# 18 Auditory Brainstem Implantation in Children with Inner Ear Malformations

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## 18.1 Introduction

First auditory brainstem implantation (ABI) was performed in 1979 in House Ear Institute (HEI) in Los Angeles, by Drs. William House and William Hitselberger after removal of an acoustic neuroma [1]. Initial ABI electrode consisted of a simple ball-type electrode which was placed into the lateral recess of the fourth ventricle over the area of the cochlear nuclei. Fayad et al. and Otto et al. [2, 3] reported that the first 25 patients implanted with the ABI prior to 1992 at HEI received a single-channel system. This was replaced by multichannel implant in 1992, which has resulted in improved performance. First multichannel ABI in Europe was performed in 1992 by Drs. Roland Laszig and Peter Sollmann [4]. In 2000 FDA approved the nucleus multichannel ABI device for implantation [1]. For the first two decades, main indication for ABI was $NF_2$ patients.

In 2001 Colletti et al. [5] reported for the first time in literature their ABI experience in two children with severe inner ear malformations and no apparent cochlear nerve. The first patient was a 4-year old child with bilateral common cavity and a narrow internal auditory canal with bilaterally absent cochleovestibular nerve. Until that time, cochlear implant (CI) surgery was contraindicated in these patients and no appropriate rehabilitation was possible. This marked the beginning of a new era where ABI surgery was started to be used in prelingually deafened children with severe inner ear malformations such as cochlear and labyrinthine aplasia or aplastic cochlear nerves. In 2006 Hacettepe University started to use ABI in prelingually deaf children with severe inner ear malformations. After a period of time, other centers also started to use ABI for habilitation of hearing loss in these children.

This chapter will focus on ABI use in prelingually deafened children with severe inner ear malformations. $NF_2$ or meningitis although occasionally may be mentioned, but they are not within the scope of this chapter.

## 18.2 Indications

ABI can be used in children with severe malformations and complete ossification of cochlea after meningitis. Inner ear malformations constitute the main group. ABI is not required in all cochleovestibular malformations. Patients with

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incomplete partition type II and III and enlarged vestibular aqueduct always have a cochlea with certain deformities and a cochlear nerve development and therefore, they can be rehabilitated with CI. In the first Consensus paper, Sennaroglu et al. [6] divided the indications into two groups. Most recently updated pediatric ABI indications can be summarized as follows [7] (please refer to Chap. 1 for detailed description of inner ear malformations and cochlear nerve deficiency):

### 18.2.1 Definite Indications

1. **Complete labyrinthine aplasia (Michel aplasia)**
   The cochlea, vestibule, vestibular aqueduct, and cochlear aqueduct are absent.
2. **Rudimentary otocyst**
   Millimetric otic capsule remnant without internal auditory canal
3. **Cochlear aplasia**
   This is absence of the cochlea. The accompanying vestibular system may be normal or there may be an enlarged vestibule.
4. **Cochlear nerve aplasia**
   This is the absence of the cochlear nerve.
5. **Cochlear aperture aplasia**
   This is the absence of the bony channel transmitting the cochlear nerve between IAC and cochlea.

### 18.2.2 Probable Indications

1. **Hypoplastic cochlea with hypoplastic cochlear aperture with deficient cochlear nerve:** Hypoplastic cochleae may have different audiological presentations. Some patients may be aided with hearing aids and they may have excellent speech and language development. If they are accompanied by hypoplastic cochlear aperture on temporal computed tomography (CT), usually cochlear nerve is hypoplastic or absent and they commonly have severe to profound hearing loss. In the latter group, the cochlear nerve entering the cochlea is hypoplastic and it may be difficult to determine accurately the functional capacity of the cochlear nerve with the present audiological tests.
2. **Common cavity and incomplete partition type I cases where cochleovestibular (CVN) and cochlear nerves (CN) are apparently missing.** In common cavity the nerve entering the cavity is termed as cochleovestibular nerve (CVN). If the CVN and CN are present in common cavity and IP-I anomalies, respectively, they are candidates for cochlear implantation. However, in situations where they are absent, ABI is the only habilitation option. It is important to note that common cavity can be easily confused with cochlear aplasia and vestibular dilatation [8]. The results of CI in cochlear aplasia and vestibular dilatation are not successful and this should be avoided.
3. **Common cavity and incomplete partition type I cases if the CVN and CN are present:** Even if the nerve is present, the distribution of the neural tissue in the abnormal cavity or cochlea is unpredictable, and ABI may be indicated in such cases if CI fails to elicit an auditory sensation.
4. **The presence of a hypoplastic CVN** is a challenge in these cases. In this situation, it is not possible to determine the amount of cochlear fibers traveling in the CVN. If there is a suspicion, a cochlear implant can be used in the first instance, and ABI can be reserved for the patients in whom there is insufficient progress with CI.
5. **The hypoplastic CN** presents a dilemma for the implant team. A hypoplastic nerve is defined as less than 50% of the usual size of the cochlear nerve or less than the diameter of the facial nerve [7, 9]. Radiology of these patients should be carefully reviewed with an experienced neuroradiologist. If sufficient amount of neural tissue cannot be followed into the cochlear space, an ABI may be indicated. In these cases, final decision is always made according to audiological findings.

Children with hypoplastic nerves or thin unbranched CVN constitute the most controversial group in decision making between CI and ABI. It must be kept in mind that children with hypoplastic nerves usually do not reach levels of those with normal cochlea and cochlear nerve, in terms of

hearing and language development. It is obvious that radiology may not predict the presence of the cochlear nerve accurately in the above mentioned five groups of challenging patients. In all these subjects audiological findings, as well as radiological findings, should be used together in order to decide between CI and ABI. If an experienced pediatric audiologist detects a slight response on either side of these cases with insert earphones, this information is very valuable in the side selection for CI. In such cases, family should be carefully counseled about the possibility of ABI surgery if insufficient progress with CI is encountered during postoperative follow-up (please refer to Chap. 32 for the decision making between CI and ABI in patients with cochlear nerve deficiency).

Depending on type of IEM, ABI can be performed unilaterally, bilaterally, or contralateral to CI side. Bilateral ABI is done consecutively but CI and ABI surgery can be done consecutively or simultaneously.

## 18.3 ABI Models

There are three ABI brands currently available for surgery. In Hacettepe University our team uses all three brands. All three brands are reported to be compatible with MRI at field strengths of 0.2, 1.0, and 1.5 T with a bandage over the implant area. **We strongly suggest to refer to individual implant manufacturer's manual in case of an MRI use in a patient with ABI**.

