

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

Hiroshi Yamazaki^{1,3*}, MD, PhD; Yasushi Naito^{1,3}, MD, PhD; Keizo Fujiwara¹, MD;
Saburo Moroto¹, AuD; Rinko Yamamoto¹, AuD; Tomoko Yamazaki¹, AuD; Ichiro
Sasaki², M.T.

¹Department of Otolaryngology and ² Clinical Laboratory, Kobe City Medical Center
General Hospital, ³Institute of Biomedical Research and Innovation, Japan

*Present address is Department of Otolaryngology Head and Neck Surgery, Graduate
School of Medicine, Kyoto University, Kyoto

Corresponding Author: Hiroshi Yamazaki

Department of Otolaryngology Kobe City Medical Center General Hospital

Postal code 650-0047 2-1-1 Minatojima Minamimachi Chuo-ku, Kobe City, Japan

Tel: +81-78-302-4321 FAX: +81-78-302-2487

*Present address: Department of Otolaryngology Head and Neck Surgery,

Graduate School of Medicine, Kyoto University

54 Shogoin-Kawahara, Sakyo-ku, Kyoto, 606-8507

Tel: +81-75- 751-3343 FAX: +81-75-3 751-7225

E-mail address: h_yamazaki@ent.kuhp.kyoto-u.ac.jp

Acknowledgements

We thank Ms. Takako Tosaka at our institute for performing the EABR testing. This study was supported by a Grant-in-Aid for Young Scientists (B): 25861607 and a Grant-in-Aid for Scientific Research (C): 22591894 from the Japanese Ministry of Education, Culture, Sports, Science and Technology.

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 programming 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 antero-inferior cavity, which was visualized inferior to the fundus of the IAC. Since the width of the midpoint of the IAC was $>2.5\text{mm}$ 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

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 μ sec 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 $>0.1 \mu$ V, (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

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,

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 severe 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, programming 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 referring 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

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

References

1. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. *Laryngoscope* 2005;115:1-26.
2. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. *Cochlear Implants Int* 2010;11:4-41.
3. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope* 1987;97:2-14.
4. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. *Laryngoscope* 2002;112:2230-41.
5. Choi BY, An YH, Park JH et al. Audiological and surgical evidence for the presence of a third window effect for the conductive hearing loss in DFNX2 deafness irrespective of types of mutations. *Eur Arch Otorhinolaryngol* 2013;270:3057-62.
6. Fitoz S, Sennaroglu L, Incesulu A et al. SLC26A4 mutations are associated with a specific inner ear malformation. *Int J Pediatr Otorhinolaryngol* 2007;71:479-86.
7. Song MH, Cho HJ, Lee HK et al. CHD7 mutational analysis and clinical considerations for auditory rehabilitation in deaf patients with CHARGE syndrome. *PloS one* 2011;6:e24511.
8. Elmaleh-Berges M, Baumann C, Noel-Petroff N et al. Spectrum of temporal bone abnormalities in patients with Waardenburg syndrome and SOX10 mutations. *AJNR Am J Neuroradiol* 2013;34:1257-63.
9. Naito Y. *Pediatric ear diseases : diagnostic imaging atlas and case reports*. Basel: Karger, 2013:p.
10. McElveen JT, Jr., Carrasco VN, Miyamoto RT et al. Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. *Laryngoscope* 1997;107:1032-6.
11. Cinar BC, Atas A, Sennaroglu G et al. Evaluation of objective test techniques in cochlear implant users with inner ear malformations. *Otol Neurotol* 2011;32:1065-74.
12. Gordon KA, Papsin BC, Harrison RV. Activity-dependent developmental plasticity of the auditory brain stem in children who use cochlear implants. *Ear Hear* 2003;24:485-500.
13. Runge-Samuelson C, Firszt JB, Gaggl W et al. Electrically evoked auditory brainstem responses in adults and children: effects of lateral to medial placement of the nucleus 24 contour electrode array. *Otol Neurotol* 2009;30:464-70.
14. Cushing SL, Papsin BC, Gordon KA. Incidence and characteristics of facial nerve stimulation in children with cochlear implants. *Laryngoscope* 2006;116:1787-91.
15. Archbold S, Lutman ME, Marshall DH. Categories of Auditory Performance. *Ann Otol Rhinol Laryngol Suppl* 1995;166:312-4.
16. Gordon KA, Papsin BC, Harrison RV. An evoked potential study of the developmental time course of the auditory nerve and brainstem in children using cochlear implants. *Audiol*

