32 Cochlear Nerve Deficiency and Current Management of Inner Ear Malformations

Levent Sennaroglu, Gonca Sennaroglu, and Esra Yücel

Special Features

1. They may pass neonatal hearing screening test done with otoacoustic emissions only. Automatic ABR can diagnose the situation at birth.
2. MRI may not demonstrate the true status of the cochlear nerve if it is hardly visible in a narrow internal auditory canal.
3. Decision about best treatment modality should be made together with radiological and audiological findings.
4. CI results, in general, not satisfactory in the majority of the cases.
5. Better audiological outcome can be obtained using bimodal stimulation with CI and ABI.

32.1 Introduction

Cochlear nerve (CN) hypoplasia presents a dilemma to the implanting teams in choosing the most appropriate habilitation method. In patients with CN hypoplasia (CN deficiency), cochlear nerve has a smaller diameter than normal and usually the results of cochlear implantation (CI) are not as good as in children with normal cochlea and CN. If CI outcome is insufficient, they may need a contralateral ABI during follow-up. Therefore, it is important to diagnose this condition preoperatively and counsel the family accordingly.

32.2 Definition

According to Casselman et al. [1] cochlear branch of the cochleovestibular nerve (CVN) is normally larger than the facial nerve (FN), although the latter can be as large or even larger. If CN is smaller in diameter than FN on parasagittal section of internal acoustic canal (IAC), it can be accepted as hypoplastic. The findings were more constant in the cerebellopontine angle, where the facial nerve and the CVN are found, and the latter was nearly always 1.5 times larger than the FN and was never smaller. Kutz et al. [2] also agree with the definition that CN hypoplasia is used if CN is smaller than the facial nerve in the mid-portion of the IAC. Morita et al. [3] reported the mean diameter of CN and CVN as 0.9 mm and 1.2 mm, respectively. They concluded that better outcome from cochlear implantation can be expected when CN and CVN are depicted on MRI, regardless of the nerve diameters. Jaryszak et al. [4] measured CN on MRI and found that normal CN has vertical diameter

L. Sennaroglu (✉)
Department of Otolaryngology, Hacettepe University, Faculty of Medicine, Ankara, Turkey

G. Sennaroglu · E. Yücel
Department of Audiology, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_32

1.4 mm, horizontal diameter 1.0 mm, and cross-sectional area 1.1 mm^2.

32.3 Classification of Cochlear Nerve Abnormalities

It is very important to classify the cochlear and cochleovestibular nerves appropriately. Sennaroglu L proposes the following classification for CN and CVN abnormalities in inner ear malformations (please refer to the Classification of Cochlear and Cochleovestibular Nerves in Chap. 1: Classification (Figs. 1.13, 1.14, 1.15 and 1.16 for more details):

32.3.1 Normal Cochlear Nerve (CN)

Normally CN can be followed between cochlea and brainstem on lower axial sections passing through the IAC (Fig. 1.13a). On parasagittal sections, there is a separate CN located in the anterior inferior part of the IAC, entering the cochlea (Fig. 1.13b). The size of the cochlear nerve is similar in size when compared with the CN on the contralateral normal side. According to Casselman et al. [1] on parasagittal view the size of the CN is larger than the ipsilateral FN.

32.3.2 Hypoplastic CN

There is a separate CN but the size is less than the contralateral normal CN or ipsilateral normal facial nerve (Fig. 1.14a, b). CN hypoplasia can be subdivided into two groups:

Type I: CN is definitely present and it can be followed easily into the cochlea but its size is smaller in diameter when compared to ipsilateral FN and contralateral normal CN. CI is definitely indicated in this situation (Fig. 1.14a, b).
Type II: CN is an extremely thin and hardly visible and on axial MRI it can be scarcely followed into cochlea (<10% of the normal CN or ipsilateral FN). These are the cases where a decision between CI and ABI has to be made (Fig. 1.14c).

32.3.3 Absent CN

There is no nerve in the anteroinferior part of the IAC (Fig. 1.15a, b). This situation is definitely present in cochlear aplasia. It can also be seen in cochlear aperture hypoplasia and aplasia.

32.3.4 Normal CVN

Normally cochlear and vestibular nerves originate at the brainstem together forming the CVN. CVN then separates into CN and superior and inferior vestibular nerves in the IAC. In cases of common cavity CVN enters the cavity without separating into individual nerves. With radiological precision at the present time, it is impossible to determine the cochlear fiber content in the CVN but if the size is 1.5–2 times as much as the ipsilateral FN or similar to contralateral normal CVN it can be accepted as anatomically normal (Fig. 1.16a, b).

32.3.5 Hypoplastic CVN

If CVN is smaller than contralateral CVN or ipsilateral FN, it can be accepted as hypoplastic (Fig. 1.16b). CVN hypoplasia is particularly important in CC.

32.3.6 Absent CVN

In case of Michel deformity with absent IAC, CVN is also absent. Only FN can be identified (Fig. 1.16c).

32.4 CI and ABI Indications

In general CI and ABI are indicated in three categories:

Group 1: Definite CI Indications
These are IEMs that are definite CI candidates. IP-II, IP-III, and EVA are situations where ABI is never indicated.
Group 2: Definite ABI Indications
In the First Consensus Meeting [5] indications for ABI were discussed and two groups of indications were identified. In Definite ABI Indications group, there is no possibility for CI surgery and definitely an ABI is the only surgical option. Rudimentary otocyst is later added to these indications. These are:

1. Complete labyrinthine aplasia (Michel aplasia)
2. Rudimentary otocyst
3. Cochlear aplasia
4. Cochlear nerve aplasia
5. Cochlear aperture aplasia

Group 3: Possible ABI Indications
In this group of candidates there is a role for CI and ABI. CI can surgically be placed into a cochlear hypoplasia (CH), cochlea with incomplete partition type I (IP-I) anomaly, common cavity (CC), or a normal cochlea with hypoplastic CN, but the outcome cannot be determined at the beginning. It is usually accepted to give a trial period with CI and in case of insufficient progress in hearing and language development, contralateral ABI should be done. These indications are:

1. CH with cochlear aperture hypoplasia: As the cochlea has less than 1.5 turns, it is difficult to expect similar outcome as in patients with normal cochlea. In addition, these cases usually have cochlear aperture stenosis with CN deficiency. ABI may be indicated in these cases. However, it is possible to have normally developed CN in some cases with CH-II, CH-III, and CH-IV and ABI is not indicated in those particular cases.
2. CC and IP-I cases if the CVN or CN is hypoplastic, respectively. If CI is performed, CVN or CN may not be sufficiently developed enough to carry information to the brainstem.
3. CC and IP-I cases even if the CVN and CN are present, the distribution of the neural tissue in the abnormal cochlea is unpredictable, and ABI may be indicated in such cases if CI fails to elicit an auditory sensation.
4. The presence of an unbranched CVN is a challenge in these cases. This is seen in CC. The nerve entering CC is more correctly termed as CVN as there is no differentiation into CN and VN. In this situation, it is not possible to determine the amount of cochlear fibers traveling in the nerve. If there is a doubt, a cochlear implant can be used in the first instance, and ABI can be reserved for patients with an insufficient response.
5. Hypoplastic CN presents a dilemma for the implant team. A hypoplastic CN is defined as when the size is less than contralateral normal CN or ipsilateral FN. CI is definitely indicated in CN hypoplasia type I patients but type II patients with hardly visible CN is a dilemma to the implanting team to choose between a CI and ABI. The radiology in these patients should be carefully reviewed with an experienced neuroradiologist. Decision should be made with audiological findings as well. If a sufficient amount of neural tissue cannot be followed into the cochlear space, an ABI may be indicated.

