Treatment Alternatives in Inner Ear Malformations

11

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Inner ear malformations (IEM) are characterized by abnormal cochlear and vestibular anatomy together with cochlear nerve deficiency. They may present with different audiological configurations, ranging from normal hearing to conductive, mixed, and sensorineural hearing loss. As a result, different treatment options are available for different malformations. At this point it is appropriate to mention that not all IEMs need cochlear or brainstem implantation for restoration of hearing. There are five options for (re)habilitating hearing loss in IEMs:

11.1 Normal Hearing

Patients with enlarged vestibular aqueduct (EVA) and incomplete partition II (IP-II) anomalies may have normal hearing at birth. They may obtain pass in hearing screening if they have normal hearing. Usually they show progressive deterioration requiring a hearing aid or a cochlear implantation later on in their life.

11.2 Hearing Aids

Hearing aids may be a sufficient option in EVA, IP-II, some cases of IP-III, and some cases of cochlear hypoplasia (CH) if they have moderate to severe mixed or sensorineural hearing loss (SNHL). There is no indication for hearing aids in definite indications of ABI [1]. Common cavity, almost all IP-I cases, majority of cochlear hypoplasia and IP-III cases do not benefit from a hearing aid trial.

11.3 Stapedotomy

It has been shown in histopathological studies that it is common to have stapedial fixation in CH cases [2]. If the patient has a cochlear nerve, with an air-bone gap in cases of CH, stapedotomy is a good option to decrease air-bone gap. Depending on the bone conduction thresholds stapedotomy may provide quite satisfactory hearing alone in pure conductive hearing loss (Case 1, Chap. 26), but in situations with mixed hearing loss a hearing aid may be necessary to obtain better hearing after closure of the ABG with stapedotomy (Case 2, Chap. 26). Stapedotomy is contraindicated in IP-II and IP-III who may also present with mixed air hearing loss.

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11.4 Cochlear Implantation

Cochlear implantation (CI) is indicated in cases with severe-profound hearing loss if they have a cochlear nerve. This may be present at birth or it may be progressive over time. It is important to remember that these patients may need special surgical approaches and electrodes for CI [3].

11.4.1 Surgical Approach

Majority of the CI operations in malformations can be done via the classical transmastoid-facial recess approach. Sometimes the presence of complex malformations makes this approach impossible and the surgeon must be ready to modify the surgical approach. In some cases with CH, and rarely in IP-I cases, it may be necessary to modify the surgical approach due to facial nerve abnormality. In patients with CH, facial nerve (FN) frequently has an abnormal course (lying on the promontory or round window) and CI was inserted via vestibule, promontory, or posterior to FN. In two patients with IP-I anomaly who had severe FN anomaly, a combined transmastoid-transcanal approach was used [4]. In addition, patients with common cavity (CC) anomaly may need transmastoid labyrinthotomy or double labyrinthotomy approach.

11.4.1.1 Facial Recess Approach

Facial recess approach was described by House [5] and is the standard approach in CI surgery in the majority of the clinics. In this approach round window identification and entry into cochlea are done through the triangular space between facial canal, fossa incudes, chorda tympani nerve, and ear canal (Fig. 11.1a). Two situations may complicate the facial recess approach:

Abnormal Location of the Facial Nerve in the Facial Recess

The course of the facial nerve may be altered in certain malformations such as CH, CC, and IP-I. The vertical segment of the FN is usually dislocated anteromedially towards the promontory; it may be lying over the oval and the round windows and the surgeon may be unable to use the facial recess approach.

Unfavorable Cochlear Anatomy Through the Facial Recess Area

In certain IEMs cochlear promontory is not fully developed. As a result, round window and other necessary landmarks may not be visualized making cochleostomy very difficult.

