EABR-based evaluation of the spatial distribution of auditory neuronal tissue in

common cavity deformities

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Abstract

Objective: In a common cavity deformity (CC), the cochlea and vestibule are confluent to form a single cavity without internal architecture and distribution of auditory neuronal tissue is unclear. The purposes of this study are to reveal the spatial distribution of auditory neuronal tissue in CC using electrically evoked auditory brainstem response (EABR) during cochlear implantation.

Study Design: Retrospective case review.

Setting: Cochlear implant (CI) center at a tertiary referral hospital.

Patients: 5 patients with CC who underwent cochlear implantation and intraoperative EABR testing.

Main Outcome Measures: Spatial distribution of electrodes which elicited an evoked wave V (eV) in EABR testing was evaluated in each CC.

Results: EABR testing demonstrated that electrodes attached on the inner wall of the antero-inferior cavity of the CC successfully elicited a reproducible eV in all cases and the latency of each eV was an approximately 4 msec, which is similar to those reported in patients without an inner ear malformation. Interestingly, in Case 1 with the lowest percentage of eV-positive electrodes (31.8%), CI-aided audiometric thresholds were changed, depending on the frequency allocation to eV-positive electrodes in the programming. CI-mediated facial nerve stimulation was observed in 3 of 5 cases and results of EABR

testing were useful for optimizing the device program to decrease facial nerve stimulation without sacrificing CI-mediated auditory performance.

Conclusions: The results of EABR testing suggested that auditory neuronal elements are distributed to the antero-inferior part of CC, mainly around or near the inner wall of the cavity. In cases with CC, EABR testing is useful to achieve the optimal electrode array placement and to adjust programming parameters of the implanted device, which might be essential to maximize CI outcomes and to decrease facial nerve stimulation.

Key Words: cochlear implant, common cavity, EABR, inner ear malformation, intraoperative, auditory nerve

INTRODUCTION

Inner ear malformations account for about 20-30% of congenital severe and profound

hearing loss and many children with an inner ear malformation are undergoing cochlear

implantation (1,2). In 1987, Jackler et al originally proposed a classification of inner ear

malformations based on the hypothesis in which termination of ordinary inner ear

development leads inner ear malformations and the type of malformations, including Michel

deformity (labyrinth aplasia), cochlear aplasia, common cavity deformity (CC), cochlear

hypoplasia, and incomplete partition, corresponds to each step of inner ear development (3).

Later, Sennaroglu developed Jackler's classification and further divided cochlear hypoplasia

and incomplete partition into cochlear hypoplasia type I-III and incomplete partition type I-III,

respectively (2,4). In addition to accumulating evidences for the association between inner

ear malformations and genetic mutations (5-8), a limited variety in the shape of a malformed

bony labyrinth between patients with the same class of malformations suggests that the

majority of inner ear malformations are a result of genetic etiology (2). CC is the second most

frequent inner ear malformation in which the cochlea and vestibule are confluent to form a

single cystic cavity without internal architecture. Cochlear implantation in children with CC

is a challenge to clinicians because of its difficulty in array placement, high risk of

cerebrospinal fluid (CSF) gusher and misinsertion into the internal auditory canal (IAC), and

high incidence of facial nerve abnormalities (1,2,9). In 1997, McElveen described a

transmastoid labyrinthotomy approach for cochlear implantation in CC, which has been

widely accepted as a surgical method for placement of the electrode array in CC while

minimizing risk of injury to the facial nerve (10).

Cochlear implant (CI) electrically stimulates cell bodies of spiral ganglion neurons

and their fibers to restore afferent input to auditory central nervous system. Effective

CI-mediated stimulation of auditory neuronal elements should be necessary to maximize CI

outcome, but the spatial distribution of spiral ganglion neurons and auditory nerve fibers is

unclear in CC, due to no differentiation between the cochlea and vestibule in addition to the

lack of a modiolus which would contain spiral ganglion neurons. Hearing performance with

CI was reported to vary widely among CC cases (1), which might be caused by the difference

in the distribution of the auditory neuronal elements. Recently, electrically evoked auditory

brainstem responses (EABR) using CI-mediated stimulus are used for objective evaluation of

auditory neuronal responses in the brainstem in patients with or without an inner ear

malformation (1,11,12). In this study, we investigated the spatial distribution of auditory

neuronal tissue in CC using EABR and examined the utility of EABR data in optimizing

programing parameters of the implanted device.

MATERIALS AND METHODS

We retrospectively examined 5 patients with CC (Cases 1-5) with congenital

profound sensorineural hearing loss who underwent cochlear implantation at Kobe City

Medical Center General Hospital from 2005 to 2013 (Table 1). The average age at

implantation was 27.4 months and the average follow-up period was 26.0 months. The type

of inner ear malformations was determined using the CT-based classification published by

Sennaroglu (2). Discrimination between CC and cochlear aplasia is sometimes difficult (2)

and in this classification cochlear aplasia is distinguished from CC by a normal or dilated

vestibule and semicircular canals. Moreover, cochlear aplasia is located at the postero-lateral

part of the IAC fundus, whereas the IAC enters at the center of a cavity in CC. Our cases

were classified as CC based on the lack of at least one semicircular canal and the presence of

a cavity in the antero-inferior direction to the fundus. Since the long axis of the CC was often

not parallel to the axial plane, coronal CT images were more effective for identifying the

anterio-inferior cavity, which was visualized inferior to the fundus of the IAC. Since the

width of the midpoint of the IAC was >2.5mm in each case, no CC was associated with a

narrow IAC (2). MRI confirmed the presence of a vestibulocochlear nerve, but an isolated

cochlear nerve bundle was not detected in all patients. The use of human subjects in this

study was approved by the Research Ethics Committee of the Kobe City Medical Center

General Hospital.

