI, the hearing level became 45 dB HL with the CI on.

Before CI, the caloric tests showed no responses in both ears, but the rotational chair tests showed normal responses to both rotations.

Before CI, VEMPs were present in both ears. After CI, VEMPs were present with the CI on.

Patient 3: 5-year-old boy with right CI. CT scan demonstrated middle ear abnormalities, narrow IAC, IP-II deformity, cystic vestibules, and LSCC. The SSCC and PSCC were normal. The hearing

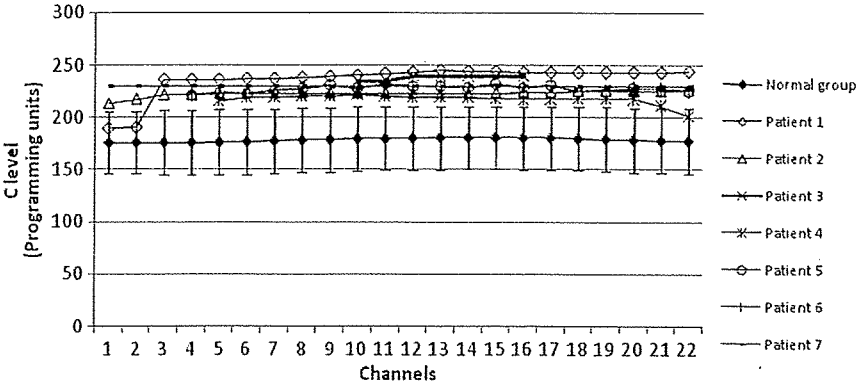

Figure 1. C levels of inner ear malformations.

