Cochlear Aplasia

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Special Features

1. Two types are present: cochlear aplasia with normal vestibule and dilated vestibule.
2. Cochlear aplasia with dilated vestibule must be differentiated from common cavity.
3. Definite indication for ABI.
4. Cochlear nucleus appears to be well developed in spite of absent cochlea and cochlear nerve.

22.1 Definition

Cochlear aplasia (CA) is the absence of the cochlea [1]. The accompanying vestibular system may be normal (CANV) (Fig. 22.1a) or it may have dilated vestibule (CADV) (Fig. 22.1b) [2]. The labyrinthine segment of the facial nerve is anteriorly displaced and usually occupies the normal location of the cochlea. It is essential to distinguish cochlear aplasia with a dilated vestibule (CADV) from common cavity (CC) [3]. If the cochleovestibular nerve (CVN) is present, cochlear implantation can be done in CC. However, CI surgery should be avoided in CADV. In spite of this, it may be very difficult to distinguish between these entities in some patients.

22.2 Histopathology and Pathophysiology

There is neither any histopathological report of a cochlear aplasia in the literature nor a specimen in the temporal bone collections of the Massachusetts Eye and Ear Infirmary and University of Minnesota.

When the cases of cochlear aplasia were investigated radiologically, their HRCT showed bony otic capsule development; usually, otic capsule formation fills that particular space left for the cochlea, and the labyrinthine segment of the facial nerve is anteriorly dislocated. It is possible that this bone consists of enchondral and outer periosteal layers.

After the development of the otic vesicle at the end of the fourth week, the membranous labyrinth develops in three areas: the cochlea, the vestibule, and the endolymphatic duct. Cochlear aplasia is the absence of the cochlear duct, where vestibular and endolymphatic structures may develop normally [4]. The time of the insult must be around the fifth week. Otic capsule development is always normal, and the facial nerve is anteriorly displaced into the usual location of the cochlea.

It has been observed that CANV cases are bilateral and almost always symmetric, with sim-

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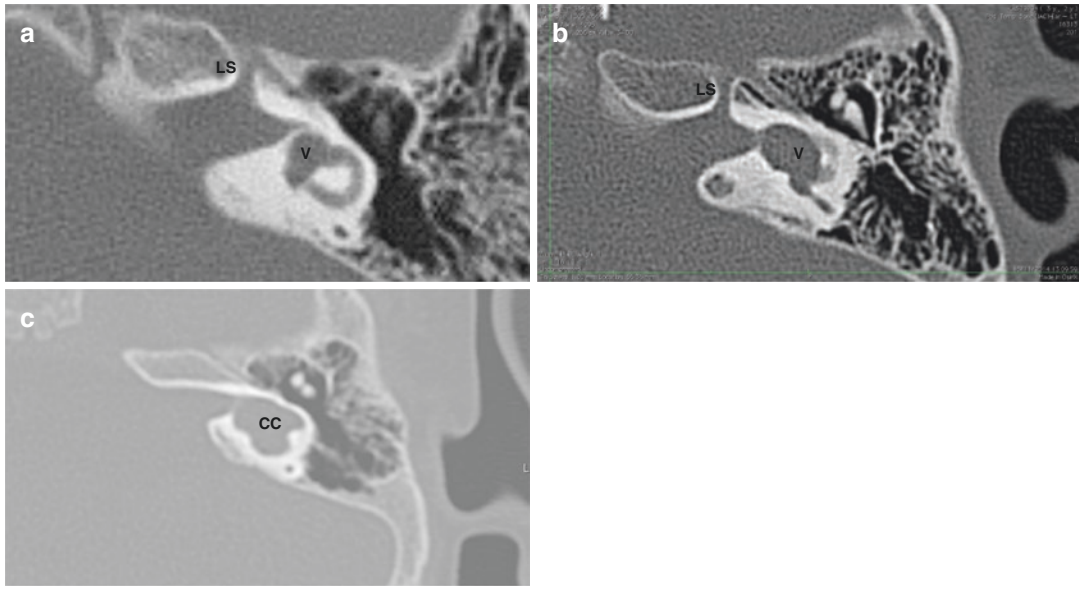

Fig. 22.1 (**a**) Cochlear aplasia with a normal vestibule (V), (**b**) cochlear aplasia with a dilated vestibule, (**c**) common cavity (CC). Please note that labyrinthine segment of the facial nerve (LS) is anteriorly dislocated to the usual location of cochlea

ilar features repeating in a similar way in different patients. It is very unlikely that an external cause would destroy only the cochlear bud completely, leaving the vestibular development normal. Therefore, there is a strong possibility that the origin in CANV is genetic. CADV, however, is usually asymmetric suggesting that it may be genetic or environmental [1].