If CN is visible and its size is roughly between 50 and 100% of FN or contralateral CN, it is advisable to use CI first and in case of unsatisfactory outcome performing an ABI.

Real challenge is the situation where CN is hardly visible and it is <50%. Depending on the patient, age, additional disability in these cases CI and ABI are indicated. If the age is around 2–3, the procedure can be done simultaneously.

32.5 Preoperative Workup

32.5.1 Radiology

Ideally CN should be demonstrated with 3 T MRI done under general anesthesia. 1.5 T MRI or MRI done without general anesthesia may fail to demonstrate the nerve and false diagnosis of CN hypoplasia/aplasia may be given. Importance of proper evaluation of the cochleovestibular nerve, especially its cochlear branch, is of extreme importance prior to cochlear implantation. This was discussed in the Second Consensus Meeting [6]. From the radiological point of view differentiation between aplasia–hypoplasia and a normal size of the cochlear branch can be very difficult and requires the highest possible resolution. If the IAC is narrow the demonstration of CN is even more difficult. Golden standard is to perform 3.0 T MRI under general anesthesia. Most appropriate method for evaluation of the nerves is heavily T2 weighted sequence in the axial plane and direct parasagittal images with the same heavily T2 weighted sequence made perpendicular on the nerves in the IAC and cerebellopontine angle. Parasagittal images should be done bilaterally to compare the two sides to find the side with better developed CN. Direct parasagittal images have a better resolution than reformatted images using the axial images. **If the images are of poor quality, it is vital to repeat MRI to obtain excellent quality images before any implant surgery**. In the future, if there is insufficient progress with CI, most appropriate treatment option for the contralateral side will be decided with a proper MRI and audiological findings. If MRI is not of standard quality, it is very difficult to make correct decision in the latter situation.

32.5.2 Audiology

For preoperative evaluation of ABI candidates, all audiological test battery should be applied [7]. This test battery includes both subjective and objective tests. It is apparent that in patients with complete labyrinthine aplasia and cochlear aplasia no response is expected. But, even in these patients sometimes a response is observed in low frequencies with maximum audiometric limits which can be accepted as vibrotactile sensation. Even this response may be important during programming and follow-up.

In subjective tests, the candidate should be evaluated with insert phones; if this is not possible, free field evaluation should be done. According to the age of the child, behavioral observation audiometry (BOA), visual reinforced audiometry (VRA), or play audiometry (PA) can be used.

For objective evaluation, it is appropriate to start with tympanometry and acoustic reflex tests to show middle ear status for all age groups, especially for infants and children. These tests should be followed by otoacoustic emissions (OAE) and auditory brainstem response (ABR) measurements. It is possible to obtain cochlear microphonics (CM) during ABR.

Subjective tests are very important even when no response is obtained by other tests, including the objective ones. Sometimes subjective tests are the only method which give information about hearing status of the patient. Some patients with hypoplastic CN demonstrate behavioral response with pure tone or speech stimulation. These patients are counseled that the ear with best response with insert phone will be selected for CI, and the patient will be followed up for 6–9 months with CI. At the end of this period an eABR is also done to see if there is any response with CI. If there is no development in speech perception and no response on eABR, ABI will be recommended to the family. In this situation we prefer to perform ABI in the contralateral ear, thereby providing bilateral amplification in these children. It is also very important to take into consideration the observations of the family.

32.5.3 Language Evaluation

Speech and language skills of children with hypoplastic CN should be evaluated by an experienced rehabilitative audiologist and/or speech and language therapist. This evaluation should consider the progress of child's functional auditory skills and effects on speech and language. Over the years our team observed that patients with hypoplastic CN can obtain thresholds with CI at 30–40 dB but may not develop functional auditory perception skills and had a slow rate in speech and language improvement. Although most of them were capable of basic auditory skills, such as detection and discrimination of both speech and environmental sounds, it was clearly observed that their identification and recognition skills have reached a plateau which could not provide adequate acoustic features for developing higher levels of language skills such as integrative thinking and auditory memory. Therefore, it is very significant to evaluate these children by an experienced speech and language therapist and share this finding with the team to decide for contralateral ABI.

32.6 Intracochlear Test Electrode

Promontory or round window stimulation is difficult to provide a response in cases with severe IEM. In Hacettepe University, we tested the possibility of using an intracochlear test electrode (ITE) to simulate a cochlear implant to make the intraoperative decision between CI and ABI with electrically auditory brainstem response (EABR) testing. The electrode was produced by Med El. ITE has 3 intracochlear contact points with 18 mm length and 1 extracochlear ground electrode. Intracochlear part is inserted into the cochlea up to the ring. It was used in 11 subjects with various inner ear malformations [8]. In cases with normal anatomy and IP-II, excellent wave morphology was obtained. If there was no eABR, decision for an ABI was made. There were two cases with conflicting results. One was an IP-I with definite CN on MRI. The test result was negative but CI surgery was done and CI provided very good language development after long-term follow-up. The second conflicting result was from a child with common cavity. He had benefit from CI but he developed facial stimulation which was present on all contacts. During revision procedure ITE was used but there was no response during surgery. In this particular patient ITE failed to produce eABR in a patient with common cavity who had already a good progress with a CI. As a result, it appears that, if there is a positive response, ITE is reliable and a decision for a CI can be made reliably. A negative response, however, has to be considered very carefully and radiology and preoperative audiological test methods should be used together to make the decision between CI and ABI.

In CN hypoplasia, it is important to choose the best treatment option at the beginning. As can be seen, there is no single test method giving correct guidance in CN hypoplasia. The best approach is to combine all information together and act according to radiological and audiological results and make the decision together with the team members.

In general results of CI in CN hypoplasia are not promising. Therefore, it is still debatable whether CI or ABI should be the appropriate treatment modality.