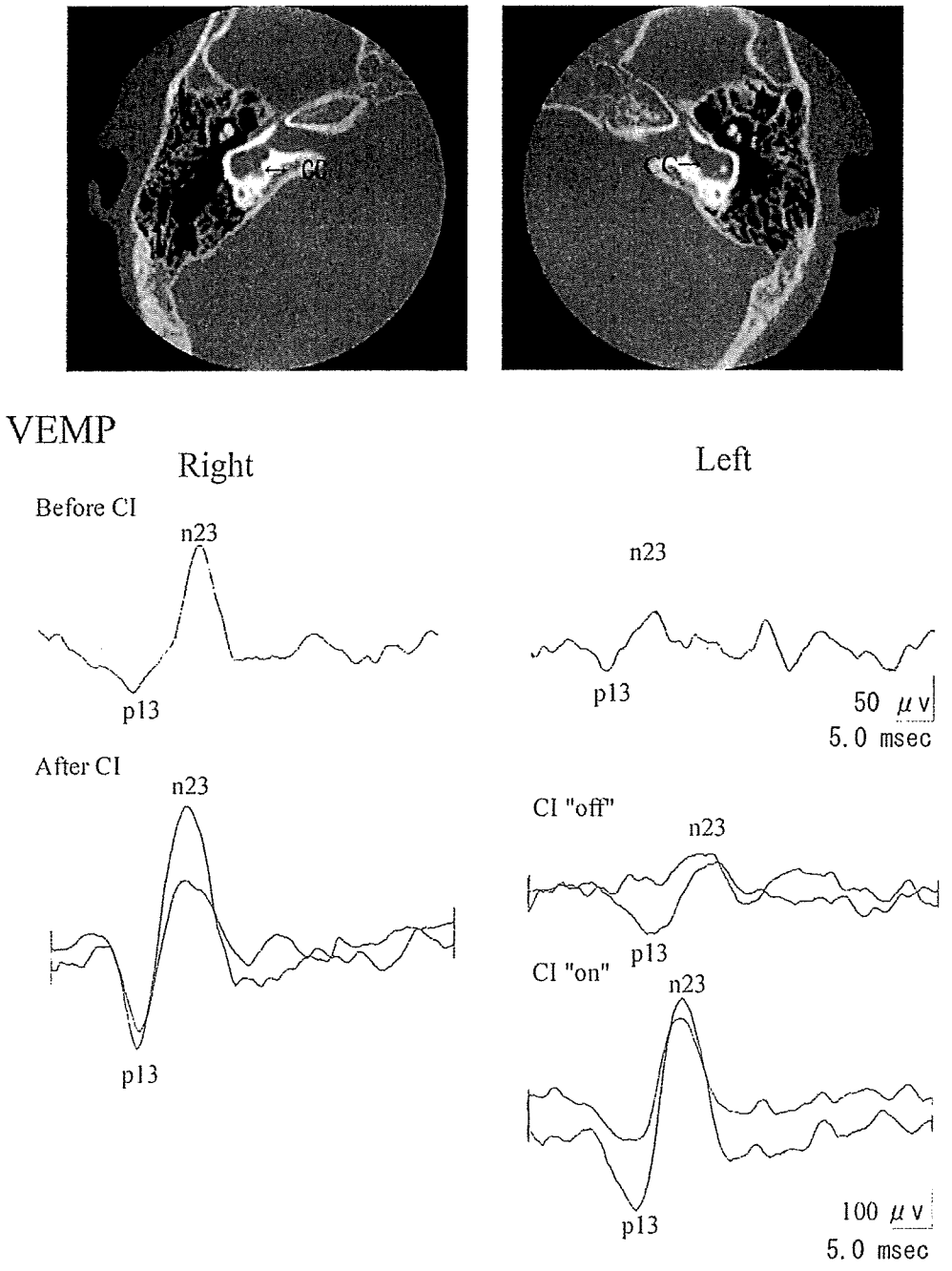

Figure 2. Patient 1: 7-year-old boy, CC deformity, left CI. CT scan demonstrated a deformity of the CC communicating with IAC. VEMPs were present in both ears before CI. VEMPs were present with the CI on.

level was 138.8 dB HL. After CI, the hearing level was off the scale even with the CI on.

Before CI, the caloric test showed no responses in the right ear (the left ear was not tested because of microtia and atresia); the rotational chair tests showed normal responses to both rotations.

Before CI, VEMPs were absent in both ears. After CI, VEMPs were present with the CI on.

Group B patients

Patient 4: 6-year-old boy with left CI. CT scan demonstrated a deformity of the CC communicating with the IAC. The LSCC was dilated, but the SSCC and PSCC were normal. The hearing level was off the scale. After CI, the hearing level was 45 dB HL with the CI on.

Before CI, the caloric test showed no responses in both ears, and the rotational chair test showed poor responses to both rotations.

After CI, VEMPs were present with the CI on.

Patient 5: 8-year-old boy with right CI (Figure 3). CT scan demonstrated IP-I deformity and cystic vestibules, but normal LSCC, SSCC, and PSCC. The hearing level was 101.3 dB. After CI, the hearing level was 45 dB HL with the CI on.

Before CI, the caloric test showed no responses in both ears, and the rotational chair test showed poor response to the right rotation and normal response to the left rotation.