1. *Cochlear Hypoplasia*: In normal cases, the usual protuberance of the promontory is provided by the normally developed basal turn of the cochlea. In patients with severe CH, promontory is underdeveloped as a result of the hypoplastic basal turn and the cochlea may be inaccessible through the facial recess. Facial recess approach may be used in CH-IV where the basal turn is normal but middle and apical turns are underdeveloped. However, it is also possible to have abnormal facial nerve in CH-IV necessitating subtotal petrosectomy (Case 3, Chap. 26).
2. *Common Cavity*: Facial nerve is expected to be in an abnormal location in common cavity (CC) because of severe cochleovestibular developmental anomaly. If HRCT of a patient with CC is examined, it can be seen that CC is located posteriorly which can be easily approached through the mastoid. During the surgery of the first CC patient in our department, facial recess was opened but there was no promontory or round window. In these cases, there is no separate cochlea on the anterior part to produce promontory. Therefore, in CC it is not advisable at all to open the facial recess to make a labyrinthotomy. Labyrinthotomy can easily be done through mastoid as described by McElveen [6]. Transmastoid labyrinthotomy [6] or double labyrinthotomy [7] approaches are used in patients with CC for electrode placement into the cavity.

11.4.1.2 Transcanal Approach

Alternative approaches (transcanal approach for the cochleostomy) were reported in standard CI surgery by Kiratzidis [8] and Kronenberg [9] (Fig. 11.1b). When the anatomy of the inner ear is not severely distorted transcanal approach can also be used for cochlear implantation. Examples are EVA and IP-II. In severe anomalies, such as CH, it may be difficult to use this approach. In

