Inner Ear Malformations

Classification, Evaluation and Treatment

Levent Sennaroglu
Editor

MOREMEDIA

Springer

Inner Ear Malformations

Levent Sennaroglu
Editor

Inner Ear Malformations

Classification, Evaluation and Treatment

Springer

Editor
Levent Sennaroglu
Department of Otolaryngology
Hacettepe University
Faculty of Medicine
Ankara
Turkey

ISBN 978-3-030-83673-3 ISBN 978-3-030-83674-0 (eBook)
https://doi.org/10.1007/978-3-030-83674-0

© Springer Nature Switzerland AG 2022
This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.
The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.
The publisher, the authors and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, expressed or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

Foreword

It was not long ago that inner ear malformations were poorly understood, little studied, and of minimal relevance to practicing otologists. Symptomatic of this neglect, all inner ear malformations, which have a rich diversity of types and subtypes, were lumped together under the omnibus term "Mondini" deformities. Recent advances, driven by the ability to visualize minute structures of the inner ear during life via progressively more sophisticated imaging, have enabled recognition and definition of patterns of malformation. Studies correlating clinical features associated with each morphology enable improved ability to predict hearing loss stability over time. Contemporary imaging also assists in identifying those at risk for CSF leak and meningitis and is important in establishing candidacy for cochlear implantation. Using modern imaging, and an impressive collection of some 700 inner ear malformations, Dr. Sennaroglu has refined and improved earlier inner ear malformation classification schemes.

I have always thought it more than a small miracle, given all the precise biological steps needed to form a human being, that so many infants are born free of imperfections. The study of inner ear malformations provides clues into the essential steps of normal embryology of the inner ear. Unraveling the mysteries of inner ear organogenesis has special relevance not only to birth defects but also to potential pathways forward to regenerating the organ of Corti in adult hearing loss. One promising strategy for regeneration is rekindling the developmental cascade through upregulation of master regulatory genes such as ATOH-1. The lessons learned via study of how embryology goes wrong provides important clues to help drive research into the steps of development and their control mechanisms.

It is a sign of maturity of the field of study of inner ear anomalies that the body of knowledge is now sufficient to merit a book-length treatment of the subject. I congratulate Dr. Sennaroglu and his coauthors for compiling such a comprehensive and well-thought-out monograph covering all aspects of inner ear malformations. Their work lays the foundation for future advances which will elucidate the crucial biological underpinnings for this family of disorders

and, in time, develop biologically based interventions to restore hearing function or, ideally, recognize and correct their genetic basis.

Robert K. Jackler
Department of Otolaryngology-Head & Neck Surgery
Stanford University School of Medicine
Stanford, CA, USA

Departments of Neurosurgery and Surgery
Stanford University School of Medicine
Stanford, CA, USA

Preface

Management of Inner Ear Malformations (IEM) constitutes a major part of the implant teams' work at the present time. With the precision of new generation computerized tomography (CT) and magnetic resonance imaging (MRI), it has been possible to diagnose IEMs more frequently than before.

When I was a research scholar in House Ear Institute in 1997, I had the opportunity to increase my knowledge about cochlear implantation and all kinds of ear surgery. In addition, I examined histopathology slides with Prof. Fred Linthicum, M.D. I learnt to examine the temporal bone slides systematically from superior to inferior. One day an important event happened which would have a big impact on my future career. Prof. Antonio De La Cruz, M.D., offered me to go to the Department of Radiology to consult the images of a difficult atresia patient. There, he introduced me to Prof. William W.M. Lo, M.D., who was the radiologist of St Vincent's Hospital opposite to House Ear Clinic with a special interest in temporal bone radiology. This was at the same time my introduction to radiology of the temporal bone. Prof. Lo was an excellent radiologist. I spent many hours with him. He not only showed me basic temporal bone CT and MRI images but also opened his archive of special cases, and we discussed them in his free time. I owe a lot to this great teacher and I remember him with great respect.

This increased my knowledge of the radiology of the temporal bone. Back in Turkey, in 1997, we started cochlear implant (CI) surgery. I noticed that the term "Mondini" was being used to define two completely different anomalies: incomplete partition type I (IP-I) and II anomalies. I checked the literature. In the excellent book by Prof. Harold Schuknecht, M.D., the term "Mondini Deformity" was used for these two completely different anomalies: one was a cystic cochlea without interscalar septa and modiolus, while the other one had only a cystic apex with almost normal modiolus. Theoretically, the majority of the spiral ganglion cells are located in the 2/3 basal part of the modiolus, and I thought it would not be correct to give the same name to these completely different anomalies.

In the meantime, we started to run into some problems with IEMs in our young cochlear implant program. Scans with IEMs were usually reported as normal. I was pointing out that IP-I anomaly was different than "Mondini deformity" but my remarks were not taken seriously. Prof. Ergin Turan, M.D. (our senior professor in the implant team), always told me to ask the opinion of radiologists to make the correct diagnosis. Therefore, I decided to make a study to show the difference between them and to send the study to

Laryngoscope, the journal which Prof. Turan had subscription at that time (I was hoping he would believe me if the article was published). After he saw the paper published in *Laryngoscope*, he was impressed with my contribution and honored me saying that "From now on you will be the only one who will operate on IEM's to accumulate the data in one hand and publish it accordingly." Whenever he had patients with IEMs, he handed them to me for surgery. This honored me greatly and I strongly thank Prof. Turan for urging me into this field and fully supporting me in this area. This was the beginning of my journey with IEMs, which would last during all my academic career.

The main outcome of this paper was to create a platform "to talk the same language all over the world." In meetings I have seen people talk about "small" and "big cochleae" or use names with no meaning to the audience. If we do not have an accepted classification, presentations and scientific work will not be understood. Classification of IEMs is important for otolaryngologists, audiologists, radiologists, geneticists, and neurosurgeons dealing with Auditory brainstem implantation (ABI). After our initial classification, it was updated in 2010 and finalized in 2013. Since 2013 there has been no more new anomalies; therefore, the time finally arrived to write a book!

We have been examining some patients with no inner ear development or cochlear nerve, and nothing could be done at the beginning for these patients. After the introduction of ABI surgery in children by Prof. Vittorio Colletti, M.D., in 2001, in 2003 we seriously thought about the first ABI candidate. At that time, there was not sufficient information in the world about ABI in children. When Prof. Richard Ramsden, M.D., started pediatric ABI surgery in Manchester, with whom we communicated frequently about the indication, we decided to start ABI in 2006. The first three children were operated together with Prof. Colletti in Hacettepe University. So far, we have done 125 pediatric ABI surgeries in IEMs, and Hacettepe University has become the most experienced clinic in pediatric ABI in the world, providing international support as well. Prof. Burcak Bilginer, M.D., from the Department of Neurosurgery and his team played a major role in this development. I think Prof. Colletti opened an era in the management of severe IEMs, and we believe it is our mission to continue this approach.

In 2006 one of the children that I operated died because of meningitis 3 months after CI surgery. She had gusher during the surgery. I repaired the leak properly, but 3 months later she developed meningitis and in spite of the extraordinary support of our hospital she could not be saved. This had a big impact on me and I designed an electrode with cork like stopper to stop CSF leakage more efficiently. I had already thought about the design before, but this made me go ahead and produce the electrodes with cork stopper to more efficiently manage CSF gusher during surgery. That is the beginning of FORM electrode series.