Case 1 initially underwent cochlear implantation with the standard transmastoid

labyrinthotomy (10), followed by reimplantation using a modified transmastoid

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labyrinthotomy with a 3.0 mm-diameter hole at the postero-lateral wall of the CC for the electrode insertion. In the standard labyrinthotomy approach, a labyrinthotomy is created at the area where the lateral semicircular canal would normally be situated. The size of the labyrinthotomy is approximately 1.0 mm which is large enough for electrode insertion. The modified labyrinthotomy approach was developed from the standard labyrinthotomy approach by changing the size of a labyrinthotomy. In the modified labyrinthotomy approach, most of the postero-superior wall of the CC was removed to make a large hole of 3.0 mm of diameter, which allowed better access into the antero-inferior part of the CC. The IAC fundus could be identified through the large labyrinthotomy and a pre-bent electrode array was inserted into the antero-inferior cavity beyond the fundus of the IAC with the curved end of the electrode array foremost to prevent intrameatal placement or undesirable folding of the tip. Immediately after electrode insertion, we gently filled the cavity of the CC with soft tissue to push the electrode array in an antero-inferior direction for attaching electrodes on the wall of the antero-inferior cavity. The hole of labyrinthotomy was then covered by small pieces of bone and sealed by a thin layer of bone pate with fibrin glue. Continuous facial nerve monitoring was used to prevent facial nerve injury during mastoidectomy and labyrinthotomy. In the other patients, this modified transmastoid labyrinthotomy approach was conducted in the initial implantation. Nucleus device with 22 active electrodes (Ch1 to 22) including CI24RST, CI24REST, or CI422 was implanted in all cases. The first two of these devices

have full-banded electrodes, while CI422 has half-banded electrodes that are originally

designed to point toward the modiolus of the cochlea. CI422 was used in Cases 4 and 5, in

which the electrode array was turned around to position half-banded electrodes close to the

inner wall of the cavity.

Intraoperative EABR testing was performed with Nucleus Custom Sound EP

software (Cochlear Corp., NSW, Australia). The biphasic electrical stimuli with a stimulus

pulse width of 50 to 100 µsec and 200 to 230 current levels were delivered at 20 Hz of pulse

rate using MP 1+2 mode. Other conditions were defaults in the autoNRT program. The

EABR was recorded by Neuropack (Nihon Koden, Tokyo, Japan) with a filter setting of 20

Hz to 3K Hz on the opposite side to minimize artifacts of the implanted device. At least 500

sweeps were averaged. All EABR testing were performed without neuromuscular blockade to

detect CI-mediated facial nerve stimulation (FNS). The presence of an evoked wave V (eV)

was determined by (i) reproducible responses with amplitude >0.1 µV, (ii) a

current-dependent increase in amplitude, and (iii) latency of the wave >3 msec, which are

developed from the criteria in the previous report (13). Once a putative eV was identified, an

intensity of CI-mediated stimulus was increased stepwise by 3 or 5 current levels to confirm

current-dependent increase in the amplitude of the putative eV. During these sessions, only a

few step-increases of current intensity sometimes resulted in emergence of an unusual large

biphasic response. This response differed from the putative eV, which showed

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current-dependent increase in the amplitude in the previous sessions with a lower intensity of stimulus, in respect of its biphasic waveform, relatively large amplitude and late latency (4 to 7 msec). These characteristic features suggest that this response might result from myogenic activity, probably facial myogenic compound action potential, even though an obvious facial twitching was not observed during the EABR testing. As reported previously, the threshold for a facial myogenic compound action potential is significantly lower than the threshold for observable facial movement, supporting this conclusion (14). Comparison of results in EABR tensing between with and without the use of a neuromuscular blocking agent would be effective to evaluate contamination of myogenic compound action potentials in EABR, but we did not conduct EABR testing under neuromuscular blockade due to the limitation in time. In Case 1, EABR tests were also performed under sedation at one year after the initial implantation. The position of the inserted electrode array was evaluated by intraoperative X-ray and postoperative CT. Hearing outcomes with the CI were evaluated by hearing thresholds, speech discrimination scores of closed-set Japanese infant words, and category of auditory performance (CAP) scores (15) at 40 months after the reimplantation in Case 1 and 29, 22, 18, and 9 months after the implantation in Cases 2, 3, 4, and 5, respectively. The speech discrimination test was performed only if the CAP score was ≥4 (Table 1).

RESULTS

EABR testing after the initial CI in Case 1

In Case 1, X-ray during the initial cochlear implantation demonstrated that most of the electrodes were located within the CC, but the CI-aided performance was still poor (CAP score 1) even after one year use of CI. EABR testing under sedation at that time elicited a reproducible eV only at 2 of 11 tested electrodes with an odd number (18.2%, Ch17 and Ch19). The latency of the detected eVs was approximately 5 msec (Fig. 1A) and considerably longer than 4.05 msec which was previously reported in patients without an inner ear malformation at one year after implantation (16). Postoperative CT indicated that the electrode array was fully inserted in the CC, but did not reach the antero-inferior cavity due to stacking at the fundus of the IAC (Fig. 2A and B). The eV-positive Ch17 and 19 appeared to locate outside but near the antero-inferior cavity of the CC (Fig. 1B and C). These results suggested suboptimal electrical stimulation of auditory neuronal elements was responsible for the inadequate auditory performance.

EABR testing during the reimplantation in Case 1

To overcome this problem, we performed reimplantation using the modified transmastoid labyrinthotomy with a larger hole for electrode insertion to achieve better access into the antero-inferior cavity. EABR testing during the reimplantation revealed a clear eV at 7 of 22 tested electrodes (31.8%, Ch10-16) and the latency of eV ranged from 3.8 to 4.1 msec (Fig.

1D), similar to 4.05 msec which was reported in patients without an inner ear malformation

(16). X-ray and CT demonstrated that these 7 electrodes with a positive eV located at the

curved end of the bent array which was inserted into the antero-inferior part of the CC (Fig.