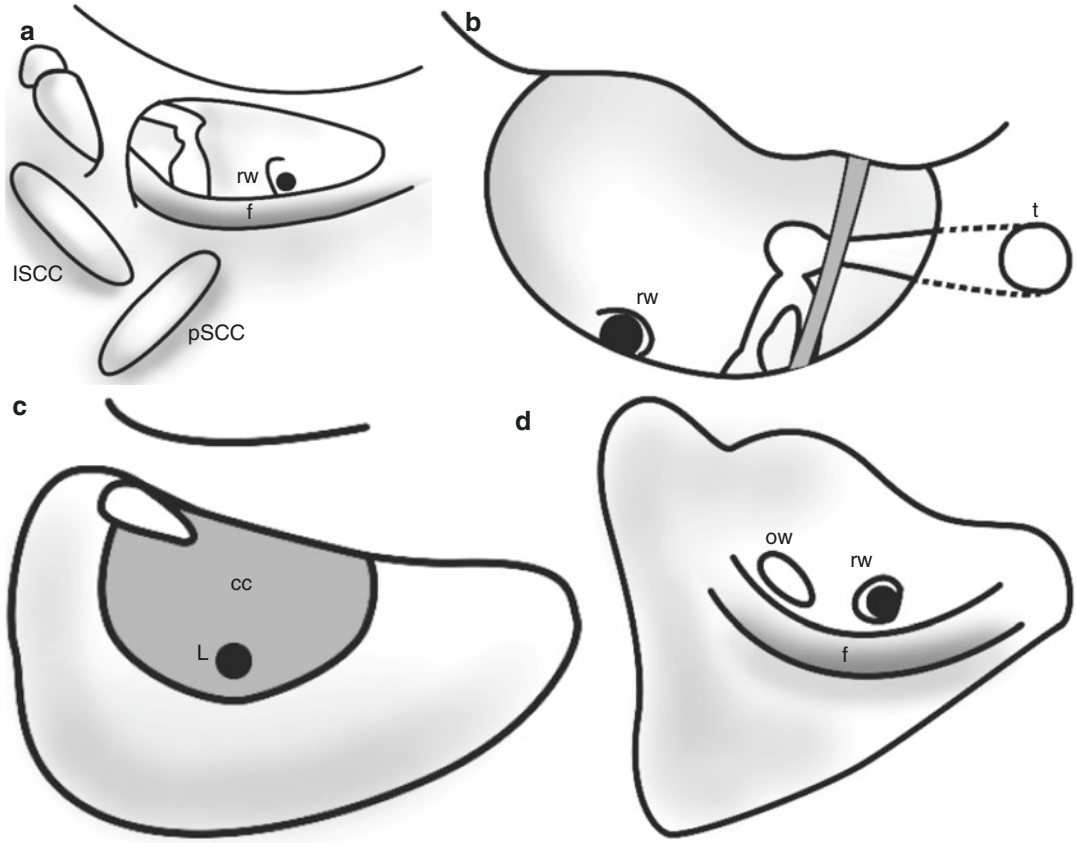

Fig. 11.1 Surgical treatment options: (**a**) Facial recess approach, (**b**) Transcanal approach, (**c**) Transmastoid labyrinthotomy approach, (**d**) Subtotal petrosectomy. *rw* Round Window; *f* Facial Nerve; *lSCC* Lateral Semicircular Canal; *pSCC* Posterior Semicircular canal; t tunnel for the electrode; *OW* Oval Window; *CC* Common Cavity; *L* Labyrinthotomy

CC it is not advisable at all to use alternative approaches. In addition, in IP-I and IP-III it is difficult to manage severe gusher and electrode misplacement into IAC. Therefore, in general alternative approaches are not advisable in IEMs.

Transcanal approach, however, can be used in combination with transmastoid approach in certain situations [4]. Because of facial nerve abnormal location, it was impossible to use the facial recess approach in two cases of IP-I and the electrode was inserted by transcanal approach (Fig. 11.2a–d). After identifying the difficult anatomy, a cut was produced in the bony ear canal with a tiny diamond bur. After insertion through the ear canal, the electrode was transferred to the mastoid cavity. The cut was covered with a thin cartilage. Both cases had IP-I anomaly and first case was reported [4].

Transcanal and facial recess combination can be used in cases with oval window fistula with CSF gusher and round window electrode insertion. Transcanal approach provides direct access to the footplate area and evaluation and management of CSF fistula at the footplate can be done better than facial recess approach. Facial recess can then be used for cochlear implantation through the round window and management of gusher around electrode. The advantage is that the electrode lead is placed in the mastoid cavity.

Weber et al. [10] also reported transcanal approach in four patients. They indicated that removal of incus greatly facilitated the vision if the promontory is flat. The fact that combination of transcanal and transmastoid approaches was extremely useful in these situations has to be in