In 2013, I received a very honoring invitation from the Department of Otolaryngology at Massachusetts Eye and Ear Infirmary at Harvard University to give the "Harold D. Schuknecht Lecture in Otology." Prof. Harold Schuknecht, M.D., is one of the otology legends. Before going to Boston, I requested to investigate the specimens with IEMs during my stay at Harvard University. After my lecture, I investigated some specimens with IEMs and I

noticed an extraordinary finding: the meaning of that finding was not noticed before even though that patient had been published three times before. I told this with great enthusiasm to the doctors around me, but naturally nobody was as interested as me in the subject. After 1 year, I took a sabbatical from my university and went to investigate that specimen. I spent one whole month in Eastern Temporal Bone Bank which consisted of a small room with more than 2000 specimens with different temporal bone pathologies, and at the end, I underlined the pathophysiology of not only IP-II (my main reason to go back to Harvard) but the whole spectrum of IEMs. This will be a great opportunity to geneticists because they will have more chance to understand the etiology of IEM. This in future may develop methods to genetically prevent these malformations.

One of the main reasons to be invited to Harvard was also to share our huge ABI experience with the team there. I gave a lecture on IEMs and outcome of pediatric ABI. During my stay we evaluated possible candidates. I learnt later that three centers in the USA used the ABI data from Verona University (Prof. Vittorio Colletti, M.D.) and Hacettepe University to apply to FDA to get approval to start ABI in children. As Hacettepe Implant Team, we are very honored that after 6 months they had FDA approval for starting ABI in children in the USA.

One of the most important factors of the success of Hacettepe Implant Team is the early setup of audiology unit in our university. Prof. İ. Nazmi Hosal, M.D., performed his residency in the USA in the 1950s and saw the importance of audiology in modern otolaryngology. After returning to Turkey, he set up the audiology department in Hacettepe University in 1967. I think this is the birth of modern otolaryngology in Turkey. This allowed performing a large number of cochlear and brainstem implants because decision-making, programming, and rehabilitation could be done more efficiently with this experienced group. In the audiological management of patients with IEMs, Prof. Gonca Sennaroglu, Ph.D., and Prof. Esra Yücel, Ph.D., and their teams made enormous contributions.

What is next? Classification is very important. If we do not classify phenotypes properly, we cannot notice the difference between them. In recent years I do not see new malformations coming up. This means we may come to the end of new anomalies. During my 1 month stay at Harvard University in 2014, my study shed light on understanding the pathophysiology of IEMs. Geneticists now have a more specific target and if they can identify the genetic defects responsible for a particular pathology it may be possible to prevent certain IEM in future.

Levent Sennaroglu
Ankara, Turkey

Acknowledgments

During my childhood I gained important fundamental behaviors from my family. This had an important influence on my personality as well as my working habits. My grandfather, who had been a teacher for 42 years, demonstrated to me and my brother the importance of education. My grandmother who was a housewife spent a lot of time with us from whom we learnt humane behavior and at the same time received warning if we did something wrong. I learnt the concept of systematic working from my father, who was a civil engineer. This not only helped me in surgery working according to certain principles but in my whole career as well. I wanted to organize and categorize things around me. Because of this systematic approach, I learnt how to keep database for everything and categorize things appropriately. This is the reason for placing different malformations into appropriate places in the classification. My mother, who is the first Turkish woman doctor in Cyprus, was very interested in our education and always created a peaceful atmosphere during our studies. I thank all of them with all my heart for their efforts in initial shaping of my character.

We had a very good education in Cyprus from primary school until the end of the high school. All our teachers built our knowledge brick-by-brick. I remember all of them with great respect.

I would like to thank my teachers in the Department of Otolaryngology in Hacettepe University. Particularly important is Professor İ. Nazmi Hosal who brought modern otolaryngology to Turkey. I learnt basic principles of otolaryngology and particularly otology during my residency and initial years of my practice in Hacettepe University. I started to work in otology with Professor Bulent Gursel and I would like to thank him for encouraging me to continue my studies in otology.

Visiting House Ear Clinic had a dramatic influence on my career. I saw the advantages of working specifically in one topic within otolaryngology. Until that time, I was practicing different aspects of otolaryngology although I was interested in otology. When I saw the House Ear specialists working in otology for more than half a century with tremendous contribution to the literature, I decided to practice only one aspect: otology and neurotology. As a result, I started to see ear-related problems more and recognize repetition of certain disorders. I thank all the specialists in the House Clinic, but particularly Dr. WWM Lo who taught me the radiology of the temporal bone.

Drawings are very important to explain certain ideas. I thank Shamkhal Jafarov who produced all the drawings in the book. He had his otolaryngology training in our department and we have seen his excellent artistic abilities.

I would like to thank my associates in otology, audiology, neurosurgery, and radiology in Hacettepe University. I think it is great to work together in harmony as a team. As Hacettepe Implant Team we organized three international pediatric ABI Consensus Meetings. We keep learning from each other continuously all the time. This is the main reason for being productive in our profession.

Finally, I would like to thank my family. Producing so much information necessitates getting some of the precious time from my family. I hope my children, Selin and Özdemir, understand their father's passion when they get older. Particularly, I would like to thank my wife, Gonca, who showed a great understanding to my work and travel. She is the only one to whom I thanked twice; as a family member and an excellent pediatric audiologist in our team.

Contents

1 Classification of Inner Ear Malformations

Levent Sennaroglu

Inner ear malformations (IEM) are diagnosed more often today. Two important reasons for this are improved imaging quality and increased awareness. There may be surgical difficulties during surgery of IEMs and outcome may not be as good as cochlear implantation in patients with normal anatomy. Therefore, preoperative diagnosis and proper classification is very important for the surgeon, audiologist, and also counseling the family. Universally agreed classification is very important so that we can understand each other better.

Morphologically congenital sensorineural hearing loss (SNHL) can be investigated under two categories. Majority of the congenital hearing loss (80%) are **membranous malformations**. Here the pathology involves inner ear hair cells. There is no gross bony abnormality and therefore, in these cases, high resolution computerized tomography (HRCT) and magnetic resonance imaging (MRI) of the temporal bone reveal normal findings. Remaining 20% have various **bony inner ear malformations** involving the bony labyrinth and therefore, can be radiologically demonstrated by CT and MRI. The latter group involves surgical challenges as well as problems in decision-making. Some cases may be managed by hearing aids. Majority of these patients have bilateral severe to profound hearing loss and are candidates for cochlear implantation. Those cases with severe malformations may require special surgical approaches for implant placement. During cochlear implantation, there may be facial nerve abnormalities, cerebrospinal fluid leakage, electrode misplacement, or difficulty in finding the cochlea itself. During the surgery for IEMs, the surgeon must be ready to modify the surgical approach or choose special electrodes for surgery. Decision-making between cochlear implantation (CI) and auditory brainstem implantation (ABI) may also be challenging in some cases of IEMs. A minority of congenital bony abnormalities can present with pure conductive or mixed-type hearing loss.