1. **Cochlear Company:** The currently used ABI electrode of the Cochlear Company (Nucleus ABI541) has an array with 21 electrodes that are embedded in a silicone carrier and connected to an implantable internal receiver/stimulator (Fig. 18.1a, b). The flexible silicone plate measures 3 × 8.5 mm, with individual electrodes 0.7 mm in diameter. It has a T-shaped Teflon mesh to keep the electrode in the lateral recess.
2. **Med-El Company**: Med-El company developed the ABI electrode from the Combi 40 cochlear implant (Med-El Company, Innsbruck, Austria). Current Med-El ABI is based on Synchrony implants where the magnet is removable. The receiver/stimulator has an array with 12 platinum electrodes with a diameter of 0.6 mm On the reverse side of the silicone carrier is a Dacron mesh that facilitates fixation in the lateral recess. There is one reference electrode. Intraoperative EABR measurements and assessment of the desired position of the active electrode can be done by a placing electrode which has four active con-

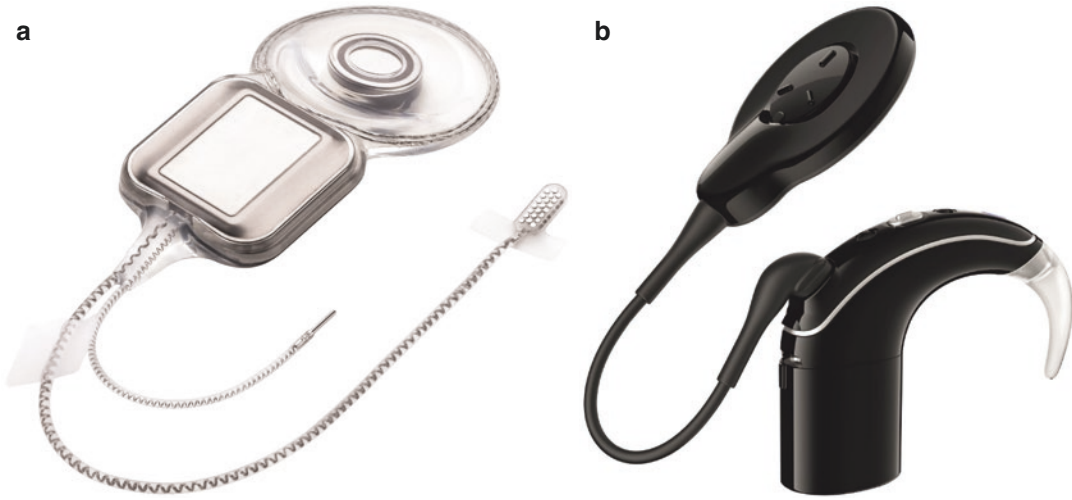


**Fig. 18.1** (**a**) Auditory brainstem implant Nucleus ABI 541 (Used with permission of the Cochlear Company), has an array with 21 electrodes that are embedded in a silicone carrier and connected to an implantable internal receiver/stimulator (**b**) Processor CP1000 working with the ABI541

tacts or the actual implant itself. The placing electrode is used only for the intraoperative testing. When the tests are finished and the correct location of the electrode is determined, placing electrode is replaced by the actual ABI implant. The actual ABI has a platinum band on the implant body. This is used during intraoperative tests and it has to be removed after the tests.

3. **The Digisonic SP ABI**: ABI of Oticon Medical Company comes with an array of 15 surface electrodes. As with other Oticon implants, Digisonic SP ABI implant body does not need an implant bed. It is fixed to the skull with two screws.

In children the size of the lateral recess is smaller than adults. In our university Teflon mesh is cut and reduced in size before insertion. The mesh is more useful in adults where the recess is larger and migration is more probable. In children our team has not encountered any migration of the electrode out of the recess in 128 patients operated so far. Electrode paddle is secured with 3–4 muscle pieces (2–3 mm in size) which are placed in the recess behind the electrode paddle pushing toward the cochlear nuclei in front.

## 18.4 Members of the ABI Team

ABI surgery is a technically demanding operation. The team has to be experienced in the surgery, audiological follow-up, and rehabilitation of cochlear implant patients. In addition, an experienced pediatric neurosurgeon is indispensable to achieve success and to avoid possible complications as much as possible. If the surgery leads to cranial nerve damage and/or brainstem injury which brings forth neurological sequels in otherwise healthy children, this would be a catastrophe both for family and the team. Besides, this might create negative impact on public opinion regarding ABI surgery. It is very important to avoid any possible complications in these children by working with an appropriate team. Placing the implant in the brainstem involves the close collaboration of an experienced pediatric neurosurgeon and pediatric anesthesiologist together with the neurootologist who is experienced in implant surgery. Occasionally, location of foramen Luschka leading to lateral recess is not apparent and careful dissection is necessary to identify the exact location. Our team experienced many situations where the foramen of Luschka was closed with mucosal folds or fibrotic tissue. In these situations, it would be impossible to identify the exact location by an inexperienced surgeon which involved opening the covering tissue to identify the foramen underneath. This is one of the most important issues to prevent malposition of the electrode which may lead to unsuccessful results and an experienced pediatric neurosurgeon is the key to avoid this complication.

## 18.5 Age Limit for ABI in Children

According to the consensus statement, age limit for ABI in children is similar to CI patients [6]. Better language outcome is expected when children are operated between 1 and 2 years of age. ABI surgery is more challenging than CI surgery because young children have less blood volume and cerebrospinal fluid in the posterior fossa. From the neurosurgical point of view, in the consensus paper optimum lower limit was determined as 18 months but, depending on the experience of the center, it was also suggested that it may be done as early as 12 months old. It is without doubt that earlier intervention will have better audiological outcome. In Hacettepe University 12 of the 128 pediatric cases were operated at the age of 12 months. Although the surgical risks may be less when the child is operated at a later age, language outcome will not be satisfactory because of the brain plasticity. Operating children with older age, however, carries the risk of discrediting the surgery, as it will be thought that this intervention will not produce good hearing and language outcome. Therefore, ideal age appears to be between 1

and 2 years of age, and with experience of the team, it has to be lowered to around age 1. As these are prelingually deafened children, this procedure should not be offered to patients older than 5 years old.

## 18.6 Preoperative Evaluation

All members of the team have to evaluate ABI candidates in detail.

Radiological workup involves CT and MRI of the temporal bone. Classification of the malformation can be done with temporal CT and MRI. Diagnosis and indication for ABI are straightforward with CT in cases such as Michel deformity and cochlear aplasia. Children with cochlear hypoplasia, hypoplastic cochlear aperture, and narrow IAC need more careful audiological and radiological evaluation with MRI. MRI demonstrates the neural structures in the IAC. As mentioned in the second consensus meeting in detail, MRI of the IAC should be direct parasagittal imaging with 3.0 T rather than reformats [10]. Any vascular abnormality around the lateral recess can be seen on MRI. If a bimodal stimulation is planned with CI on one side and ABI on the contralateral side, better audiological side should be chosen for CI. For unilateral ABI, side with more developed inner ear or the cochleovestibular nerve should be preferred. As stated in the preceding paragraphs MRI has limitations in the diagnosis.