22.3 Literature Review

Cochlear aplasia is a definite indication for ABI [5]. Although it is known that presence of a cochlear nerve is required for CI application, there are some cochlear aplasia cases reported to have benefit from CI surgery even though no separate cochlear branch has been identified in MRI scans. Jeong and Kim [6] reported two pediatric cases who had CI electrode insertion into the vestibule (one with dilated and the other with normal vestibule). The case with normal vestibule had aided thresholds at 25 dB HL at post-op 4 years. Speech perception scores also showed improvement in this period; score of 5 on CAP and 61%, 44%, and 43% scores on monosyllabic word test for phonemes, words, and on sentence test were obtained, respectively. Similar to the case with normal vestibule, patient with dilated vestibule also showed aided thresholds at 25 dB HL, score of 4 on CAP and 76% and 50% scores on monosyllabic word test for phonemes and words at post-op 3 years, respectively. Kontorinis et al. [7] have reported outcomes of five cases of CA who did not have a separately identified CN branch. Four cases had CI and one case had ABI. The CAP scores of all users reached 4–5 in the follow-up period; meaning that speech sounds and common phrases can be discriminated without lip reading. It is interesting that these patients have derived benefit from CI although a separate CN could not be identified. In fact, the authors have discussed that this may be related to the possibility that CI electrode has contact with residing nerve fibers in inner ear structures, thereby providing stimulation. When the images are examined it is possible that cases who showed benefit may be CC rather than CA. In spite of these literature findings, the authors of the present book strongly advise to be extremely cautious to suggest a CI surgery in a very clear cochlear aplasia case as shown in Fig. 22.1a, b.

22.4 Clinical Findings

They present with nonprogressive profound SNHL.

22.4.1 Radiology

According to the IEM database of Hacettepe University Department of Otolaryngology out of 776 patients with various IEMs 49 of 1652 ears had CA (3%). Fourteen of these were CANV (30%) and 33 were CADV (70%).

Cochlea normally occupies anterolateral part of IAC, whereas vestibule is located in the posterolateral part. In cochlear aplasia IAC development is normal, vestibular system is in its normal posterolateral location but cochlea is absent in the anterolateral part of the IAC. There are two types:

1. **Cochlear aplasia with normal vestibule (CANV):** vestibule and semicircular canals are normally developed in their usual location (Fig. 22.1a).
2. **Cochlear aplasia with dilated vestibule (CADV):** IAC formation is normal as well as the location of the vestibule. The only difference is the dilatation of the vestibule (Fig. 22.1b). This can be misdiagnosed as common cavity. A common cavity represents an ovoid or round structure with cochlear and vestibular neural tissue (Fig. 22.1c). IAC is usually posteriorly rotated and opens directly into the center of this deformity. In this way it is possible to differentiate between CADV and CC. This is very important to avoid an unsuccessful intervention in CADV with a CI surgery.

In both subtypes labyrinthine segment of the facial nerve is anteriorly displaced occupying the usual location of the cochlea.

The width and length of the IAC are normal in these patients. This is an interesting finding because in cochlear aplasia cochlear nerve is absent. In other cases of absent or hypoplastic cochlear nerve IAC is usually narrower than normal.

MRI demonstrates normal sized IAC with normal or dilated vestibule (Fig. 22.2a, b). It is also possible to have CANV and CADV in the same patient (Fig. 22.2c). Characteristically, in a CA patient only three nerves are present in sagittal oblique section passing through IAC (Fig. 22.2d).