32.7 Literature Review: CI in Hypoplastic CN

There is a controversy regarding the type of implant to be used in the treatment of patients with hypoplastic CN [7]. Occasionally it is possible to obtain good hearing and language development in certain cases with hypoplastic CN. Majority of the reports indicate insufficient or no hearing and limited language development with CI [7]. These patients become candidates for ABI. It is important to correctly diagnose this subset of children and proceed with ABI directly when required; however, as indicated before, for the present time, preoperative and intraoperative audiological tests are not precise enough to enable correct diagnosis. Controversies between CI and ABI in children was published by our group before [7].

Bradley et al. [9] reported their long-term experience in six children with hypoplastic CN. Preoperatively they observed clear response to sound with hearing aids. IAC was narrow in four while two had normal width on imaging. With initial programming, all children demonstrated auditory thresholds within normal range that is obtained in children with normal anatomy. However, after using CI for 2–6 years they demonstrated unsatisfactory outcome: five were at CAP level 2 and one was at level 4. They concluded that even if they obtained thresholds similar to other CI users, the benefit of CI in children with hypoplastic CN is very limited.

Warren et al. [10] reported CI results in three cases with narrow internal auditory canals. Patients had two visible nerves on MRI; one entering the vestibule in each case, while other was assumed to be functioning FN. Preoperatively two of the families had reported responses to auditory stimuli with amplification over time. Promontory stimulation testing with electrical stimulation provided positive response in the three cases. They all underwent CI surgery. Although only early postoperative results were available (4, 5, and 9 months, respectively), they all showed responses to auditory stimuli. They provided explanation to sound transmission without any CN on MRI with two possibilities. The first explanation was that extremely narrow distal IAC made the identification of a thin CN impossible. The second possible explanation was that the nerve turned towards cochlea after entering the vestibule. Our group observed similar cases, where there is response with CI, sometimes obtaining similar thresholds to CI users with normal anatomy but progress usually reaches a level, and language development usually does not reach the level of CI in normal cochlea.

Valero et al. [11] investigated eABR responses in patients with hypoplastic CN using CI. They observed atypical amplitude and latencies in these patients which suggested to be due to nonauditory generators. They cautioned that they should not be accepted as typical EABR peaks. Long-term CI stimulation of the hypoplastic nerve did not promote normal auditory brainstem maturity and did not discourage uncharacteristic development in the brainstem. There was no relationship between the severity of the hypoplastic CN, bony cochlear nerve canal, and IAC and the type of abnormal evoked response. It was not possible to determine whether they will be good CI candidates with these structural defects. Auditory outcome could not be predicted from the observed evoked responses. They also reported similar outcome that in spite of initial limited improvement in speech perception outcomes, children with hypoplastic CN did not obtain comparable behavioral results with their CIs compared to children with normal CN. After long-term CI use this difference was still present; children with hypoplastic CN obtained scores of 24 months of those with normal CN at their 120th follow-up month. These data, along with abnormal electrophysiological findings, suggest that children with hypoplastic CN do not reach hearing levels obtained by children with uncompromised CN after CI use. This should be included in the counseling of the families to modify their expectations after CI use in terms of auditory and spoken language expectations.

Buchman et al. [12] reported CI results in patients with IEMs. Their observation is similar to other centers with poor speech perception in children with CN deficiency after CI. They concluded that this is most probably due to insufficient peripheral nerve populations in patients with CN deficiency preventing the development of synchronized auditory stimulation. In these cases, they suggested initial trial of CI before ABI. They were able to find association between intracochlear eight nerve compound action potential (ECAP) testing results and the development of speech perception abilities. However, they emphasized the requirement of further electrical stimulation technologies prior to placement of CI.

Recently Birman et al. [13] reported better outcomes of auditory performance with CI in patients with aplastic/hypoplastic CN. Pediatric CI surgery in CN aplasia/hypoplasia is associated with variable outcomes. Their study found that approximately 50% of children used sign language and 50% used verbal language as their main mode of communication. Overall, approximately 75% of children were able to use some verbal language. After CI nearly 50% of those with CN aplasia and 90% of those with CN hypoplasia gained some speech understanding (CAP score 5–7). CN aplasia/hypoplasia is commonly associated with developmental delay and syndromes, particularly CHARGE syndrome. Their findings are encouraging and useful for preoperative counseling regarding the likelihood of CI outcomes in CN aplasia/hypoplasia. However, a comment that mentions "50% of cases with CN aplasia obtains CAP scores between 5 and 7" must be taken very cautiously.

Kutz et al. [2] also reported their results after CI in children with hypoplastic CN. Seven children underwent CI in an ear without any CN on MRI. One child developed early closed-set speech recognition. The other six children developed only speech detection or pattern perception. Two children with hypoplastic nerve were also implanted. One developed consistent closed-set word recognition and the other developed early closed-set word recognition. They concluded that CN deficiency is a common cause for profound sensorineural hearing loss and children with a deficient but visible CN on MRI can expect to show some speech understanding after cochlear implantation. However, these children do not develop speech understanding to the level of implanted children with normal CN. Children with an absent CN determined by MRI can be expected to have limited sound and speech awareness after CI surgery.

Song et al. [14] reported their results of intracochlear evoked auditory brainstem response (EABR) versus extracochlear EABR in predicting long-term outcomes of patients with narrow IAC. They concluded that intracochlear EABR measured either intraoperatively or in the early postoperative period may play an important role in deciding whether to continue with auditory rehabilitation with a CI or to switch to an ABI so as not to miss the optimal timing for language development. They also compared evoked compound action potential (ECAP) and EABR measurements and found that in cases with IEMs, including narrow IAC where the number of auditory nerve fibers that can be stimulated is limited, intracochlear EABR can be more successfully recorded than can ECAP. They concluded that for these cases in which CI has been performed initially, considering the limited prognostic value of preoperative extracochlear electrophysiologic testing or imaging, intracochlear EABR measured either intraoperatively or in the early postoperative period may provide valuable prognostic information to predict long-term outcomes.

Song et al. [15] argued that promontory stimulation test may not predict the long-term outcome accurately in cases with hypoplastic CN. They correlated the diameter of IAC on HRCT with the presence of CVN during surgery. These findings suggest that the presence or absence of the CVN could not be accurately predicted by the diameter of the IAC measured on temporal bone CT, although cases with narrow IACs, measuring less than 1.5 mm, may be more frequently associated with the absence of the CVN. Despite the fact that MRI findings were often correlated with the surgical findings regarding the presence or absence of the CVN, a very thin CVN identified during ABI operation was not detectable on MRI of one patient. Promontory EABR failed to show any consistent response in any of the patients. Despite the lack of response on promontory EABR in any of these patients, a CVN was identified during ABI surgery in four patients. Although intracochlear EABR is considered to be more precise than promontory stimulation, it also bears certain problems of its own. Intracochlear EABR was shown to have limitations in precisely predicting the presence or absence of the CVN in this study, too. In particular, it was not possible to acquire any auditory response in a patient due to artifacts induced by muscle potentials resulting from stimulation of the facial nerve.