Side selection is very important in ABI surgery. The team should try to choose the side where more information can be provided to the cochlear nucleus. Therefore, side with more developed neural structures (e.g., facial nerve presenting unilaterally, or more prominent CVN or vestibular nerve) may imply better developed cochlear nucleus area. If equal under all conditions, more developed inner ear is preferred (if there is a cochlear aplasia on one side and a hypoplastic cochlea on the other side, the latter can be preferred). In addition, side where the entrance of the lateral recess is more favorable and the lateral recess is more accessible (where cerebellar retraction will be less) can be chosen.

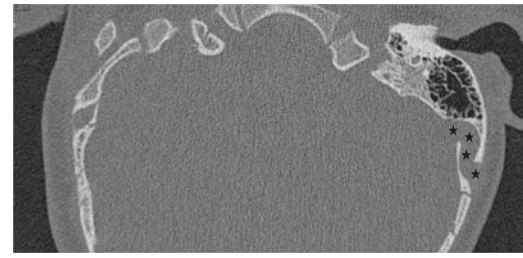

**Fig. 18.2** Temporal CT showing a huge emissary vein (black stars) at the craniotomy area

Certain situations may be a contraindication to retrosigmoid approach. An example is huge emissary vein (Fig. 18.2) coming from intracranial space and located in the area of retrosigmoid craniotomy. In two such cases our team was not able to perform retrosigmoid approach and retrolabyrinthine approach was used to place the ABI.

## 18.7 Anatomy of the Cochlear Nuclei

Anatomy of the brainstem relevant to ABI surgery is complex discussed in details before [11].

(a) **Anatomy of the Cochlear Nuclei**

The target for placement of the ABI electrode array is the cochlear nucleus complex, consisting of dorsal and ventral cochlear nuclei [11]. Colletti et al. [12] indicated that the cochlear nucleus complex in humans is located on the dorsal surface of the brainstem, immediately rostral to the pontomedullary junction. It consists of three subnuclei: the dorsal cochlear nucleus (DCN), the inferior ventral cochlear nucleus (IVCN), and the superior ventral cochlear nucleus. The DCN and IVCN have exposed surfaces in the floor of the lateral recess of the fourth ventricle, whereas the superior ventral cochlear nucleus is located deep in relation to the middle cerebellar peduncle and is not directly accessible when a conservative approach is used. The surfaces of the DCN and IVCN, which are contiguous to each other, measure on average 3 × 8 mm.

(b) **Anatomy of the Foramen of Luschka**

The foramen of Luschka is the lateral termination of the fourth ventricle and is found between the roots of the cochleovestibular nerve and glossopharyngeal nerves. The choroid plexus, which covers the foramen of Luschka, lies within a triangle formed by the eighth nerve, the ninth nerve, and the lip of the foramen of Luschka [13]. Klose and Sollmann [14] dissected 100 specimens under surgical conditions and found that the exits of the nerves VII, VIII, and IX formed a triangle of about 5 × 6 mm. The taenia of the choroid plexus was present in 92% and had to be cut in 51% in order to enter the foramen of Luschka. The foramen of Luschka has a mean size of 3.5 × 2.0 mm. It was wide open in 24%, open only after incision of the arachnoid in 53%, functionally closed but opened by extensive dissection in 18%, and anatomically occluded in 5% of the specimens. In addition, they identified the presence of a typical straight vein at the cochlear nucleus leading to the entrance of the foramen of Luschka in 76% of specimens. Our team experienced similar findings; in majority of the pediatric ABI cases foramen of Luschka was open. In less than 10% of cases it was completely closed by mucosal folds and opening these folds made it possible to identify foramen of Luschka. Ninth cranial nerve was the most important landmark in these cases as the choroid was not initially visible.

(c) **Stimulation of the Cochlear Nucleus by ABI**

Abe and Rhoton [15] pointed out that it is still controversial whether the dorsal or ventral nucleus should be the site of the implantation. Both nuclei have advantages and disadvantages in terms of placement and stimulation via ABI. The dorsal cochlear nucleus (DCN) has the advantage that it is located more medially and this makes it less likely to be damaged by the pressure of a tumor in the cerebellopontine angle or by the operative removal of an acoustic neuroma. In addition, the DCN is easier to identify than the ventral cochlear nucleus (VCN) because it underlies a smooth prominence, the auditory tubercle, in the lateral recess.

Many authors consider the DCN as the preferred target for implantation [12, 15–17]. Brackmann et al. [17] recommended electrode placement entirely within the lateral recess, where it would stimulate the DCN and the intraventricular part of the ventral cochlear nucleus. This position results in optimal auditory stimulation and the least stimulation of adjacent structures, including cranial nerves V, VII, and IX, or the overlying flocculus of the cerebellum. Also, placement completely within the lateral recess provides better stabilization of the electrode minimizing the chances of migration.

Toh and Luxford [1] indicated that the VCN is the main relay nucleus for nerve VIII input, and its axons form most of the ascending pathway. Abe and Rhoton [15] described VCN having a somewhat irregular shape, sitting at the junction of the cerebellopontine angle cistern and foramen of Luschka, and often having the taenia of the rhomboid lip crossing its surface, making it difficult to find a stable position for the stimulating electrode. According to Laszig et al. [18] the VCN might have advantages over the DCN. First, the VCN has a greater input of primary auditory neurons than the DCN. Second, the VCN has fewer inhibitory circuits than the DCN and, finally, projects more strongly onto the inferior colliculus. According to Abe and Rhoton [15] because of the close proximity of the ventral nucleus to other cranial nerves and tracts, ABI may cause nonauditory side effects during stimulation. It also extends deeper into the brainstem than the dorsal nucleus and full activation of ventral nucleus may cause greater stimulation of adjacent areas (such as the activation of the facial, glossopharyngeal, vagus, or accessory nerves, vestibular nuclei, brainstem tracts, inferior cerebellar peduncle, and the flocculus).

Terr et al. [19] stressed the importance to avoid the extraventricular part of the VCN for the implant to avoid the side effects. One advantage of including the intraventricular part of the VCN is that it is a richer source of efferent connections to higher centers than the DCN.

## 18.8 Cranial Nerve Monitorization

Neural integrity of the seventh and ninth cranial nerves is monitored constantly with electromyography throughout the procedure [20]. Monitorization is more important in ABI surgery done in NF2 cases, where there is tumor removal in addition to implantation. Main target is protection of the lower cranial nerves. Facial nerve is slightly superior and deeper when compared to glossopharyngeal nerve. Foramen is closer to glossopharyngeal nerve root entry point rather than FN. We typically do not work around the FN entry point. Therefore, less dissection is done for FN. FN may be affected from traction if cerebellum is retracted too much for exposure during dissection around ninth nerve.

