Chapter 8
EABR of Inner Ear Malformation and Cochlear Nerve Deficiency After Cochlear Implantation in Children

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Abstract When cochlear implantation has been performed in a case involving inner ear malformations, it is particularly important to perform objective physiological measurements of the cochlear implant. The inner ear malformations can be divided into categories according to the observation of modiolus deficiency and/or cochlear nerve deficiency (CND). CND severity can be categorized in one of three ways, according to the MRI findings: (1) a hypoplastic cochlear nerve, (2) the absence of cochlear nerve, and (3) the absence of vestibulocochlear nerve. EABR is a reliable and effective way of objectively confirming device function and implant responsiveness of the peripheral auditory neurons up to the level of the brainstem in cases of inner ear malformation. EABR can often be recorded in cases in which the presence of excessive stimulus artifacts precludes the successful acquisition of ECAP, such as in cases with modiolus deficiency cochlea. This chapter presents cases with or without modiolus deficiency, depending on the severity of cochlear nerve deficiency, and describes their EABR characteristics. Vestibular simulated EABR is also shown, demonstrating the interactions between vestibular and auditory pathways.

Keywords EABR • Modiolus deficiency • Cochlear nerve deficiency

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8.1 Introduction

When inner ear malformations are present, it is particularly important to perform objective measurements of the cochlear implant (CI), as these measurements will show whether the electrodes are appropriately positioned and whether there is initial failure of the device during surgery. These measurements are also useful for predicting the audiological outcomes after CI implantation, for assisting the speech processor fitting when behavioral results are difficult to obtain, and for characterizing the pathophysiology of hearing loss. The different ways of objectively measuring CI function can be divided into those that measure nonphysiological variables and those that measure physiological variables. Objective nonphysiological assessment tools include those that measure electrode-specific voltage, impedance, and electrical field patterns across the array. These provide insights into the properties of the surrounding tissue, the electrode–tissue interface, and the path of current flow and help to identify electrode failures [1]. However, these tools are not used to assess the physiological function of the auditory pathway. Physiological objective assessment tools measure various aspects of the auditory responses to electrical stimulation through a CI. These include electrically evoked stapedial reflexes [2], electrically evoked compound action potentials (ECAPs) [3], electrically evoked auditory brainstem responses (EABRs) [4], electrically evoked auditory middle latency responses [5], and electrically evoked auditory cortical potentials [6]. ECAP can be recorded quickly and easily without the need for surface or scalp electrodes and is probably the most widely used measure in clinical settings. In contrast, while EABR recordings require the placement of surface electrodes, they can provide information about the auditory pathway up to the level of the brainstem [7].

The inner ear malformations can be categorized according to the type of modiolus deficiency and/or cochlear nerve deficiency (CND), if any. The modiolus present type includes enlarged vestibular aqueduct (EVA), incomplete partition type II (IP-II), and cochlear hypoplasia type III (CH-III). The modiolus absent type includes common cavities (CC) and incomplete partition type I (IP-I). CND can be divided into three categories, according to the MRI findings (Fig. 8.1a–d): (1) a hypoplastic cochlear nerve; cochlear nerve can be identified, but is smaller than facial nerve; (2) the absence of cochlear nerve; vestibulocochlear nerve can be identified but cochlear nerve cannot be separated; and (3) the absence of vestibulocochlear nerve; vestibulocochlear nerve cannot be confirmed at all. The present chapter shows cases of each type, describing their EABR characteristics.