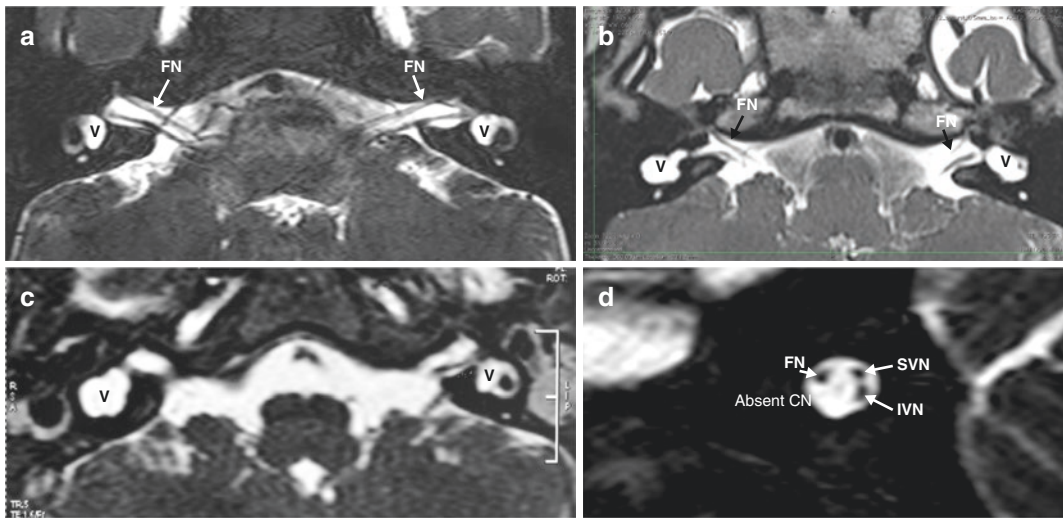

Fig. 22.2 Axial MRI demonstrating bilateral symmetric cochlear aplasia with normal vestibule (**a**) and dilated vestibule (**b**). It is also possible to encounter cochlear aplasia with normal vestibule (left) and dilated vestibule (right) in the same patient (*V* vestibule, *FN* facial nerve) (**c**). Parasagittal section perpendicular to the IAC showing absent cochlear nerve (*FN* facial nerve, *SVN* superior vestibular nerve, *IVN* inferior vestibular nerve) (**d**)

22.4.2 Audiological Findings

During audiologic evaluation, these patients will have no response at all or profound hearing loss at low frequencies (Fig. 22.3a, b). When evaluated together with complete labyrinthine aplasia, otocyst deformity, and cochlear aplasia, profound hearing loss at low frequencies demonstrates that this is purely a vibrotactile response and should not be interpreted as hearing in CI candidates with other pathologies. Nevertheless, most frequently observed audiometric profile for these cases is no response at the upper limits of the audiometer. Sometimes, they show auditory response at low frequencies in the free field testing and this is mostly regarded as vibrotactile stimulation.

Management: As there is no inner ear development, ABI is the only feasible surgical option to provide hearing sensations in children with cochlear aplasia. This is a definite indication for ABI.

In our department between 2006 and September 2018, 2646 patients underwent CI and ABI operations. One hundred and twenty-five children had ABI surgery for IEMs. Thirteen patients with cochlear aplasia underwent ABI surgery. Ten had CADV and 3 had CANV.

Similar to ABI users with other IEMs, aided thresholds reach the intensity levels of daily