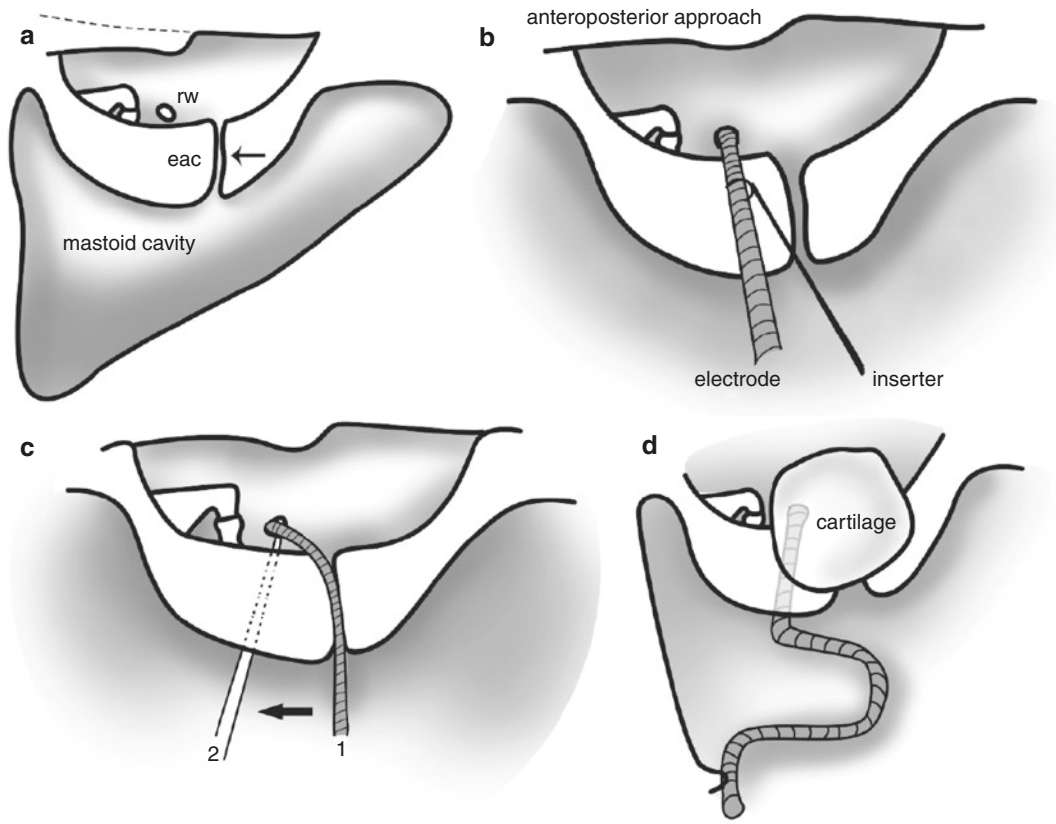

Fig. 11.2 Split ear canal technique. (**a**) A cut was produced in the bony ear canal with a tiny diamond bur. (**b**) Electrode insertion through the ear canal, (**c**) Transfer of the electrode through the opening into the mastoid cavity. (**d**) The cut was covered with a thin cartilage. (Modified from the paper Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. Otol Neurotol. 2002 Jan; 23(1): 39–42) rw = round window, eac = external ear canal

surgeon's armamentarium. The surgeon must be ready to modify the surgical approach in complex IEMs such as CH.

11.4.1.3 Transmastoid Labyrinthotomy

This approach was first done by McElveen [6] and reported by Molter et al. [11]. As already pointed out facial recess approach is not the appropriate approach in CC deformity. The authors reported aberrantly coursing facial nerves in 1/3 of patients with CC undergoing cochlear implantation. By taking a direct transmastoid labyrinthotomy approach to the CC and avoiding the facial recess and promontory dissection, one may be able to implant the electrode array with maximum visualization and with minimal risk to the facial nerve. It is advisable not to attempt to open up the facial recess or expose the facial nerve. McElveen et al. [6] suggested to make a labyrinthotomy at the location of lateral semicircular canal. In our department we made a slight modification and we create the opening anywhere along the cavity away from the facial nerve (Fig. 11.1c) (Video 11.1). Usined into the common cavity. Recently, we had some cases who had inadvertent entry of the electrode into the IAC. Therefore, it is advisable to check the position of the electrode intraoperatively in all patients with CC and gusher. In case of electrode misplacement into IAC, double labyrinthotomy can be done easily and the electrode is located in the CC as described by Beltrame et al. [12] (Video 11.2).

11.4.1.4 Canal Wall-Down Mastoidectomy with Blind Sac Closure of the External Auditory Canal

In situations of difficult anatomy where the FN prevents the standard facial recess approach, using a canal wall-down procedure, better visualization of the promontory, oval and round windows can be obtained (Fig. 11.1d). In patients with uncontrollable gusher and recurrent meningitis this may also be necessary in addition to proper control of leakage point. There is a disadvantage of this procedure. There is a possibility of leaving some squamous epithelium in the cavity becoming cholesteatoma within a period of few months. This may create a surgical problem because in patients with CI, as MRI cannot be done for differentiation of cholesteatoma from other soft tissue mass. Therefore, it is difficult to follow up the mastoid for cholesteatoma.

In cases of gusher it is very important to properly control the point of cerebrospinal fluid (CSF) leakage. The FORM electrode with the silicon stopper is particularly developed to more efficiently control the CSF leakage in gushers. The electrode is passed through a tiny piece of fascia and both are inserted together. **IT IS THE SURGEON'S RESPONSIBILITY NOT TO LEAVE OPERATION THEATER WITHOUT FULLY CONTROLLING CSF GUSHER**. Continued CSF lumbar drainage for 4–5 days after surgery is very important to keep the fascia and electrode in place. In our department we find this method quite sufficient to control CSF leakage. Once the leakage is controlled fully the surgeon may perform subtotal petrosectomy where the cavity is obliterated with abdominal fat and the Eustachian tube closed after blind sac closure of the ear canal. This may provide additional barrier to prevent meningitis. **The latter should not be done if there is still leakage around the electrode.** The safest situation is to control the leakage point efficiently.