- Neurootol* 2006;11:7-23.
17. Kelley MW. *Development of the inner ear*. New York: Springer, 2005:xii, 240 p.
 18. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. *J Laryngol Otol Suppl* 2000;25:1-14.
 19. Ahn JH, Oh SH, Chung JW et al. Facial nerve stimulation after cochlear implantation according to types of Nucleus 24-channel electrode arrays. *Acta Otolaryngol* 2009;129:588-91.
 20. Buchman CA, Copeland BJ, Yu KK et al. Cochlear implantation in children with congenital inner ear malformations. *Laryngoscope* 2004;114:309-16.
 21. Gordon KA, Papsin BC, Harrison RV. Toward a battery of behavioral and objective measures to achieve optimal cochlear implant stimulation levels in children. *Ear Hear* 2004;25:447-63.
 22. Merkus P, Free RH, Sanna M. Auditory brainstem implant indications. *Auris Nasus Larynx* 2013;40:113-4.
 23. Colletti V, Fiorino F, Sacchetto L et al. Hearing habilitation with auditory brainstem implantation in two children with cochlear nerve aplasia. *Int J Pediatr Otorhinolaryngol* 2001;60:99-111.
 24. Jin Y, Shinjo Y, Akamatsu Y et al. Vestibular evoked myogenic potentials of children with inner ear malformations before and after cochlear implantation. *Acta Otolaryngol* 2009;129:1198-205.
 25. Black FO, Pesznecker SC. Vestibular adaptation and rehabilitation. *Current opinion in otolaryngology & head and neck surgery* 2003;11:355-60.
 26. Ahn JH, Lim HW, Lee KS. Hearing improvement after cochlear implantation in common cavity malformed cochleae: long-term follow-up results. *Acta Otolaryngol* 2011;131:908-13.
 27. Fishman KE, Shannon RV, Slattery WH. Speech recognition as a function of the number of electrodes used in the SPEAK cochlear implant speech processor. *Journal of speech, language, and hearing research : JSLHR* 1997;40:1201-15.
 28. Garnham C, O'Driscoll M, Ramsden et al. Speech understanding in noise with a Med-El COMBI 40+ cochlear implant using reduced channel sets. *Ear Hear* 2002;23:540-52.

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).