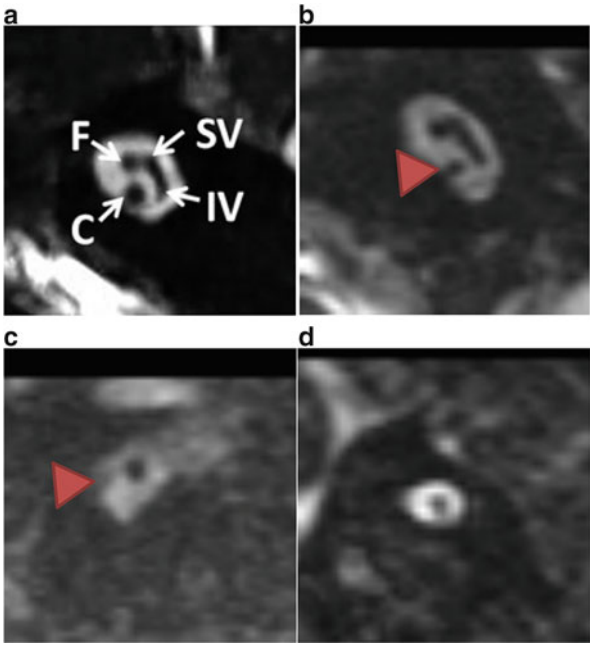

Fig. 8.1 Reformatted parasagittal oblique MRI images. (**a**) A normal cochlear nerve of larger diameter than the facial nerve. *F* facial nerve, *C* cochlear nerve, *SV* super. (**b**) A hypoplastic cochlear nerve of smaller diameter than the facial nerve (*red triangle*). (**c**) Facial and vestibulocochlear (*red triangle*) nerves are identified, but cochlear nerve is not separated. (**d**) Absence of vestibulocochlear nerve. Only facial nerve is recognized

8.2 Measurement and Reading of EABR

8.2.1 Measurement of Intracochlear EABR

EABRs were recorded on electrodes within the cochlea. The responses were recorded with the Neuropack (Nihon Kohden Co., Tokyo, Japan) electrodiagnostic system and were triggered externally by the stimulus output of each CI company's software and the interface unit. The interface unit was also connected to a stock speech processor and the subject's headpiece; the stimulus signal was transmitted across the skin to the implanted device. The electrically evoked brainstem potentials were recorded by using needle electrodes placed on the forehead (different electrode), the nape of the neck (indifferent electrode), and the contralateral earlobe (reference electrode). The recording of electrical activity included two or three replications of 1000 sweeps at each stimulus level with a time window of 10 ms for each stimulus condition. Frequency cutoffs of 100 and 1000 Hz were used. The pulse duration was set to 30 ms and the stimulation amplitude for a single recording fell from high to low current. If no response was detected, pulse duration was increased.