## 18.9 Surgery

ABI surgery can be performed through translabyrinthine, retrosigmoid, or retrolabyrinthine approaches [11]. In children main approach for auditory brainstem implantation (ABI) has been retrosigmoid approach. Temporal bone is much smaller in a child of 2–3 years of age when compared to an adult. As a result, translabyrinthine approach will provide much limited surgical exposure than retrosigmoid approach in a child. In addition, drilling of the temporal bone takes more time to expose the brainstem in comparison to retrosigmoid approach. Therefore, for the placement of ABI in a child, retrosigmoid approach appears to be advantageous. In addition, retrosigmoid approach makes it possible to bypass the mastoid air cells so that intracranial contamination by the middle ear flora can be prevented.

However, translabyrinthine approach has been utilized for ABI in a child by Helge Rask Andersen and his team (not published, personal communication), and the electrode was successfully placed into the recess.

Bento et al. [21] described the extended retrolabyrinthine approach (RLA) for ABI placement which was performed consecutively in three children without any further complications. They stressed the importance of radiological examination both in evaluation of the etiology and to choose the side to be operated on for RLA based on the size of the jugular bulb. They advised that side with less prominent jugular bulb should be chosen. They stated that approach is more familiar to the otologist. After a postauricular incision and mastoidectomy, they identified jugular bulb as the main landmark for access to the dura. It was exposed by removing bone from its entire circumference. Only the intracranial portions of the seventh and eighth cranial nerves were exposed. Then cerebellar flocculus and lower cranial nerves were identified. After retracting the choroid plexus, they identified foramen of Luschka and placed the ABI electrode. RLA was chosen due to their extensive experience in using this technique for vestibular schwannoma surgery in patients with useful hearing. RLA allowed direct visualization of the foramen of Luschka through a limited approach. There was no requirement for cerebellar retraction or even for opening the internal auditory meatus and semicircular canals. The disadvantage of this approach in children is that it cannot be used in a very young child with an extremely large jugular bulb.

As a result, all three approaches can be used in ABI surgery of children but retrosigmoid approach is still being the most widely used technique when compared to the other two methods. With any preferred method, it should be noticed that distorted anatomy at the cerebellopontine angle, at the cranial nerve entry zones, and brainstem due to absence of the cochleovestibular nerve makes surgery more difficult at certain cases [1].

**Surgical approaches in pediatric ABI surgery** [11]:

### 18.9.1 Retrosigmoid Approach

This approach is preferred by neurosurgeons and some neurotologists. Main advantage of retrosigmoid approach (RS) is the duration of surgery. As the craniotomy step is more rapid, it is more preferable to translabyrinthine or retrolabyrinthine approach. This approach makes it possible to pre-

serve inner ear structures. In addition, mastoid air cells are bypassed in RS approach and this prevents intracranial contamination with the middle ear flora. Children frequently have otitis media and it is more important to bypass mastoid in this age group where the surgery is done around age of 1. Watertight closure of the dura avoids the need to seal the temporal bone cavity with abdominal fat. In this route facial and cochlear nerves are identified at their entry zone and at the distal end in the internal auditory canal. As a result, in children with severe inner ear anomalies RS approach is the preferred route.

There are two different positions used for this approach: lateral oblique and semi-sitting positions. In children with severe inner ear malformations, lateral oblique position is preferred. In this position the patient's neck is slightly flexed and the ipsilateral shoulder of the patient is taped down and forward. In adults with NF2, Behr et al. [22] preferred the semi-sitting position with the head inclined and turned 30° toward the side of the tumor and then fixed in a Mayfield clamp. They used a question mark-shaped retroauricular skin incision.

Behr et al. [22] indicated that sometimes blood or CSF may interfere with safe placement of the device; this may cause damage to the caudal cranial nerves by suction or manipulation. According to their experience semi-sitting position provides easier removal of blood and CSF from the surgical field; this aids in fixation of the array by fibrin glue in almost dry surroundings.

In Hacettepe University, in nontumor cases, ABI has been placed via RS approach while the patient is in lateral oblique position (Video 18.1). A straight vertical skin incision about 7–8 cm in length is performed behind the ear, incision extends from 1 cm above asterion to a point inferior and posterior to the mastoid tip. A RS craniotomy is performed where the superior and anterior limits are transverse and sigmoid sinuses, respectively. In order to enable less cerebellar retraction, bone removal is slightly enlarged inferior toward the jugular foramen. It is important to make the implant bed before opening the dura to avoid bone dust entering the intracranial space. The implant bed is positioned vertically above the surgical field as far away from the incision as possible. One suture hole is drilled inferior to the implant bed to fix the device. If a Digisonic SP ABI is used, no implant bed is prepared but the implant is positioned away from the incision.

Then standard RS approach is performed. Here the first step is the opening of the cerebellopontine cistern to drain excessive amount of cerebrospinal fluid. This will allow the surgeon work easier without using any retractor. With opening of the cerebellopontine cistern more superiorly, the anatomic structures in the cerebellopontine angle are identified. Lower cranial nerves are first exposed (Fig. 18.3a). In prelingually deafened children with malformations hypoplastic vestibulocochlear nerves, the facial and the lower cranial nerves are identified (Fig. 18.3b).

The next step is identification of the flocculus to reach the lateral recess. The choroid plexus protruding from the foramen of Luschka and the cochlear vein are landmarks for this step. The choroid plexus, which covers the foramen of Luschka, lies within a triangle formed by the eighth nerve, the ninth nerve, and the lip of the foramen of Luschka [13] (Fig. 18.3c). To approach the lateral recess, arachnoid over the foramen is cut, and the flocculus and choroid plexus are retracted either by suction or bipolar coagulator. The choroid plexus projecting from the lateral recess and overlying the cochlear nucleus complex is followed and the entrance to the lateral recess is found. The dorsal cochlear nucleus, which is the most accessible portion of the cochlear nucleus complex for electrical stimulation, is identified since it bulges in the floor of the lateral recess [13].

In certain situations, lower cranial nerves cannot be identified. In three children operated in Hacettepe University, severe fibrosis made the identification of the nerves impossible. In order to avoid damage to the cranial nerves, individual nerves were not dissected. Instead, in these cases choroid plexus was identified close to the root entry zone of the ninth nerve and used as a landmark for the foramen of Luschka.

