6 Preoperative Audiological Evaluation

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Special Features

1. Asymmetric hearing loss, fluctuation, and sudden HL may be important signs of inner ear malformations.
2. Testing with insert earphones is important for choosing the appropriate ear for the implantation.
3. Observing cochlear microphonics in auditory brainstem response testing can be a sign of cochlear nerve deficiency.
4. Preoperative audiological evaluation in inner ear malformations should be done carefully with two experienced pediatric audiologists.

Subjective evaluation was a widely used method of evaluation prior to the 1960s, when the objective evaluation method was not as effective as it is today. Behavioral testing represents a key aspect of audiological evaluation. In terms of the difference between subjective and objective testing, objective testing methods evaluate only a part of the auditory system, while behavioral testing, which is subjective, can evaluate the entire auditory system. Due to the upper limits of objective measurements, it may not prove possible to obtain responses in certain situations, although it would be possible to observe a response using behavioral testing methods.

6.1 Preoperative Evaluation Process

The preoperative evaluation process involves the otological, radiological, and audiological evaluation of the patient. The audiological evaluation helps to make a connection between the otological and radiological findings. Together with radiology, an audiological diagnosis enables the implant team to choose the most appropriate management strategy, such as cochlear implantation or auditory brainstem implantation, for children with inner ear malformations (IEMs). To diagnose the specific type of IEM, it is necessary to plan the radiological evaluation as soon as possible so as to allow for an effective audiological follow-up, including hearing aid fitting, auditory rehabilitation, and decision-making regarding auditory implants.

Kimura et al. [1] studied the relationship between vestibular function and gross motor development in children with IEMs using the rotational chair test. They concluded that IEMs are related to vestibular dysfunction as well as to delayed motor development. Indeed, both hearing loss and vestibular impairment can be seen in

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patients with IEMs. It is, therefore, important to preoperatively evaluate the vestibular systems of children with IEMs.

6.2 Audiological Evaluation

6.2.1 History of Hearing Loss

The anamnesis of a given patient consists of his/her medical history, developmental milestones, and family status, as well as observations on the part of the audiologist. The audiologist should carefully observe the physical status and facial appearance of the child, taking note of the occurrence of eye contact, vocalization, any response to environmental stimuli, and age-appropriate developmental milestones. The prenatal, perinatal, and postnatal factors that can cause hearing loss should be considered when taking the patient's history. It is difficult to identify a clear risk factor for an IEM. Most children with IEMs do not exhibit any obvious perinatal or postnatal risk factors. However, it is important to note that the most common prenatal risk factors are consanguineous marriages and a family history of hearing loss. These findings suggest that the etiology of the hearing loss in cases of IEMs could be genetic.

During the diagnosis and follow-up of children with IEMs, it is important to examine the initial diagnosis of hearing loss (HL), the duration of the HL, any history of sudden HL, progressive HL, attacks of HL, as well as the duration of use and benefit derived from hearing aids during daily life. Each of these points must be considered when beginning the preoperative audiological evaluation.

6.2.2 Behavioral Testing

The aims of the behavioral testing process are (1) to identify HL for medical follow-up; (2) to diagnose HL for auditory rehabilitation; (3) to assess the degree, type, and configuration of the HL; (4) to determine the difference between the ears; and (5) to determine the most suitable ear for implantation.

Thai-Van et al. [2] published audiological results concerning a child that appeared to conflict with the radiological diagnosis. The child exhibited behavioral responses at the level of 50 dB HL despite having cochlear nerve aplasia. This finding highlighted the importance of the audiological evaluation as well as the limitations of imaging in children with IEMs.

Lim et al. [3] retrospectively evaluated the medical records and radiological images of 42 children under 13 years old with unilateral sensorineural HL and bony cochlear nerve canal stenosis. They found that the degree of HL varied from moderate to severe/profound despite no correlation being identified between the pure-tone thresholds and the diameter of the bony cochlear nerve canal.

The review study conducted by Freeman and Sennaroglu [4] also emphasized the significance of subjective audiological testing, even when no response can be observed using other electrophysiological testing methods.

The identification of any audiological response is critical in terms of selecting the ideal ear for implantation, since it allows for the better ear to be considered during the pre-implantation counseling process. When the behavioral testing is performed using insert earphones, it is possible to select the better ear. Importantly, insert earphones are more readily accepted by children when they are used with their own ear molds.

In the study by Weiss et al. [5], the threshold estimation with insert earphones for children aged 18–24 months was determined to be more accurate than that achieved with supra-aural earphones. The subjective rating of the acceptance of the insert earphones was also higher when compared with the acceptance of the supra-aural earphones due to the reduced negative behaviors.

The evaluation of the bone-conduction thresholds is also necessary in order to identify the air-bone gap. In the case of some cochlear malformations, such as incomplete partition type II (IP-II), IP-III, enlarged vestibular aqueduct (EVA) syndrome, and certain cases of cochlear hypoplasia, it is possible to encounter an air-bone gap. In fact, the various audiological characteristics of incomplete partition malformations have been reported in a prior study [6].

Behavioral testing should be performed by an experienced pediatric audiologist. The pediatric audiologist should be aware of the normal development of the child, identify the developmental responses, and observe the child effectively. It is also necessary to determine which testing methods are most appropriate for a child given his/her developmental age and cognitive status. Age-appropriate testing includes behavioral observation audiometry, visual reinforcement audiometry, and play audiometry.

6.2.2.1 Behavioral Observation Audiometry

Behavioral observation audiometry (BOA) is based on the observation of behavioral responses to acoustic stimuli (i.e., eye blinking, head turning, head movement, changes in respiration, voice and suction). It is difficult to obtain a response to the pure tone in the case of babies aged 0–4 months. To better observe the responses, it is more appropriate to evaluate them using speech sounds (Ling sounds), frequency modulation (FM), or narrowband noise rather than using pure-tone sounds. It is recommended that the test be performed in a sound field using an ascending method. This method should always be evaluated alongside the objective testing. Since radiological evaluation is not recommended for babies under the age of 6 months, it is not possible to determine whether there is an IEM or not.