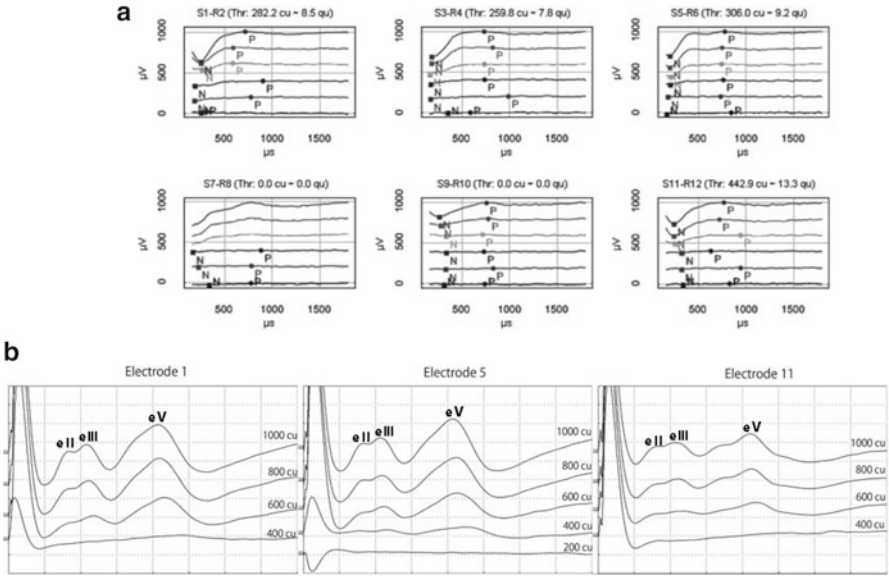

Fig. 8.2 (**a**) ECAP waves of case no. 1. (**b**) EABR waves of case no. 1

8.2.2 EABR Waves of Patients Without Inner Ear Malformation

8.2.2.1 Case No. 1

In this case, hearing loss was found by newborn hearing screening. Congenital cytomegalovirus infection was confirmed by polymerase chain reaction for cytomegalovirus DNA in his umbilical cord. CT and MRI studies showed normal inner ear structure. He was fitted with hearing aids bilaterally, but his hearing loss progressed to profound sensorineural hearing loss. At the age of 3 years, he underwent implantation with a CONCERTO Flex28 (MED-EL, Innsbruck, Austria). All electrodes were inserted, and further assessment via telemetry showed good ECAP and EABR responses via the cochlear implant (Fig. 8.2a, b). After cochlear implantation, his hearing recovered well, and he achieved an IT-MAIS score of 34 at 6 months after implantation.

8.2.2.2 Comment

The basal electrodes have higher thresholds and longer wave eV latencies than the apical and middle electrodes. It may be that the higher thresholds and longer wave eV latencies of the most basal electrodes are the result of the greater distance from

the neural elements compared to the more apical electrodes, which are located further along the scala tympani. According to our series, the mean wave eIII latencies of the ears without a malformation for apical and basal electrodes were 2.29 ± 0.22 and 2.40 ± 0.24 ms, and the mean wave eV latencies of the ears without a malformation for apical and basal electrodes were 4.26 ± 0.40 and 4.55 ± 0.32 ms [8]. All children without malformations had EABR wave eV latencies of less than 5 ms. The patients were divided into three groups according to their EABR responses. The *typical* response group included all patients showing reproducible wave eV responses with EABR eV latencies of less than 5 ms. The *atypical* response group was defined as those patients who presented with reproducible wave eV responses that were measured in only a limited number of electrodes and/or that showed EABR eV latencies of more than 5 ms pulse duration. In the *no* response group, no identifiable wave eV responses could be seen in any of the electrodes, even with longer pulse duration.

8.3 EABR Waves of Patients with Modiolus Present Type of Inner Ear Malformation

8.3.1 Modiolus Present and Cochlear Nerve Present Type

8.3.1.1 Case No. 2

This is a case of congenital progressive hearing loss with bilateral enlarged vestibular aqueduct (EVA) (Fig. 8.3a, b). SLC26A4 mutations were confirmed. At the age of 3 years, she underwent implantation with a CONCERTO Flex soft (MED-EL) in her right ear. All electrodes were inserted, and further assessment via telemetry showed good ECAP and EABR responses via the cochlear implant (Fig. 8.3c). After cochlear implantation, her hearing recovered well.

8.3.1.2 Comment

The presence of enlarged vestibular aqueduct (EVA) in the presence of normal cochlea, vestibule, and SCCs is a typical case of modiolus present and cochlear nerve present type of inner ear malformation. This type of inner ear malformation shows as good EABR and CI performance as those without malformation. In the cochlear malformation cases in which the modiolus was present, the basal electrodes have higher thresholds and longer wave eV latencies than the apical and middle electrodes. These are similar threshold and latency patterns to those observed in the patients without malformations.