1E), attaching on the inner wall of this area (Fig. 2C and D).

EABR-based programing in Case 1

After the reimplanation, frequency-specific hearing thresholds in Case 1 changed

depending on the frequency-to-electrode allocation in the programming. When frequencies of

188 to 7938 Hz were allocated to all 22 electrodes, the hearing thresholds were 40 dB at 1000

and 2000 Hz, for which Ch10-16 with a positive eV were responsible, but over 80 dB for

other tested frequencies. On the other hand, when frequencies from 188 to 5063 Hz were

allocated to 8 sequential electrodes, including eV-positive Ch10-16, the hearing thresholds

ranged from 30 to 50 dB at all tested frequencies (Fig. 3). Even after the reimplantation,

CI-mediated FNS was induced by electrical stimulation of Ch10-12 and Ch16-20. Since

Ch17-20 failed to elicit eV and seemed not to contribute to auditory perception, these

electrodes were deactivated. Regarding Ch10-12 and Ch16 with a positive eV, the maximum

stimulation level was set to a value below the threshold of CI-mediated FNS.

EABR testing and EABR-based programing in the other CC cases

In the other four patients who underwent the modified transmastoid labyrinthotomy at the

initial implantation, postoperative CT showed the optimal position of the electrode array

(data not shown). Even though the size and shape of each CC differed among the cases (Fig.

4A-D), electrodes inserted in the antero-inferior cavity successfully elicited eVs in all 4 cases,

similarly to Case 1 (Fig. 4E-H). As shown in Fig. 4I-L, reproducible eVs were detected in 9

of 11 tested electrodes with an even number (81.8%) in Case 2, 8 of 11 with an odd number

(72.7%) in Case 3, 12 of 22 electrodes (54.5%) in Case 4, and 5 of 11 with an even number

(45.5%) in Case 5. The latency of the detected eVs was approximately 4 msec in all cases

(Fig. 4I-L). Using program in which several electrodes without eV at the most distal and/or

proximal part of the electrode array were deactivated, their pure tone hearing thresholds were

40 or 45dB at 500, 1000, and 2000 Hz except for 50dB at 2000Hz in Case 5 who had used his

CI for only 9 months. Cases 2 and 4 suffered from CI-mediated FNS and almost all

electrodes were responsible for this aversive symptom. In both patients, deactivation of some

electrodes without eV in addition to a decrease of current levels of the other electrodes

including eV-positive ones was effective to reduce FNS.

CI outcomes and other CI-related problems in CC cases

Before implantation, no patient could detect sounds, indicating that their preoperative CAP

score was zero, but auditory perception improved after activation of the CI in all patients

(Table 1). The postoperative CAP score reached to 6 in Cases 1 and 2 who had utilized their

CI for over 2 years, indicating that they understood common phrases without sign language

or lip-reading. Speech discrimination scores of closed-set infant words were 76% and 80% in

Cases 1 and 2, respectively. The other 3 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 (Table 1).

Minor CSF leakage occurred during implantation in Case 4, which was easily stopped by

gently packing several pieces of soft tissue inside the cavity followed by sealing the

labyrinthotomy site with periosteum and bone pate. After implantation, Cases 1 and 2

suffered from dizziness, which spontaneously disappeared within a week (Table 1).

DISCUSSION

The present study demonstrated that reproducible eVs were elicited by electrical

stimulation of electrodes which were located at the antero-inferior part of the CC in all

patients. Even though the electrode array was almost fully inserted in each CC using the same

surgical procedure, the percentage of electrodes with a positive eV varied widely between

patients, ranging from 31.8% to 81.8%. Although the percentage of eV-positive electrodes

seems to be influenced by the size of the antero-inferior part of CC, it is difficult to predict

the exact number and position of eV-positive electrodes on the basis of radiographic findings,

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suggesting the importance of EABR testing in cases with CC. Interestingly, in Case 1 with

the lowest percentage of eV-positive electrodes, CI-aided audiometric thresholds were

changed, depending on which frequency was allocated to the eV-positive electrodes in the

program of the implanted device. Both these electrophysiological and audiometric data

indicate that auditory neuronal elements are mainly distributed in the antero-inferior part of

the CC. CC is thought to be caused by an arrest in differentiation of the otic vesicle during

the fourth gestational week (3,4). In the normal development of an inner ear, the ventral

portion of the otic vesicle elongates in the ventral direction, initiating cochlear development

(17); therefore, the antero-inferior part of CC might be programmed to differentiate to a

cochlea. These findings support our conclusion regarding the antero-inferior distribution of

auditory neuronal tissue in CC.

In Case 1, moving the electrode array closer to the inner wall of the antero-inferior part of

the CC shortened the eV latency from approximately 5.0 msec to 3.8-4.1 msec, which is close

to 4.05 msec measured in patients without inner ear malformations (12). Given that changes

in stimulus intensity do not significantly affect latencies of evoked waves in EABR testing

(12), the eV latencies of this study are comparable to those of the previous study, even though

an amplitude and pulse width of EABR stimulus are slightly different between studies. Thus,

the eV latency of Case 1 at the reimplantation suggests that auditory neuronal elements

mainly distributed around or near the inner wall of the antero-inferior part of the CC. The

longer distance between a stimulating electrode and neuronal tissue at the initial implantation

might have caused ineffective auditory stimulation in Case 1. This conclusion is supported by

the previous histological study which reported that neural elements are likely to lie in the wall

of CC (18). Another possible explanation for the change in the latency of the waves after the

reimplantation is that the detected waves in the initial EABR were not auditory neural

responses, but the myogenic compound action potential caused by facial nerve stimulation.

However, the Ch17- or Ch19-mediated facial twitching was not observed in the EABR

testing and electrical stimulation of other electrodes located more near the facial nerve, such

as Ch11 and Ch13, elicited no obvious response, suggesting the waves elicited by stimulation

of Ch17 and Ch19 in the initial EABR testing may be auditory neural responses rather than

myogenic responses.