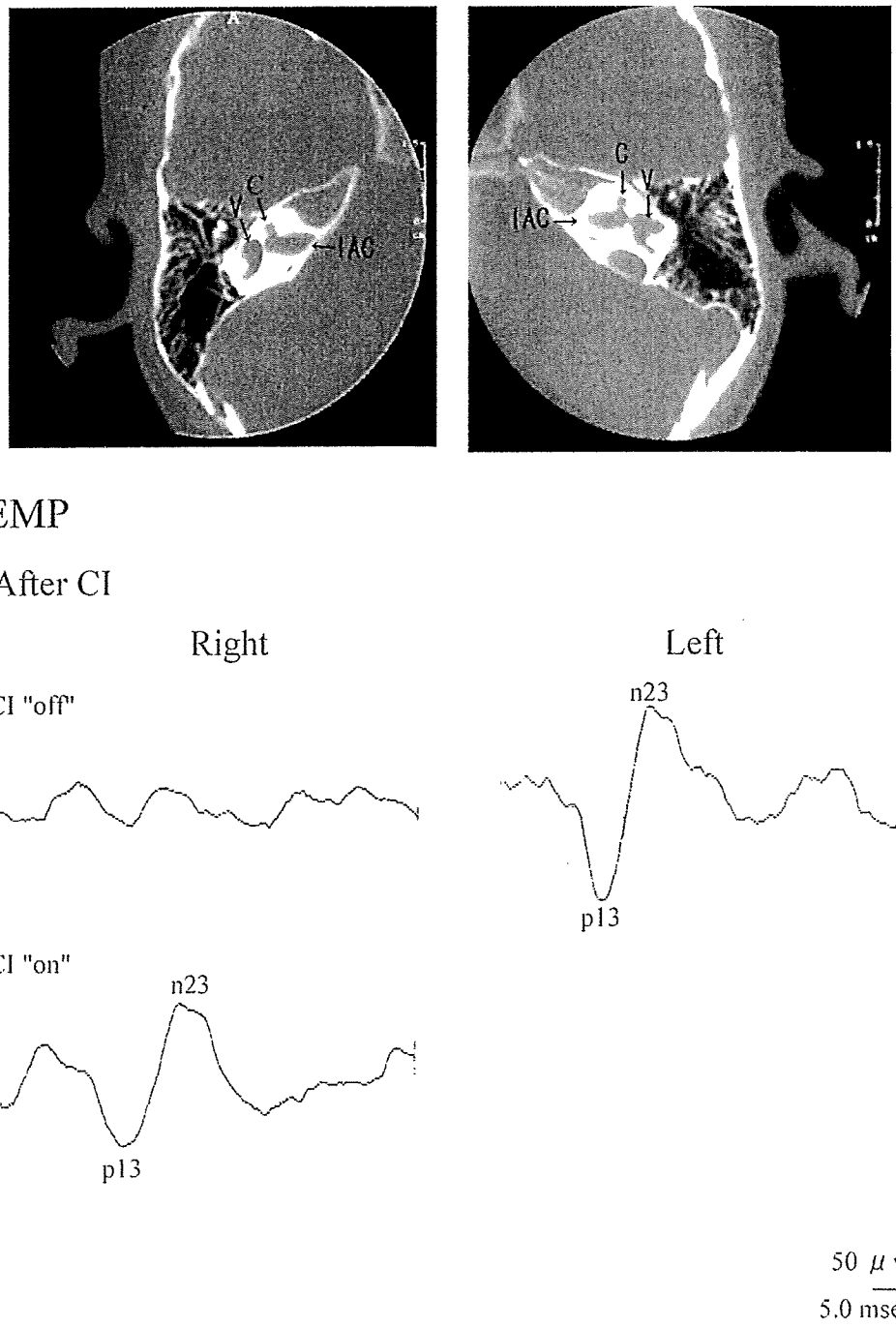

Figure 3. Patient 5: 8-year-old boy, IP-I deformity, right CI. CT scan demonstrated IP-I deformity. After CI, VEMPs were present with the CI on.

After CI, VEMPs were present with the CI on.

Patient 6: 7-year-old boy with right CI. CT scan demonstrated IP-I deformity, cystic vestibules, and normal LSCC, SSCC, and PSCC. The hearing level was off the scale. After CI, the hearing level was 35 dB HL with the CI on.

Before CI, the caloric test showed no responses in both ears, and the rotational chair test showed poor response to the right rotation and normal response to the left rotation.

After CI, VEMPs were present with the CI on.

Patient 7: 5-year-old boy with left CI. CT scan demonstrated narrow IAC, but normal cochlea, vestibules, LSCC, SSCC, and PSCC. The hearing level was off the scale. After CI, the hearing level was 50 dB HL with the CI on.

After CI, VEMPs were present with the CI on.

Summary of VEMPs in seven patients

The results of VEMPs are summarized in Table II and Table III. Before CI, two patients showed VEMPs, but one patient showed no VEMPs. After CI, all the patients showed VEMPs with the CI on. The peak latencies of p13 and n23 in patients with the CI on were markedly delayed compared with those in controls.