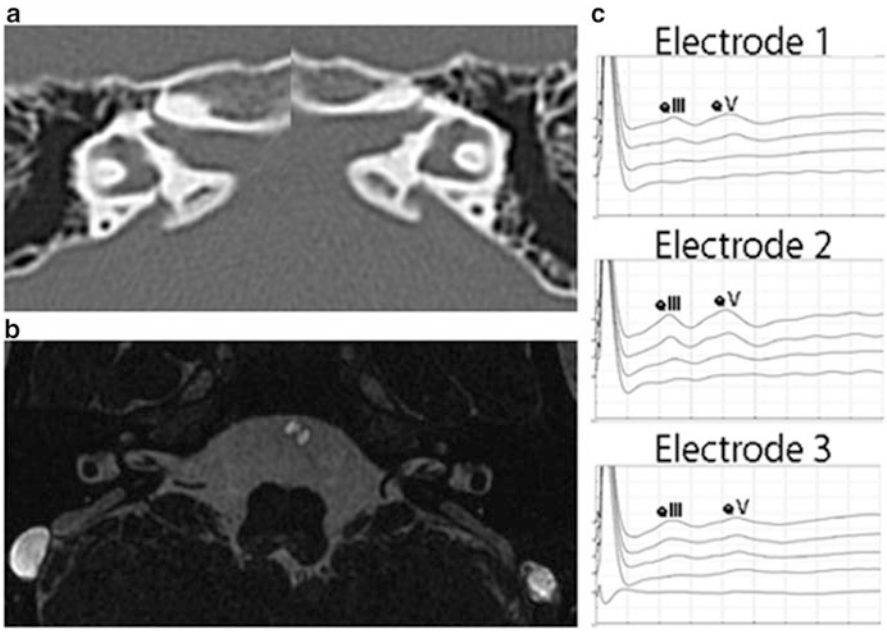

Fig. 8.3 (**a**) Axial computed tomography imaging study showing enlarged vestibular aqueduct malformation. (**b**) Axial MRI study showing enlarged vestibular aqueduct malformation and normal cochlear nerves. (**c**) EABR waves of case no. 2

8.3.2 Modiolus Present and Cochlear Nerve Deficiency Type

8.3.2.1 Case No. 3

This child was 10 years old and had progressive hearing loss (Fig. 8.4a). She has a very thin cochlear nerve canal in CT (Fig. 8.4b), and her cochlear nerves could not be seen on MRI (Fig. 8.4c), but she had obvious auditory response on both ears. She had cochlear implantation (MED-EL CONCERTO Flex soft) on the left ear. Her ECAP showed a threshold at 600 CU (current unit) with a 30-ms pulse duration (Fig. 8.4d), which is the usual pulse width. Meanwhile, her EABR threshold was 800 CU with a 55-ms pulse duration (Fig. 8.4e), which means that we need to nearly double the intensity to obtain the EABR threshold as compared with ECAP. Now, her category of auditory performance (CAP) score is 6, and she is very satisfied with CI.

8.3.2.2 Comment

Even the patient has cochlear nerve deficiency, if she has obvious auditory response with hearing aids, she can be a good indication for cochlear implantation. Because obvious auditory response implies that the cochlear nerve is functionable. Cochlear nerve canal stenosis cases have normal spiral ganglion cells, so ECAP shows good