In cases of previous mastoid surgery or chronic otitis media blind sac closure of the ear canal should be done together with complete removal of the skin in the ear canal. In these cases, and also if the procedure is done for difficult anatomy, it may be a better option not to obliterate mastoid cavity and the Eustachian tube as described by El-Khaslan et al. [13]. In the postoperative period an air filled cavity will be seen in the middle ear and mastoid area on HRCT. If there is an expanding soft tissue mass on repeated HRCT, this is likely to be a cholesteatoma and exploration should be planned. If these cases are obliterated with fat, soft tissue will make the investigation of the mastoid cavity for residual cholesteatoma in the postoperative period almost impossible. MRI is contraindicated in patients with CI and without MRI, a soft tissue in the mastoid cavity cannot be differentiated from cholesteatoma.

11.4.1.5 Oval Window

Kim et al. [14] reported that they had to use the oval window for electrode insertion in two patients with CH. Preoperative imaging studies showed that the children had very small cochlear buds. When they opened the facial recess, they noticed that the stapes were present, but no round window niche was identified. In spite of their efforts to open the small cochlear bud, it was not possible to find the cochlear lumen. They removed the stapes and inserted electrodes through the oval window into the vestibule.

11.4.2 Electrode Choice

It is evident from the classification of IEMs, there are many varieties of cochlear malformations with considerable structural differences. Radiology is the method for diagnosing the type of IEM. It is advisable to choose the electrode according to the type of cochlear malformation on HRCT and MRI. When choosing the particular type of electrode, it is important to keep in mind to place the electrode in appropriate location to provide maximum stimulation of the neural tissues, obtain full insertion, prevent CSF leakage around the electrode, and finally make it possible to revise the situation in patients with high risk of complications. Therefore, preoperative HRCT is extremely important to accomplish these goals.

11.4.2.1 Special Electrodes for Malformations

FORM Electrodes

After having a fatal complication following a severe CSF leakage in an IP-I case in 2006, Sennaroglu L developed the idea of a progressive silicon stopper in the shape of a "cork" to more effectively stop the CSF leakage after electrode insertion [15]. This idea was developed into a special electrode and FORM electrode series were produced by Med El. The electrode has a "cork" like silicon stopper which marks the end of insertion (Fig. 11.3) [15]. It is thought that this will effectively block the cochleostomy preventing CSF leakage.

There are two lengths for FORM electrode:

(a) FORM 24: The length of this electrode is 24 mm and it was calculated using the formula $2\pi r$ after measuring the diameter of malformed cochleae in IP-I, IP-II, and IP-III [16] (Fig. 11.3a, b). A previous radiological study [16] was used to determine the length of this electrode so that it will make only one full turn around the cochlea [15]. A longer electrode has more chance to enter IAC, particularly in IP-III. This electrode can be used in large CC patients as well. As it has contacts on both surfaces it may provide better stimulation than electrodes with contacts on one surface. Sennaroglu L proposed to measure the diameter of the CC and estimate the perimeter of CC using the formula $2\pi r$ [3] (Fig. 11.4). This measurement will roughly give the electrode length to make one full turn around CC.

(b) FORM 19: The length of this electrode is 19 mm (Fig. 11.3c, d). There is a large group of CH where the dimensions are much smaller in relation to normal cochlea. CH-II is a cystic hypoplasia where there is a risk of CSF gusher. If we insert a long electrode into a small cochlea there is a risk that the electrode will not be fully inserted into the cochlea. Therefore, the silicon cork may not be at the level of the cochleostomy. After experiencing partial insertions with FORM 24 in CH, Sennaroglu L urged the Med El company to produce this shorter version of the electrode (FORM 19) to make full insertion into smaller hypoplastic cochleae. Therefore, it is not advisable to use FORM 24 in CH cases. Likewise, FORM 19 will be too short for incomplete partition cases and this may result in insufficient stimulation. FORM 19 may be used in small CC as well.