Friedland et al. [16] indicated that endoscopes may be useful in identification of the foramen of

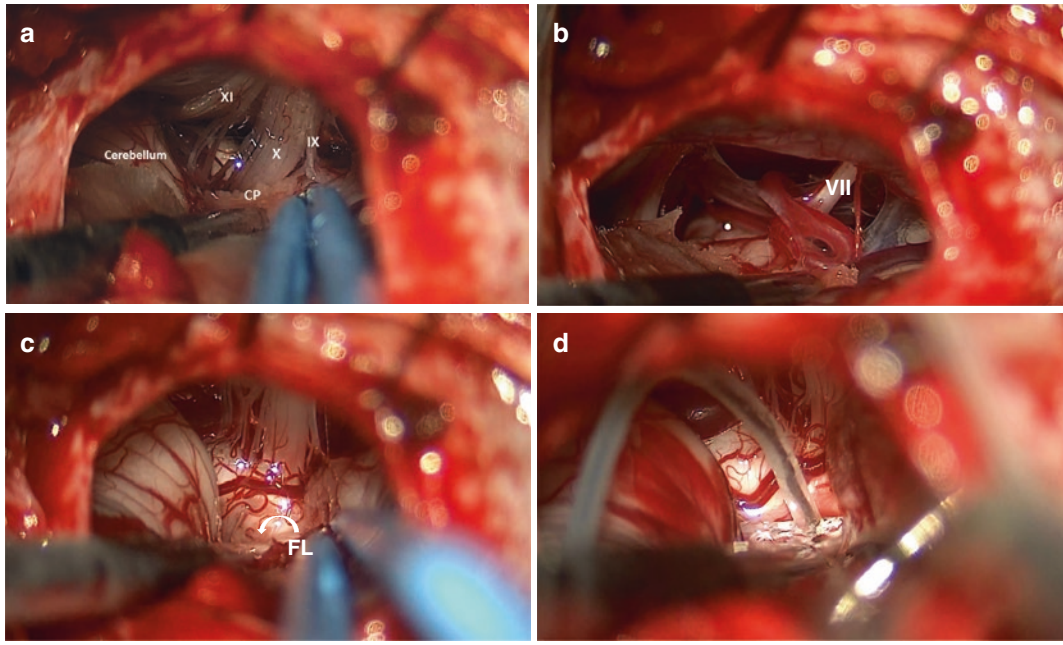


**Fig. 18.3** (**a**) Retrosigmoid approach, showing lower cranial nerves (IX = glossopharyngeal nerve, X = Vagus, XI = nervous accessories, CP = choroid plexus), (**b**) facial nerve (VII), (**c**) foramen of Luschka (FL), (**d**) electrode in position

Luschka. The use of the 30° angled endoscope allows visualization anterior to the flocculus and glossopharyngeal root entry zones prior to any retraction. This allows preservation of the delicate taenia, which has been shown to be a useful landmark for the cochlear nuclei. Furthermore, the foramen of Luschka can be easily distinguished from other reported "false" passages by direct visualization into the recess. Using endoscopes may also avoid strong cerebellar and flocculus retraction in case of large tumors in identification of the foramen of Luschka. The small diameter of the endoscope and ability to advance the scope to the implant site allow less retraction of the cerebellum. Further, craniotomy size can be reduced when endoscopes are used for approaches to the cerebellopontine angle. They claim that with experience the electrode may be inserted with less retraction when a 30° endoscope is used.

At this moment, CSF pressure is raised by anesthesiologist to force CSF outflow from the lateral recess and this also helps to determine the foramen of Luschka accurately. The width of the recess is controlled with a blunt hook or dissector, but it is not always easy to open the entrance of the foramen Luschka because of the underlying veins and sometimes small arteries. Particularly in patients with a history of meningitis, the arachnoid which covers the entrance of the Luschka will be an important problem for the surgeon After opening and controlling the recess, the receiver-stimulator is placed into the implant bed and fixed. The electrode is inserted gently into the recess (Fig. 18.3d). Care should be taken to avoid injury to numerous vessels around this area feeding the brainstem. If a small branch is bleeding it has to be controlled with surgicel application or fine tipped bipolar cautery before undertaking insertion of the electrode paddle. It is very important to place the contact surfaces facing the cochlear nuclei. In our institution, the mesh around the electrode paddle is reduced in size as the recess is not as large as in adults. Final position of the electrode is verified with the help of electrically evoked auditory brainstem responses. According to test results electrode paddle can be advanced vertically slightly in or

out of the recess. It may also be moved slightly to the front and backwards. Usually, it is sufficient to see the outer rim of the electrode paddle. If we do not see the outer rim of the paddle it usually indicates to much insertion. To stabilize the electrode, two or three millimetric muscle tissue are placed into the recess behind the electrode pushing the electrode anteriorly to create better contact with the cochlear nuclei. Then dura is then closed tightly.

Sometimes ABI surgery cannot be performed and this should be mentioned preoperatively to the patient and the family during counseling. In two of our patients lateral recess was too narrow for the ABI to be placed. Both of these patients were 1-year-old children with malformations. ABI could be inserted after the recess was slightly enlarged. In one adult patient with NF2 the foramen was too narrow and ABI could only be done to the contralateral side. Behr et al. [22] also reported a patient where ABI could not be performed safely because of a large vein inside the lateral recess.

#### 18.9.1.1 Disadvantages

As Lenarz et al. [23] pointed out, the disadvantage of a reduced view into the lateral recess in RS approach can be overcome by retraction of the cerebellum and the optimization of electrode placement with the help of precise intraoperative monitoring. In Hacettepe University, at the beginning we used cerebellar retraction in pediatric ABI cases. With experience ABI is placed without any cerebellar retraction. However, in NF2 cases retraction is necessary during surgery. In addition facial nerve is not optimally exposed in the fundus of the IAC. In nontumor patients undergoing ABI surgery RS approach is advantageous.

### 18.9.2 Translabyrinthine Approach

This is the initial approach used by House and Hitselberger after removal of acoustic neuroma [1]. Behr et al. [22] and Laszig et al. [18] indicated that the route to the lateral recess is more straightforward in the translabyrinthine (TL) approach, because the opening of the skull is more lateral than the RS approach. The TL approach provides a wide angle of view posterior to the eighth nerve and the lateral recess [3]. It is preferred by the majority of the otologists [24]. Sollmann et al. [25] and Otto et al. [3] preferred TL approach in ABI surgery. This approach allows early and safe identification of the facial nerve during the NF2 surgery [20]. Facial nerve and the fundus of the IAC are better controlled with this approach and therefore may be the best approach in NF2 cases where the tumor is located laterally in the IAC. In addition TL approach avoids cerebellar retraction [18]. The taenia of the choroid plexus in the lateral recess might have to be divided in order to facilitate insertion.