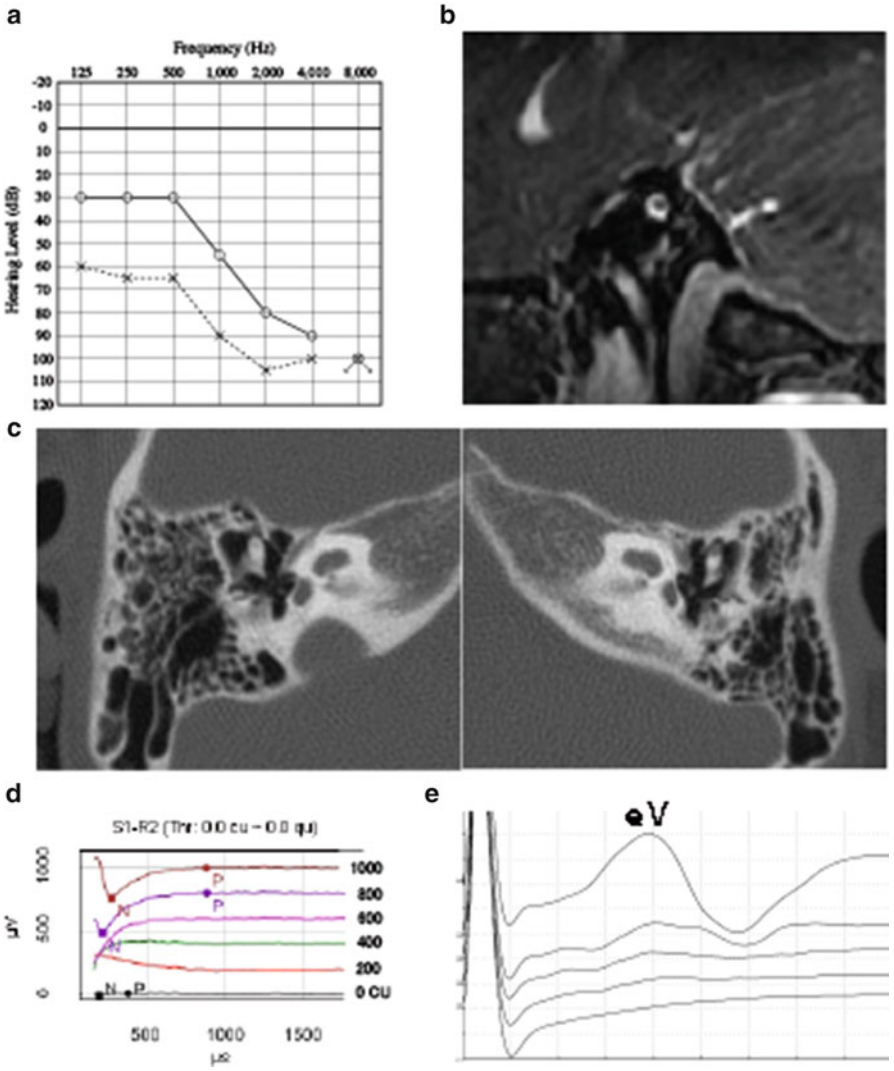

Fig. 8.4 (**a**) Pure tone audiometric result for case no. 3 before cochlear implantation. (**b**) Parasagittal oblique MRI study showing the absence of cochlear nerve. (**c**) Axial computed tomography imaging study showing cochlear nerve canal stenosis. (**d**) ECAP waves of case no. 3. (**e**). EABR waves of case no. 3

responses with the usual intensity. However, because a high intensity is needed to go through the thin auditory nerve, the EABR threshold is high. It may be better to use modiolar-hugging electrodes, because peri-modiolar electrode placement reduces the spread of excitation of CI stimulation. These reduced nerve stimulation thresholds may result in improved speech discrimination by implant users with modiolus presence and cochlear nerve deficiency.

8.4 EABR Waves of Patients with Modiolus Absent Type of Inner Ear Malformation

8.4.1 Modiolus Absent and Vestibulocochlear Nerve Present Type

8.4.1.1 Case No. 4

This child has congenital profound hearing loss with bilateral common cavity (CC) malformation (Fig. 8.5a). He underwent cochlear implantation with PULSAR Standard (MED-EL) at 2 years old in his right ear and CONCERTO Standard (MED-EL) at 4 years 8 months old in his left ear. In the right ear, no ECAP response and variable EABR responses were obtained; in the left ear, variable ECAP and EABR responses were obtained (Fig. 8.5b, c). His IT-MAIS score was 35 at 1 year after first implantation.

8.4.1.2 Comment

The type of cochlear malformation characterized by modiolus absence and vestibulocochlear nerve presence is CC or incomplete partition type I (IP-I) with open fundus of the internal auditory canal (IAC). ECAP recordings depend largely on spinal