6.2.2.2 Visual Response Audiometry

Visual reinforcement audiometry (VRA) is based on the reinforcement of the behavioral response to sound accompanied by a visual stimulus. This method can be used in babies after the age of 5–6 months, when head movements start alongside the development of indirect localization abilities. The child can be positioned in the parent's lap, in a high chair, or in a baby seat to see the visual reinforcement. Light boxes, moving toys, or video VRA screens can be used to present the reinforcement. The repetition of speech sounds (i.e., /ba/, /sh/, /s/), a frequency-specific pure-tone stimulus, a warble tone, or narrowband noise (NBN) can be used as the auditory stimulus. As the behavioral responses are usually observed at higher intensity levels, speech stimuli that provide near-threshold information are most commonly used. Shaw et al. [7] found that when the VRA procedure was applied using NBN, babies aged 6–30 months responded better than when frequency-modulated tones, such as FM, were used. They hence recommended the use of NBN when conditioning babies by means of VRA.

At the beginning of the testing, the first stimulus should generally be presented above the threshold level (i.e., 70 dB HL). For children with severe IEMs (such as cochlear aperture stenosis or cochlear nerve hypoplasia), it is not usually possible to observe any response at this level. The ascending method is used, with the stimulus increasing in 10 dB increments, after providing the conditioning. During the test, the aim is to condition the baby by matching the light to the sound. Once the baby has been conditioned to the test and begins to turn his/her head toward the light, the threshold is determined by increasing the intensity, starting with lower levels (e.g., 30 dB). It is important to use supra-aural or insert earphones, since the intention is to elicit an ear-specific response. It is also important to use a bone-conduction vibrator, since an air-bone gap may be encountered in certain types of IEMs. During the test, one experienced pediatric audiologist should be in the testing room with the child, and he/she should focus his/her attention on the midline.

6.2.2.3 Conditioned Play Audiometry

The intention behind the play audiometry procedure is to pair the auditory stimulus with an interesting game, such as overlapping blocks, throwing a cube into a box, or inserting rings onto bars. Children with IEMs are usually diagnosed during the first year of life. Due to this early diagnosis, play audiometry is only rarely used in the preoperative audiological evaluation of IEMs. For children with progressive HL or sudden HL (i.e., EVA, IP-II), the HL tends to be diagnosed later, and the decision concerning implantation made at an older age, when compared to other IEMs.

Generally, the test should be started at a level of 70 dB for conditioning. The intensity of the stimulus is increased if no response is observed. Once the child has learned the game, the threshold is determined by increasing the intensity, starting from a low intensity (e.g., 30 dB). The ascending method is used, with the stimulus being increased in increments of 10 dB at the suprathreshold level. If necessary, the use of clinical masking is important in children who are cooperative. Due to the asymmetric audiological characteristics of EVA and IP-II, as well as in the case of unilateral deafness, masking should be used when determining the air- and bone-conduction thresholds.

Supra-aural earphones, insert earphones, or bone vibrators can be used during the testing. Two audiologists should work together to ensure that the test is performed safely, in a short time, and correctly. During the test, the audiologist nearest to the child should encourage the child, reward him/her with applause when a correct response is given, and check the reliability of the test.

6.3 Electrophysiological Measurements

In terms of the objective testing methods, electroacoustic immitancemetry, otoacoustic emission testing, auditory brainstem response testing, and electrical ABR testing should be performed during the preoperative evaluation.

6.3.1 Electroacoustic Immitancemetry

Tympanometry and acoustic reflex measurements should be performed during a routine audiological evaluation to assess the status of both the middle ear and the cochlear nerve. The use of these tests is even more critical in the case of IEMs due to the possible air-bone gap. In some types of IEMs, such as EVA, IP-II, and IP-III, it is possible to observe an air-bone gap without any middle ear pathology. Due to the third window phenomenon, it is not possible to explain this air-bone gap by means of tympanometry. Despite the presence of an air-bone gap, a type A tympanogram is usually observed, while acoustic reflexes can be detected according to the degree of the HL. This finding suggests the need to move away from the middle ear pathology. In some types of cochlear hypoplasia, it is possible to observe stapes fixation, which is characterized by the absence of an acoustic reflex in the presence of an air-bone gap. Electroacoustic immitancemetry testing is a fast and reliable method that can assist with the correct diagnosis of an IEM.

6.3.2 Otoacoustic Emission Testing

Otoacoustic emission (OAE) testing is an important test method that evaluates the function of the outer hair cells in the cochlea. Positive OAE results indicate normal outer hair cell function rather than cochlear nerve function. It is, therefore, important to interpret the OAE results together with the auditory brainstem response findings. In patients with cochlear nerve deficiency, testing with OAE can show positive responses where cochlear microphonic (CM) responses were observed in the auditory brainstem response testing without any repeatable waves. Testing using only OAE during neonatal hearing screening (NHS) can lead to false positive responses in children with cochlear nerve deficiency. Such cases can pass the OAE testing on one occasion, while they can fail on another occasion during test repetitions. Hence, automatic auditory brainstem response testing should be used routinely rather than relying on only OAE during NHS.

The second consensus meeting on the management of IEM and decision-making between cochlear implantation (CI) and auditory brainstem implantation (ABI) highlighted the importance of the preoperative audiological evaluation. Sennaroğlu et al. [8] stated that the use of OAE with the CM responses in auditory brainstem response testing might provide an indication of cochlear nerve aplasia/hypoplasia. The clinician should thus be careful when interpreting the audiological results.

James et al. [9] evaluated three children with unilateral cochlear nerve (CN) aplasia whose OAE responses were bilaterally positive and whose CM responses were observed by means of auditory brainstem response testing. After recording the OAE responses, broadband noise was applied to the contralateral ear with normal hearing at a level of 60 dB SPL. The authors reported that the suppression of the OAE was detected in all three children, which suggests an intact efferent neural function despite the finding of CN aplasia via magnetic resonance imaging (MRI).

6.3.3 Auditory Brainstem Response Testing

Auditory brainstem response (ABR) testing provides information about the function of the auditory pathway from the distal portion of the cochlear nerve to the lateral lemniscus. In cases of cochlear nerve deficiency (CND), the CM could be seen in the ABR testing. When observing the CM in ABR testing, the possibility of CND should be borne in mind and early radiological evaluation is advisable. In cases of Michel deformity, cochlear aplasia, and rudimentary otocyst malformations, no replicable waveform could be seen in ABR testing. Although in cases of IP-I, cochlear hypoplasia with CND, and cochlear aperture anomalies, the CM will be an important indicator of the inner ear malformation. In relation to these conditions, a radiological evaluation should be performed as soon as possible to ensure an appropriate diagnosis.