The operation is performed with the patient in the supine position with the head turned away from the surgeon [20]. Fayad et al. [2] indicated that a postauricular "C" shaped incision is preferred for this approach. The C-shaped incision extends 1–1.5 cm above the pinna. This modification allows the placement of the internal receiver and magnet under the scalp. Care must be taken so that the incision does not directly cross the area where the receiver/stimulator is to be placed. Failure to do this may cause device extrusion. Kuchta et al. [20] also modified the standard TL incision by placing a postauricular incision far enough posteriorly to allow sufficient flap coverage of the implant. After TL removal of the temporal bone and the tumor, landmarks for the foramen of Luschka are identified.

The taenia choroidea is the lateral limit of the ependyma of the lateral recess [3]. Lying directly beneath the taenia choroidea is the target cochlear nucleus. Fayad et al. [2] indicated the importance of the ninth cranial nerve to identify the foramen of Luschka. The ninth nerve is generally in a fixed anatomic position leading to foramen of Luschka in almost every case. In the surgical setting, where there is almost always distortion of the brainstem from the tumor, the foramen of Luschka is located superior to the ninth nerve. In addition, Laszig et al. [18] indicated that whenever possible following the eighth nerve leads the surgeon to the cochlear nucleus complex. CSF can be seen emerging from the foramen; this

might be enhanced by asking the anesthesiologist to raise the intracranial pressure. The receiver-stimulator is secured before placement of the electrode. The ABI electrode is then gently inserted into the lateral recess.

The most favorable position for the ABI electrode array was decided by stimulating through the electrode array and monitoring auditory evoked potentials and electromyographic activity from the seventh and ninth cranial nerves. If there is electromyogenic activity, slight adjustments are made in the position of the electrode to decrease the postoperative side effects. After the electrode array was properly positioned, it was held in place by 2–3 pieces of muscle and surgicel. Proper fixation allows better contact with cochlear nuclei and decreases the possibility of migration. The ground electrode is placed under the temporalis muscle. The wound was closed in layers by using abdominal fat to obliterate the mastoid defect.

#### 18.9.2.1 Disadvantages

It may be difficult to provide exposure of the lateral recess in cases where the sigmoid sinus is anteriorly located or the jugular bulb is located in a high position. In children temporal bone is smaller when compared to adults, and TL approach results in a much smaller surgical exposure than the RS approach. Due to drilling of the temporal bone it may also take more time to expose the brainstem in children when compared to RS approach. Therefore, RS approach is preferred in children. In addition, the RS approach makes it possible to bypass the mastoid air cells preventing intracranial contamination with the middle ear flora. Watertight closure of the dura avoids the need to seal the temporal bone cavity with abdominal fat.

### 18.9.3 Retrolabyrinthine Presigmoid Approach

This is done in situations where RS approach was not possible and TL approach was not necessary. In tumor cases TL approach is very valuable to remove the tumor from lateral part of the IAC with direct visualization of the facial nerve. In children with nontumor indications for ABI there is no necessity to expose the IAC for that purpose. We had two children with severe vascular abnormalities preventing RS approach. One was observed on temporal CT and the other one was seen during surgery. There was wide continuous bleeding between the dural layers in RS incision area. As there was no vessel identified which can be ligated or coagulated, the procedure had to be stopped. Both cases were operated by retrolabyrinthine presigmoid approach.

## 18.10 Intraoperative Monitoring

After placement of the electrode, electrical ABR is utilized to identify the localization of the cochlear nucleus. Different electrodes and electrode groups are stimulated one by one to check the position of the ABI electrode in relation to the cochlear nucleus. This will help to position the electrode array to maximize auditory stimulation while nonauditory stimulation is minimized. In children, the recess is not very large; therefore, after placement, usually only slight movements in and out of the recess are possible. If the electrode is too deeply inserted, there will be response only on the lateral contacts. This necessitates pulling out the electrode until response is observed from the medial contacts. Similarly, if the response can only be obtained from the electrodes localized at the tip, it should be slightly inserted deeper into the recess. In adults, we encountered a few cases where the width of the lateral recess was twice the size of electrode. In these cases the electrical ABR is very useful in confirming the exact placement of the array. Slight adjustments in the position of the array should be made according to electrical auditory responses. The surgeon and the audiologist should be familiar with the numbers of individual active channels on the electrode array. A diagram showing the channels for both left and right sides should be kept in the operating room to avoid confusion about electrode orientation. Position of an individual active channel of an already inserted electrode on the left side is completely opposite on the right side.

In patients undergoing ABI surgery, an intraoperative eABR demonstrating III and V. waves is a valuable finding (Fig. 18.4a). This shows that the electrode is in the correct location. Sometimes there may be no response (Fig. 18.4b) or myogenic activity (Fig. 18.4c). Myogenic activity shows a possible future side effect. In this situation the position of the electrode array is adjusted according to the findings.

## 18.11 Surgical Complications

Majority of the complications so far are related to adult ABI surgery for NF2 cases. According to Toh and Luxford [1] CSF leak, electrode migration, and nonauditory side effects are the most common complications in ABI surgery. Fatal complications are rare. Laszig et al. [18] reported that one of their patients died in the perioperative period following tumor removal and ABI insertion as a result of pulmonary embolism and pneumonia. Grayelli et al. [26] also reported one fatal embolism. Both cases can be accepted as a complication of posterior fossa surgery rather than ABI surgery.

CSF leaks may be due to passage of CSF along the electrode lead, from the subarachnoid space to the subcutaneous plane. It is very important to close the dural incision tightly to avoid this complication. Usually the leaks respond well to conservative management, such as pressure dressing. Reexploration is rarely necessary for control of the leak. Otto et al. [3] reported two CSF leaks as a complication of tumor removal (in 61 patients) that resolved after the application of