We would also emphasize that MRI failed to identify an isolated cochlear nerve

bundle, but the vestibulocochlear nerve contained sufficient cochlear nerve fibers to transmit

the CI-mediate auditory signals to the brainstem in all CC cases. Since CC is thought to be

caused by developmental arrest before differentiation between the cochlea and vestibule (3,4),

it is possible that a cochlear nerve is not separated from vestibular nerves to constitute a

single vestibulocochlear nerve as the cochlea and vestibule are represented by a single

chamber.

In the present study, 3 out of 5 cases (60%) suffered from CI-mediated FNS, which is

consistent with the previous study reporting the high frequency of FNS among implanted

patients with inner ear malformations (14,19,20). Since electrical stimulation close to the

facial nerve as well as exceeding or leakage electrical current causes FNS, deactivating the

responsible electrodes or reducing current levels in these electrodes is usually effective to

reduce FNS. On the other hand, in cases with a sever inner ear malformation, high current

level and/or increased pulse width are often required to achieve good auditory performance

(11,20), suggesting a necessity to adjust the current level to an appropriate value which is

high enough to provide sufficient auditory input, but lower than the threshold for FNS.

Usually, programing parameters of the device is adjusted based on the patient's auditory

behavioral responses. Cases 1, 2, and 4, however, suffered from FNS before development of

their auditory performance and furthermore, many electrodes were responsible for the FNS,

showing difficulty in making an appropriate program of their CI. Previous studies

demonstrated that nonbehavioral measures including EABR are useful in the determination of

useful cochlear implant stimulation levels, particularly in young children and infants with

limited auditory experience (21). In this study, although precise EABR thresholds were not

examined, we gradually and carefully increased a current level and pulse width of electrical

stimulus in each electrode by refereeing to the amplitude and pulse width of the electrical

stimulus that had evoked eV in intraoperative EABR testing and if FNS was observed, we

decreased the current levels (amplitudes) at the responsible electrodes below the threshold of

FNS or sometimes deactivated these electrodes. This method might be useful to decrease FNS without sacrificing CI-mediated auditory responses especially before the patients show clear auditory responses.

Based on the experience of Case 1, we used the modified transmastoid labyrinthotomy with a large hole in the other 4 CC cases at the initial implantation. Although the shape of the CC varied among these cases, the optimal electrode array placement was achieved and a reproducible eV with approximately 4 msec of latency was elicited by electrical stimulation of electrodes in the antero-inferior cavity in all cases. These results suggest the effectiveness of this approach for cochlear implantation in CCs, especially when the antero-inferior cavity is small like the CC in Case 1. Regarding this modified transmastoid labyrinthotomy approach, however, three points require attention: (i) damage to the auditory neuronal tissue in the CC, (ii) probable difficulty in control of CSF leaks, and (iii) damage to the vestibular system. Atraumatic round window insertion techniques have attracted recent attention for preservation of residual hearing (22,23). In cases with CC, the patients are usually deaf before implantation, but it is possible that less trauma in auditory neuronal elements in CC leads to better CI-aided outcomes. In the present study, the results of EABR testing in Case 1 clearly demonstrated improved auditory brainstem responses after reimplantation with the modified transmastoid labyrinthotomy, suggesting that optimal array placement attached on the inner wall of the antero-inferior cavity might be more important than atraumaticity at least

in this patient. In Case 4, the IAC and CC were connected through a small defect of the IAC

fundus and CSF leakage after the labyrinthotomy was stopped by plugging the IAC fundus

with pieces of soft tissue. However, if CC widely communicates with the IAC, a risk of a

postoperative CSF leakage would increase because the larger labyrinthotomy leads the more

difficulty in achieving a complete seal to manage a CSF leak. The third issue is that the

posterior part of the CC, which may correspond to a primitive vestibule, is destroyed in the

modified transmastoid labyrinthotomy. A previous study showed vestibular evoked myogenic

potentials in some cases with CC (24), suggesting that destruction of the posterior part of the

cavity might impair vestibular function on the operated side. In fact, Cases 1 and 2 showed

mild disequilibrium after surgery, but their symptom   disappeared within a few weeks,

probably due to vestibular adaptation (25) or recovery of some vestibular function. However,

the long-term effect and safety of the modified transmastoid labyrinthotomy approach has not

been established, we have to closely monitor auditory and vestibular performance of our 5

CC cases for a long time.

Postoperative CAP scores varied widely between patients in this study. Since the

follow-up period was different between patients, the short duration of CI use might be

responsible for immature auditory development especially in Cases 4 and 5. Long-term

observation for up to 4 years will be required to lead a definite conclusion as described by the

previous study (26). What is noteworthy here is that Case 1 who showed eV only at 7 of 22

electrodes (31.8%), 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 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 the long-term use of the CI with an appropriate program. Previous studies using CI patients without an inner ear malformation demonstrated that the speech discrimination saturated around 8 electrodes and did not improve when more electrodes were activated (27,28). In Case 1, at least 7 electrodes successfully activated auditory neurons in the brainstem and this number might be enough to achieve sufficient CAP and infant word discrimination scores, although spatial distribution of auditory neuronal elements might be different between the CC and a cochlea without a malformation. Since we examined only 5 CC patients and the follow-up duration was short in this study, further investigation should be necessary to reveal a relationship between the number of eV-positive electrodes and CI-mediated auditory performance.

CONCLUSION

The present study using EABR testing demonstrated that auditory neuronal tissue is distributed in the antero-inferior part of CC, mainly around or near the inner wall of the cavity in all cases, regardless of the fact that a shape of the CC was widely different between

patients. In cases with CC, EABR testing is useful to achieve the optimal electrode array

placement and to adjust programming parameters of the implanted device, which might be

essential to maximize CI outcomes and to decrease facial nerve stimulation.