6.3.4 Electrical Auditory Brainstem Response Testing

Electrical ABR (eABR) testing is a useful evaluation technique that shows the activity of the auditory system, especially the cochlear nerve. The determination of the expected wave V latency is important during a preoperative evaluation. The eABR results are associated with the postoperative audiological outcomes following implantation. eABR waveforms are generally correlated with the neural integration, and they are an important indicator of the reaction of the cochlear nerve to electrical stimulation [10].

Cinar et al. [11] studied the electrically evoked ABR using an intracochlear test electrode (ITE) and a cochlear implant electrode in different inner ear malformations. They emphasized the importance of intraoperative eABR in patients with inner ear malformations, although they also reported that when a positive behavioral response is observed during the preoperative evaluation, CI can be performed even if there is no response in eABR testing. Further, it was observed that some cases exhibited no response in eABR testing even though the cochlear nerve was present on the MRI in inner ear malformations. Therefore, when using ITE, a finding of no response during eABR testing should be interpreted with caution, and the final decision should be made after taking into account both the audiological and MRI findings.

Ehrmann-Müller et al. [12] reported the results of the audiological evaluation of children with cochlear nerve deficiency prior to CI. The audiological assessment battery included subjective and objective tests, such as ABR testing and auditory steady-state response (ASSR) testing. They also performed promontorium stimulation testing or eABR testing when there was no response during the free-field testing. They emphasized how eABR serves as a predictive tool during the preoperative evaluation and, despite the cochlear nerve aplasia observed via MRI, the presence of cochlear nerve fibers can be indirectly demonstrated via eABR testing. Auditory brainstem implantation can be recommended for patients who do not exhibit any response to electrical and/or acoustic stimuli.

Based on our clinical experience, eABR is not always the most efficient tool for predicting the functionality of the cochlear nerve in patients with IEMs. The decision-making process should, therefore, consist of a preoperative audiological evaluation, the radiological findings, and the intraoperative eABR results. Despite good responses being observed in both behavioral tests and daily life, in some cases it is not possible to observe any waveform in the eABR testing. Hence, a correct decision can only be made in consultation with the implant team and after the available options have been discussed with the family.

6.4 Follow-Up with Hearing Aids

When HL has been diagnosed, the habilitation process should begin with a hearing aid trial prior to the radiological evaluation. Although the radiological evaluation will be performed at around 6–9 months, and the presence of an anomaly in the cochlea and auditory nerve will be defined at that point, it is possible to recommend bilateral hearing aids for a child from the age of 3 to 6 months when the diagnosis of HL is confirmed. There could be directive indicators for the audiologists to observe during the follow-up. There might be indications that the child is benefiting from the use of hearing aids, such as a request to wear hearing aids by the child, the presence of satisfaction with the hearing aids, and the observation of auditory reactions during daily life by the parents. If there is a suspected response, such children should be followed up with bilateral hearing aids despite the absence of the cochlear nerve on the MRI. Although the use of hearing aids may seem unnecessary in the presence of severe IEMs prior to auditory implantation, it will contribute to helping the child become accustomed to wearing a device and preparing the family for the habilitation process. Getting used to the earmold by using hearing aids during this process will help to facilitate the use of insert earphones during behavioral testing.

6.5 Case Studies

Case 1: ZÇ, A Two-Year-Old Female

The parents applied for an audiological evaluation after she failed to pass the NHS in both ears. During her first evaluation, she exhibited no response to any sound, including narrowband noise, warble tone, and pure tone in a free field. ABR testing was planned as the next step. In terms of the ABR testing, there were no remarkable waves at the level of 99 dBnHL, although CM responses were observed in both ears (Fig. 6.1a). Following the audiological evaluation, the patient was evaluated by means of high-resolution computed tomography (HRCT) and MRI. Bilateral cochlear aperture stenosis and cochlear nerve hypoplasia were identified. During the follow-up, bilateral hearing aids were recommended for both ears when she reached the age of 7 months. The threshold testing with hearing aids and with insert earphones revealed the benefit of the devices, with thresholds between 55 and 60 dB HL being observed in low and middle frequencies (Fig. 6.1b). Despite good speech perception scores, her speech development was not as good as in children with normal cochlear anatomy. Bilateral CI was recommended.

Case 2: AMİ, A Three-Year-Old Female

She failed the NHS in both ears. She had no prenatal, natal, or postnatal risk factors. She was diagnosed with bilateral CM responses with negative OAE. Her behavioral testing was performed using insert earphones (Fig. 6.2). Her radiological evaluation revealed bilateral cochlear aperture stenosis and a narrow internal acoustic canal with CN aplasia. She exhibited good responses to environmental sounds as well as an improvement in language development with the use of hearing aids. Bilateral CI was recommended.

Despite the presence of CN aplasia, in the case of good auditory responses and language development appropriate for the child's chronological age, CI should be recommended. During the initial counseling, the possibility of ABI during follow-up should also be mentioned to the family. If limited progress is observed with the cochlear implant during the audiological follow-up, the patient can be evaluated with regard to the suitability of ABI.

Case 3: LG, A Two-Year-Old Female

She failed the NHS in both ears. She was born in the 25th gestational week and diagnosed with developmental delay. Bilateral hearing loss was