It is very important to classify the IEMs properly and have a universally accepted system. There are eight groups of IEMs and a proper universally accepted correct classification is as important as a common language [1]. If we do not have a common language it is very difficult to understand the findings or results of another clinician or a researcher. Universally accepted classification of cochlear malformations is particularly important in the field of cochlear implantation. Otolaryngologists, audiologists, speech and language specialists, geneticists, etc., should be familiar with this system; otherwise, it will be very difficult to understand and compare the outcome after CI surgery in this particular patient group. The author has examined and analyzed temporal bone images of more than 700 subjects with IEMs. Since 2013 there has been no

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_1

different abnormalities necessitating the creation of a new category of IEM. However, it should be noted that recognizing a new kind of IEM which repeats itself will make it necessary to update the present classification scheme.

Jackler et al. [2] provided the first classification of the IEMs where they separated inner ear anomalies into five categories:

1. Michel deformity
2. Cochlear aplasia
3. Common cavity
4. Cochlear hypoplasia
5. Incomplete partition

"Mondini" malformation has been and unfortunately occasionally still used today to define many different IEMs. Carlo Mondini described a very specific pathology consisting of a triad of anomalies [3]. Jackler et al. [2] in their excellent classification were successful in distinguishing common cavity and cochlear hypoplasia cases from "Mondini" malformation. However, their classification was based on polytomography which is a technique completely replaced today by HRCT and MRI of the temporal bone. Over the years with the technical developments in radiology, HRCT and MRI were able to provide better images of the temporal bone abnormalities. In addition, it is very difficult to compare the present-day images with the polytomography images in the paper by Jackler et al. [2]. As a result, Sennaroglu and Saatci [4] refined this classification in 2002 by defining the radiological features of two completely different types of incomplete partition (IP) anomalies of the cochlea, as IP-I and IP-II. Later the cochlear pathology observed in X-linked deafness has been recognized as the third type of incomplete partition [5, 6]. Over the years four cochlear hypoplasia subtypes were clearly identified and added to the classification [1, 7]. It has been observed that cochlear aperture (bony canal for cochlear nerve) may be hypoplastic or aplastic in an otherwise completely normal cochlea and may have a serious effect on the outcome with CI surgery. With the addition of cochlear aperture abnormalities, classification system has moved one more step to become more precise [8, 9]. According to the latest classification, there are eight categories for IEMs [1]:

1. Complete Labyrinthine Aplasia (Michel Deformity)
2. Rudimentary Otocyst
3. Cochlear Aplasia
4. Common Cavity
5. Cochlear Hypoplasia
6. Incomplete Partition Anomalies
7. Enlarged Vestibular Aqueduct
8. Cochlear Aperture Abnormalities

In this chapter you will also find updated classification of Cochlear Nerve Abnormalities.

IEMs were grouped into categories and subgroups according to their radiological appearance. Within a group, they usually demonstrate similar clinical features. It is very important to remember their clinical behavior when a clinician encounters a specific image. The surgeon may then expect specific problems during surgery or an audiologist may observe a hearing loss pattern. This forms the basis of the present classification system. It is natural to have certain variability within an individual group.

As can be seen, we have used the initial framework. It is very important not to create a lot of new names and categories. Placement of a new category into appropriate place is very important. We tried to avoid creating new names but placing the new anomalies into appropriate subcategories. Only when absolutely necessary a new category is created as in the situation of "cochlear aperture abnormalities."

It is very important to avoid names like "Mondini" but rather to use descriptive names. This will help achieving the goal of universal language.

One of the missing elements in previous classification systems was measuring the dimensions. This is necessary to differentiate between incomplete partition anomalies and cochlear hypoplasia. Complete labyrinthine aplasia, rudimentary otocyst, common cavity, and cochlear aplasia can be diagnosed by visual observation of the image. IP-II and IP-III, together with CH-I and CH-IV,

can be diagnosed by their shape; therefore, measurement is not necessary. However, IP-I can be confused with CH-II. Proper diagnosis is necessary for choosing the correct length of the electrode. CH-III may be confused with a normal cochlea. In the present system cochlear dimensions necessary for the distinction between the mentioned abnormalities are also provided (see Chap. 26).

The classification system of Jackler et al. [2] was based on embryogenesis and developmental arrest where they tried to explain each malformation by arrest of development at a different stage of embryogenesis. Therefore, they concluded that arrest of development at certain periods produced different IEMs. The author after investigating images of more than 700 patients with IEMs and observing repetition of certain malformations came to believe that majority of inner ear anomalies may be a result of genetic etiology. Sometimes there are two different abnormalities on either side making environmental factors responsible for developmental error. In other words, developmental arrest is more likely in asymmetric anomalies.

1.1 Normal Cochlea

Before discussing the characteristics of IEMs, the clinician has to be familiar with the normal anatomy of the cochlea as seen on HRCT and MRI of the temporal bone. Schematic representation of normal cochlea and cochlear anomalies is given in Fig. 1.1. Corresponding HRCT images of the inner ear malformations are provided in Figs. 1.2, 1.3, 1.4, 1.5, 1.6, 1.7, 1.8, 1.9, 1.10, 1.11, 1.12, 1.13, 1.14, 1.15, and 1.16.

Cochlea has 2½ or 2¾ turns [6]. Midmodiolar section is the most important section to determine the normal cochlear architecture and incomplete partition anomalies (Fig. 1.2a). Midmodiolar view demonstrates the modiolus as a quadrangular or pentagonal structure in the center of the basal turn and between the basal and middle turns of the cochlea [10]. Interscalar septa are thicker partitions between the inner wall of the cochlea and the modiolus, which appear to separate the normal cochlea into 2½ or 2¾ turns: the basal, middle, and apical turns. The cochlear aperture (bony canal for cochlear nerve) is the central bony passage at the base of the modiolus transmitting the cochlear nerve and blood vessels.

The section inferior to the midmodiolar view passes through the area of the round window niche (Fig. 1.2b). This section shows the basal, middle, and apical cochlear turns. Basal turn is in continuity at this section. It is important to see the interscalar septum between the middle and apical turns.

MRI provides additional information. Modiolus can be seen in more detail in Fig. 1.13a. It is possible to distinguish between scala tympani and vestibuli. Most important is the demonstration of the cochlear nerve (Fig. 1.13a). Therefore, they are complementary in the correct diagnosis of IEMs and planning the appropriate treatment. It is very important to see the cochlear nerve on direct parasagittal section perpendicular to the IAC (Fig. 1.13b). Normally there are four different nerves in the IAC, and the cochlear nerve is located in the anterior inferior part of IAC.

1.2 Cochlear Malformations

1.2.1 Complete Labyrinthine Aplasia (CLA, Michel Deformity)

Complete Labyrinthine aplasia is the absence of the cochlea, vestibule, semicircular canals, vestibular and cochlear aqueducts (Fig. 1.3a–c). The petrous bone may be hypoplastic whereas the otic capsule may be hypoplastic or aplastic [11]. In the majority of patients, the internal auditory canal (IAC) consists only of the facial canal and the labyrinthine, tympanic, and mastoid segments of the facial nerve can be followed in the temporal bone. In some patients, however, it may not be possible to observe the facial canal in the temporal bone in spite of normal facial functions. Cochleovestibular nerve is absent. Middle ear ossicles are usually present.