Discussion

In this study, we found that all the patients with inner ear malformation showed VEMPs with the CI on. In our previous study, we reported that 42% of patients with a normal inner ear on CT showed no VEMPs with the CI on [7]. The appearance rate of eliciting normal VEMPs seemed to be higher in patients with inner ear malformation than in patients with a normal inner ear on CT. The rate of detection of facial nerve stimulation in inner ear malformation has been reported to be higher in patients with inner ear malformations than in patients with a normal inner ear on CT [10]. The surgical procedures, current intensity, stimulation schemes, and testing paradigms may affect the appearance rate of VEMPs. In our study, the C levels were higher in patients with inner ear malformation than in the patients with a normal inner ear. This suggests that C levels may affect the appearance of VEMPs.

In our study, two patients showed VEMPs before CI and also showed VEMPs after CI with the CI on. This suggests that in these cases the sensory cells of both saccule and inferior vestibular neurons may be present. One patient showed no VEMPs before CI, but showed VEMPs with the CI on. This suggests that in this case the sensory cells of saccule may be absent, but the inferior vestibular neurons may be present. However, the possibility that these responses are of nonvestibular origin cannot be ruled out.

CC deformity has been reported to occur in 2% of patients with congenital profound sensorineural hearing loss [11]. In this inner ear malformation, the cochlea and vestibules form a common cavity, usually lacking an internal architecture. This deformity is regarded as hypoplasia of the cochleovestibular nerve or complete aplasia of the cochleovestibular nerve [12]. In embryos of approximately 5 weeks or less, the saccule is demarcated from the remainder of the vesicle; it sends out a single ventral evagination, the primordium of the cochlear duct. CC deformity most probably results from an arrest in otocyst development during the fourth gestational week [1]. In the human early developmental stage, neuroblasts of the cochlear ganglion separate from the otic epithelium at approximately the fourth gestational week. The vestibulocochlear nerve (CVN) develops at approximately 9 weeks of gestation [13]. Sennaroglu et al.

Table II. Summary of results of VEMPs.

			VEMP						
			Before CI			After CI			
Group	Patient no.	Type of inner ear malformation	Age at recording (years)	Operated ear	Nonoperated ear	Age at recording (years)	CI off	CI on	Nonoperated ear
Group A	1	Common cavity	5	+	+	6	+	+	+
	2	IP-II VLAS	5	+	+	5	−	+	No recording
	3	IP-I, narrow IAC	5	−	−	5	−	+	−
Group B	4	Common cavity	No recording	No recording	*	6	−	+	+
	5	IP-I	No recording	No recording	*	8	−	+	+
	6	IP-I	No recording	No recording	*	7	−	+	−
	7	Narrow IAC	No recording	No recording	*	5	−	+	−

Table III. Latency of VEMPs in patients and controls.

	Latencies of VEMPs (ms)									
	Before CI				After CI					
	Operated ear		Nonoperated ear		CI off		CI on		Nonoperated ear	
Patient no.	p13	p23	p13	p23	p13	p23	p13	p23	p13	p23
1	9.1	15.1	12	19.8	15	23	13.5	19	11	17
2	9.9	14.7	11	17.8	–	16.6	23.8	*		
3	–	–	–	15.8	21.3	–				
4	*	*	–	16.6	21.6	12.6	16.5			
5	*	*	–	15.5	22.1	12.4	18.1			
6	*	*	–	17.3	24.5	–				
7	*	*	–	17.8	23.2	–				

The peak latencies of p13 and n23 in patients with the CI on are markedly delayed compared with those in controls.
*Controls ($n=9$): mean latency of p13 = 10.5 ± 0.5 ms; mean latency of n23 = 16.0 ± 1.4 ms.
*No recording.

reported that a patient with a common cavity, who had a 'common CVN' without branching into the cochlear and vestibular nerves, showed good benefit from CI, but showed nystagmus after CI [14]. This suggests that the cochlear and vestibular nerve fibers must be present in their CVN. However, the function of the inferior vestibular nerve remains unknown.