Common Cavity Electrode

In CC the electrode is inserted into a cavity. There is a possibility that the electrode may go into IAC particularly in cases of CSF gusher. This is due to the fact that electrode is not inserted into bony scala which normally guides it towards the apex. In CC it may go in any direction. To minimize this unwanted effect, Beltrame et al. [7] described a special electrode for CC, which has a non-active tip to be seized through another opening. Two labyrinthotomy openings are done. A superior

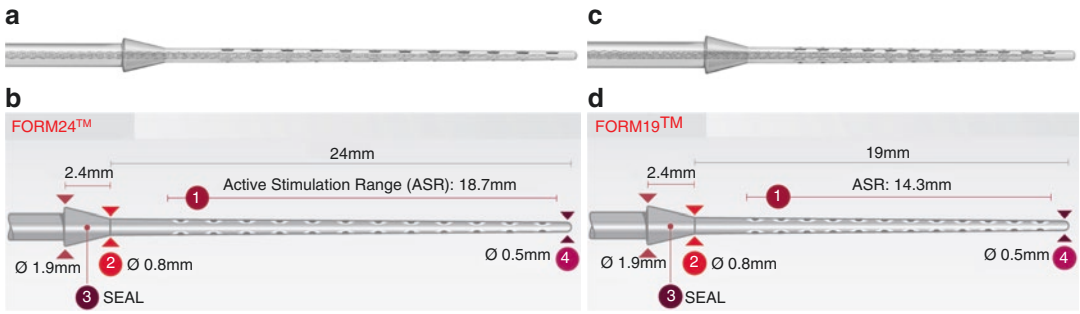

Fig. 11.3 FORM electrode series. (**a** and **b**) FORM 24 electrode. (**c** and **d**) FORM 19 electrode. (With permission from Med-El company)

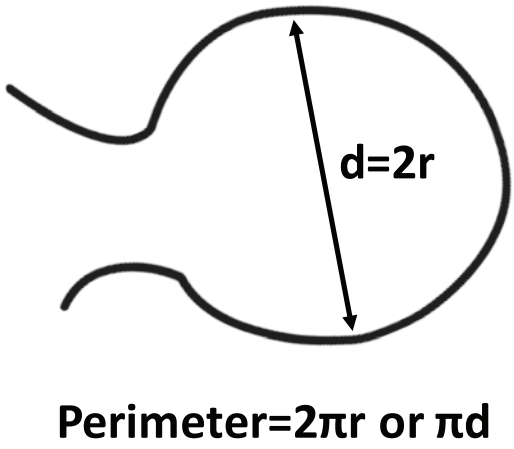

Fig. 11.4 Measuring the diameter of common cavity and using the formula perimeter = $2\pi r$ (or $d\pi$) gives the length of the electrode that makes one full turn in the cavity

labyrinthotomy is made in an area close to where the non-ampullated end of the lateral semicircular canal would normally be seen. A second labyrinthotomy of the same size is made 3–4 mm inferiorly to the first one. The terminal non-active part of the electrode array ends with a small ball, which is needed to hook the electrode array. This non-active part of the implant is pushed into the superior labyrinthotomy until it is seen and hooked using a 0.5 mm hook through the inferior labyrinthotomy. Then the two arms are advanced together pushing the electrode array along the inner wall of the cavity. In this way the tip is prevented from going towards the IAC. But as we do not know the exact location of the neural tissues, there is possibility to damage the delicate neural tissue around the common cavity while pushing the electrode outward. However, this double labyrinthotomy approach is valuable to avoid the tip entry into IAC with all kinds of straight electrodes.

Standard Electrodes

Standard electrodes can be used in some malformations such as EVA, CH-III, and CH-IV. Electrodes with full contact rings (Oticon EVO and Standard, Nucleus straight, slim straight 422 or 522 series), or contact on both surfaces (Med El standard, Medium) are more appropriate than modiolar hugging electrodes in majority of the situations.