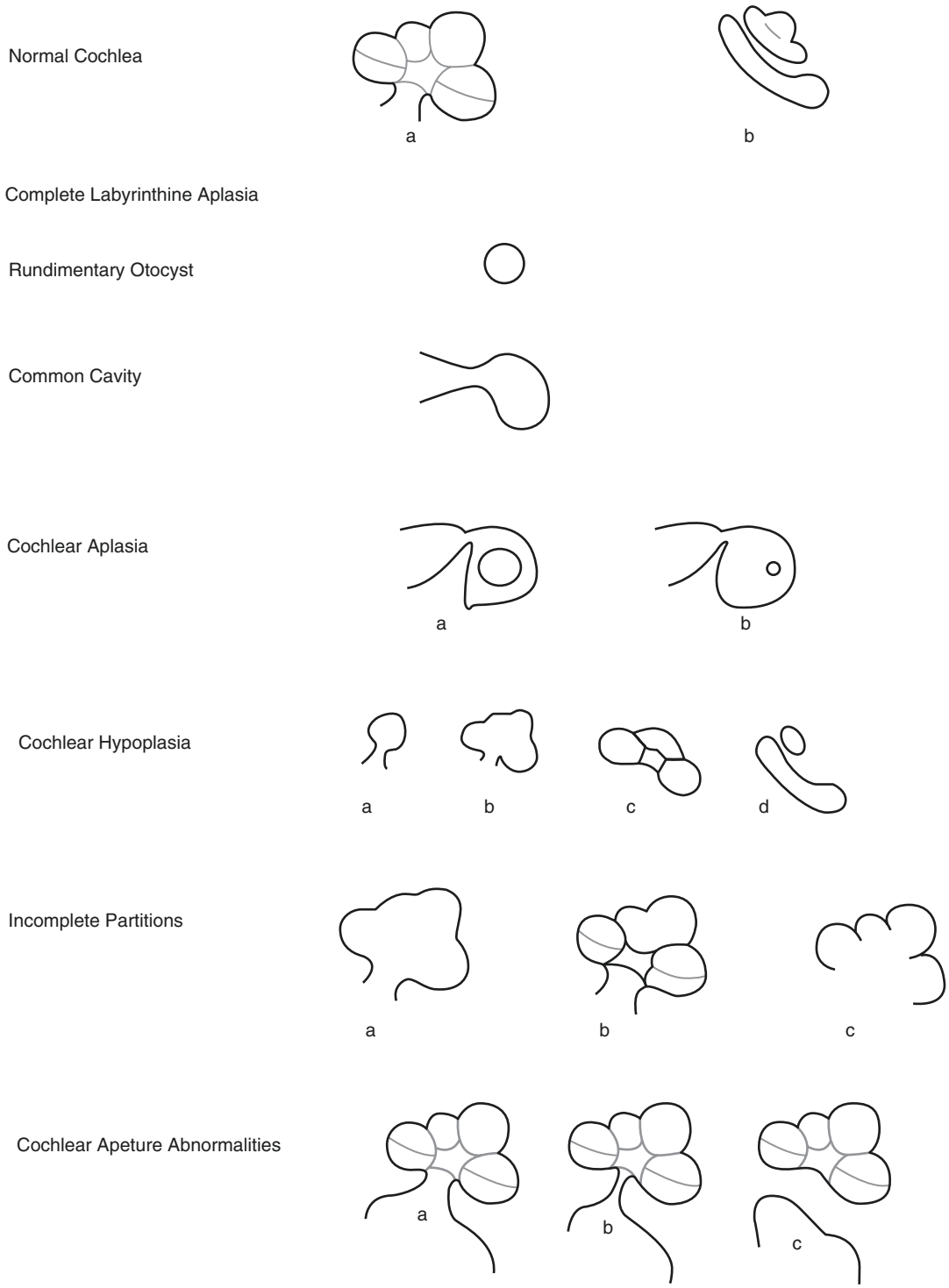


Fig. 1.1 Schematic representation of normal cochlea and inner ear malformations

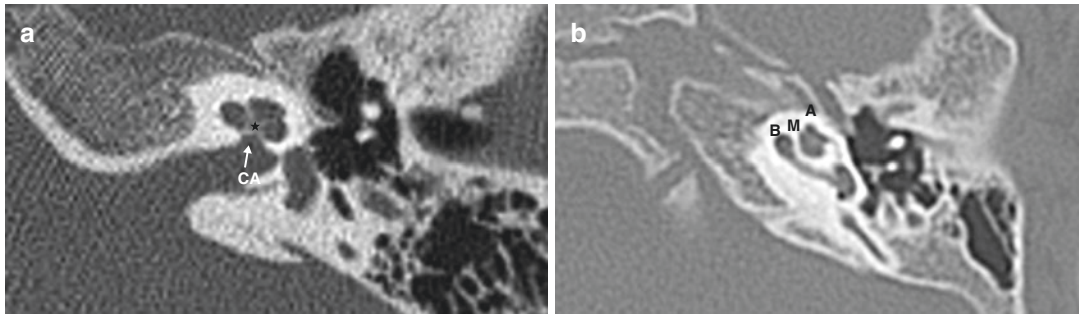


Fig. 1.2 Normal cochlea. (**a**) Midmodiolar section (black star = modiolus, CA = cochlear aperture), (**b**) Section through round window niche (B = Basal, M = Middle, A = Apical turns)

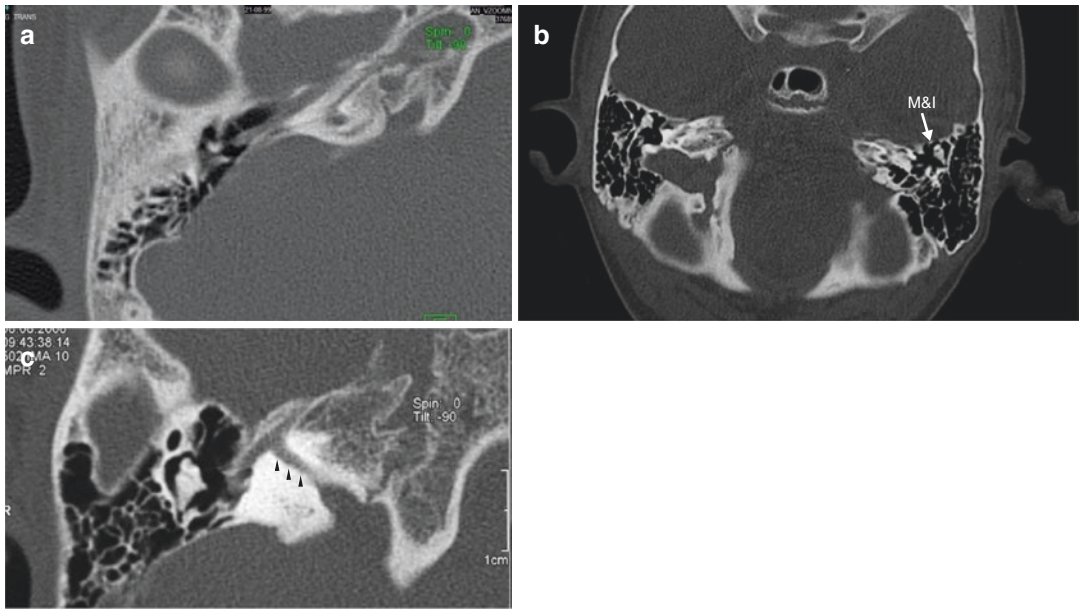


Fig. 1.3 Complete Labyrinthine aplasia (CLA). (**a**) CLA with aplastic petrous bone, (**b**) CLA without otic capsule, (**c**) CLA with otic capsule (arrowheads = labyrinthine segment of facial nerve)