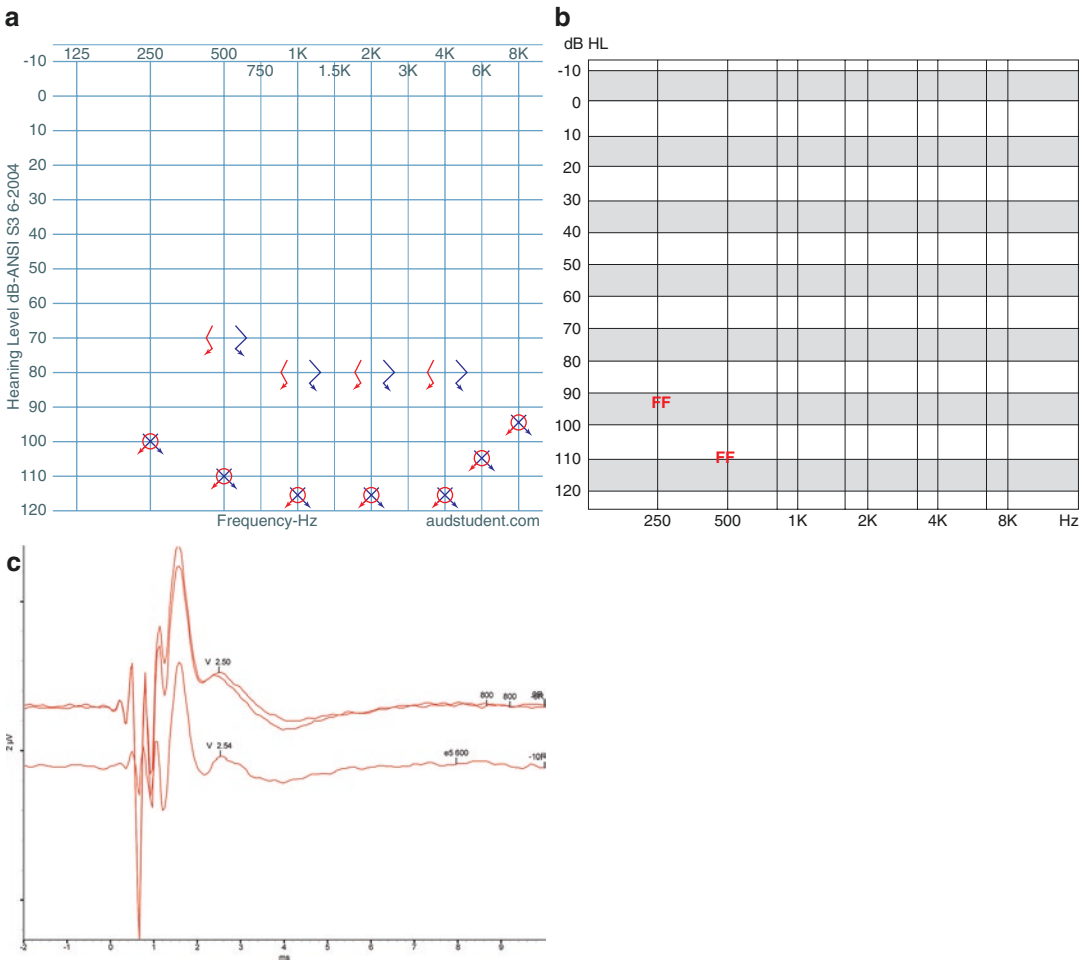

Fig. 22.3 Audiological findings in cochlear aplasia; No response (**a**), profound hearing loss at low frequencies (**b**). An example of intraoperative eABR wave from ABI (**c**)

speech sounds in time [8]. Cochlear nuclei in cases with cochlear aplasia can be stimulated as observed in intraoperative eABR recordings (Fig. 22.3c). Aided threshold examples are also given in figures below. Even though benefit from ABI shows variations between subjects, it can be concluded that adequate stimulation is provided to ABI users with cochlear aplasia. These findings show that cochlear nucleus can be stimulated for hearing despite the absence of cochlea, possibly indicating well developed cochlear nuclei in these patients.

During the activation of ABI, it is very important to have electrocardiographic monitoring of the patient to observe critical side effects. During follow-up sessions different side effects can be observed; the patient is observed closely with one of the parents and another audiologist. Parents are informed about the changes that can be expected. Audiologist should also have information about patient's typical habits (such as different facial expressions, eye blinking, etc.) and continuous behaviors. It is important to differentiate these behaviors from side effects. Active and inactive electrodes should be identified at initial activation but it is very important to keep as much electrode active as possible at secure limits.

It is necessary to have at least two audiologists during follow-up sessions. Depending on impedance changes and also child's improved sound experience, active and inactive electrodes may change with time. If an electrode causes a side effect, it is advisable to change all stimulation parameters, using longer duration/pulse width and smaller amplitudes, etc. If side effects disappear with these changes, then this electrode should be active.