Song et al. [15] finally concluded that residual response on pure tone audiometry and behavioral response to environmental sounds appeared to be more accurate markers for predicting the presence or absence of the CVN compared to imaging or electrophysiologic testing because all three patients who showed a response to sound stimuli demonstrated thin CVNs during surgery. Our team also reached a similar conclusion, that is, audiological tests seemed to be more important in decision making between CI and ABI. However, in our series, there are a few patients who showed certain progress initially with CI, but could not carry on when more sophisticated learning processes were required.

As can be seen, children with hypoplastic cochlear nerve present a dilemma to the implant team and it is still a problematic issue to decide between CI and ABI in patients with narrow IAC and hypoplastic CN. It is still difficult to determine whether a CI will be a good solution for the hearing loss. Intracochlear eABR might be a better indicator compared to preoperative electrophysiological tests. Generally, different groups report poor outcome of CI in children with CN hypoplasia.

32.8 ABI in CN Hypoplasia

Second Consensus Meeting on ABI in Children With Complex Inner Ear Malformations [6] was organized to discuss the long-term results ABI in this group of patients. These are cases with hypoplastic/aplastic CN and pathologies like cochlear and labyrinthine aplasia. Hacettepe results showed that majority of the children with complex IEMs who had an ABI obtained CAP scores around category 5 [16]. With better thresholds (25–40 dB) it was possible to obtain CAP scores 6, 7, and 8. The speech intelligibility was, in general, poor. SIR scores were around 2 out of 5. With better thresholds, it was possible to obtain scores up to 4. Patients with common cavity obtained scores better than other type of malformations in all categories. This was most probably due to the presence of cochlear nerve fibers in the CVN. Therefore, when compared to other papers where only CI is used, it may be better to combine the treatment with bimodal stimulation using CI and ABI.

Therefore, in cases of hypoplastic or aplastic CN, ABI provides acceptable auditory performance. But in general, it is not as good as CI in normal anatomy. Therefore, our team looked into options to provide better hearing and hence language outcome in these children. More specifically we looked into options to provide bilateral stimulation in these children.

32.9 Sequential CI and ABI in Hypoplastic CN

In Hacettepe University 125 children underwent ABI surgery between July 2006 and September 2018. One hundred cases are using only ABI for hearing restoration. Twenty patients are using CI and ABI together. These are patients with hypoplastic CN, CH or IP-I cochleae, and CC. After their insufficient progress with CI for 1-year decision for a contralateral ABI was made in 14 cases. Some of these children have thresholds around 40 dB with CI, but because of the hypoplastic CN they show insufficient progress in language development. Therefore, it should be kept in mind that, even with actual CI after a year, it may be necessary to have ABI even with acceptable thresholds with CI. This also shows the difficulty of intracochlear test electrode (ITE) to determine the appropriate modality of CI vs ABI intraoperatively. Even with actual CI it takes a year to make this decision. Apart from these patients, three children are bilateral ABI users.

In Hacettepe University there are 14 patients who had ABI surgery after insufficient progress with CI. In 2014 there were six patients who used their ABI more than a year and long-term results of CI and ABI of these six patients were presented in 12th European Symposium Pediatric Cochlear Implant, in Toulouse in 2015 [17]. Average duration between CI and ABI was 1.5–2 years. This is due to the fact that it is possible to obtain acceptable thresholds after CI surgery. In the beginning, they demonstrated a progress but after a certain period the language development comes to a plateau. Therefore, there is a long period between CI and ABI surgery even though a careful follow-up is done. Their auditory performance and intelligibility scores were also presented. Although they had similar CAP scores in CI only and ABI only situations, auditory performance showed a dramatic increase when CI and ABI were used together. Average CAP score before ABI was 1, but after ABI surgery it was 4.8. Same improvement was observed in SIR scores as well. Before and after ABI average SIR scores were 1.2 and 3.3, respectively. This is now an acceptable method of treatment in these cases. Recently our paper regarding bimodal stimulation using ABI and CI for patients with inner ear malformations was submitted and it is still under review.

Therefore, it appears that the most acceptable treatment option is CI on the side with implantable cochlea, hypoplastic CN, or hearing response with insert ear phones and ABI on the contralateral side with worse anatomy and hearing. Latest

Table 32.1 CAP and SIR scores of 12 patients before and after ABI

CASE	CAP		SIR	
	Before ABI	After ABI	Before ABI	After ABI
1	2	7	2	5
2	1	6	1	4
3	1	5	1	2
4	1	5	1	2
5	0	4	1	2
6	1	2	1	2
7	2	5	1	2
8	2	6	1	4
9	1	5	1	2
10	0	5	1	2
11	1	4	1	2
12	1	5	1	2

CAP and SIR scores of 12 patients who have used their both devices for more than a year are presented in Table 32.1. Please note the increase in CAP and SIR scores after ABI.

In patients with insufficient progress with CI, our team never removed the CI and performed ABI in the same ear. At the beginning, we choose the better side for CI and we performed later ABI on the contralateral worse side to provide bilateral stimulation. We strongly believe that these cases need bilateral stimulation more than other CI candidates.

32.10 Simultaneous CI and ABI

Our team observed the important gap between CI and ABI in cases of insufficient progress in children with hypoplastic cochlear nerve. Therefore, the option of simultaneous CI and ABI was proposed in some cases. Between 2015 and 2018, six children had simultaneous CI and ABI. This is indicated in the following situations:

1. One side definite-one side possible indications: There is no need to wait for the outcome of CI on the side with possible indications and to perform ABI on the side with definite indications. To obtain best audiological outcome both procedures can be done in the same setting.
2. If the outcome of CI looks very limited with hardly visible CN on both sides and patient's age is between 2 and 3, it may be a better option to perform CI and ABI simultaneously to avoid loss of time.

This is done in situations where poor outcome with CI is expected. The CI side has a CN which is barely visible on 3 T MRI and there is a very limited response with insert earphones while the other side has a definite ABI indication. Simultaneous CI and ABI surgery has two advantages. In children who are relatively late for surgery between 2 and 3 years of age, particularly with additional disabilities, waiting for the result of CI surgery may result in a late ABI surgery. With the disability it is more difficult to decide whether the child is making progress with CI. As indicated before, it usually takes 1.5–2 years by the time the child receives contralateral ABI. Then the child becomes 3 and 3.5 years old and the benefit from ABI decreases. In order to avoid this, CI and ABI can be done in the same setting. If the child benefits from CI, then he will have bilateral stimulation at the beginning. In cases where CI is not beneficial, the child will not lose valuable time waiting for ABI. This approach was used in six patients so far in Hacettepe University and the first case was presented in CI 2015 meeting [18].