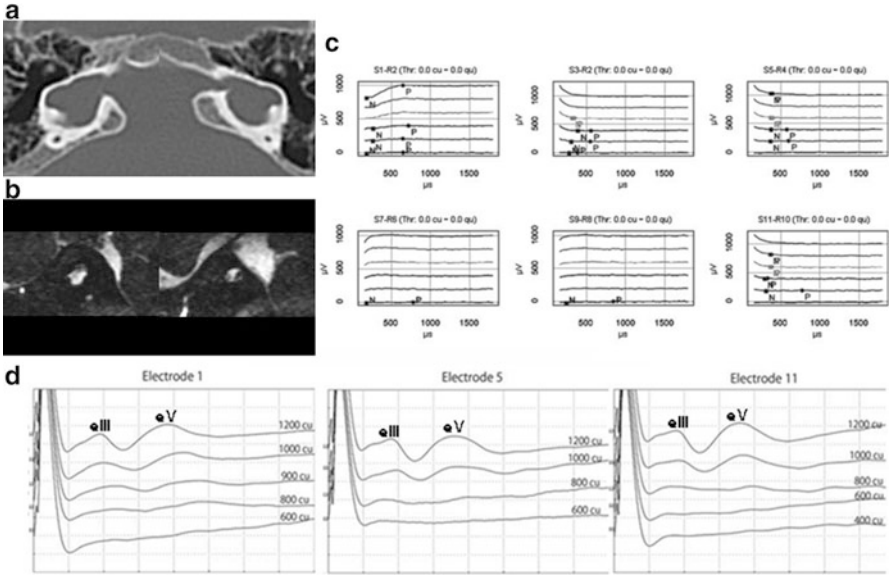

Fig. 8.5 (**a**) Axial computed tomography imaging study showing common cavity malformation. (**b**) Parasagittal oblique MRI study showing vestibulocochlear and facial nerves. (**c**) ECAP waves of case no. 4. (**d**) EABR waves of case no. 4

ganglion cells, which are very often defective in modiolus deficiency-type malformed cochlea. EABR can be obtained in modiolus deficiency-type implant users because the measures are not dependent on the implant having telemetry capabilities and because the wave eV of EABR, which occurs at a later latency than ECAP, is easier to isolate from the stimulus artifacts. The cochlear malformation cases with modiolus deficiency did not exhibit threshold and latency differences between electrodes. The auditory nerve tissues in modiolus deficiency malformations are supposed to be in the inner ear wall, and so the distances from each electrode to the auditory nerve tissue should not be different in modiolus deficiency-type malformations.

8.4.2 Modiolus Absent and Vestibulocochlear Nerve Deficiency

8.4.2.1 Case No. 5

This patient was 1 year and 6 months old and has common cavity with very narrow internal auditory canal on both sides (Fig. 8.6a). Only the facial nerves were recognized by the MRI (Fig. 8.6b). The auditory response with hearing aids was vague, but the damped-rotational chair test (DRCT) showed normal vestibular function (Fig. 8.6c). She had cochlear implantation in the left ear. The intracochlear EABR during surgery showed typical good responses at all electrodes (Fig. 8.6d). Now, she shows obvious auditory response with CI and takes auditory verbal education.

8.4.2.2 Comment

This type of malformation is challenging. Some doctors may say this is a case of cochlear aplasia with enlarged vestibule. We think good vestibular functions in the cases of comorbidity of common cavity and narrow internal auditory canal can be an indication for CI. In the case of internal auditory canal stenosis, vestibular evaluation helps us to determine the neural connection between the inner ear and the brain. It is possible that vestibular nerves can obtain the function of auditory nerve via auditory stimulation plasticity. Amphibians and reptiles are able to hear without cochlea. Smith reported interactions between the vestibular nucleus and the dorsal cochlear nucleus [9]. The next case (no. 6) demonstrated the possibility of these interactions.

8.5 Vestibular Simulated EABR

8.5.1 Case No. 6

This patient suffered bilateral profound hearing loss at age 3 as a result of meningitis. He underwent cochlear implantation with Concerto Flex28 (MED-EL) at 20 years old in his left ear. He had stage I cochlear ossification; all electrodes were