Declaration of interest

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Figure Legends

FIG. 1. Results of EABR testing in Case 1 before and after the reimplantation. A: EABR

testing after the initial implantation with the standard labyrinthotomy approach shows eVs in

Ch17 and Ch19 among the 11 tested electrodes with an odd number. The latency of these eVs

is approximately 5 msec (arrowheads). B: A maximum intensity projection image of

T2-weighted MRI of the CC on the implanted side. The antero-inferior part of the CC (AI) is

smaller than the postero-superior part (PS). C: X-ray at the initial implantation demonstrates

that Ch17 and Ch19 with a positive eV in EABR testing (circles) seem to be outside but near

the antero-inferior part of the CC (dotted line). D: EABR testing after the reimplantation with

the modified labyrinthotomy approach shows a distinct eV in 7 of 22 electrodes. The latency

of these eVs ranges from 3.8 to 4.1 msec (arrowheads). E: X-ray after the reimplantation

demonstrates that electrodes with a positive eV (circles) are located at the curved end of the

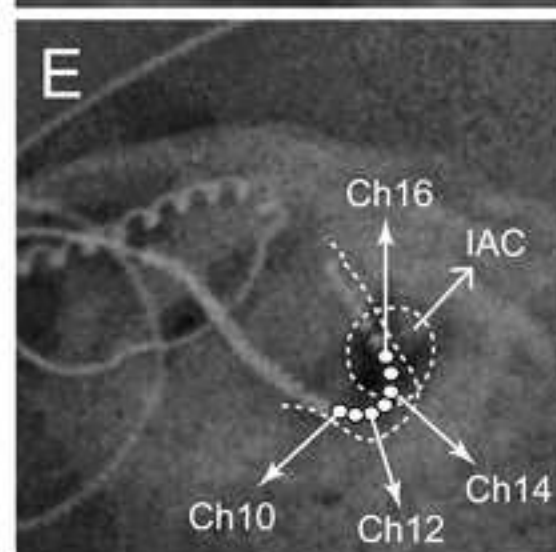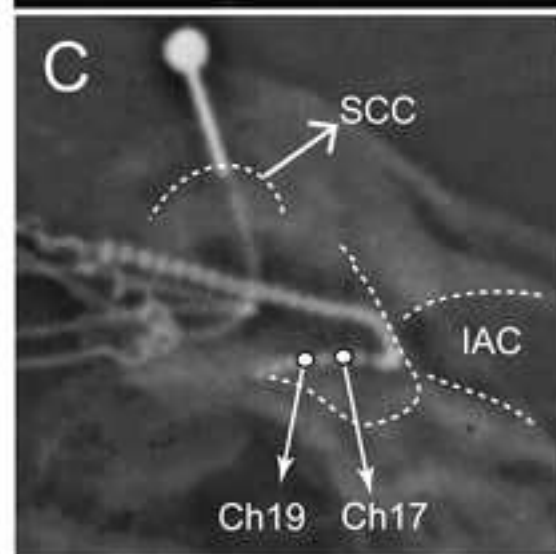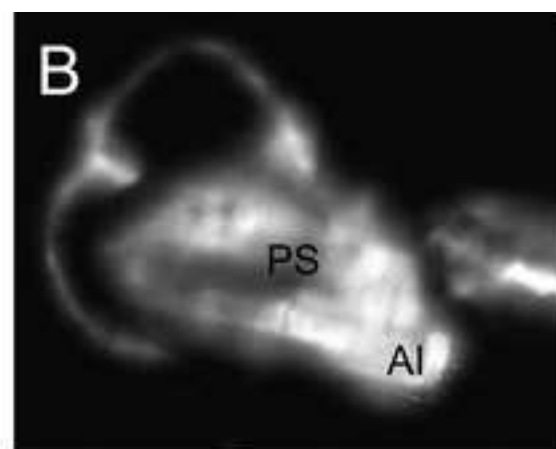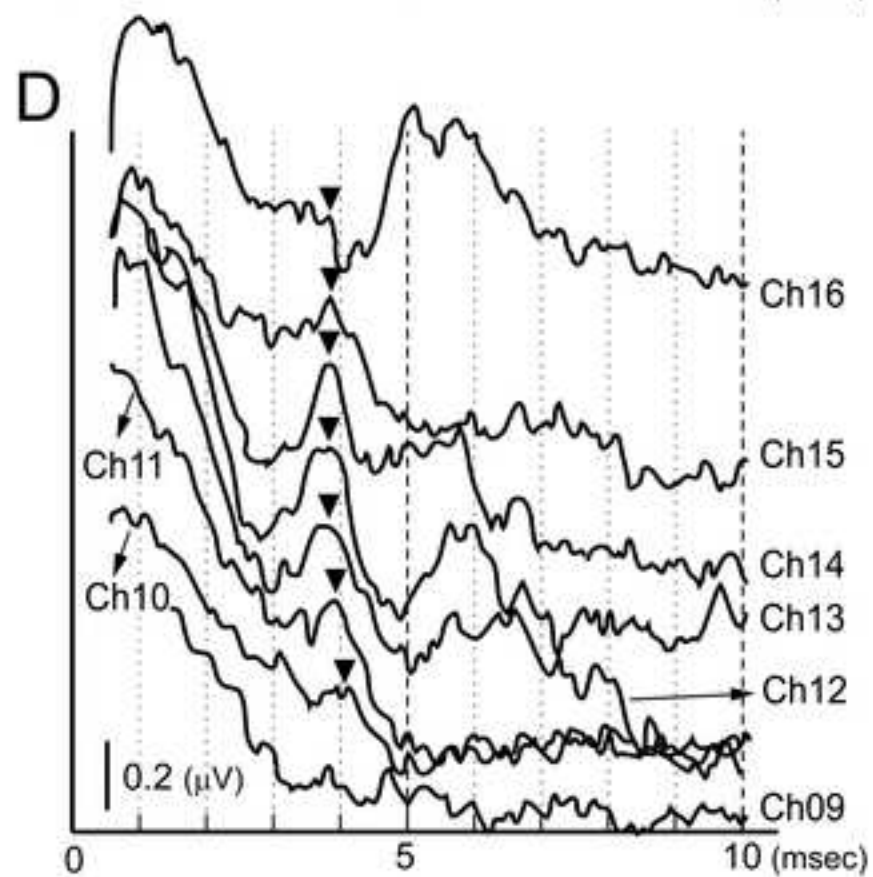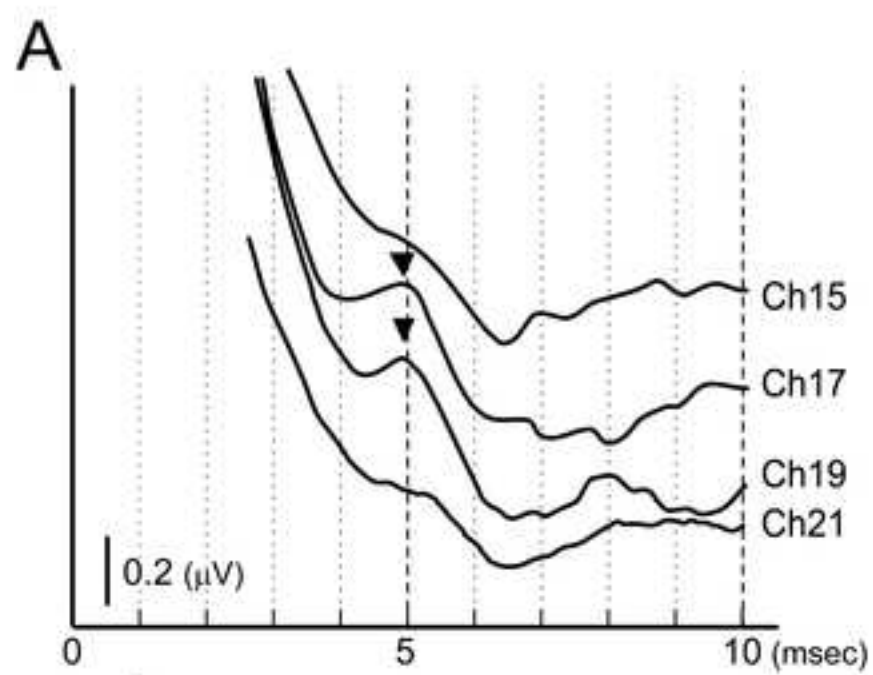pre-bent electrode array inserted in the antero-inferior part of the CC (dotted line).