32.11 Bilateral ABI

In Hacettepe University, three bilateral ABI surgeries were performed. This situation is definitely indicated in bilateral definite indications. We know that in general, ABI results are not as good as CI in normal anatomy. Therefore, severe IEMs are true indications for bilateral stimulation. In addition, if, in the future, there is a device problem and a revision is necessary, ABI may not be easily inserted into the same correct location. ABI revision is not like CI revision. In one child with total ossification after meningitis, ABI electrode could not be removed from the brainstem. Therefore, a contralateral ABI was done. If this procedure becomes necessary at later ages, when,

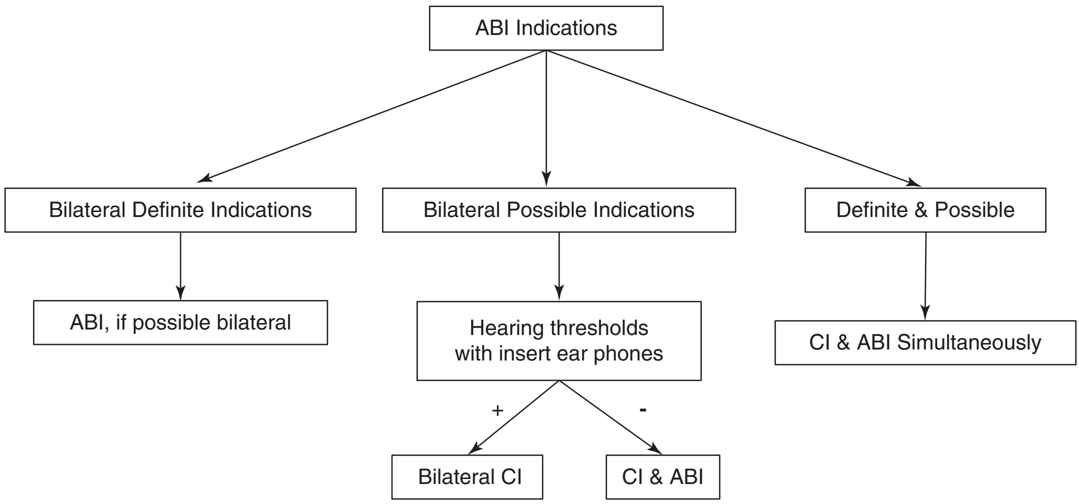

Fig. 32.1 Current management strategy in severe inner ear malformations with cochlear nerve deficiency

for example, at the age of 20, the person definitely will not have the same benefit from the side which has not been stimulated since birth. Therefore, the best option is bilateral ABI in definite indications.

32.12 Current Management Strategy

Children with CN deficiency may show different progress and get varying degrees of benefit from cochlear implantation. Children with hypoplastic nerve show adequate performance only in discrimination and/or identification of speech sounds. Although this improvement seems as a success of cochlear implantation, it is not efficient and does not convey highly qualified electrical information for other important structures that will help develop speech and auditory-oral language system. For developing an auditory-oral language system which also covers understanding and production of speech, spelling, phonological awareness, writing, reading comprehension, and auditory memory skills, they need a summation of temporal and spectral information by ABI and CI. This is also apparent for children who have aplastic CN and are unilateral ABI users. Although ABI provides speech sounds in a wide frequency range, the resolution of the signal is not fine enough to affect the intelligibility of their speech and related language skills as mentioned above. By using CI and ABI, or bilateral ABI, advantage of summation, loudness, and intensity is obviously obtained in functional speech perception and intelligibility.

Based on our experience with 259 CI, 100 ABI, 20 CI, and ABI, 3 bilateral ABIs in total of 404 cases with IEMs following algorithm are provided (Fig. 32.1):

Bilateral Definite Indications: ABI is indicated. It is advisable to make it bilateral.
Bilateral Possible Indications: Depending on the results of audiological evaluation, if there are auditory responses with insert earphones, bilateral CI should be done. If there is no response in one ear and good responses on the other with insert earphones: CI and ABI are the best management strategies. Some of these cases can be done simultaneously.
One side Definite-One side Possible Indications: Simultaneous CI and ABI.

32.13 Cases

Case 1 NO 2-year-old female, operated with CI (May 2011) then with ABI (May 2013).

She had bilateral CH-III where CN was hypoplastic on the left side and aplastic on the right side. Her audiogram with insert ear phones revealed a response on the left side (Fig. 32.2a). She had CI surgery in May 2011 on the left side. Initially she showed some progress with CI especially on basic auditory skills such as detection and pattern identification of speech sounds. Her speech and language development then came to a plateau after which point she did not demonstrate a progress on recognition and higher level of language skills at the end of 2 years. Our team decided to make an ABI on the contralateral side. In May 2013 she underwent right ABI surgery.

During the audiological follow-up, initially she was stimulated with bipolar mode in order to eliminate any possible side effects (SPEAK strategy, 250 rate and 100 pulse width). Thirteen electrodes were activated at the first activation, remaining were deactivated with the reason of inadequate auditory response and side effects. The first audiogram with ABI was given in Fig. 32.2b. After reaching the maximum limit of stimulation for ABI at the end of the fourth year, the stimulation mode was changed from bipolar to monopolar (MP1 + MP2). After monopolar stimulation, E20–E22 were deactivated due to inadequate stimulation and E3–E4 were deactivated because of the side effects such as lateral sways with the stimulation. She showed a very good progress after ABI both in functional auditory skills and language development rate. Figure 32.2c shows postoperative thresholds with CI and ABI after 5 years. Her latest auditory perception and speech and language evaluation scores, while she is almost 10 years old, are shown in Tables 32.2 and 32.3.

We performed first CI and there was some progress initially. It is possible to see some development with CI in patients with hypoplastic CN. It comes to a plateau after a follow-up period. Therefore, the team must be very careful in following these children. They can learn simple information with CI but when it comes to more complex information, it is not possible to develop clear speech. Due to the limited progress with CI, ABI was performed on the contralateral side. After ABI her speech development and auditory skills showed a great improvement. However, it took almost 2 years between CI and ABI.

Case 2 DED 2-year-old female, operated June 2016, simultaneous CI and ABI.

She was referred for an ABI surgery to our clinic. Her CT and MRI demonstrated bilateral common cavity with a well-developed CVN on the right side. On the left side there was extremely hypoplastic CVN without visible connection with the common cavity. During preoperative audiological evaluation, auditory responses were observed on the right side where there was a common cavity with a well-developed CVN (Fig. 32.3a). With these findings the family was offered the option of simultaneous CI and ABI surgery. Simultaneous CI and ABI were performed in June 2016 (CI on the right side with well-developed CVN, and ABI on the left side).