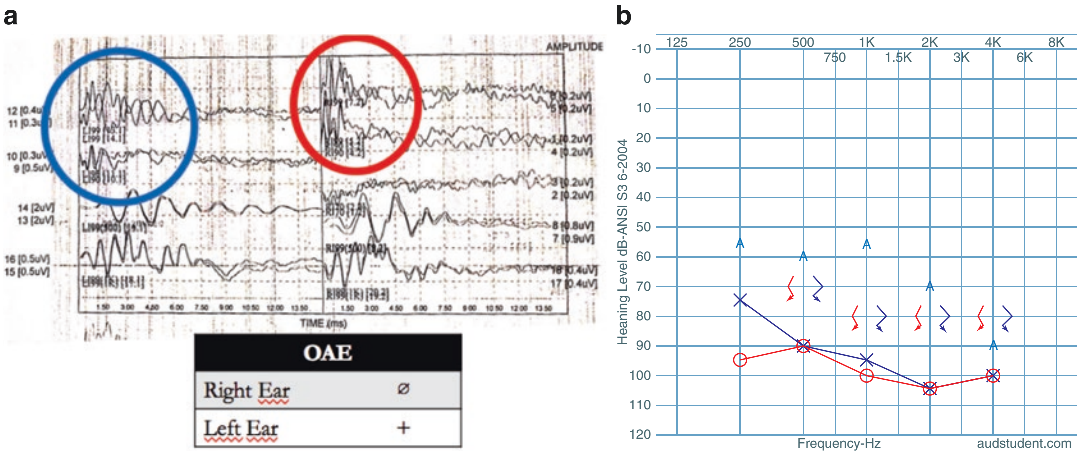

Fig. 6.1 Case 1: (**a**) Auditory brainstem response (ABR) showing cochlear microphonics without any response at the level of 99 dBnHL on both ears and otoacoustic emission (OAE) test results. (**b**) Audiogram with insert earphones presenting responses on both ears and thresholds with hearing aids revealing good responses between 55 and 60 dB hearing level in low and middle frequencies (A = aided threshold with hearing aids)

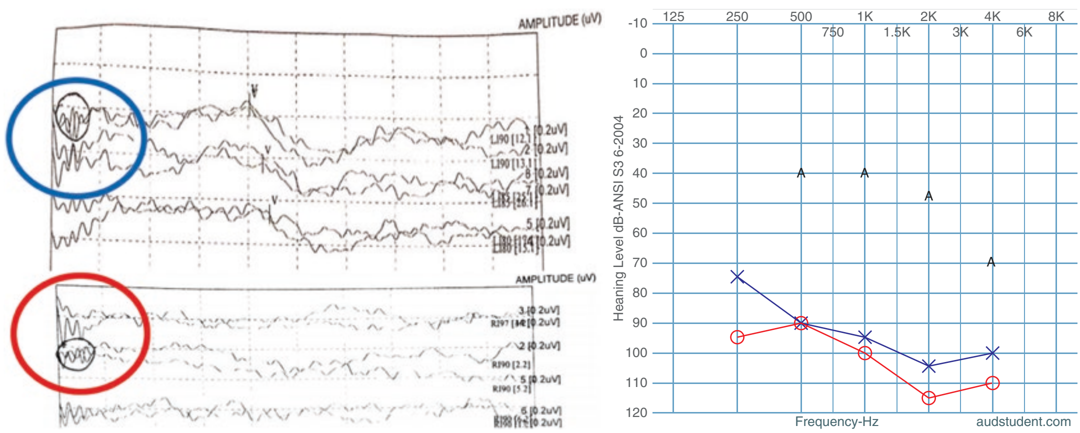

Fig. 6.2 Case 2: ABR test result showing bilateral cochlear microphonics with repeatable wave V on the left ear and the audiogram with insert earphones indicating good responses on both ears

identified at the age of 14 months and hearing aids were recommended on the both side. During the ABR testing, bilateral CM responses were observed without any repeatable waveform, especially with larger amplitudes on the left side. During the behavioral testing with VRA using insert earphones, responses were observed on only the right side (Fig. 6.3). The radiological evaluation revealed bilateral cochlear aperture stenosis and a narrow internal acoustic canal with CN hypoplasia in the right ear and CN aplasia in the left ear. Simultaneous CI in the right ear and ABI in the left ear were recommended.

In patients with CND, it is necessary to recommend bilateral amplification prior to surgery to prepare the child and his/her family for the rehabilitation period.

Case 4: NO, A Nine-Year-Old Female
She passed the NHS in both ears. She had the risk factor of prematurity. During the first audiological evaluation, which was performed at the age of 6 months, the ABR results showed bilateral CM responses with positive OAE in both ears, in which the amplitude of the CM responses was wider on the right side (Fig. 6.4a). These findings

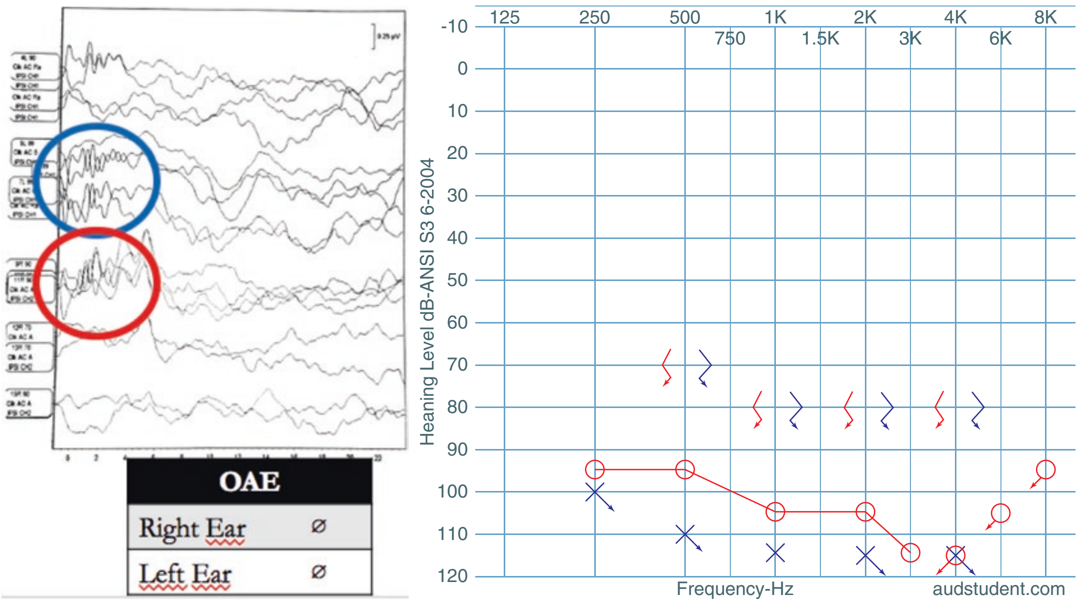

Fig. 6.3 Case 3: ABR indicating cochlear microphonics bilaterally with negative OAE and audiogram with insert earphones indicating good responses at the level of 95–110 dB from low to high frequencies on the right ear and no response on the left ear