FIG. 2. Evaluation of the electrode array placement by postoperative CT in Case 1. A and B:

Coronal CT images after the initial implantation with the standard labyrinthotomy approach.

The electrode array inserted from the hole of the standard labyrinthotomy (black arrows) is

stuck at the fundus of the IAC (arrowheads) and does not reach the antero-inferior cavity

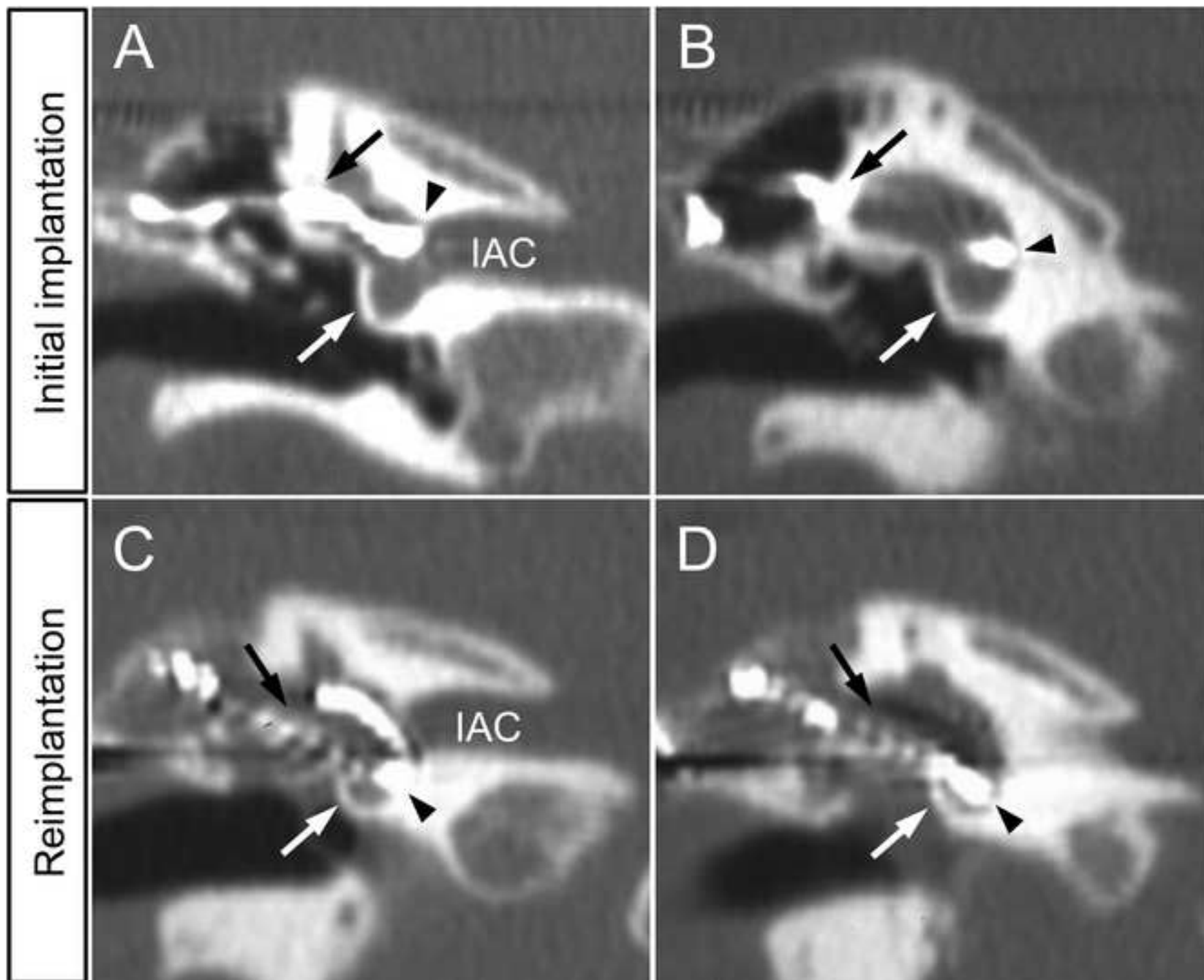(white arrows). C and D: Coronal CT images after reimplantation with the modified