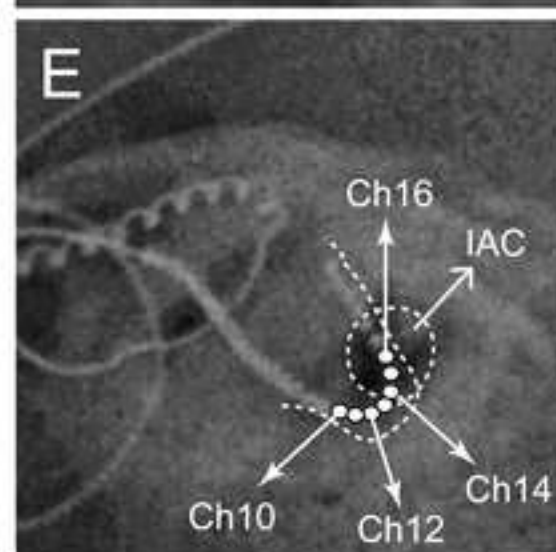
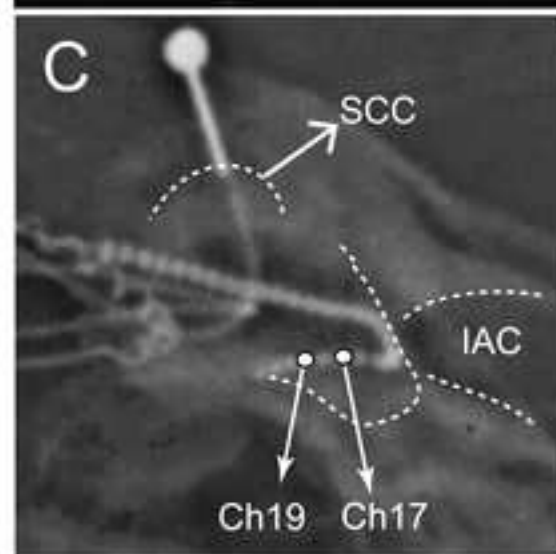
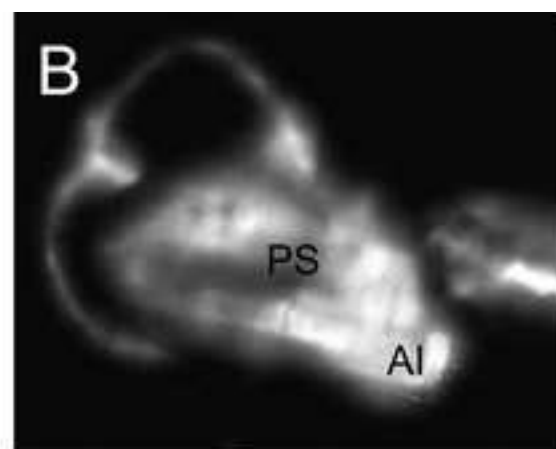
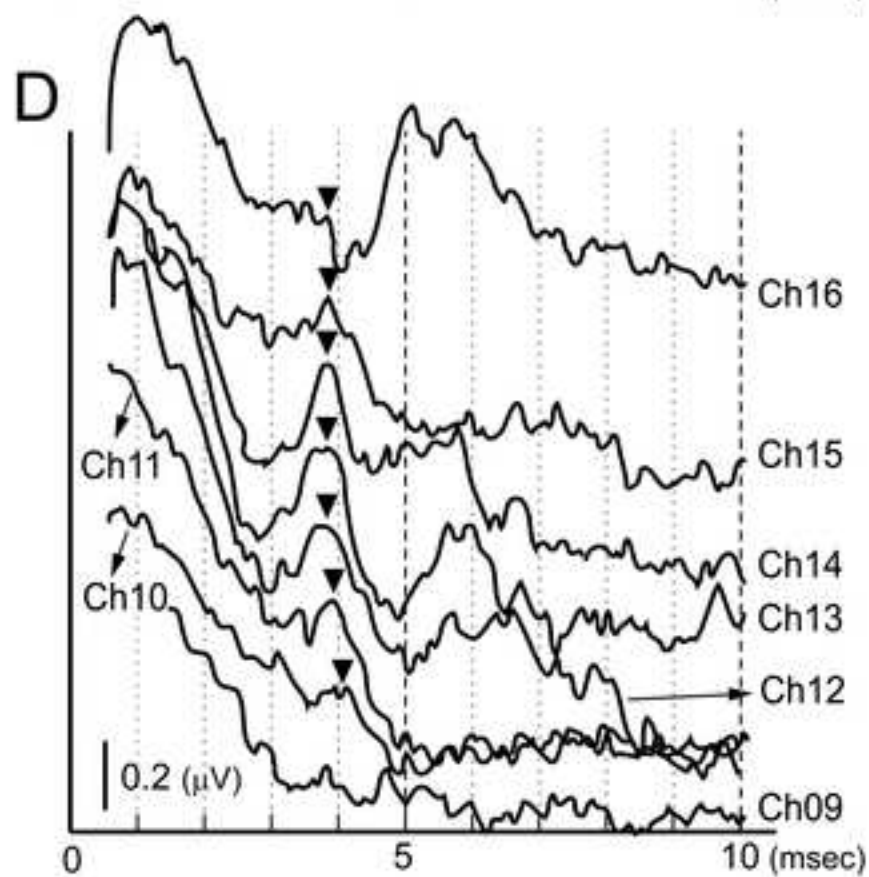
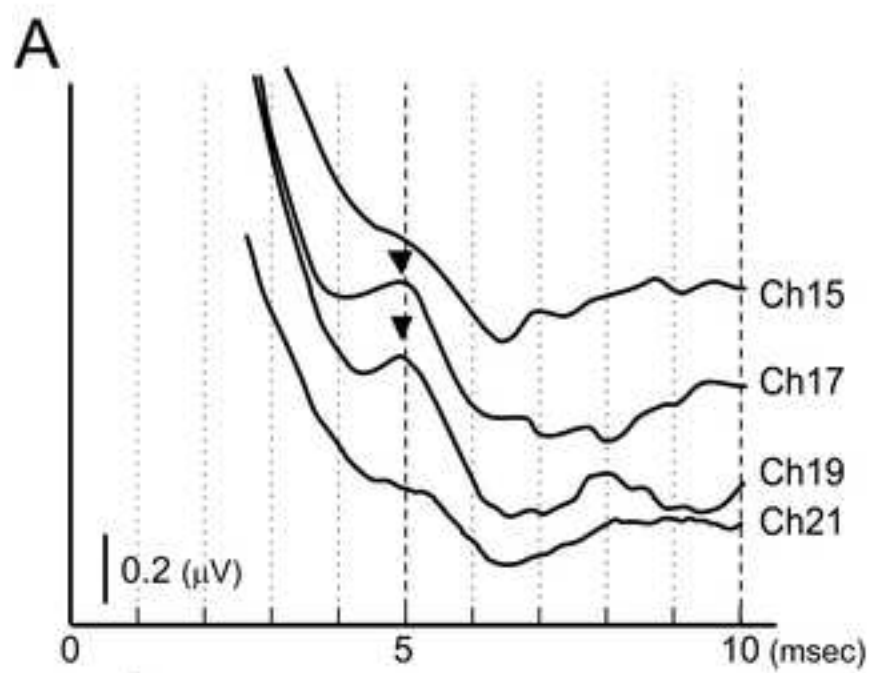
FIG. 2. Evaluation of the electrode array placement by postoperative CT in Case 1. A and B: Coronal CT images after the initial implantation with the standard labyrinthotomy approach. The electrode array inserted from the hole of the standard labyrinthotomy (black arrows) is stuck at the fundus of the IAC (arrowheads) and does not reach the antero-inferior cavity (white arrows). C and D: Coronal CT images after reimplantation with the modified