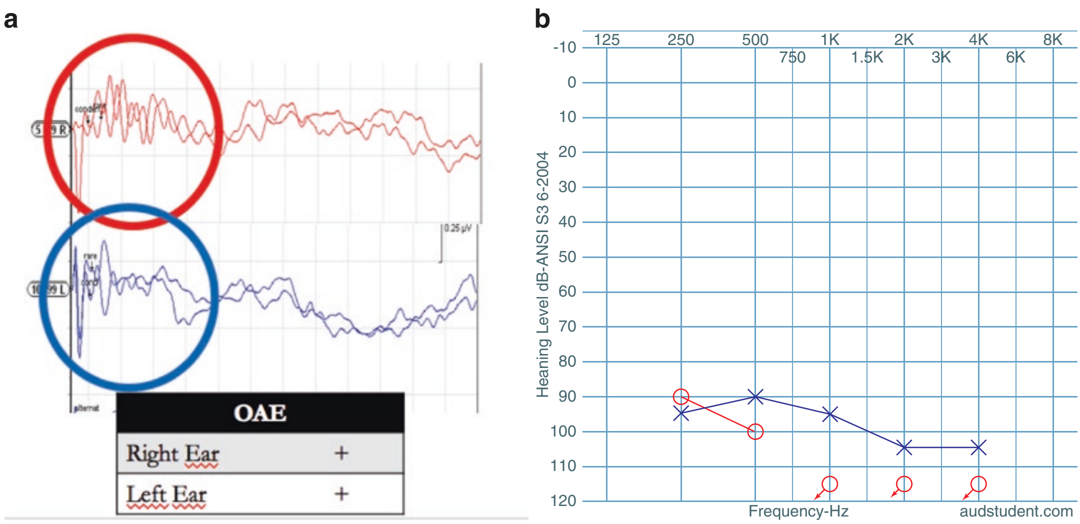

Fig. 6.4 Case 4: (**a**) ABR showing bilateral cochlear microphonics without any repeatable waveform, in which amplitude of the cochlear microphonics was wider on the right side. OAE test result revealed bilateral positive responses. (**b**) Audiogram with insert earphones indicating good responses on the left ear in all tested frequencies whereas no response on the right ear except vibrotactile stimulus at the low frequencies

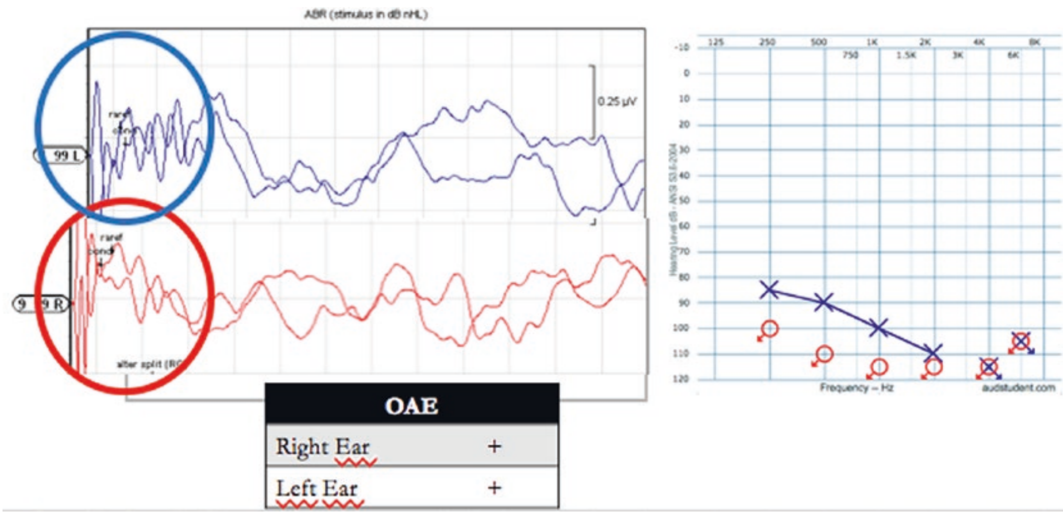

Fig. 6.5 Case 5: ABR presenting bilateral cochlear microphonics with positive OAE responses test result and audiogram with insert earphones indicating good responses on the left ear in the frequency range of 250–2000 Hz despite no response to the sound on the right ear

were suggestive of bilateral auditory neuropathy spectrum disorder. Bilateral hearing aids were recommended and an auditory rehabilitation program was planned for during the follow-up period. The family reported good responses to environmental sounds when using the hearing aids in daily life. During the follow-up, she was tested using insert earphones. The audiological findings suggested asymmetric hearing loss and no response was observed in the right ear, except in relation to a vibrotactile stimulus at low frequencies (Fig. 6.4b). The radiological assessment showed bilateral cochlear hypoplasia type III and hypoplastic aperture with CN aplasia in the right ear and CN hypoplasia in the left ear. A cochlear implant was recommended in the left ear. Her language development showed an improvement following left ear CI, although she reached a plateau after a while and showed no further development. In agreement with her family, ABI was recommended for the contralateral ear so as to provide bimodal stimulation.

Case 5: AE, A Six-Year-Old Male

He passed the NHS in both ears without any risk factors. He had a motor defect in his left hand. His family noticed that he exhibited no response to sounds during daily life and, therefore, applied to another center for the investigation of this complaint. An audiologist performed OAE testing and informed the family that the boy had bilateral normal hearing. The family was not satisfied and so applied to our clinic for a second opinion. An audiological evaluation was performed when he was 15 months old. ABR testing was performed and CM responses were observed bilaterally without any repeatable wave V. He was also evaluated using insert earphones and good responses were observed in the left ear (Fig. 6.5). He was recommended to undergo radiological evaluation. The radiological evaluation showed bilateral cochlear aperture stenosis with CN hypoplasia in the left ear and CN aplasia in the right ear. Right ABI and left CI were recommended.

If the signs suggest the presence of auditory neuropathy spectrum disorder, a radiological evaluation should be planned as soon as possible. To achieve better speech recognition and language development, it is important to provide more information for patients with IEMs.

Case 6: ŞO, A Two-Year-Old Female

At the age of 9 months, she was diagnosed with bilateral cochlear hypoplasia type I, with cochlear nerve hypoplasia on the left side, while the right cochlear nerve was aplastic. Her first test using insert earphones showed auditory responses in the left ear (Fig. 6.6a). ABI was performed in her right ear when she was 16 months old. Her behavioral responses with the left hearing aid showed an improvement during that period. She routinely used her left hearing aid together with the right auditory brainstem implant when she woke up in the morning (Fig. 6.6b). An example of her behavioral testing using insert earphones is provided in Video 6.1. She underwent CI surgery on the left side.