labyrinthotomy approach with the 3.0mm-diameter hole on the postero-lateral wall of the

cavity (black arrows). The curved end of the pre-bent electrode array (arrowheads) is attached

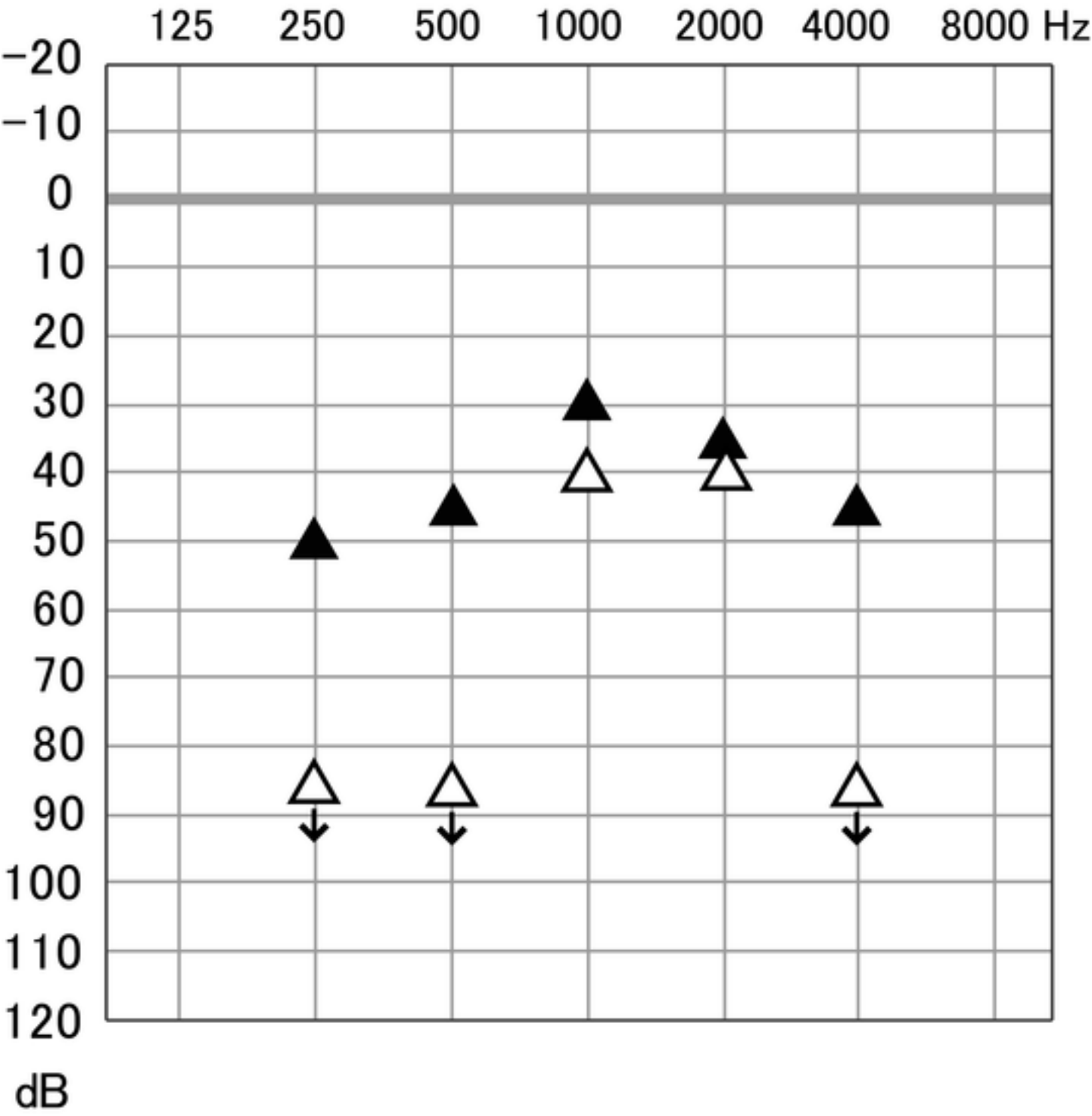on the inner wall of the antero-inferior part of the CC (white arrows).

FIG. 3. Change of CI-aided hearing thresholds in Case 1, depending on the

frequency-to-electrode allocation in the programming of the device. When frequencies from

188 to 7938 Hz are allocated to all 22 electrodes, the hearing thresholds are 40 dB for 1000

and 2000 Hz, for which eV-positive Ch10-16 are responsible, but over 80 dB for other tested

frequencies (white triangles). However, when frequencies from 188 to 5063 Hz are allocated

to only 8 sequential electrodes including Ch10-16 with a positive eV in the EABR testing, the

hearing thresholds range from 30 to 50 dB for all tested frequencies (black triangles).