22.5 Cases

Case 22.1: EK, 1.5-Year-Old Female Patient with CADV, Bilateral Sequential ABI User

During her audiological evaluation, she showed no response on electrophysiological and behavioral tests. According to CT and MRI results, she had bilateral cochlear aplasia with dilated vestibule. She was implanted with ABI on the left side (30.05.2013) and 28 months later she had second ABI on the right side (30.09.2015). She has all electrodes active bilaterally. Figure 22.4a and b show her intraoperative eABR recordings. Figure 22.4c shows free field thresholds with bilateral ABI. Her MAIS, with only left implant, was 26/40 after 2 years. Bilateral MAIS was 36/40 1 year after second implant. CAP score was 3 and SIR score was 2 with only left implant, and they were improved to 5 and 4, respectively, after second implant. After 1 year bilateral use, word identification was 6/12 and sentence identification was 5/10.

This patient demonstrates the importance of bilateral stimulation. In this particular case ABI was the only option for bilateral stimulation which improved her situation.

Case 22.2: BS, 3.5-Year-Old Female with CADV, Right ABI User

She had bilateral CADV. On preoperative testing, there was no response on electrophysiological and behavioral tests. She was implanted with an ABI at 3.5-year-old on the right ear. She had device failure and she had reimplantation on the same side. She has been using ABI for almost 6.5 years. After reimplantation, her free field thresholds were between 30 and 35 for 500 and 4000 Hz but later on threshold levels started to increase and she had some inactive electrodes due to side effects (Fig. 22.5).

She is a unilateral ABI user. The fact that thresholds increased on the right side, decreased her benefit from ABI. If she had been a bilateral user, this would have lesser impact on her daily life.

Case 22.3: FT 4-Year-Old Male Patient with CADV, Right ABI User

He was implanted at the age of 3 years and 9 months on the right ear. With his ABI he has been using auditory verbal communication and can detect and identify Ling's sounds. His free field thresholds are shown in Fig. 22.6. He has been using ABI for almost 10 years and has also ADHD. Despite the comorbidities, he makes benefit from the implant.