According to radiological findings [7], three subgroups of CLA are present:

1. **CLA with Hypoplastic or Aplastic Petrous Bone**

   In these cases CLA is accompanied by hypoplasia or aplasia of the petrous bone. Middle ear may be adjacent to posterior fossa (Fig. 1.3a).
2. **CLA Without Otic Capsule**

   In this group of CLA, formation of the petrous bone is normal, but the otic capsule is hypoplastic or aplastic. Otic capsule has three layers. According to Donaldson [12], the inner endosteum receives its vascular supply from the IAC, and middle enchondral and outer periosteal layers get their vascular supply from the middle ear mucosa. CLA without otic capsule may be due to abnormal vascular supply from the IAC and middle ear, resulting in the absence of all three layers of the otic capsule (Fig. 1.3b).
3. **CLA with Otic Capsule**

   Formation of the petrous bone and the otic capsule is normal. It can be speculated that vascular supply from the middle ear is normal as

the otic capsule normally develops. The pathology may be due to absent vascular supply from IAC, so that only membranous labyrinth and endosteum are completely absent. Facial nerve canal can be seen (Fig. 1.3c). Only in this group of CLA with otic capsule development the facial canal is in its normal location. This shows that otic capsule formation is essential for the facial canal to obtain its normal position.

Management

It is not possible to perform cochlear implant (CI) surgery in these children as there is no inner ear development. Auditory brainstem implantation (ABI) is thus the only surgical option for hearing habilitation [13, 14].

1.2.2 Rudimentary Otocyst

A rudimentary otocyst is used to describe incomplete millimetric representations of the otic capsule (round or ovoid in shape) without an IAC (Fig. 1.4). Parts of the semicircular canals may accompany rudimentary otocyst. This pathology represents an anomaly between a complete labyrinthine aplasia (CLA) and common cavity (CC). In CLA, there is no inner ear development, while in CC, there is an ovoid or round cystic space instead of a separate cochlea and vestibule. The CC communicates with the brainstem via the nerves in the IAC. The rudimentary otocyst is a few millimeters in size without the formation of an IAC.

The inner ear is in the form of an otocyst (otic vesicle) between the third and fourth week [7]. The insult probably occurs at the beginning of the formation of the otocyst and results in rudimentary otocyst deformity.

Management

The fact that there is no connection between the otocyst and the brainstem makes this group a contraindication to CI surgery. As a result, these patients are candidates for ABI.

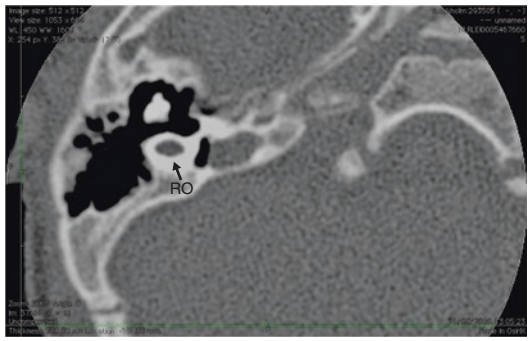


Fig. 1.4 Rudimentary otocyst (RO)

1.2.3 Cochlear Aplasia

Cochlear aplasia is the absence of the cochlea. There are two subgroups according to accompanying vestibular system:

1. **Cochlear aplasia with normal vestibule (CANV):** vestibule and semicircular canals are normally developed (Fig. 1.5a).
2. **Cochlear aplasia with a dilated vestibule (CADV):** vestibule and semicircular canals show dilatation (Fig. 1.5b). This must be

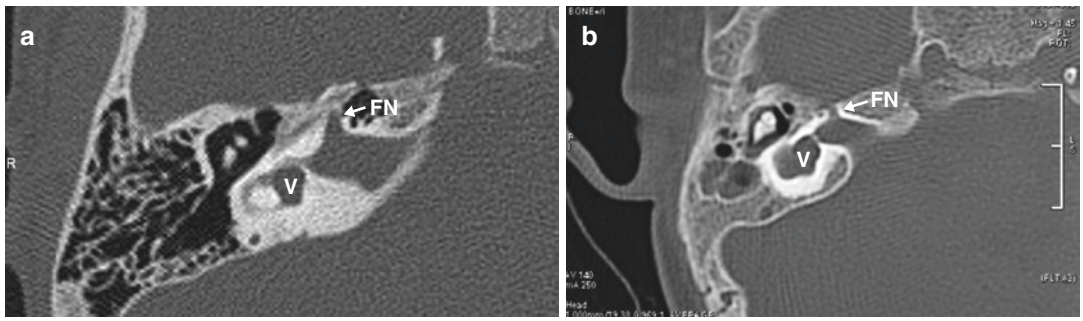


Fig. 1.5 Cochlear aplasia. (**a**) Cochlear aplasia with normal vestibule, (**b**) Cochlear aplasia with a dilated vestibule (V = Vestibule, FN = anteriorly dislocated labyrinthine segment of the facial nerve)

differentiated from a common cavity (CC) deformity.

IAC development is normal. Normally cochlea occupies anterolateral part and vestibular system is on the posterolateral part of IAC. In cases with cochlear aplasia, cochlea is absent in its anterolateral position.

The labyrinthine segment of the facial nerve is anteriorly displaced and occupies the normal location of the cochlea. In CC cochlear implantation can be done, if cochleovestibular nerve (CVN) is present. **However, CI surgery should not be done in CADV**. In some patients, it may be very difficult to distinguish between these entities. Audiological findings are very important in choosing the right method of implantation.

CANV is usually symmetrical. The fact that similar findings are present in different patients suggests genetic etiology. In CADV, however, asymmetric development may be present; pathology may be due to genetic or environmental factors. Otic capsule development is always normal.

After the development of the otic vesicle at the end of the fourth week, the membranous labyrinth develops in three areas: cochlea, vestibule, and endolymphatic duct [7]. Cochlear aplasia is the absence of the cochlear duct, where vestibular and endolymphatic structures may develop normally. The time of the insult must be around the fifth week. It is possible that genetically cochlear development may be defective right from the start of embryological development.

As there is no inner ear development, ABI is the only feasible surgical option to provide hearing in children with cochlear aplasia.

1.2.4 Common Cavity

A **common cavity** is defined as a single chamber, ovoid or round in shape, representing cochlea and vestibule (Fig. 1.6). Theoretically, this structure has cochlear and vestibular neural structures. There may be accompanying semicircular canals (SCC) or their rudimentary parts. IAC usually enters the cavity at its center. Cases with vestibular dilatation are occasionally termed as "vestibular common cavity"; however, this is not a correct term.