In the present study, patient 1 showed VEMPs in both ears before CI, and patient 4 showed VEMPs in only the nonoperated ear. This suggests that sensory cells of saccule and inferior vestibular neurons are present in patients with CC deformity. In patients 1 and 4, VEMPs were also present with the CI on. This suggests that inferior vestibular neurons are present in our patients with CC deformity, although the cochlea and vestibule form a common cavity.

Finally, we would like to emphasize that VEMPs are a useful tool for diagnosing the function of the saccule–inferior vestibular nerve system in inner ear malformations. Even in patients with inner ear malformation, there are patients with sensory cells of saccule and inferior vestibular neurons and patients with vestibular neurons only but no patients without sensory cells of saccule and inferior vestibular neurons in inner ear malformations.

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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Vowel perception of Cochlear Implant Users

- Listening vs. Lip reading -

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Introduction

As speech processing devices or coding strategies have progressed, speech perception in subjects using cochlear implant (CI) has become significantly improved. Some CI users, however, use signs or lip reading to communicate with others because of the limited auditory information. Comparison between speech perception with and without lip reading will provide information on degree of visual dependence, but it is difficult to unify the test conditions. In Japanese, most syllables end with vowels and thus vowel perception is highly involved in speech intelligibility. We prepared vowel perception test to analyze the contribution of visual and auditory inputs during vowel processing in CI uses. Results in normal hearing (NH) participants were also obtained and compared with those in CI users.

Method

We enrolled 30 adults (54.3±16.9 years old, 47.3±38.5 months of implant use) and 40 children(9.2±3.5 years old, 50.6±24.5 months of implant use) who received CI. 10 adults (37.0±11.9 years old) and 11 children (9.45±1.6 years old) who have NH also participated as controls. The auditory stimuli were 5 Japanese vowels (/a/, /e/, /i/, /o/ and /u/) separately uttered by a female speaker. The visual stimuli were apparent motion that was succession of pictures of woman's face, and 6 images were used: neutral lip form without any movement, and 5 lip forms articulating Japanese vowels. These stimuli were sequenced by the SuperLab software. The following three

test conditions were used: 1) AV, the matched pairs of visual and auditory stimuli (e.g. lip form /a/ and voice /a/); 2) AV-mismatch, the inconsistent pairs of stimuli (e.g. lip form /a/ and voice /i/); and 3) A-only, neutral lip form and auditory stimuli. Each test condition was divided into two blocks and 80 pairs in each conditions were presented in a random order within two blocks. The order of presentation of six blocks in total was counterbalanced across subjects. During the session, the subjects were instructed to judge which vowel were said, and to press as fast as possible one of the five plastic buttons that show /a/, /e/, /i/, /o/ and /u/. Correct rate and reaction time were measured.

Results

Correct rate in each condition was significantly higher in adult NH subjects compared to adult CI users ($P<.01$). In contrast, the correct rate of NH children was significantly higher in AV-mismatch ($P<.001$) and A-only($P<.05$) condition than that of CI children, but no significant difference of correct rate was observed in AV condition between them.

Significant main effects of presented condition were observed in both adult and pediatric CI users. Correct rate of AV-mismatch condition was significantly lower than that of AV ($P<.05$) and A-only ($P<.05$) conditions .

Reaction time of each condition was significantly longer in CI users than that of NH subjects in both age groups. Moreover, CI users took longer reaction time in AV-mismatch condition compared to other conditions ($P<.05$).

In adult CI group, strong correlation was found between the monosyllable test score and the correct rate in AV-mismatch condition ($r=.79$) (fig.1a). Inverse correlation was also found between the monosyllable score and the "lip reading rate" in AV-mismatch condition, in which they responded according to the lip shape ($r=-.78$). The same correlation, albeit weaker, was present in pediatric CI group ($r=.44$ and $r=-.48$) (fig.1b). No correlation was found between correct rate in AV-mismatch condition and age or duration of CI use in CI users of both age groups.