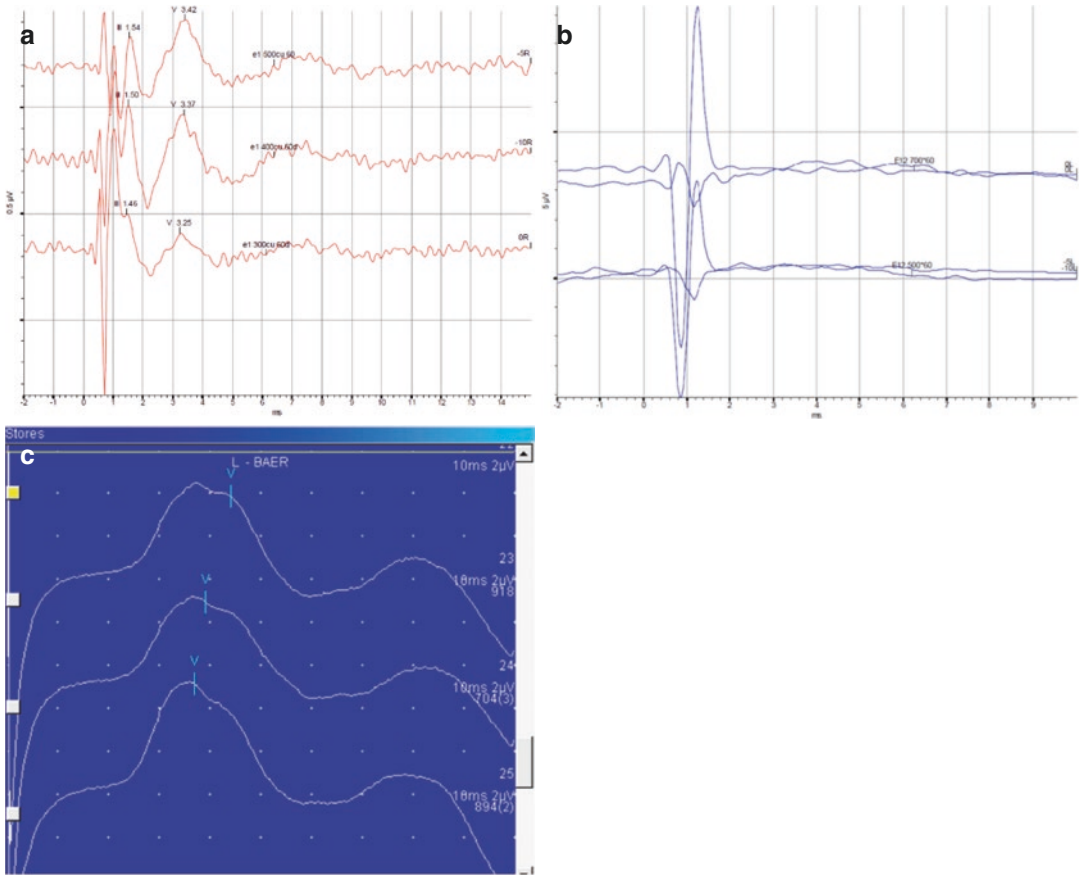


**Fig. 18.4** Intraoperative electric auditory brainstem response (eABR): (**a**) eABR recording with waves eIII and eV, (**b**) eABR recording without response, (**c**) eABR recording with both auditory and nonauditory stimulation

a pressure dressing in one, and after lumbar drainage in the other. Infectious complications (meningitis) developed in one patient. These were attributed to translabyrinthine surgery and are not directly result of electrode implantation. Grayelli et al. [26] also reported 2 cases of CSF leaks after 31 ABI surgeries. Sennaroglu et al. [27] reported a postoperative rhinorrhea in one of the initial children who underwent retrosigmoid ABI placement. She was immediately taken to the operating theater and the leakage point in the mastoid air cells was repaired.

Migration of the electrode may occur as a result of unstable positioning or changes in shape and position of the brainstem after tumor removal. Electrode position may be confirmed on high resolution CT scans. Two cases were reported by Nevison et al. [4]. Grayelli et al. [26] reported that majority of their patients had an uneventful postoperative course (83%). One patient had CPA hematoma displacing the array secondary to a head trauma 2 months after surgery. Behr et al. [22] reported a case of electrode dislocation. The postoperative CT scan suggested that the electrode was in the correct position. When the transmitter coil was fitted no auditory sensation was perceived, no side effects sustained, and there were normal electrode impedance measurements. A second CT scan showed a small lateral displacement of the array when compared with the first scan. At revision, 8 months after the first operation, electrode array was found to be located 4 mm lateral to the correct position. After repositioning, as in the first operation, E-ABRs were recorded by stimulation of each electrode of the test array. Laszig et al. [18] also reported a case of device migration.

None of the 128 children with inner ear malformations who had ABI surgery in Hacettepe University had device migration. There may be two reasons for this. As they had no tumor preoperatively, no shift in the brainstem occurred in the postoperative period as may occur in NF2 patients. In addition, lateral recess is smaller when compared to adults and the electrode tightly fits into the recess. As a result electrode migration in children is rarer when compared to adult NF2 cases. In our series electrode migration was experienced in a child with meningitis. It was not possible to remove the electrode plate which was attached tightly to the brainstem.

Toh and Luxford [1] indicated that nonauditory side effects have occurred in 42% of multichannel implant users and seem to be related to electrode position. Symptoms related to glossopharyngeal nerve stimulation are typically a sense of tingling or constriction in the throat. Some patients have nausea and shoulder contraction related to vagal and accessory nerve stimulation, respectively. There may be facial twitching due to stimulation of the intact facial nerve. A mild sense of jittering of the visual field also has been reported, possibly related to activation of the flocculus of the cerebellum. Nonauditory side effects in the multichannel device generally occur with stimulation of the more medial or lateral electrodes. They can usually be reduced by switching reference electrodes, increasing the duration of the stimulus pulse, or turning off the electrode. The severity of the nonauditory sensations often decreases over time, sometimes allowing for reactivation of electrodes previously turned off.

Otto et al. [3] reported that postoperatively, 6 of the 61 patients who received implants did not report useful auditory sensations. This is a very important finding that should be included in the informed consent. One of those patients received a contralateral ABI during subsequent second-side tumor surgery and made use of his implant. No patient underwent surgery specifically for bilateral implantation, or only for repositioning of an ABI electrode array.

Colletti et al. [28] reported the complications of ABI surgery in their series composed of adults and children. They had no mortality. One child had a slow recovery after surgery, a computed tomographic scan revealed an intracerebellar clot. Revision surgery was performed, and clot was evacuated. He had a full neurologic recovery. Another child developed meningitis. This resolved uneventfully with medical treatment. As a minor complication they observed temporary asymptomatic cerebellar edema in the postoperative computed tomographic scans in nine children. They were all treated successfully with steroids

and diuretics. Four children developed postoperative wound seroma which was successfully treated with aspiration and pressure dressing. Apart from these, infection of the incision, temporary dysphonia, and balance disorders occurred in certain patients but resolved after treatment. The authors concluded that the surgery bares less complications when compared to ABI operation of NF2 patients and overall complication rate of ABI is not much greater than that of CI and comparable to neurovascular decompression.

Bayazit et al. [29] reported two cases of postoperative cerebrospinal fluid (CSF) leakage following ABI surgery in five children. Attention was drawn to possible long term complications such as device failure, infection, biofilm formation, or extrusion, about which still knowledge is limited.

In our series of children, one of the initial three patients had postoperative rhinorrhea. He was revised immediately and the defect in the mastoid was repaired. Four patients had transient facial nerve palsy which resolved completely in three cases within 2 weeks. The fourth child had grade II facial nerve recovery. This was attributed to the cerebellar retraction.