Case 7: EES, A Six-Year-Old Male

He was evaluated following a complaint of hearing loss, and ABR testing was performed when he was 2 years old. He had a family history (his cousin) of hearing loss. During the initial ABR testing, a wave V was observed at a level of 50 dBnHL in the right ear and at 70 dBnHL in the left ear (Fig. 6.7a). Due to a lack of cooperation with the use of supra-aural earphones, the first audiogram was performed in a free field (Fig. 6.7b). The radiological evaluation was recommended for the asymmetric hearing loss, and the HRCT demonstrated a bilateral IP-II deformity. The family was informed about the risks of sudden and progressive hearing loss. He was advised about needing protection from head trauma. Two months later, the family applied with complaints of sudden HL and dizziness, nausea, and vomiting. The ABR testing was repeated and no wave was observed at the level of 90 dBnHL with a click and tone-burst stimulus bilaterally. He was hospitalized for medical treatment (Fig. 6.7c). Although his family did not want CI, a fluctuation in his hearing was determined during the audiological follow-up with hearing aids. He had experienced three sudden HL attacks in the right ear. After the sudden hearing loss (Fig. 6.7d), he was hospitalized for medical treatment with steroids and dextran for 10 days. His hearing improved following the hospitalization (Fig. 6.7e). Two weeks later, the family again applied with sudden HL (Fig. 6.7f) on both sides. As a result, his hearing deteriorated and CI was recommended (Fig. 6.7g).

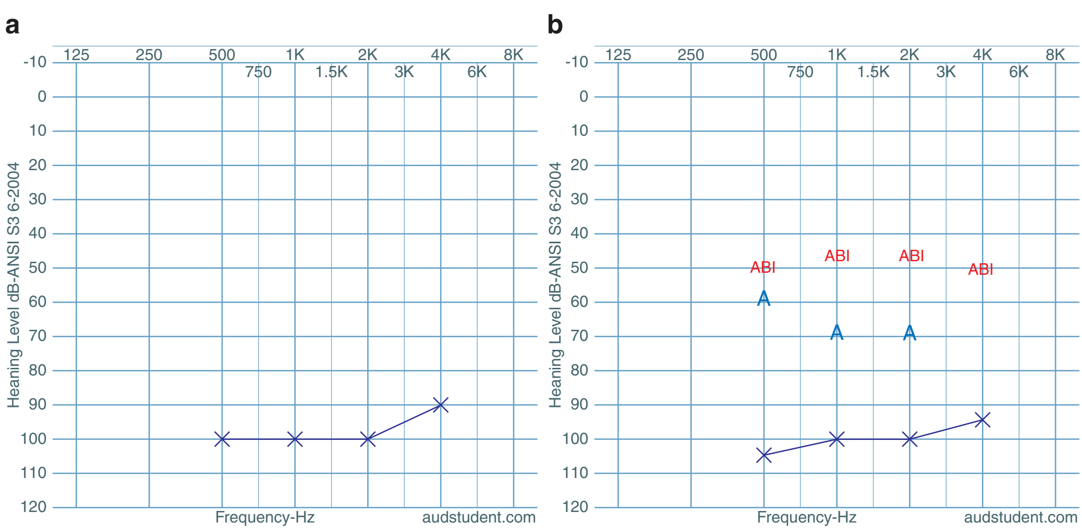

Fig. 6.6 Case 6: (**a**) Hearing thresholds of the left side with insert earphones indicating behavioral responses on the left ear when she was at the age of 9 months. (**b**) Hearing thresholds with left hearing aid (A) presenting responses in 0.5–2 kHz and right auditory brainstem implant (ABI) showing good responses in the area of speech banana (45–50 dB HL)