Discussion

It has been reported that the speech perception in NH subjects is influenced by speech-read cues. AV-mismatch condition of our test, however, was too easy for the NH subjects regardless of the age to influence on their speech perception because lip movement in AV-mismatch condition was processed as negligible information. On the

other hand, these plain tasks extracted particular response pattern in CI users. Lower correct rate in AV-mismatch condition of CI group indicated that the visual information significantly influence on their speech perception. We found that the degree of visual dependence of adult CI users was correlated with speech discrimination score, but not age or duration of CI use. Similar tendency was observed in pediatric CI users, although the correlation was weaker. This might be due to that there were some outlier subjects who depended highly on visual information in this task in spite of their good speech intelligibility. Their age at implantation or communication mode before implantation might be responsible for this result.

In the current study, the control group was not age-matched, which may explain why average reaction time was longer in CI group than in controls, but could not explain the prolonged reaction time in AV-mismatch condition within CI subjects. Among hearing impaired people, the processing visual information in speech comprehension might be so complicated that they took longer to process incongruent combination of visual and auditory stimuli.

Conclusion

The test we use in the current study was simple task even for children, and it could show numerically the degree of visual dependence. The degree of visual dependence was correlated with speech intelligibility, although some exceptions were observed in children. This task is useful to examine children who undergo CI at very young age and also to follow the developmental change of degree of visual dependence in speech perception.

Cochlear implants in children with inner ear malformations

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Summary

Nineteen children with inner ear malformation who underwent cochlear implantation (CI) were retrospectively reviewed. High-resolution CT (HRCT) scans demonstrated a common cavity deformity (CC) in 2 patients, an incomplete cochlear partition (IP) in 12, an enlarged vestibular aqueduct (LVA) in 4, and partial semicircular canal aplasia in 1. Four patients also exhibited bilateral narrow internal auditory canals (nIAC). A cerebrospinal fluid (CSF) gusher was encountered in 4 patients. No patients experienced postoperative complications. Patients with bilateral nIAC, and one with CC had poor language acquisition. All but one patients with partial semicircular canal aplasia, LVA, and simple IP showed a significant improvement in speech perception.

Introduction

Cochlear malformations have been reported to occur in approximately 20% of the patients with congenital sensorineural hearing loss[1)]. Previously, such case was regarded as a contraindication for CI [2)] because of the increased risk of surgery and the uncertain performance. Here we report our surgical experience and the performance of CI in patients with cochlear malformation.

Material and Methods

Between 1999 and 2008, 109 cochlear implant procedures have been done in our department. During this period, we had 19 patients (17%) with inner ear anomalies. In these patients, data concerning surgery, postoperative follow-up, and audiometric findings before and after implantation, including audiometric thresholds, meaningful auditory intergration scale (MAIS), meaningful use of speech scale (MUSS), scores with open set monosyllable, open set word, and closed set sentence, were analyzed.

Results

HRCT findings, surgical procedures, and surgical complications are shown in Table 1. In all patients, electrodes were successfully inserted. Insertion of the electrode into the IAC did not occur in any patient. CSF gusher was encountered in 3 patients with IP and one with LVA, in all of which the CSF leakage was stopped by packing the cochlear cavity with soft tissue followed by fibrin glue application. In no patient was a spinal drain required. Facial nerve stimulation in postoperative NRT examination was seen in

8 (42%) patient, but it was successfully managed by arranging the electrode programming. One patient (patient 6) had recurrent meningitis postoperatively despite the absence of CSF gusher at the surgery. It was proved 2 years later that the cause of meningitis was CSF leakage from the defect of the stapes footplate in the opposite ear.