FIG. 4. Results of EABR testing in Cases 2 to 5. The upper, second, third, and bottom

columns show data in Cases 2 to 5, respectively. A-D: Maximum intensity projection images

of T2-weighted MRI of the CC on the implanted side. AI and PS indicate the antero-inferior

and postero-superior parts of the CC, respectively. E-H: Electrodes with a positive eV

(circles) are located at the curved end of the pre-bent electrode array inserted in the

antero-inferior part of the CC. I-L: EABRs for 3 representative electrodes at the curved end

of the pre-bent electrode array. The latency of these eVs is approximately 4 msec in all cases
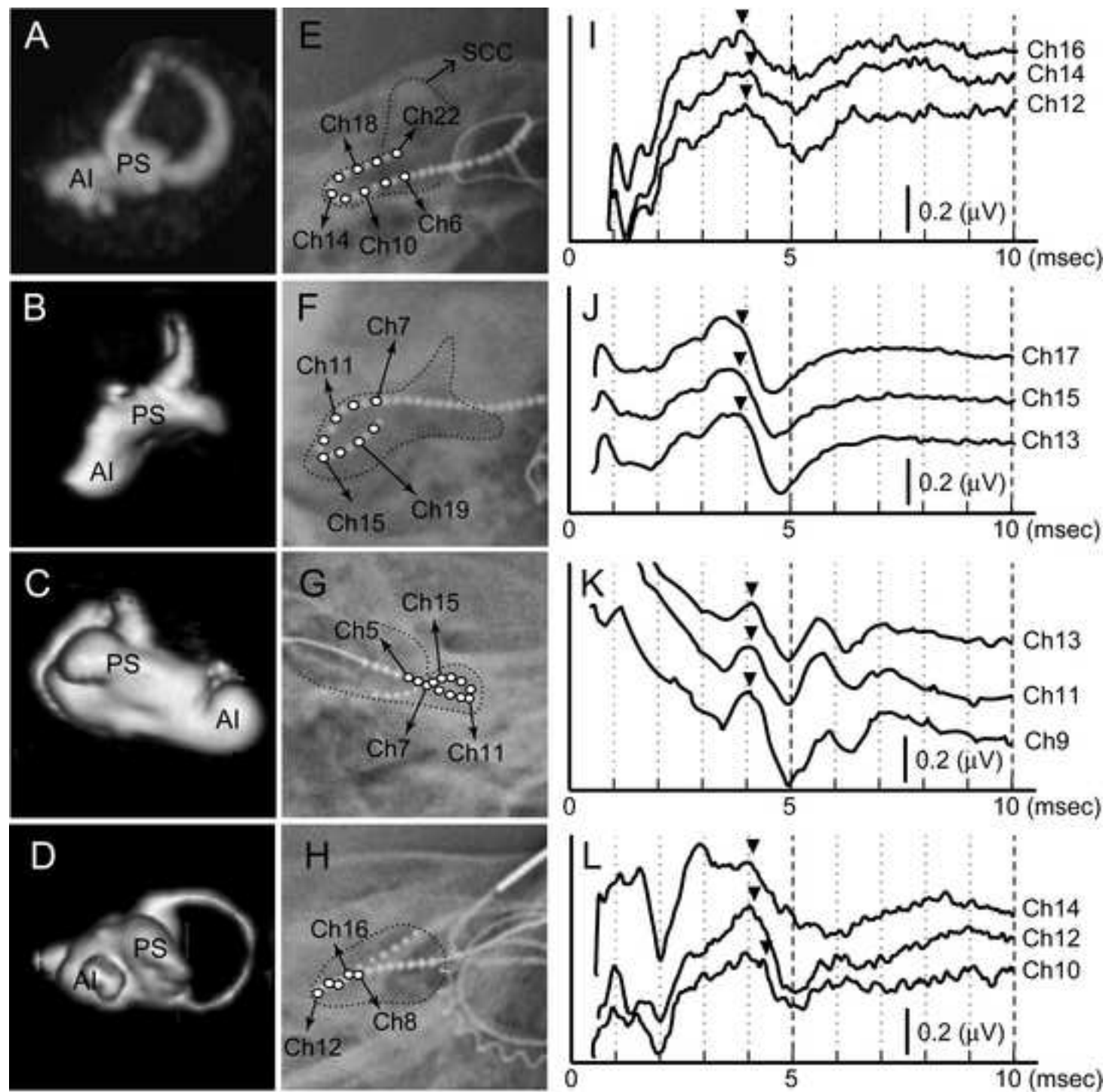
(arrowheads).

A

Ch15
Ch17
Ch19
Ch21
0.2 (µV)
0   5   10 (msec)

B

PS
AI

C

SCC
IAC
Ch19   Ch17

D

Ch16
Ch15
Ch11
Ch14
Ch13
Ch10
Ch12
0.2 (µV)
Ch09
0   5   10 (msec)

E

Ch16
IAC
Ch10
Ch12
Ch14

Table

Table 1. Clinical data for 5 CC patients

	Age at CI (month)	Side of CI	Device	Follow-up duration (month)	Electrodes with a positive eV	Pre-OP CAP score	Post-OP CAP score	Infant word discrimination score (%)	CI-related problems
Case 1 initial CI	27	R	CI24R (ST)	12	2/11 (18.2%)	0	1*	N.E.	FNS
Case 1 re-CI	39	R	CI24R (ST)	40	7/22 (31.8%)	1*	6	76	FNS Dizziness
Case 2	29	L	CI24RE (ST)	29	9/11 (81.8%)	0	6	80	FNS Dizziness
Case 3	23	L	CI24RE (ST)	22	8/11 (71.7%)	0	4	40	
Case 4	29	R	CI422	18	12/22 (54.5%)	0	3	N.E.	CSF gusher FNS
Case 5	28	L	CI422	9	5/11 (45.5%)	0	3	N.E.	

*The pre-OP CAP score of Case 1 re-CI is same as the post-OP CAP score of Case 1 initial CI.

Re-CI, reimplantation of CI; N.E., not examined; FNS, facial nerve stimulation; CSF, cerebrospinal fluid