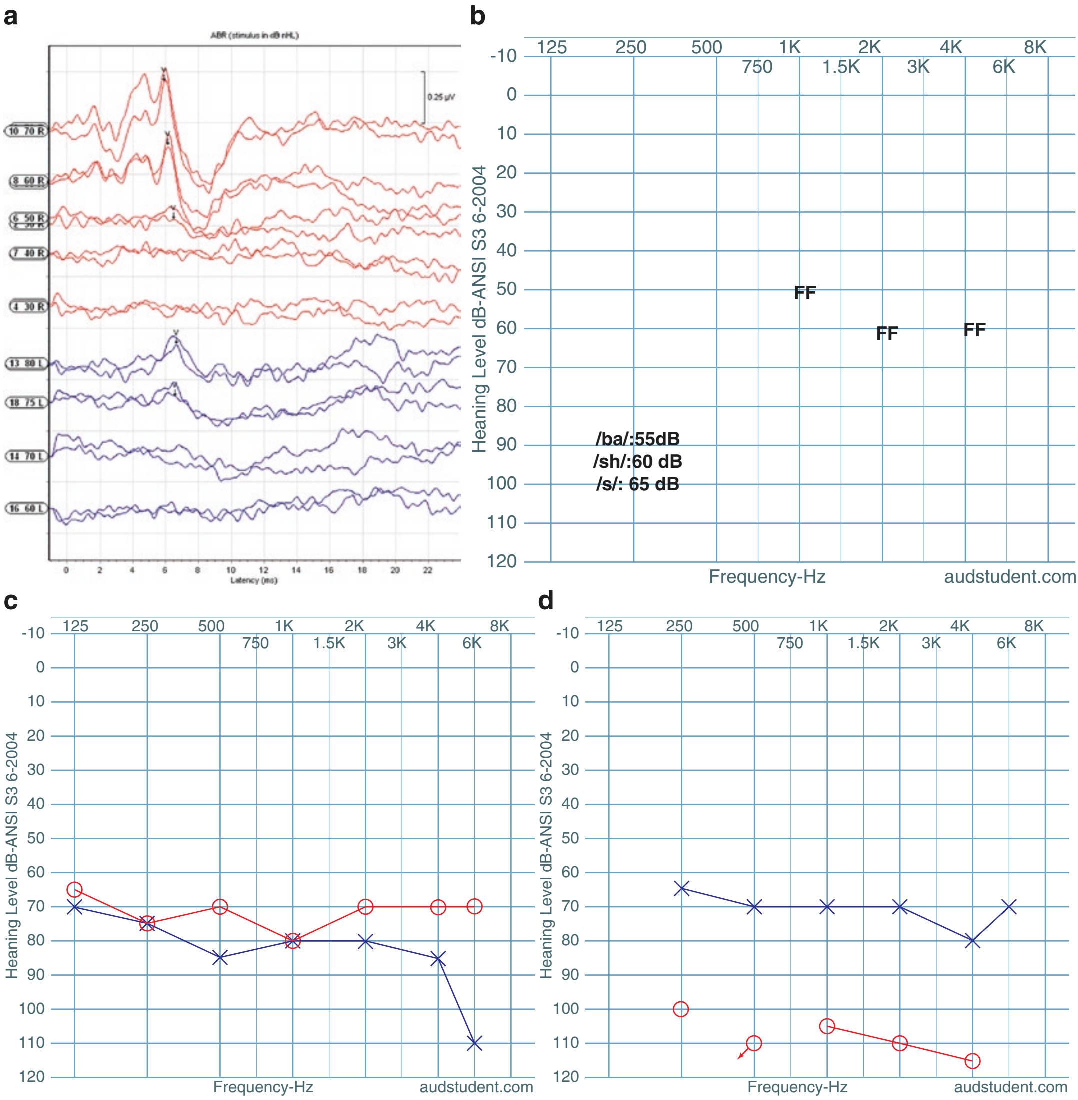

Fig. 6.7 Case 7: (**a**) First ABR test results when he was 2 years old presenting asymmetric hearing loss in which wave V was observed at the level of 50 dBnHL on the right ear and 75 dBnHL on the left ear with click stimulus. (**b**) First audiogram in free field showing hearing thresholds (FF: Free Field) without hearing aids and responses to the /ba/, /sh/, and /s/ speech stimuli. (**c**) Bilateral severe hearing loss (HL) was diagnosed after hearing loss attack. (**d**) Right side profound HL after recurrent sudden HL attacks which occur three times. (**e**) Hearing recovery on the right side after medical treatment for 10 days and his hearing thresholds improved after hospitalization up to the levels of 70 dB HL. (**f**) Sudden SNHL on both sides 2 weeks after recovery resulting with severe hearing loss on both ears. (**g**) No recovery after last attack. Due to the profound HL cochlear implantation was recommended (A: Aided thresholds with hearing aids)

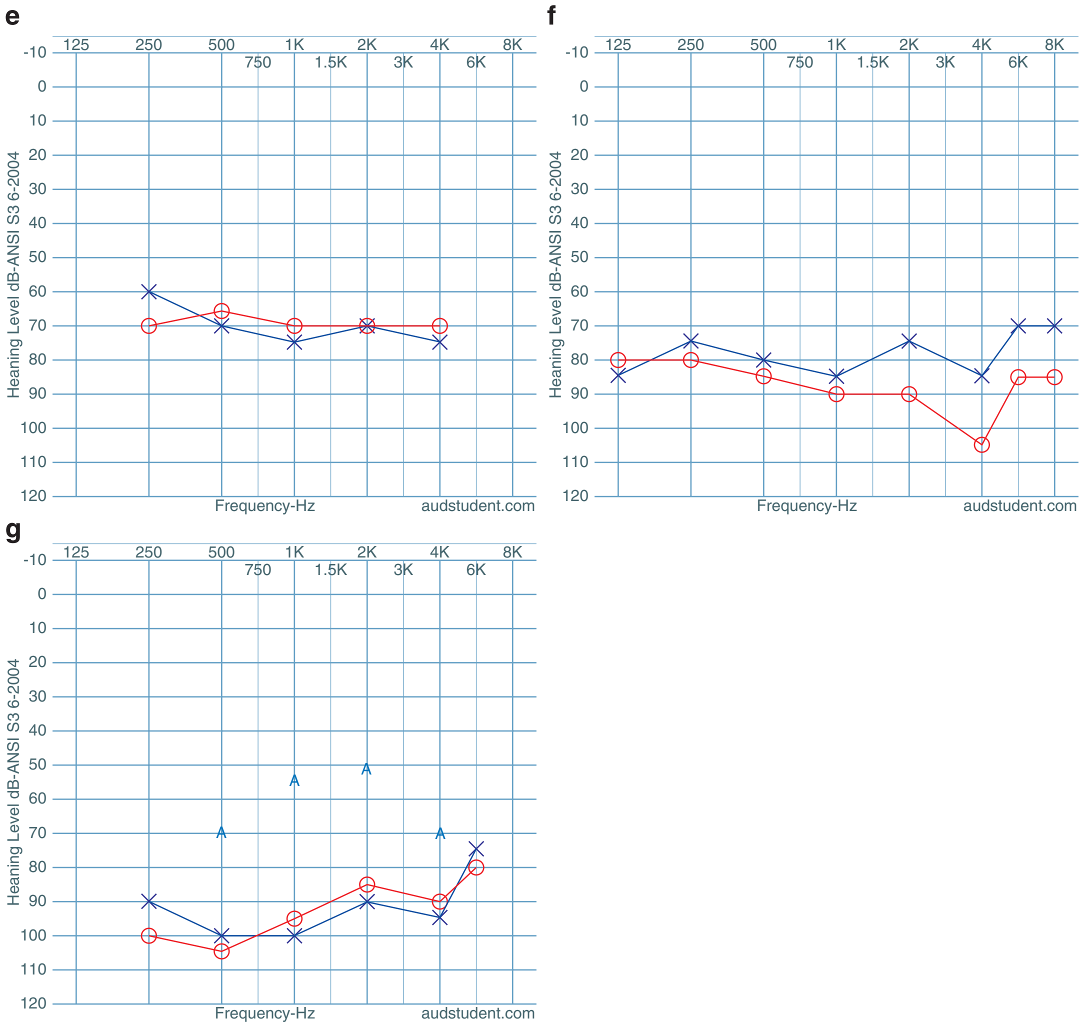

Fig. 6.7 (continued)

A history of HL attacks and sudden HL, fluctuations, or dizziness should raise the possibility of an inner ear malformation (particularly IP-II) in the minds of audiologists, and a radiological evaluation should be performed as soon as possible to ensure an early diagnosis.