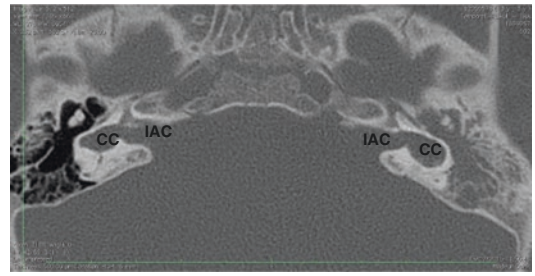


Fig. 1.6 Common cavity (CC) (IAC = internal auditory canal)

Common cavity (CC) needs to be differentiated from cochlear aplasia with dilated vestibule [6]. **Cochlear aplasia with dilated vestibule (CADV)** (Fig. 1.5b) has a dilated vestibule and semicircular canals at the posterolateral part of the IAC fundus, which is their usual location. External outline resembles the normal labyrinth. The vestibule is at its expected location. The accompanying SCCs may be enlarged or normal. A **CC** (Fig. 1.6), on the other hand, is an ovoid or round structure. SCCs or their rudimentary parts may accompany a CC. The IAC is usually deformed; narrow or enlarged, usually angled posteriorly. It enters the cavity usually at its center. The location of a CC may be anterior or posterior to the normal location of the labyrinth. Usually it is posteriorly located. It is very important to differentiate these malformations from each other, because cochlear implantation may provide hearing in a CC, whereas in CAVD, no functional stimulation will occur with CI. In spite of this, it may sometimes be difficult to differentiate between the two malformations.

The nerve entering the CC should be termed as cochleovestibular nerve (CVN). Theoretically, CVN contains cochlear and vestibular nerve fibers. CVN has to be demonstrated by 3 T MRI in candidates undergoing evaluation for CI candidacy. With the present radiological investigations, it is impossible to determine the amount of cochlear fibers within the CVN. Audiological evaluation is very

important to determine hearing present in CC which indirectly gives an estimate of the cochlear fibers within the CVN. If a behavioral audiometric response or language development is present with hearing aid use, it can be assumed that a meaningful population of cochlear fibers exists and the patient may benefit from a CI. If the CVN cannot be demonstrated with MRI or there is a very narrow or long IAC, where the presence of cochlear fibers is questionable, an ABI may be a more appropriate option from the outset. As the postoperative hearing cannot be accurately predicted before CI surgery, it is advisable to counsel the family that contralateral ABI may be necessary in case of limited language development with CI. This decision should be done as early as possible.

CC contains cochlear and vestibular neural elements. This represents development arrest before there is a clear differentiation into cochlea and vestibule: it is in between rudimentary otocyst and cochlear aplasia, and usually occurs around the fourth to fifth week [7].

At the time of insult, the CC is only millimetric in size, as a developed otocyst. The CC may have small or large dimensions: usually, a CC with a diameter of 1–3 cm is encountered. This shows that its capacity to differentiate into cochlea and vestibule may terminate but it can still enlarge; so a CC larger than an initial otocyst may be encountered. IAC may be normal or narrow in a large CC. It appears that there is no relationship of the size of the IAC (length and width) and the size of the CC.

If a CVN is present together with audiological response CI is the preferred method. If CVN is absent, ABI is the only option.

Cochlear Hypoplasia and Incomplete Partition Anomalies

In these groups of malformations there is **a clear differentiation between cochlea and vestibule**. **Cochlear hypoplasia** is the group of malformations where external dimensions of the cochlea are smaller than normal. The term **incomplete partition** anomalies is used if external dimensions of the cochlea are normal, but there are various internal architecture defects.

1.2.5 Cochlear Hypoplasia

In this deformity, there is clear differentiation between cochlea and vestibule. **Cochlear hypoplasia (CH)** represents a group of cochlear malformations with external dimensions less than those of a normal cochlea with various internal architecture deformities. In smaller cochlea, it is usually difficult to count the number of turns with CT and/or MRI. But the definition "cochlea with 1.5 turns" should be used for CH (particularly type III), rather than for IP-II cochlea. **Four** different types of CH have been defined:

1.2.5.1 Types of Cochlear Hypoplasia (CH)

1. **CH-I (Bud-like cochlea)**
   Cochlea is like a small bud, round or ovoid in shape, arising from the IAC (Fig. 1.7a). Internal architecture is severely deformed; modiolus and interscalar septa cannot be identified.
2. **CH-II (Cystic hypoplastic cochlea)**
   Cochlea has smaller dimensions with defective modiolus and interscalar septa, but with normal external outline (Fig. 1.7b). There may be complete absence of modiolus creating a wide connection with the IAC, making gusher and misplacement of CI electrode into IAC possible. The vestibular aqueduct may be enlarged and the vestibule may be dilated.
3. **CH-III (Cochlea with less than two turns)**
   Cochlea has fewer turns (i.e., less than two turns) with a short modiolus. The overall length of the interscalar septa is reduced. The internal (modiolus, interscalar septa) and external outline are similar to that of a normal cochlea, but the dimensions are smaller and number of turns are fewer (Fig. 1.7c). Most extreme variant consists of only a small basal turn and a modiolus, where middle and apical turns are absent. The vestibule and the semi-

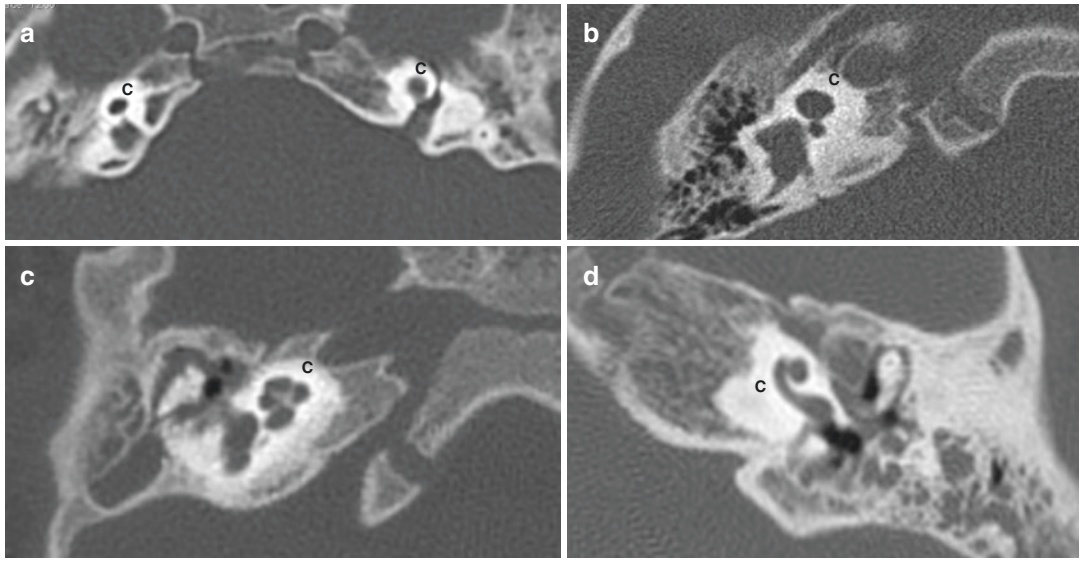


Fig. 1.7 Cochlear hypoplasia (CH). (**a**) CH-I (Bud type) with absent internal architecture (modiolus and interscalar septa), (**b**) CH-II (Cystic type) with defective internal architecture (modiolus and interscalar septa), (**c**) CH-III with smaller dimensions but normal internal architecture (modiolus and interscalar septa), (**d**) CH-IV with normal basal turn but small middle and apical turns (C = cochlea)

circular canals are usually hypoplastic. The cochlear aperture may be hypoplastic or aplastic.