Pre and post implantation audiometric findings are summarized in Table 2. All patients except one (patient 6) with simple IP, LVA, or partial semicircular canal aplasia showed a significant improvement in speech perception. Patient 6 had relatively poor score, which is considered due partly to insufficient auditory rehabilitation because of the recurrent episodes of meningitis. Among 4 patients with bilateral nIAC, 2 demonstrated the poorest performance in the postoperative speech perception tests and thus were encouraged to use sign language for language development. The other 2 patients with bilateral nIAC had relatively better sound perception, but their language development was too limited to obtain open speech perception. A nIAC (<2mm in diameter[3]) implies the aplasia of the cochleovestibular nerves, which is known to result in very poor speech perception and language development after CI. Constructive interference with steady state MRI, albeit still imperfect, may provide information regarding the presence or absence of the cochlear and vestibular nerves in such cases. Among 2 patients with CC, one (patient 18) who underwent CI at age of 3 started to perform well in speech perception and language development postoperatively. Another (patient 17) who underwent CI at nearly 6 years of age showed only marginal improvement. This suggests that better language acquisition might be achieved by earlier implantation in patients with CC. In patients with severely malformed cochlea, necessity for frequent reprogramming has been reported because of the migration of the implant [2]. None of our patients encountered fluctuation of thresholds requiring frequent reprogramming.

Conclusions

CI can be successfully performed in children with inner ear malformations. Many of these children are expected to achieve significant auditory benefits from this intervention. However, the postoperative performance is commonly poor in most patients with nIAC and CC, which should be informed to their parents during preoperative counseling.

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COCHLEAR IMPLANTATION FOR PATIENTS WITH MITOCHONDRIAL DNA A3243G MUTATION

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Summary

Maternally inherited diabetes and deafness (MIDD) is known to be associated with an A to G point mutation at nucleotide pair 3243 in mitochondrial DNA. We performed cochlear implantation (CI) in four patients with MIDD. One patient was diagnosed as having also cardiomyopathy. The A3243G point mutation was confirmed in all patients, with the heteroplasmy ranging from 6 to 30 %. All patients underwent CI without any trouble intra- or postoperatively. The open-set monosyllable, word, and sentence recognition scores were found to be better than the average scores in adult CI users with deafness of other causes. Literature review also demonstrated that patients with mitochondrial deafness are good candidates for CI.

Introduction

Sensorineural hearing loss (SNHL) occurs in approximately half of patients with mitochondrial disorders. To date, genetic testing has identified mutations in mitochondrial DNA (mtDNA) in more than 3% of persons with SNHL in Japan. Mitochondrial SNHL may be nonsyndromic or syndromic (associated with multiple disabilities ranging from diabetes to strokelike episodes), and commonly involves the cochlea, predominantly the stria vascularis, hair cells, and spiral ganglion cells [1-4)], implying that deaf patients with mitochondrial disorders may be good candidates for CI. In fact, Sinnathuray et al. [5)] have demonstrated by reviewing literatures that patients with mitochondrial deafness can be successfully rehabilitated by CI. Here we report our experience of CI for 4 adult patients with MIDD associated with mitochondrial A3243G mutation.

Material and Methods

Four unrelated patients with deafness associated with an A-to-G transition at np 3243 in the mtDNA who underwent CI were enrolled. Clinical features and postoperative audiometric findings were evaluated in these patients.

Results

Clinical features of the patients are shown in Table I. All of them were female and had diabetes and mellitus. One patient (No 4) had also cardiomyopathy. Family pedigrees showed maternal inheritance of diabetes and/or hearing impairment over two or three generations. None of their family members had clinical symptoms characteristic of mitochondrial encephalomyopathy. Age at the onset of hearing loss ranged from 28 to 40 years. The percentage of mutant mtDNAs in the peripheral leukocytes ranged from 6 to 30 %; hearing loss appeared at a younger age in patients with higher heteroplasmy.

All patients underwent CI without any trouble intra- or postoperatively. Since activation of the implant, they used CI successfully and continuously. Postoperatively, all patients showed good performance (Table 2). They had good speech recognition all in open-set monosyllable, word, and sentence tests. The average of these tests was 52, 77, and 71, respectively, in the hearing-only condition and 68, 80, and 90, respectively, in the visual-plus-hearing condition. These scores were better when compared to adult subjects with deafness of other causes such as meningitis.