Case 8: DS, A Six-Year-Old Male

He failed the NHS together with his twin. ABR testing was performed and a wave V was observed at a level of 80 dBnHL with a click stimulus bilaterally. He was diagnosed with moderate to severe mixed-type hearing loss (Fig. 6.8a). His radiological evaluation revealed a bilateral IP-III malformation. His big brother and his twin were also diagnosed with IP-III and they started the rehabilitation period with bilateral hearing aids. During the follow-up, CI was recommended because of a decrease in his hearing thresholds, the limited benefit he obtained from the hearing aids, and the inadequate improvement in his auditory skills loss (Fig. 6.8b). He underwent CI on the right ear, and he prefers to use a hearing aid in the contralateral ear. In Fig. 6.8c, his hearing thresholds with right CI and a left hearing aid are presented.

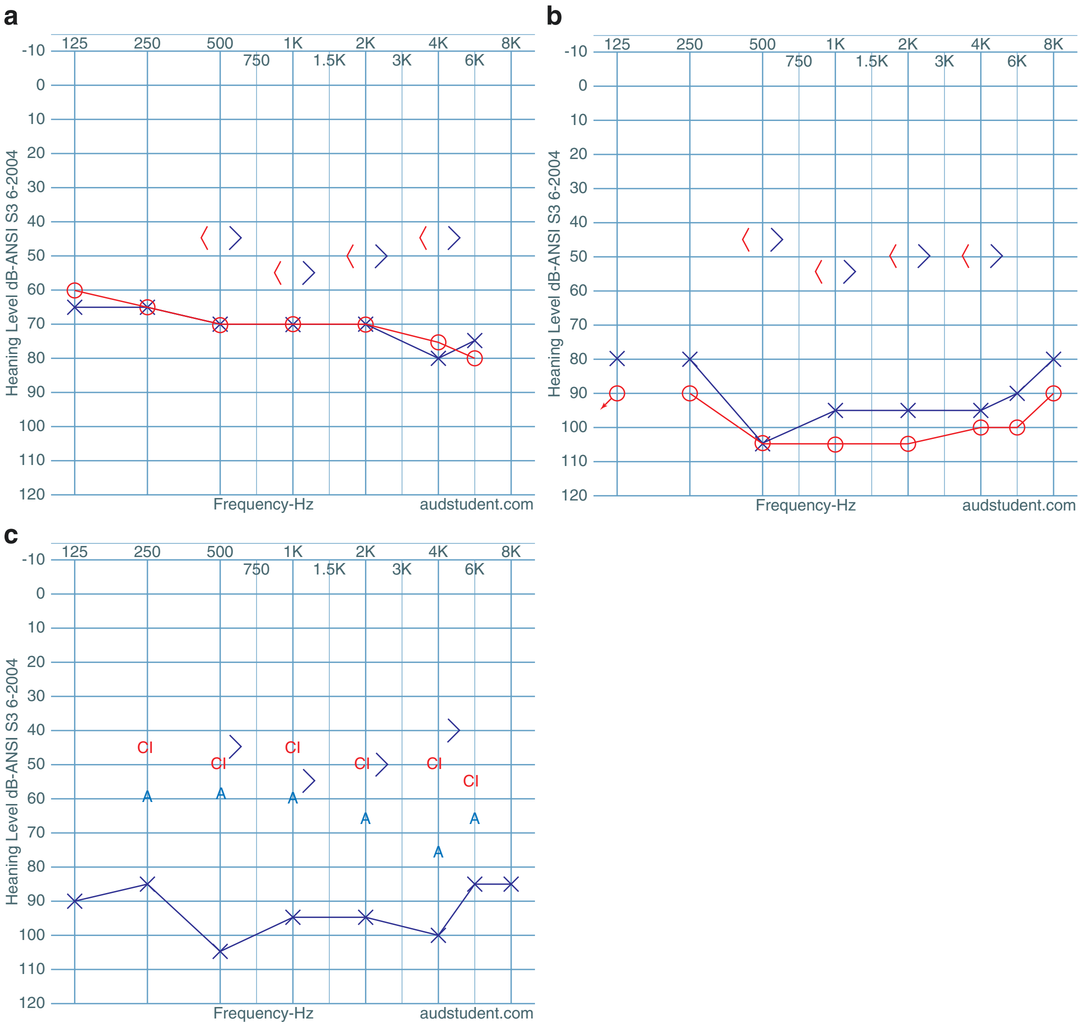

Fig. 6.8 Case 8: (**a**) First audiogram with insert earphones showing bilateral severe mixed-type hearing loss. (**b**) Follow-up audiogram requiring cochlear implantation due to the bilateral profound mixed-type hearing loss. (**c**) Hearing thresholds with right cochlear implant (CI) and left hearing aid (A) after 2 years follow-up presenting better hearing on the right ear

An incomplete partition type III malformation is characterized by severe to profound mixed-type hearing loss. It is necessary to evaluate the bone-conduction thresholds in children to determine the presence of an air-bone gap. Nearly all our patients with IP-III were rehabilitated with cochlear implants, and most of them prefer to use hearing aids in the contralateral ear. Despite the profound HL, patients with IP-III can benefit from hearing aids in the other side in terms of bimodal stimulation.

6.6 Take-Home Message

If any of the signs listed in Table 6.1 are present, children should be evaluated with regard to an inner ear malformation as soon as possible by means of a radiological evaluation.

Table 6.1 Signs suggesting cochlear malformation for audiologists

Signs suggesting cochlear malformation for audiologists
Asymmetric hearing loss
Sudden hearing loss
Progressive hearing loss
Fluctuations in hearing
Unilateral hearing loss
Air-bone gap (especially in a low frequency) without any middle ear pathology
Cochlear microphonic responses in ABR testing
Limited progress with hearing aids despite an appropriate amplification

6.7 Putting the Pieces Together

In the case of hypoplastic cochlear nerve and inner ear malformations, the patients' performance using hearing aids and auditory implants was found to negatively influence the outcomes. However, these conditions should not be considered as absolute contraindications for CI. All the cases presented in this chapter received a benefit from CI in the presence of auditory responses. We have personal experience with the recipients in each of these categories, who still exhibit a remarkable benefit from the use of a cochlear implant. Finally, the patients' performance will be influenced by the presence of additional handicaps, the age at implantation, the level of family support, as well as the cognitive and developmental status.

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