4. **CH-IV (Cochlea with hypoplastic middle and apical turns)**

   Cochlea has a normal basal turn, but middle and apical turns are severely hypoplastic and located anteriorly and medially rather than in their normal central position (Fig. 1.7d). The labyrinthine segment of the facial nerve is usually located anterior-superior part of the cochlea rather than in its normal posterior location [15] (see Chaps. 13 and 26).

In CH-I and CH-II there is arrested development of the internal architecture in addition to a small-sized cochlea. In CH-I cochlear duct length must have stopped earlier than normal. Defective modiolar and endosteal development is most probably due to defective vascular supply from the IAC. The main cochlear artery must be defective, resulting in defective endosteal development with an absent modiolus and ISS.

CH-II is better developed than CH-I. The outline of CH-II resembles that of a normal cochlea. It is round or ovoid with a partial modiolar defect. The modiolar base is normal, showing that only the internal radiating arteriole from the main cochlear artery may be defective, while the cochlear ramus of the vestibulocochlear artery supplies the base of the modiolus.

Most probably developmental arrest of membranous labyrinth in CH-III occurs between 6 and 8 weeks, resulting in a cochlea whose dimensions are smaller than normal, with normal internal architecture. In CH-IV arrest in the membranous labyrinth must be between tenth and 20th week, after the basal turn reaches full size, but before the middle and apical turns enlarge to their normal dimensions.

Management

Decision-making in patients with cochlear hypoplasia may be challenging. They may present with a range of different thresholds on audiometric testing. Decision-making about the amplification options may be difficult, particularly in patients with a hypoplastic cochlear nerve. Patients with mild to moderate SNHL can be habilitated with hearing aids and have near nor-

mal language development. The majority of CH patients have severe to profound hearing loss where a CI would be a reasonable option, if they have a cochlear nerve. Some patients have cochlear aperture aplasia with cochlear nerve aplasia and thus, an ABI would be the best hearing habilitative option. Other patients with cochlear hypoplasia have hypoplastic cochlear nerves. The best option in these cases is to perform CI in the side with better cochlear nerve. If there is limited hearing and language development, an ABI should be considered for the contralateral side. A number of these patients may be candidates for simultaneous CI and ABI.

Some cases of hypoplasia (particularly hypoplasia type IV) may have pure conductive or mixed hearing loss in which the conductive component is due to stapedial fixation. They may benefit from stapedotomy.

1.2.6 Incomplete Partition Anomalies of the Cochlea

Incomplete Partition Anomalies represent a group of cochlear malformations with normal external dimensions and various internal architecture defects. Incomplete partitions constitute 37% of inner ear malformations according to the database of Hacettepe University Department of Otolaryngology. There are three different types of incomplete partition groups according to the defect in the modiolus and the interscalar septa.

1.2.6.1 Types of Incomplete Partition Groups

1. **Incomplete partition type I (IP-I)**

This is the type of the cochlea described in "**cystic cochleovestibular malformation**" by Sennaroglu and Saatci [4] in 2002. They represent approximately 11.5% of inner ear malformations. In this anomaly there is a clear differentiation between cochlea and vestibule. Cochlea is located in its usual location in the anterolateral part of the fundus of the IAC and lacks the entire modiolus and interscalar septa (Fig. 1.8a), giving the appearance of an empty cystic structure. External dimensions (height and length) of an IP-I cochlea are similar to normal cases [16]. Cochlea is accompanied by an enlarged, dilated vestibule (Fig. 1.8b). Vestibular aqueduct enlargement is very rare. There may be a defect between the IAC and the cochlea due to developmental abnormality of the cochlear aperture and absence of the modiolus, and CSF may completely fill the cochlea.

A recent histopathology study suggests that IP-I may be due to endosteal development abnormality as a result of defective vascular supply coming from the IAC [7].

The majority of IP-I patients have severe to profound SNHL. They are almost always candidates for CI if they have a CN. In case of CN aplasia in IP-I, an ABI is indicated. Four patients with IP-I and an aplastic CN have received ABI in our department.

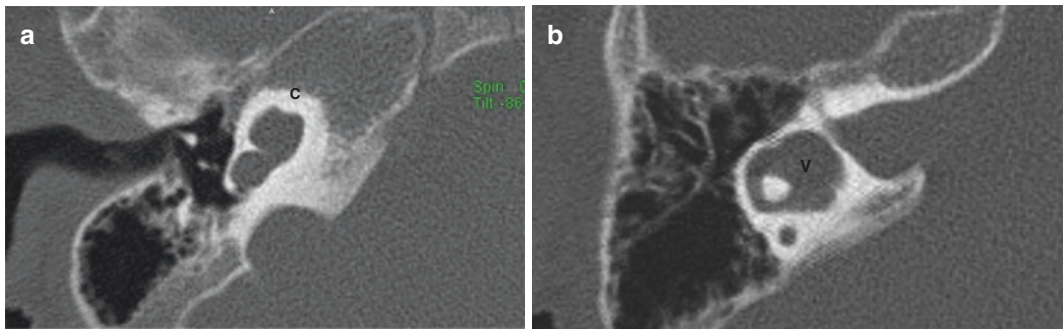


Fig. 1.8 Incomplete partition type I. (**a**) Cochlea (C) without modiolus and interscalar septa. (**b**) enlarged, dilated vestibule (V)

Recurrent meningitis can occur in IP-I patients even prior to their CI surgery or in their non-operated ear. This due to defective stapes footplate and CSF filling the cochlea is easily infected during an attack of otitis media. This is very characteristic for IP-I. Spontaneous CSF fistula and recurrent meningitis can be seen although less frequently in cochlear hypoplasia type II. This is because both IP-I and CH-II have endosteal developmental anomaly leading to defective footplate development.

All patients with IP-I and recurrent meningitis who have normal tympanic membranes but fluid filling the middle ear and mastoid should have an exploration of the middle ear with special attention to the stapes footplate.

2. **Incomplete partition type II (IP-II)**

In IP-II, the apical part of the modiolus is defective, giving rise to a cystic cochlear apex with normal external dimensions (Fig. 1.9a). This anomaly was originally described by Carlo Mondini and together with a minimally dilated vestibule and an enlarged vestibular aqueduct (EVA) (Fig. 1.9b) constitutes the triad of the **Mondini Deformity**. The term "Mondini" should be used only if the above-mentioned triad of malformations is present [4, 6, 17, 18]. The apical part of the modiolus and the corresponding interscalar septa are defective, giving the apex of the cochlea a cystic appearance due to the confluence of middle and apical turns. The external dimensions of the cochlea (height and diameter) are similar to that seen in normal cases [16]. As already pointed out by Sennaroglu and Saatci, it is not correct to define this anomaly as a cochlea with 1.5 turns [16]. The term "cochlea with 1.5 turns" is more appropriate for cochlear hypoplasia (particularly CH-III). They represent 22.5% of IEMs in our database.