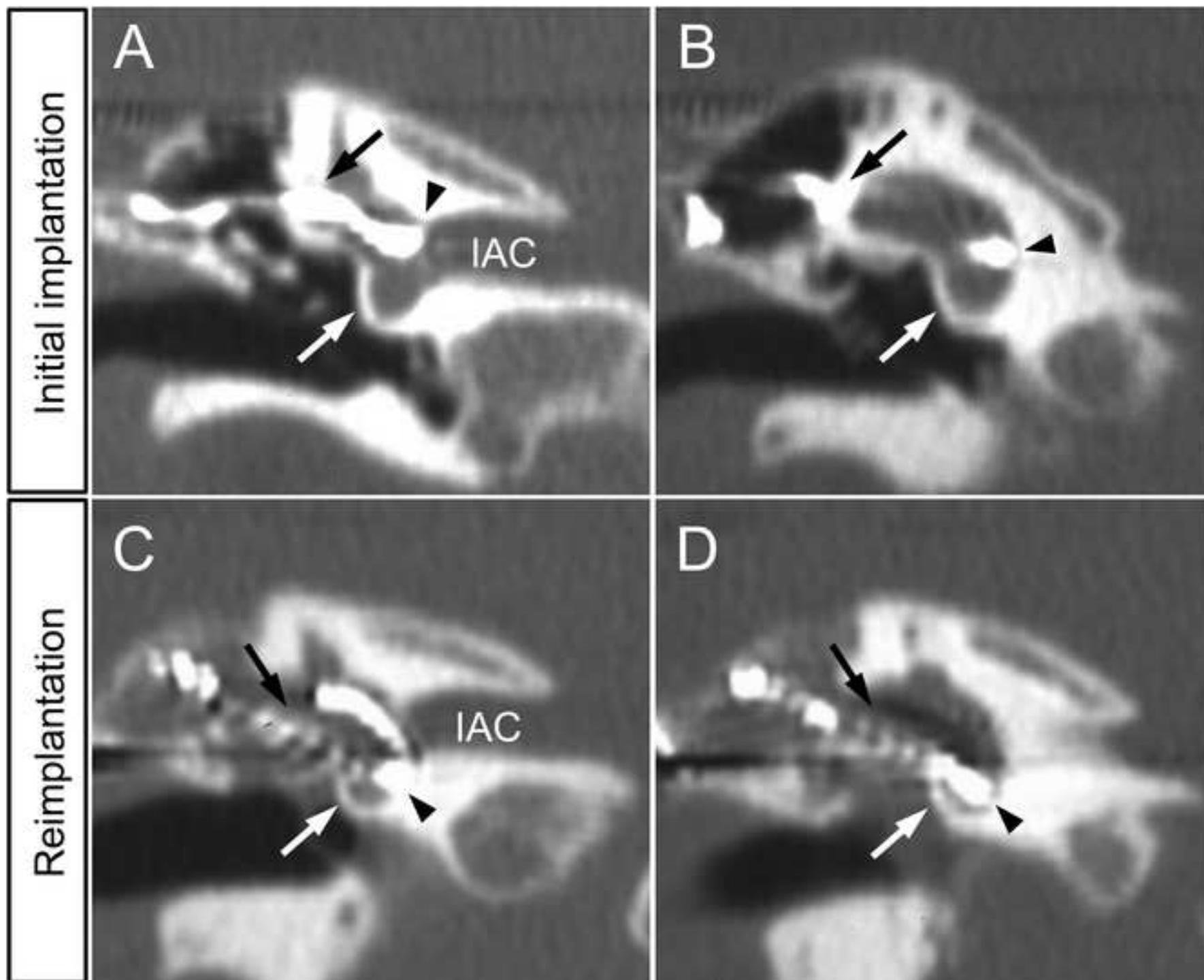
labyrinthotomy approach with the 3.0mm-diameter hole on the postero-lateral wall of the cavity (black arrows). The curved end of the pre-bent electrode array (arrowheads) is attached on the inner wall of the antero-inferior part of the CC (white arrows).