On the third day after the surgery CI was activated. ABI was activated 20 days after surgery. On the ABI side, electrode impedance values were within normal limits. Eleven electrodes were activated at the initial stimulation; others were deactivated due to side effects. First responses with ABI were at the range of 60 dB HL for speech stimulus. After a while the stimulation mode was changed to monopolar stimulation for better progress at the end of the second year. Her latest audiogram was given in Fig. 32.3b. Her audiological and communication skills were followed up by Hacettepe Audiology Team regularly. Her functional hearing was evaluated by Meaningful Auditory Integration Scale and recorded as 25/40 in the latest visit. She had the lowest scores on deriving meaning from speech sounds in both quiet and in noisy environment. However it is clearly observed that she has been developing her communication skills in auditory-verbal situations. Auditory only tasks are challenging for her especially when new acoustical information is presented. It was determined that she needs to use lip reading and acoustical ques. while she is exposed to a new concept and vocabulary. It was advised to become

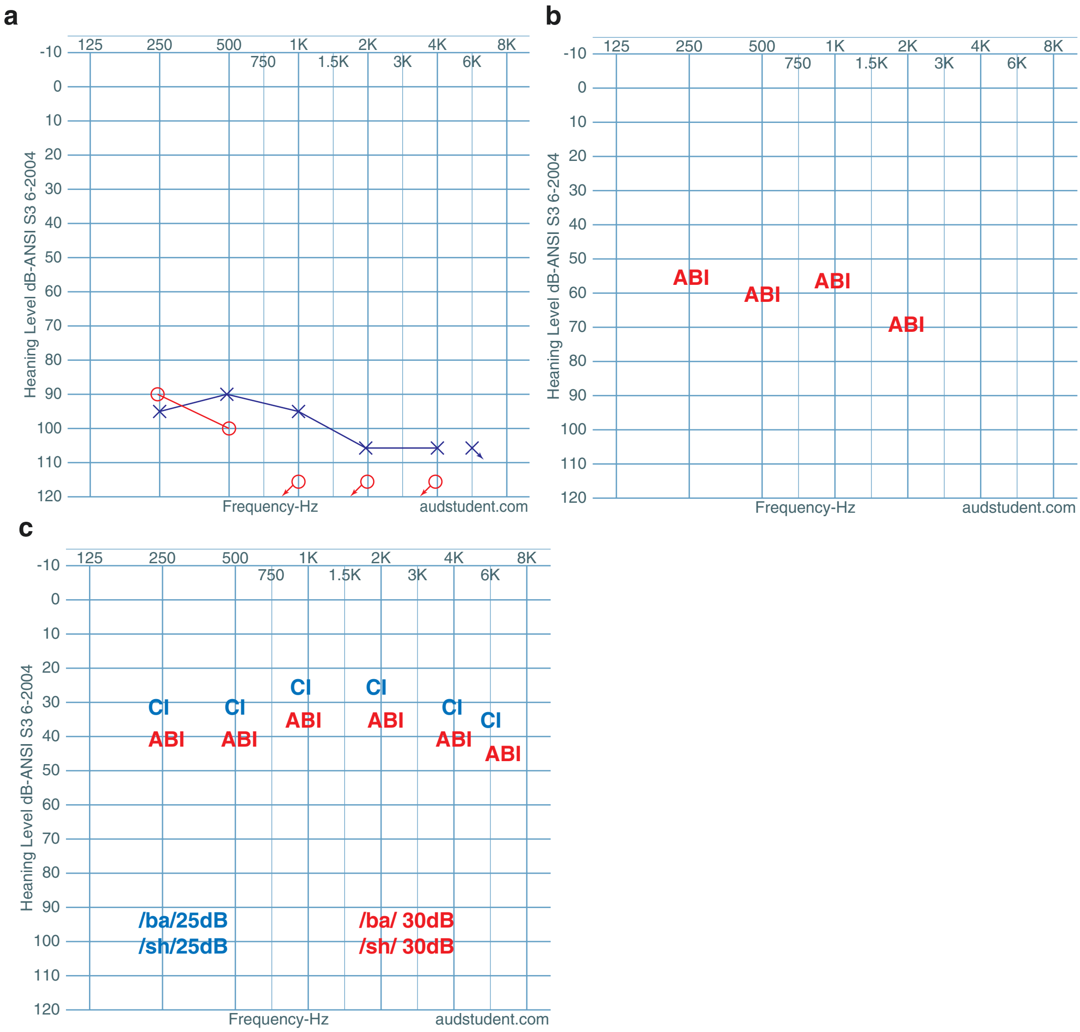

Fig. 32.2 Case 1. (**a**) Preoperative audiogram with insert ear phones showing a response on the left side, (**b**) first audiogram with ABI, (**c**) postoperative thresholds with CI and ABI after 5 years

familiar by different kinds of visual information (signing, drawings, books, games, activities in life, etc.) before listening the new concepts and vocabulary.

Although her hearing thresholds with both CI and ABI are within speech banana, and we assume that she hears most of the speech sounds it does not guarantee that she can discriminate and identify the vowels and consonants in words because of restrictions of temporal and spectral resolution. For that reason, in her educational program phonological awareness, central auditory skills and short-long term memory activites are involved. Her auditory perception and speech and language evaluation scores are shown in Tables 32.4 and 32.5.

Simultaneous CI and ABI provided the faster bilateral hearing stimulation where there is definite indication on the left side. Therefore, the time loss observed in Case 1 was avoided in this case.

Case 3 EK 1.5-year-old female operated May 2013 and September 2015 with sequential bilateral ABI.

Table 32.2 Case I: auditory perception outcomes

Tests	MAIS		Pattern perception test % (close-set)				Word identification test % (close-set)				Daily Turkish sentence recognition test % (open-set)			
Case I	CI (pre-ABI)	Bimodal CI + ABI	CI (pre-ABI)	CI (post-ABI)	ABI	Bimodal CI + ABI	CI (pre-ABI)	CI (post-ABI)	ABI	Bimodal CI + ABI	CI (pre-ABI)	CI (post-ABI)	ABI	Bimodal CI + ABI
	16/40	40/40	20	100	100	100	0	100	100	100	0	60	80	100

Table 32.3 Case I: CAP, SIR, and language outcomes

Tests	CAP		SIR		Expressive language (months)		Receptive language (months)	
Case I	CI	ABI	CI	ABI	CI	ABI	CI	ABI
	2	7	2	5	18	108	18	108

She had bilateral cochlear aplasia with dilated vestibule with no response during audiological evaluation (Fig. 32.4a). Left side was operated in May 2013 with an ABI. Left ABI was activated 3 weeks after the surgery and 4 electrodes were deactivated because of local voltage errors, high