In one patient, severe cerebellar edema occurred intraoperatively which impeded rest of the surgery. Therefore, operation was stopped and completed in a second session uneventfully 3 weeks later. Seroma occurred in five patients due to CSF leakage. In four patients it was easily controlled in a few days, with lumbar drainage and serial dressings. However, in one patient, CSF leak continued despite these measures and prolonged the hospitalization period markedly. None of our patients had to be revised due to seroma; mentioned conservative treatment was successful enough to manage this complication. In these patients CSF leakage was thought to occur around the electrode lead from subarachnoid space to subcutaneous tissue. It is important to place pieces of soft tissue around the electrode at the level of dura in order to attain effective sealing and lumbar drainage is used now routinely to avoid CSF leakage. Both of these measures were successful and this complication was not experienced in the rest of the group.

One patient had a serious postoperative complication. She was operated via retrolabyrinthine approach. She had intermittent confusion leading to coma. Intracerebral CSF flow was disturbed. Initially she was managed with drainage but as the situation recurred, permanent intraperitoneal shunt was placed and she had more stable outcome.

Overall results showed that this procedure can be performed with minimum risks in centers with experienced otology, neurosurgery, and anesthesia facilities.

## 18.12 Initial Stimulation and Follow-Up

In the first 3 patients, initial stimulation was done 3 months after the surgery. But now the device is switched on 4–5 weeks after the surgery. General anesthesia is not required; monitoring the child is sufficient.

Most comfortable levels (MCL) are found by increasing the current level step by step. During this time behavioral responses and side effects are observed. After MCLs are determined, all MCLs are decreased by 5 or 10 current unit (CU), and speech processor is activated. This decrement is done because the integrated level of all channels can be annoyingly loud for the first stimulation.

Initially the channels in the center of the electrode are activated. If there are no side effects, then it is possible to proceed to neighboring ones. Usually 6–7 channels are activated in the first visit. The rest of the channels are activated during the second visit which occurs usually 1 month after initial programming. If there is a side effect, the current level is lowered until hearing sensation without any side effects is achieved. If this is not possible, the channel leading to the side effect is closed. A few months later, the channel(s) causing side effects are activated once again. It has been observed that in many occasions, the channels initially causing side effects start to produce only auditory stimulation without any adverse reaction (Fig. 18.5a, b). The ones prompting side effects can be kept closed permanently.

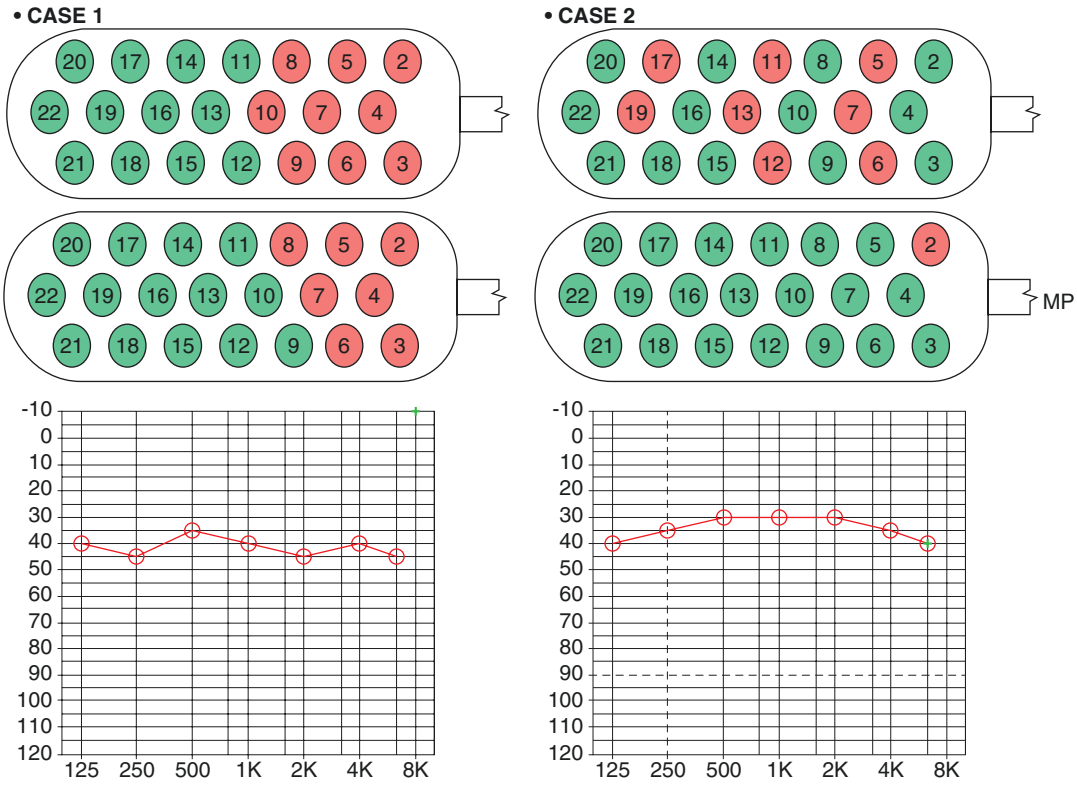


**Fig. 18.5** ABI mapping showing side effects. Case 1: mapping showing very little change in the number of active channels causing side effects over time (green = active electrodes without side effects, red = electrodes with side effects) Case 2: Follow-up of a patient which showed decrease in the number of electrodes causing side effects over time

Fitting infants and young children is a complex work, due to fact that no adult like clear responses can be obtained. But in most of the cases they perform some behaviors with sound stimulation. These may be cessation of activity, looking at mother, holding or showing the implant side or crying. These programing sessions must be done by experienced pediatric audiologists. Side effects must be observed and monitored particularly during the first stimulation. These can vary from single cough, to stimulation of vagus nerve which organizes heartbeat. So it is essential to perform this section in the presence of a medical doctor in case of cardiac arrhythmia. The initial program gives very important information for follow-up. These are all noted for future programming.

In Hacettepe University we have done eABR before initial stimulation for the first patients. It has been observed that this does not add more information than the intraoperative eABR measurements. Today eABR is not performed anymore. We use intraoperative findings for the first programming section.

## 18.13 Conclusion

ABI in children provides auditory sensation when properly placed into lateral recess. Side effects due to the stimulation of the neighboring cranial nerves are common which can be overcome by decreasing current level or closing the channel permanently. Every effort should be shown to decrease the intracranial complications by working in collaboration with an experienced otologist, pediatric neurosurgeon, and anesthesiologist. Satisfactory audiological outcome with

language development is possible but handicaps impede success of outcomes. Probable indications still continue to be challenge for the implant team.

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