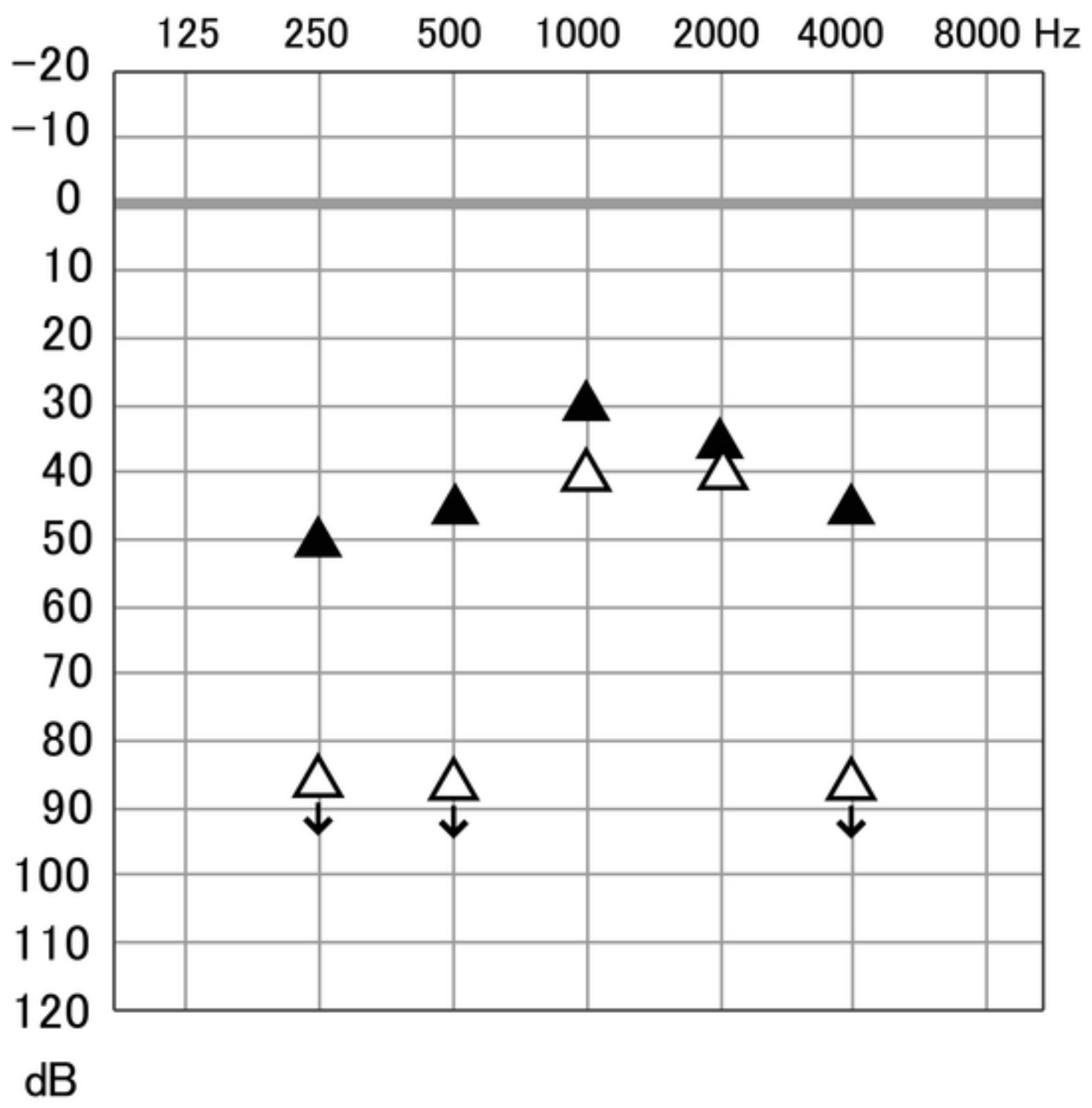
FIG. 3. Change of CI-aided hearing thresholds in Case 1, depending on the frequency-to-electrode allocation in the programming of the device. When frequencies from 188 to 7938 Hz are allocated to all 22 electrodes, the hearing thresholds are 40 dB for 1000 and 2000 Hz, for which eV-positive Ch10-16 are 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).