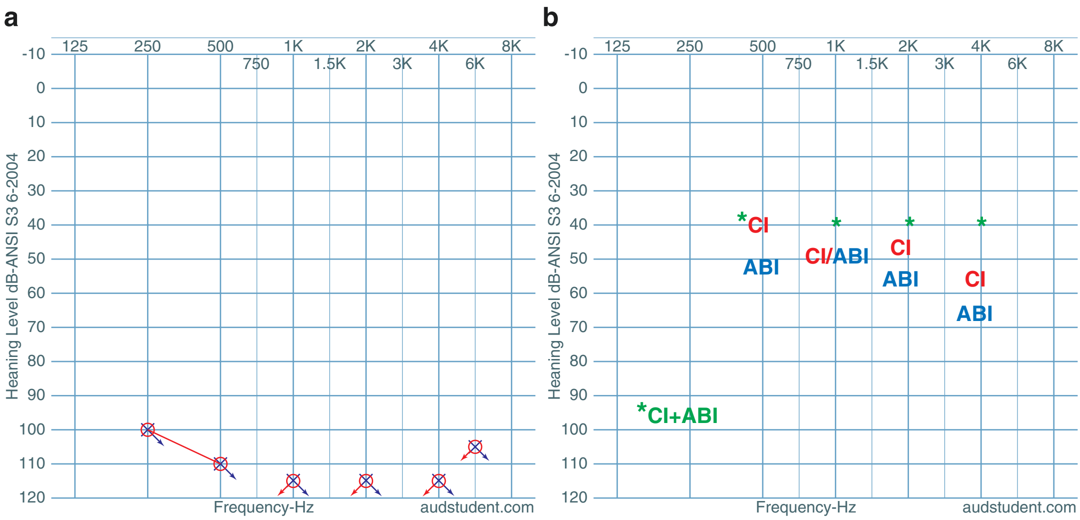

Fig. 32.3 Case 2. (**a**) Preoperative audiogram showing auditory responses on the right side where there was a common cavity with a well-developed cochleovestibular nerve, (**b**) Postoperative audiogram after 2 years

Table 32.4 Case II: auditory perception outcomes

Tests	MAIS	Pattern perception test % (close-set)			Word identification test % (close-set)			Daily Turkish sentence recognition test % (open-set)		
Case II	Simultaneous CI + ABI	CI	ABI	Simultaneous CI + ABI	CI	ABI	Simultaneous CI + ABI	CI	ABI	Simultaneous CI + ABI
	25/40	25	41,6	50	16,6	29,1	41,6	10	10	10

Table 32.5 Case II: CAP, SIR, and language outcomes

Tests	CAP	SIR	Expressive language (months)	Receptive language (months)
Case II	Simultaneous CI + ABI	Simultaneous CI + ABI	Simultaneous CI + ABI	Simultaneous CI + ABI
	4	3	24	24

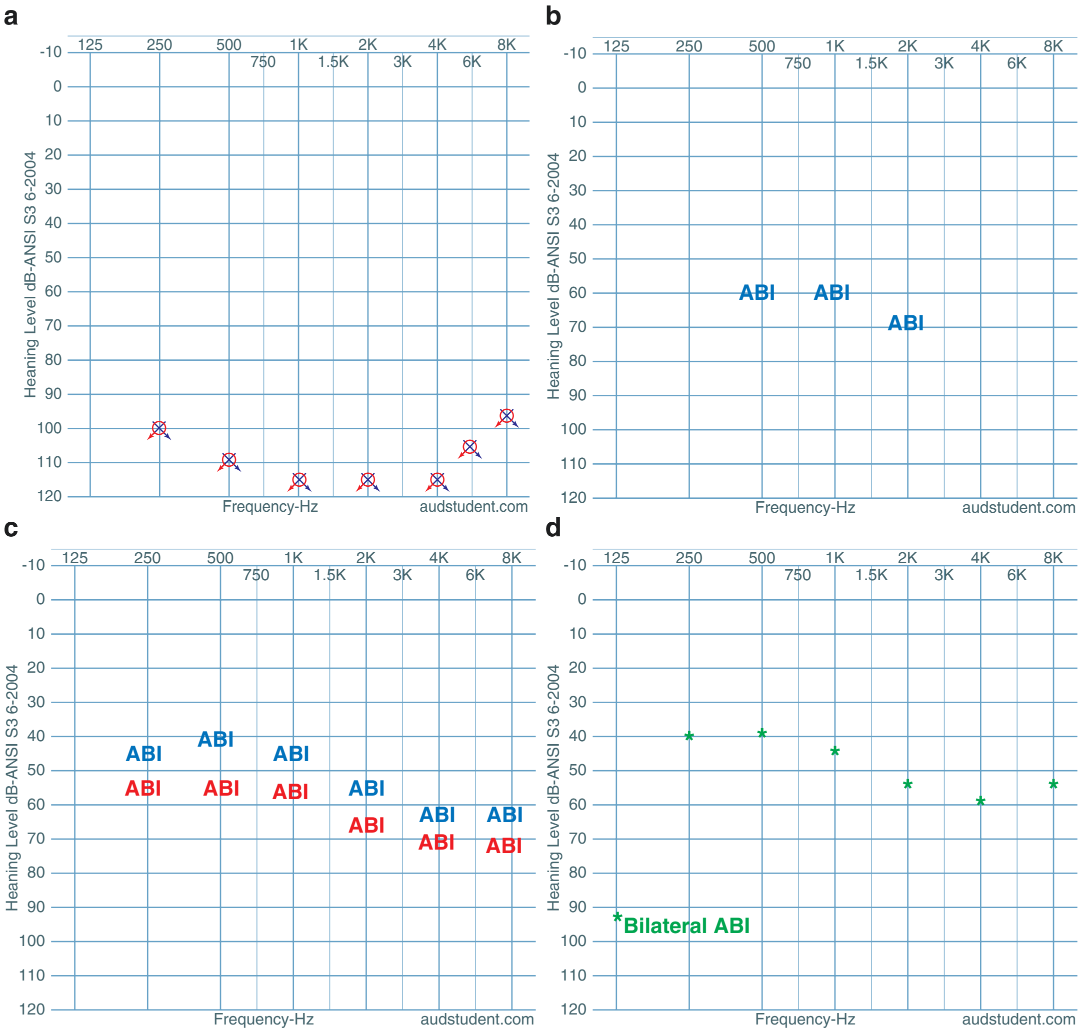

Fig. 32.4 Case 3. (**a**) Preoperative audiogram showing no auditory response, (**b**) auditory responses with left ABI, (**c**) thresholds with both ABIs, (**d**) better auditory response in the bilateral condition

impedance, and side effect (vestibular effect) At the initial stimulation of the first ABI, she had good responses to the speech stimulus at the level of 70 dB HL. During the follow-up, her auditory responses with left ABI showed improvement (Fig. 32.4b). At the end of 2 years with unilateral ABI experience, she consistently developed functional auditory skills in all closed-set informal tasks only in daily situations. Her speech intelligibility was barely improved. Her MAIS score with first ABI was 26/40 after 2 years.