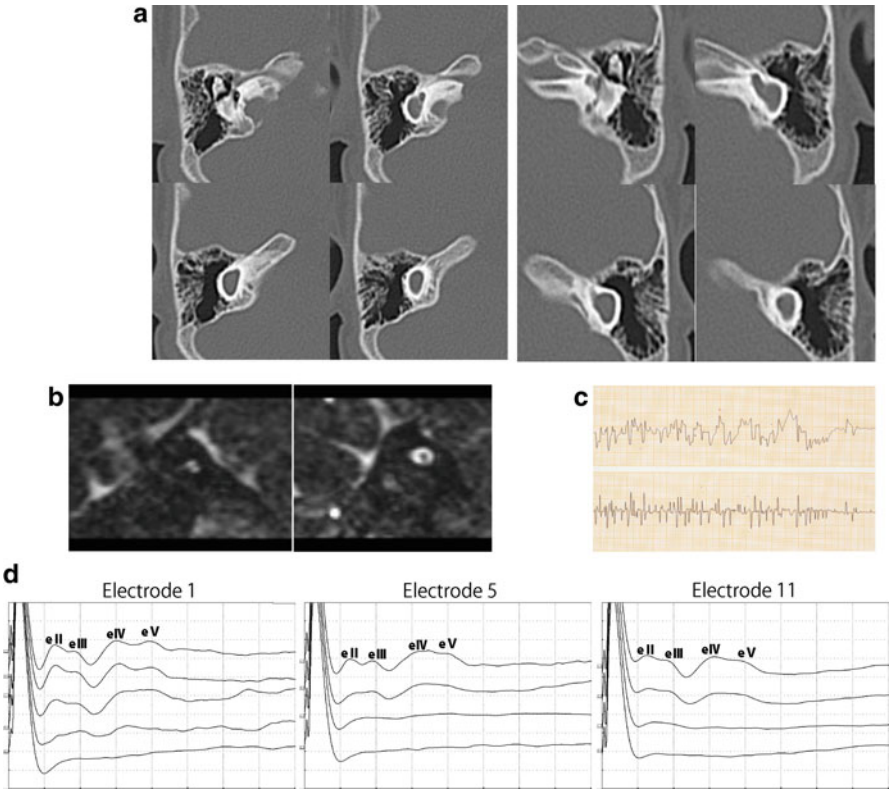

Fig. 8.6 (**a**) Axial computed tomography imaging study showing common cavity malformation and severe internal auditory canal stenosis. (**b**) Parasagittal oblique MRI study showing only facial nerves. (**c**) DRCT response of case no. 5. (**d**) EABR waves of case no. 5

wrongly inserted to vestibule and semicircular canals (Fig. 8.7a). After we found the wrong insertion, he underwent reoperation, and his hearing recovered well. Figure 8.7b showed vestibular simulated EABR in wrong insertion.

8.5.2 Comment

This is an unexpected case. The EABR in the vestibule and semicircular insertion showed reproducible wave eIII, eIV, and eV responses with similar latencies to case no. 5. Previous studies showed direct projections from the vestibular nerve to the dorsal cochlear nucleus (DCN) [9, 10]. These results suggest that the lateral vestibular nucleus (LVN) projects directly to the DCN, some of which may also receive direct projections from the vestibular nerve. Thus, vestibular and auditory information processing may be intimately connected.

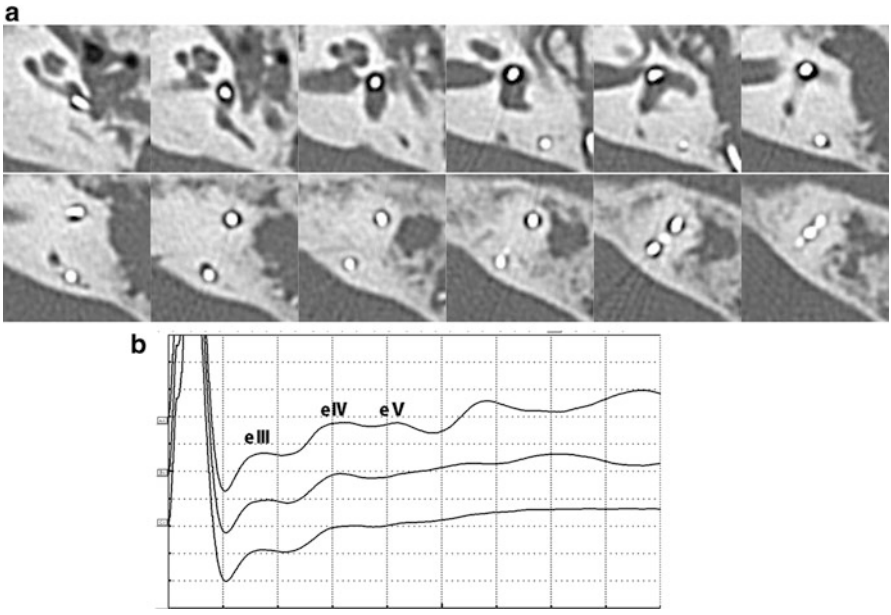

Fig. 8.7 (**a**) Axial computed tomography imaging study showing wrong insertion to vestibule and semicircular canals. (**b**) EABR waves of case no. 6 in wrong insertion