11.4.3 Type of Malformation and Electrode Choice

11.4.3.1 Common Cavity

In CC the exact location of the neural tissue is not precisely known. It is assumed to be located in the peripheral part of the cavity. Electrodes with complete contact rings or contacts on both surfaces can be used in these cases. Special electrode designed by Beltrame can be used in CC [7]. FORM electrodes can be used in CC particularly if there is a CSF leakage. If modiolar hugging electrodes are used they will curl in the center part of the CC and therefore will not provide sufficient auditory stimulation. Therefore, it is not advisable to use modiolar hugging electrodes in CC. Transmastoid labyrinthotomy approach described by McElveen et al. [6] or double labyrinthotomy approach by Beltrame [7] are ideal approaches for CC. The length of the electrode should be decided according to the size of the common cavity. If it is a large cavity we can use a long electrode. Likewise, a shorter electrode should be preferred in the presence of a small cavity. Sennaroglu [3] proposed to measure the diameter of CC on HRCT and then calculate the perimeter of the common cavity by the formula perimeter = $2\pi r$ (Fig. 11.4). In this way the surgeon can have an estimate about the length of the electrode that can be used to make one full turn around CC. Then the appropriate electrode can be chosen from Med El® standard (31 mm), Med El® Medium 28 mm, FORM 24 (24 mm), FORM 19 (19 mm), Nucleus® CI 24 RE (17 mm) or Med El® compressed (13 mm), Oticon® EVO or Classical.

11.4.3.2 Incomplete Partition Type I

In this type of cochlea, there is no modiolus, resulting in a wide connection with IAC. As a result, the location of the ganglion cells is not exactly known. Here, electrodes with complete rings or contacts on both surfaces are preferred to stimulate as much neural tissue as possible. Because of the defect at the lateral end of IAC, gusher occurs during cochleostomy in 50% of these cases. FORM24 with a "cork" type silicon ring is ideal for these cases. Oticon® and medium

Med El® electrodes can also be used. If there is no gusher, Nucleus® CI 24 RE is another option. Because of the risk for migration into IAC modiolar hugging electrodes like Nucleus® Contour electrode are not advised to be used.

11.4.3.3 Incomplete Partition Type II

In these patients, the basal part of the modiolus is normal and the apex is cystic. Normally, spiral ganglion cells are located in the basal part of the modiolus and no ganglion cells are found in the apex [17]. Theoretically, we should be able to provide considerable stimulation to the inner ear in a way similar to normal cochlea with CI. As the basal part of the modiolus is normal, the basal turn is also normal. In these cases, all kinds of electrodes (modiolar hugging and straight) can be used. 7% of these cases may have severe gusher. FORM 24 is ideal in IP-II in case a gusher occurs. Recently various modiolar defects were reported in IP-II and it is advisable to investigate imaging before decision making for the type of electrode [2].

11.4.3.4 Incomplete Partition Type III

The differences between IP-I and IP-III are the interscalar septa at the lateral wall of the IP-III cochlea and larger defect between cochlear base and IAC in IP-III. Electrodes with complete rings or contacts on both surfaces are ideal to stimulate neural tissue. Severe CSF gusher occurs in 100% of IP-III cases. Therefore, FORM series are preferred as they can effectively block the opening. Ideally FORM 24 makes a full turn around the basal turn in IP-III but sometimes interscalar septa are very thick and they decrease the intracochlear volume. Therefore, in these situations FORM 19 has more chance to make one full turn around cochlea and also stay within cochlea without migrating into IAC.

The probability of the longer electrodes entering the IAC is more than the shorter electrodes. Therefore, a full ring electrode that will make only one turn around the cochlea appears to be sufficient.

There is a high risk that modiolar hugging electrodes can go into IAC as a result of completely absent modiolus. **Modiolar hugging electrodes should be avoided in IP-III**. They can go into IAC. If noticed in the postoperative period removal of a modiolar hugging electrode in IP-III can damage the facial or cochlear nerves. A straight electrode has less chance to migrate into IAC but if that occurs it can easily be removed and repositioned without damaging CN and FN.

11.4.3.5 Enlarged Vestibular Aqueduct

Cochlea is normal and all kinds of electrodes can be used.