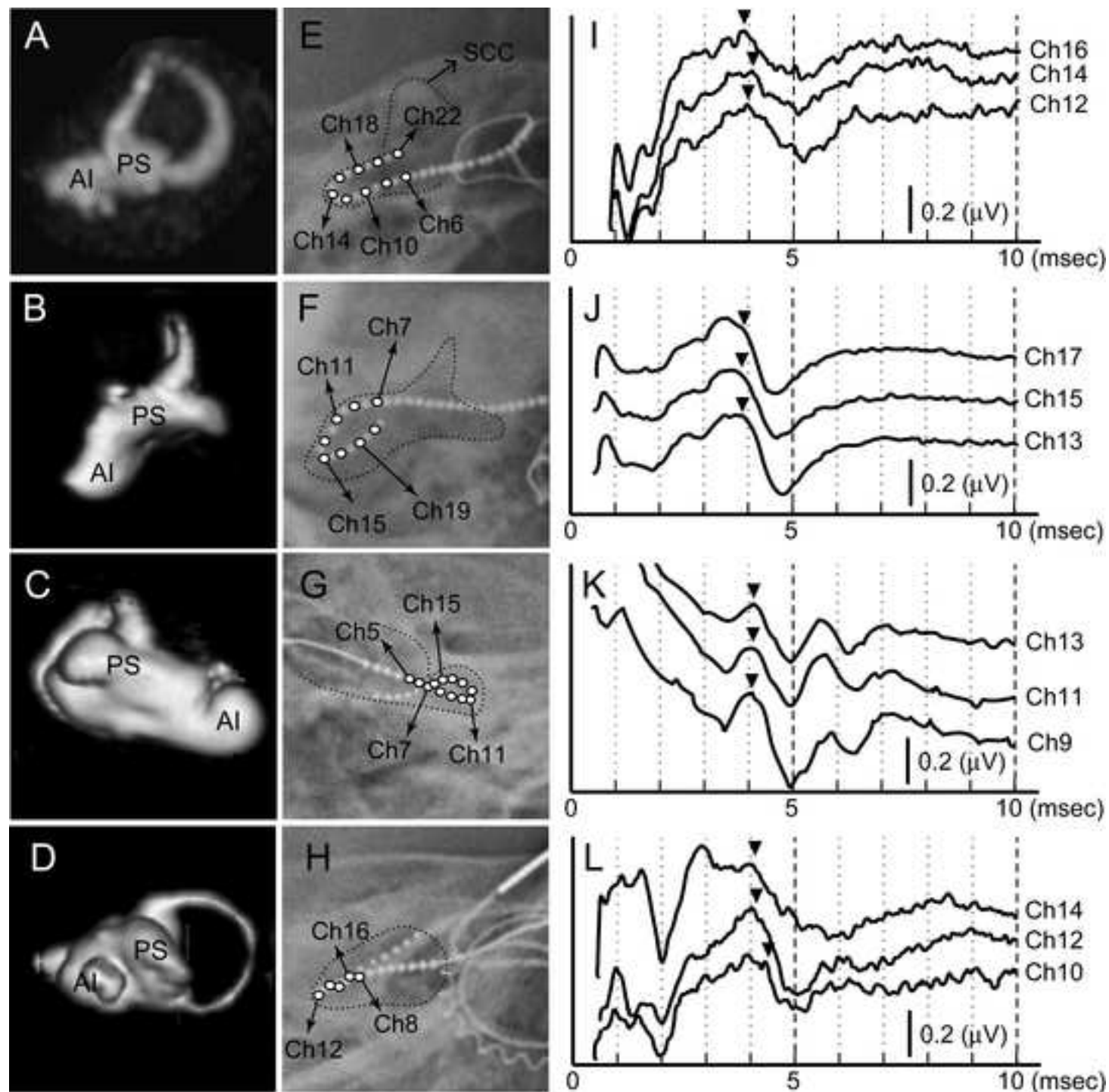


Table 1. Clinical data for 5 CC patients

	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