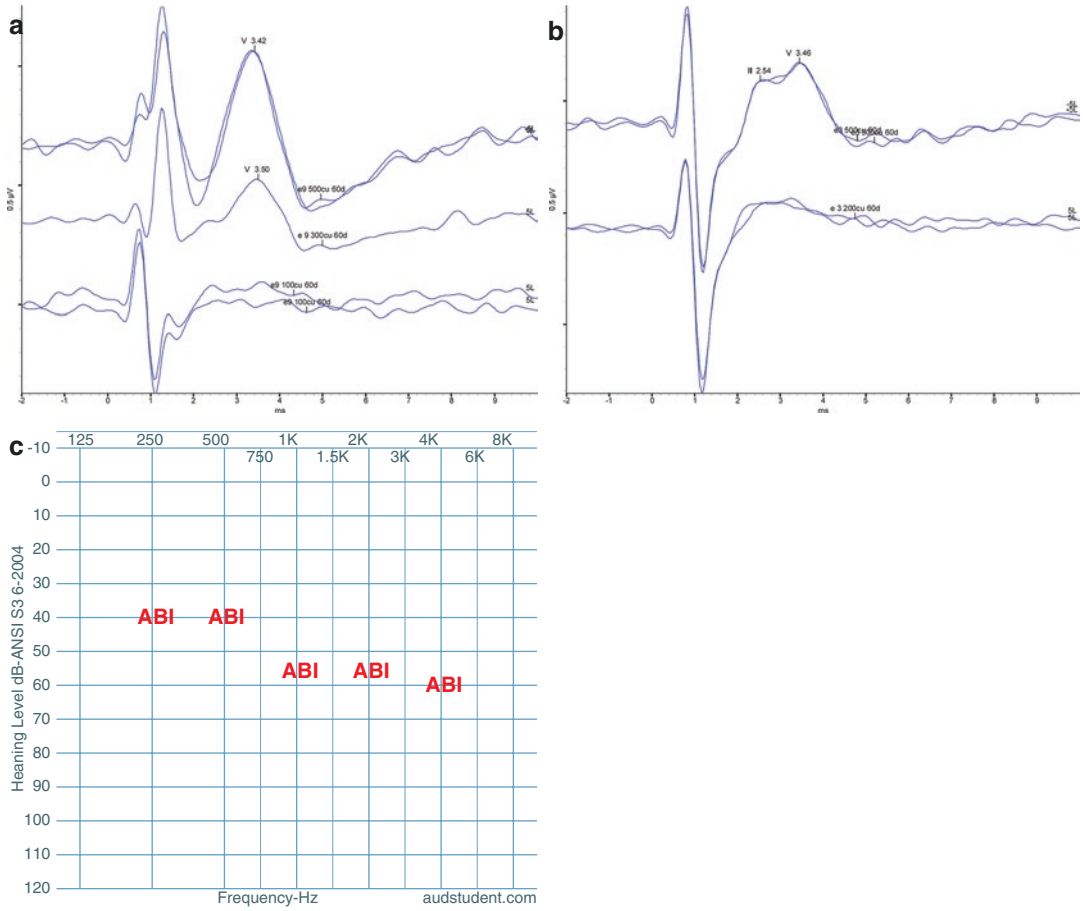

Fig. 22.4 (**a**) and (**b**) Case 22.1. Intraoperative eABR recordings from ninth (*A*) and third (*B*) electrodes. (**c**) Free field thresholds with ABI

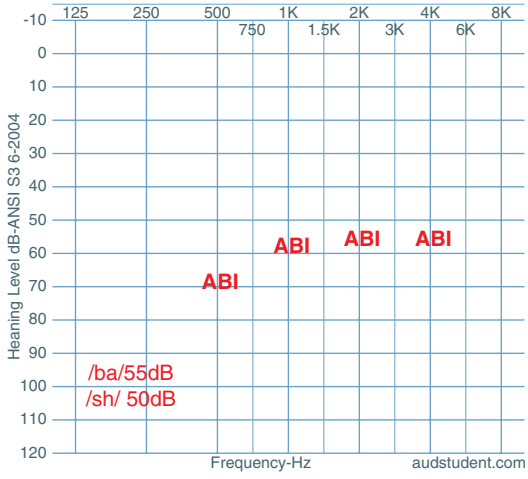

Fig. 22.5 Case 22.2. Free field thresholds with ABI

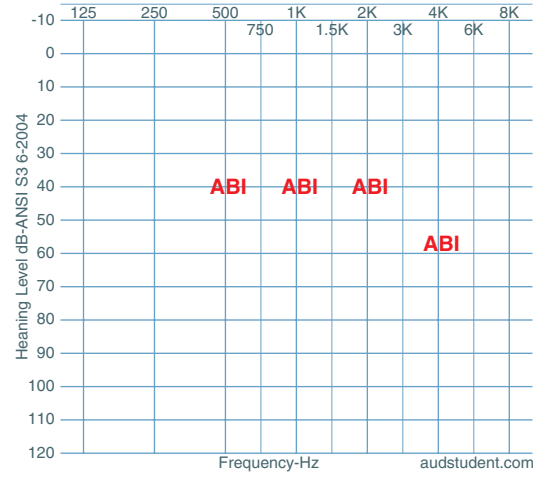

Fig. 22.6 Case 22.3. Free field thresholds with ABI

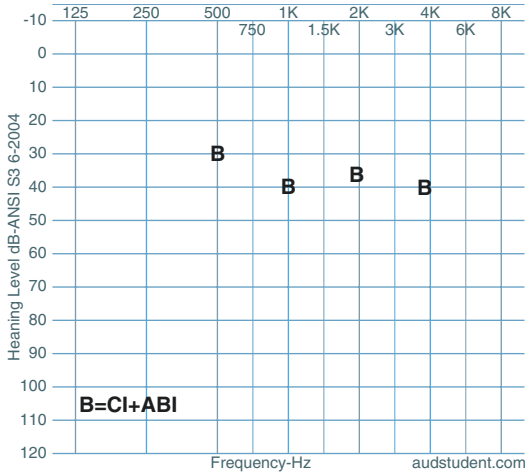

Fig. 22.7 Case 22.4. Free field thresholds with CI and ABI

Case 22.4: MD 2-Year-Old Male Patient with Cochlear Hypoplasia (Right Side) and CANV (Left Side), Bimodal User

He had different IEMs on both ears, cochlear hypoplasia on the right side and CANV on the left side. During preoperative testing, he had response to sound at 250 and 500 Hz with insert earphones and he was first implanted with CI on the right side. One and half years later, he was implanted with ABI on the left ear. Due to device failure he had a revision ABI surgery on the left side. His latest free field thresholds are shown in Fig. 22.7.

This patient shows the importance of binaural advantage by bimodal stimulation.

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