11.4.3.6 Cochlear Hypoplasia

The dimensions of the cochlea are less than normal. FORM19 is ideal for all cases of cochlear hypoplasia. Long electrodes should be avoided because of the risk of incomplete insertion. In addition, if there is no risk for gusher, such as CH-III and CH-IV, a short electrode (Nucleus® Straight, Nucleus® 522, or Med El® compressed) can also be used.

11.5 Auditory Brainstem Implantation

Auditory brainstem implantation (ABI) is also indicated in certain IEMs. These are usually severe IEMs where the cochlea, complete labyrinth, or cochlear nerve is aplastic. It may also be indicated in cochlear nerve hypoplasia.

11.5.1 Side Selection

Side selection is very important in ABI surgery. ABI is usually indicated in complex IEMs and the aim of the team should be to try to provide more hearing to both temporal cortex. If there is a hypoplastic CN on one side and CN aplasia on the other side, CI should be planned on the side with deficient CN while ABI performed on the side with aplastic CN. The aim must always be to provide bilateral stimulation. If there are definite indications on both sides, bilateral ABI is the only option to provide bilateral hearing

habilitation. If unilateral ABI is planned, side with more developed neural structures (e.g., facial nerve presenting unilaterally or more prominent CVN) 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.

11.5.1.1 Indications

In the first consensus paper on pediatric ABI, Sennaroglu et al. [1] divided the indications into two groups:

Definite Indications

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

Probable Indications

1. Cochlear hypoplasia with hypoplastic cochlear aperture: CH may have different audiological presentation. If they are accompanied by hypoplastic cochlear aperture on HRCT, usually CN 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 is difficult to determine accurately the functional capacity of the cochlear nerve with the present tests.
2. CC and IP-I cases where cochlear nerve is apparently missing. If the CN is present they are candidates for cochlear implantation. It is important to note that common cavity can be easily confused with cochlear aplasia and vestibular dilatation. The results of CI in cochlear aplasia and vestibular dilatation are not successful and this should be avoided [3].
3. CC and IP-I cases if the cochlear nerve is present: Even if the nerve is 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 cochleovestibular nerve (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 CI 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 CN is defined as less than 50% of the usual size of the cochlear nerve or less than the diameter of the FN. 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.

   Children with hypoplastic CN or thin unbranched CVN constitutes the most controversial group in decision making between CI and ABI. It must be kept in mind that children with hypoplastic CN and CVN usually do not reach levels of those with normal cochleae, in terms of hearing and language development. It is obvious that radiology may not predict the presence of the cochlear nerve accurately in these mentioned challenging five groups of 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 during behavioral testing, this information is very valuable in the side selection for CI. In such cases, family should be carefully counseled about the possibility of contralateral ABI surgery if insufficient progress with CI is encountered during postoperative follow-up.

11.5.1.2 Surgical Approach

ABI can be done via retrosigmoid, translabyrinthine, and retrolabyrinthine approaches [18]. In children main approach for auditory brainstem

implantation (ABI) has been retrosigmoid approach. The advantages are [19]:

1. 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 a much smaller 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, the retrosigmoid approach makes it possible to bypass the mastoid air cells preventing intracranial contamination with the middle ear flora.
2. Translabyrinthine approach has been utilized for ABI in a child by Helge Rask Andersen and his team (personal communication) and the electrode was successfully placed into the recess.
3. Bento et al. [20] 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 also to choose the side to be operated on for RLA based on the size of the jugular bulb. The 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 because of 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 is no requirement for cerebellar retraction or even for opening the internal auditory canal 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. This approach has been used in two patients in our department.

As a result, all three approaches can be used for ABI in children but retrosigmoid approach has been used much more widely when compared with the other two methods.

11.6 Cochlear and Auditory Brainstem Implantation

Finally, there is also an indication of bimodal stimulation with CI on one side and an ABI on the other side. These are cases of probable indications and CI is used on that side and ABI is reserved for insufficient progress on the contralateral side if there is a hypoplastic nerve or can be applied directly if there is a definite indication. This procedure can be staged or performed in the same setting under certain circumstances (see Chap. 32).

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