After seeing her good progress with the ABI, her right ear was also operated with ABI in September 2015. On the right ABI side, all electrodes were activated without any side effects. After 1 year of bilateral ABI experience, she reached full score in pattern perception (24/24), developed word identification (6/12), and sentence recognition (5/10). Binaural MAIS was 36/40 at the end of 1 year. After second ABI, her speech intelligibility and listening behavior were improved and spontaneous word learning and combining (3–4 words sentences) was started.

Table 32.6 Case III: auditory perception outcomes

Tests	MAIS		Pattern perception test % (close-set)		Word identification test % (close-set)		Daily Turkish sentence recognition test % (open-set)	
Case III	ABI	Bilateral ABI	ABI	Bilateral ABI	ABI	Bilateral ABI	ABI	Bilateral ABI
	26/40	36/40	NA[a]	100	NA[a]	50	NA[a]	50

[a]*NA* not available

Table 32.7 Case III: CAP, SIR, and language outcomes

Tests	CAP		SIR		Expressive language (months)		Receptive language (months)	
Case III	ABI	Bilateral ABI	ABI	Bilateral ABI	ABI	Bilateral ABI	ABI	Bilateral ABI
	3	5	2	4	12	54	18	60

CAP score was 3 and SIR score was 2 with one ABI, and they were improved to 5 and 4 after second ABI. She was not ready to carry out the formal speech perception tests by her first ABI.

Thresholds with right ABI and left ABI were given in Fig. 32.4c. Her auditory responses showed better in the bilateral condition as we expected (Fig. 32.4d). After 3 years of bilateral ABI experience her receptive and expressive language scores have reached, respectively, from 18 to 60 months and from 12 to 54 months (Tables 32.6 and 32.7).

As there are bilateral definite indications on both sides this approach is the most appropriate management option in these cases. These are the cases who need bilateral stimulation more than any other CI indication and bilateral ABI is their only surgical option.

32.14 Conclusion

With today's technology it is very difficult to predict the best treatment modality in patients with hypoplastic CN. Radiological and audiological evaluation methods are not yet precise enough to let the clinician decide between CI and ABI. CI surgery is usually not promising resulting in poor outcome with CAP scores less than 5. ABI appears to provide better outcome. When possible these candidates should be given the option of CI first and then ABI. This has always provided better auditory performance and speech intelligibility. A minority of patients with late age (around 2–3) may be candidates of simultaneous CI and AB.

References

1. Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers T, D'Hont G. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. Radiology. 1997;202(3):773–81.
2. Kutz JW Jr, Lee KH, Isaacson B, Booth TN, Sweeney MH, Roland PS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotol. 2011;32(6):956–61.
3. Morita T, Naito Y, Tsuji J, Nakamura T, Yamaguchi S, Ito J. Relationship between cochlear implant outcome and the diameter of the cochlear nerve depicted on MRI. Acta Otolaryngol. 2004;124(sup551):56–9.
4. Jaryszak EM, Patel NA, Camp M, Mancuso AA, Antonelli PJ. Cochlear nerve diameter in normal hearing ears using high-resolution magnetic resonance imaging. Laryngoscope. 2009;119(10):2042–5.
5. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, Ramsden R, Sarac S, Freeman S, Andersen HR. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
6. Sennaroğlu L, Colletti V, Lenarz T, Manrique M, Laszig R, Rask-Andersen H, Göksu N, Offeciers E, Saeed S, Behr R. Consensus statement: long-term results of ABI in children with complex inner ear malformations and decision making between CI and ABI. Cochlear Implants Int. 2016;17(4):163–71.
7. Sennaroglu L, Sennaroglu G, Atay G. Auditory brainstem implantation in children. Curr Otorhinolaryngol Rep. 2013;1(2):80–91.
8. Cinar BC, Yarali M, Atay G, Bajin MD, Sennaroglu G, Sennaroglu L. The role of eABR with intracochlear test electrode in decision making between

cochlear and brainstem implants: preliminary results. Eur Arch Otorhinolaryngol. 2017;274(9):3315–26.
9. Bradley J, Beale T, Graham J, Bell M. Variable long-term outcomes from cochlear implantation in children with hypoplastic auditory nerves. Cochlear Implants Int. 2008;9(1):34–60.
10. Warren FM III, Wiggins RH III, Pitt C, Harnsberger HR, Shelton C. Apparent cochlear nerve aplasia: to implant or not to implant? Otol Neurotol. 2010;31(7):1088–94.
11. Valero J, Blaser S, Papsin BC, James AL, Gordon KA. Electrophysiologic and behavioral outcomes of cochlear implantation in children with auditory nerve hypoplasia. Ear Hear. 2012;33(1):3–18.
12. Buchman CA, Teagle HF, Roush PA, Park LR, Hatch D, Woodard J, Zdanski C, Adunka OF. Cochlear implantation in children with labyrinthine anomalies and cochlear nerve deficiency: implications for auditory brainstem implantation. Laryngoscope. 2011;121(9):1979–88.
13. Birman CS, Powell HR, Gibson WP, Elliott EJ. Cochlear implant outcomes in cochlea nerve aplasia and hypoplasia. Otol Neurotol. 2016;37(5):438–45.
14. Song MH, Bae MR, Kim HN, Lee WS, Yang WS, Choi JY. Value of intracochlear electrically evoked auditory brainstem response after cochlear implantation in patients with narrow internal auditory canal. Laryngoscope. 2010;120(8):1625–31.
15. Song MH, Kim SC, Kim J, Chang JW, Lee WS, Choi JY. The cochleovestibular nerve identified during auditory brainstem implantation in patients with narrow internal auditory canals: can preoperative evaluation predict cochleovestibular nerve deficiency? Laryngoscope. 2011;121(8):1773–9.
16. Sennaroglu L, Sennaroglu G, Yücel E, Bilginer B, Atay G, Bajin MD, Mocan BÖ, Yaral M, Aslan F, Çınar BÇ, Batuk MÖ. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.
17. Sennaroglu G, Atay G, Bajin MD, Batuk M, Cicek-Cinar B, Ozkan B, Sennaroglu L, Yaralı M, Yucel E. Bimodal stimulation: one side cochlear implant and contralateral auditory brainstem implant. In: 12th european symposium pediatric cochlear implant, toulouse, France. 2015.
18. Sennaroglu L. Simultaneous CI & ABI. Paper presented at the American Cochlear Implant Alliance CI 2015 Symposium, Washington D.C. USA. 15–17 October 2015; 2015.