Discussion

The current case presentation demonstrated that speech recognition was good in adult female with MIDD associated with A3243G point mutation in mtDNA. Sinnathuray et al. [5] reviewed reports of CI for patients with mtDNA mutations in a variety of countries and found that despite the variety in mitochondrial mutations, this heterogeneous group of cochlear implantees have done well, with 58% conversing on the telephone and the remainder mostly having good open-set speech recognition and no quoted complications. These results suggest that pathologic changes in mtDNA mutations primarily involve a cochlear site, which is in good agreement with the histopathology reported in patients with deafness due to mitochondrial disorder, such as Kearns-Sayre syndrome and MIDD [2-4].

In conclusion, the current study, together with a literature review, indicates that CI is a valuable choice of therapy for patients with deafness associated with the A3243G

mutation in mitochondrial DNA. The excellent auditory performance with CI suggests that hearing loss associated with this mutation is primarily caused by insult to the cochlear tissue containing rich mitochondria (i.e., hair cells and stria vascularis).

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Table 1

No	Age at CI (y)	Gender	Heteroplasmy (%)	Onset HL (y)
1	53	F	30	30
2	46	F	29	28
3	44	F	9	35
4	65	F	6	40

Table 2

	Monosyllable (%)		Word (%)		Sentence (%)	
No	A	A+V	A	A+V	A	A+V
1	36	48	66	74	44	82
2	40	54	68	76	65	89
3	60	82	88	76	88	94
4	70	88	86	95	86	95

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人工内耳装用症例における静寂下・騒音下での補聴器装用効果の検討

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要旨：両耳聴取の利点は様々認められており，海外では両耳人工内耳により，聴取成績の向上が報告されている。片耳装用しか認められていない本邦では，人工内耳の対側耳に補聴器を装用することが一般的である。今回我々は，対側補聴器併用の利点について，日本人患者で検討した。静寂下条件では，片側人工内耳装用と，人工内耳と対側補聴器併用の場合との間に，聴取成績に有意な違いはなく，装用期間等ほかの要因を考慮しても違いはなかった。騒音負荷条件でも，補聴器併用による有意な正答率の向上は認められなかった。今回の研究で，対側補聴器の効果が認められない理由として，人工内耳の適応条件が厳しいこと，症例数不足により有意差が出なかった可能性，日本語と欧米言語の言語的な特徴の違いによる要因などが考えられた。今回の結果から，人工内耳装用者の聴取能力を向上には，両側人工内耳装用などにより対側耳をより積極的に活用することが必要と考える。

ーキーワードー
両耳補聴，小児人工内耳，補聴器，言語獲得，カクテルパーティー効果

はじめに

両耳聴の効果は，正常者においても，音の方向感の認知やカクテルパーティー効果[1)]による騒音下の聴取能力など，さまざまな利点があげられている。難聴者においては両側補聴器装用による聴取能力の向上が認められている[2)]。

人工内耳は1973年に3Mにより商品化されて以来，聴覚を失った患者に対して広く用いられている。人工内耳装用症例において，海外では両耳装用についての報告が増加しており，一般的になりつつある[3)]。これらの報告では，両耳装用による騒音下での聴取の向上など，さまざまな効果が認められている[4)]。本邦では，人工内耳は，1991年に輸入認可，1994年に保険適応となり，一般的に行える治療法となったが，現在まで片耳装用しか認められていない。海外では片耳装用の場合でも人工内耳の対側耳に補聴器を装用することにより，聴取能の向上が認められている[5)]。本邦でも人工内耳対側耳への補聴器装用は一般的に行われているが，その効果に関する報告は少なく，本邦でのエビデンスは確立しているとはいえない[6)]。我々の施設においても，海外の文献に基づいて聴取能の向上を期待して人工内耳対側耳に補聴器装用を行っているが，今回，我々の施設の症例に対し，人工内耳単独装用と比較して補聴器併用時に有意な聴取能向上が得られるかにつき，静寂下および騒音負荷下の条件において検討を行った。

対象と方法

1997年から2008年にかけて，東京大学医学部附属病院耳鼻咽喉科において人工内耳埋込術を施行され