8.6 Our Series of Cochlear Nerve Deficiency

Table 8.1 shows our 20 cases of CND who had CI surgery. We evaluated our 20 cases of CNDs by CT and MRI, vestibular functions (damped-rotational chair test), and intracochlear EABR during CI surgery. 65 % of CNDs had comorbidity of cochlear malformation, 25 % incomplete partition (IP)-I, 20 % cochlear hypoplasia, 15 % common cavity, and 5 % IP-II. On MRI one case showed a thin cochlear nerve, and 60 % showed the absence of cochlear nerves but the presence of vestibulocochlear nerves. The absence of vestibulocochlear nerves is found in 25 % of CNDs. With vestibular function tests before CI surgery, 60 % of CNDs showed normal, 25 % poor, and 10 % no response. In the cases with vestibulocochlear nerves found on MRI, 67 % showed typical EABR, while in the cases with no vestibulocochlear nerve, just one showed typical EABR. 64 % of good vestibular function cases showed typical EABR, while only 29 % of poor or no vestibular function cases showed typical EABR. In the cases with thin or absent cochlear nerves, the vestibulocochlear nerves found on MRI and obvious auditory responses with hearing aids are possible indications for CI. Even if the imaging studies show an absence of vestibulocochlear nerves, the cases with good vestibular functions can be indicated for CI, because vestibular evaluation helps us to determine the neural connection between the inner ear and the brain. Not only imaging evaluations but also evaluations by auditory response and vestibular function are important for CI

Table 8.1 Our 20 cases of cochlear nerve deficiency

Pt	CI age (years)	Cochlear malformation	Modiolus	MRI	DRCT pre-CI	EABR
1	4	No	Present	Absence of cochlear nerve	Normal	Atypical
2	2	IP-I	Absent	Absence of cochlear nerve	Normal	Typical
3	5	Cochlear hypoplasia	Present	Absence of cochlear nerve	N/A	Typical
4	2	IP-I	Absent	Absence of cochlear nerve	No response	Typical
5	2	No	Present	Absence of cochlear nerve	Normal	Typical
6	2	Common cavity	Absent	Absence of cochlear nerve	Poor	Typical
7	3	IP-I	Absent	Absence of cochlear nerve	Poor	Atypical
8	2	Cochlear hypoplasia	Present	Absence of cochlear nerve	Normal	Typical
9	2	Cochlear hypoplasia	Present	Absence of cochlear nerve	No response	Atypical
10	10	No	Present	Absence of cochlear nerve	Normal	Typical
11	6	IP-II	Present	Hypoplastic cochlear nerve	Normal	Typical
12	2	IP-I	Absent	Absence of cochlear nerve	Normal	N/A
13	4	Common cavity	Absent	Absence of cochlear nerve	Normal	Typical
14	2	No	Present	Absence of cochlear nerve	Normal	Atypical
15	4	No	Present	Absence of vestibulocochlear nerve	Poor	No response
16	1	Common cavity	Absent	Absence of vestibulocochlear nerve	Normal	Typical
17	11	No	Present	Absence of vestibulocochlear nerve	Normal	No response
18	1	IP-I	Absent	Absence of cochlear nerve	Poor	Atypical
19	2	No	Present	Absence of vestibulocochlear nerve	Normal	Atypical
20	26	Cochlear hypoplasia	Present	Absence of vestibulocochlear nerve	Poor	Atypical

indication of CNDs. Even in typical EABR cases, some cases show poor auditory performance with CI because of developmental disability. It is difficult to evaluate developmental disability in early childhood, and so we should pay considerable attention to this comorbidity with developmental disability.

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