A recent study on histopathology demonstrated that modiolar defects may be due to high CSF pressure transmission into the inner ear as a result of EVA [7]. An enlarged endolymphatic sac and duct appears to be the genetic abnormality that is causing other abnormalities allowing high CSF pressure to be transmitted into the inner ear. This results in a mild dilatation in the walls of the vestibule. However, no hydropic changes were observed in the endolymphatic space. Depending on the severity and timing of the insult, the pathology may stay at this stage and cause EVA only, or with the transmission of CSF pressure into the cochlea, it may cause a spectrum of anomalies ranging from scala vestibuli dilatation, scala communis, superior (cystic apex), to partial, subtotal, and in some cases complete modiolar defects [7]. The high pressure in the SV causes bulging of the ISS upwards. This is a constant finding in all cases, showing that cochlear pathology may be the result of high pressure in the SV and that it happened during the developmental phase; otherwise high pressure would have caused fracture of the osseous spiral lamina. If there is higher pressure, it is natural to expect

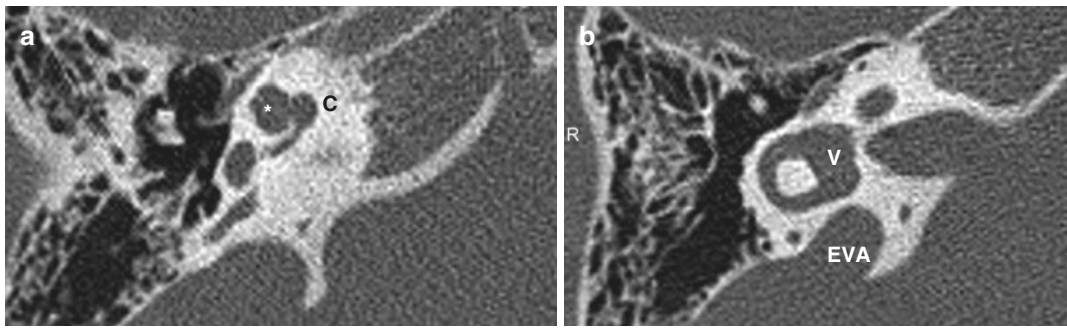


Fig. 1.9 Incomplete partition type II. (**a**) A cystic cochlear apex (white star) with normal external dimensions, (**b**) minimally dilated vestibule (V) and an enlarged vestibular aqueduct (EVA)

more destruction at the upper, and possibly the lower part of the modiolus. This is the reason for the CSF oozing and gusher sometimes observed during CI surgery.

Audiological Findings

These patients do not have a characteristic hearing level, as their audiometric threshold testing varies from normal to profound. The hearing loss can be symmetric or asymmetric, but it is usually progressive. It is also possible to have sudden SNHL. Audiological findings may be due to pressure transmission via EVA into the inner ear.

Management

At a young age, these patients may have near normal hearing and usually do not require amplification initially. With progressive hearing loss, they become candidates for hearing aid. Usually progression in hearing loss continues, ultimately creating a need for CI at some point in the future. High pulsating CSF pressure may be responsible for the progression of hearing loss. A role for head trauma has been suggested, and these patients are advised to avoid trauma by wearing helmets when playing sports and avoiding contact sports completely.

3. **Incomplete partition type III (IP-III)**

Cochlea in IP-III has interscalar septa but the modiolus is completely absent (Fig. 1.10a). The external dimensions of the cochlea (height and diameter) are not different from the normal (12). IP-III cochlear malformation is the type of anomaly present in X-linked deafness, which was described by Nance et al. [19] for the first time in 1971. Phelps et al. [20] described the HRCT findings associated with this condition for the first time, and this characteristic deformity was included under the category of incomplete partition deformities for the first time by Sennaroglu et al. in 2006 [5].

This anomaly is the rarest form of incomplete partition cases. IP-III constitutes 2.9% of the IEMs in the database in Hacettepe University Department of Otolaryngology.

HRCT demonstrates that in IP-III, the otic capsule around the cochlea is thinner than a normal cochlea and follows the outline of the membranous labyrinth as if it is formed by a thick endosteal layer. Instead of the usual three layers, probably the second and third layers are either absent or very thin. The innermost endosteal layer appears to be thickened without enchondral and outer periosteal layers.

In IP-III there may be mixed type HL or profound SNHL. Conductive component may be due to thin otic capsule. Stapes surgery should be avoided in this group. It may lead to gusher and further SNHL. They have excellent cochlear nerves (Fig. 1.10b). Therefore, ABI is not indicated in this group of incomplete partitions.

Management

Mixed hearing loss gives the impression of stapedial fixation. Stapedotomy results in severe gusher and further SNHL, and thus, should be avoided. Patients with severe HL are candidates for CI.

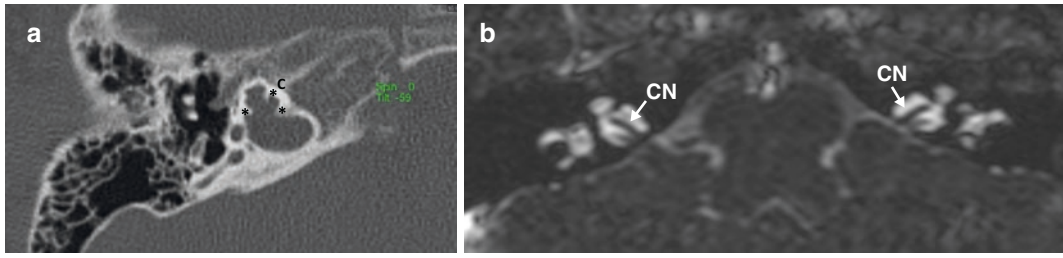


Fig. 1.10 Incomplete partition type III. (**a**) Cochlea (C) with interscalar septa (black stars) and absent modiolus. (**b**) Bilateral normal cochlear nerves (CN)

Because of the absent modiolus in IP-III, all patients have severe gusher during CI surgery and there is a chance of electrode misplacement into IAC. The position of the electrode should be checked intraoperatively in all cases. Modiolar hugging electrodes have more chance to enter into IAC and they should be avoided in the surgery of IP-III. Spontaneous CSF fistula through the stapes footplate and recurrent meningitis is very rare in IP-III in spite of high volume CSF leak during CI surgery. This is most probably due to normal endosteal development (hence a normal footplate) in IP-III. This is because in IP-III pathology is in the outer two layers of the otic capsule and endosteum is normal. Therefore, a defect in the footplate is very unlikely.

1.2.7 Enlarged Vestibular Aqueduct (EVA)

This describes the presence of an enlarged vestibular aqueduct (i.e., the midpoint between posterior labyrinth and operculum is larger than 1.5 mm) in the presence of a normal cochlea, vestibule, and semicircular canals (Fig. 1.11a). Audiological presentation and management is similar to that of IP-II. Recently Sennaroglu et al. [1] also added the importance of vertical measurement of the size of the EAV in addition to axial dimensions (Fig. 1.11b).

Management

Initially they may have near-normal hearing. With progressive hearing loss, hearing aid use may be necessary. Hearing loss is progressive and they may be candidate for CI later in their life. As in IP-II high pulsating CSF pressure may be responsible for the progression of hearing loss. These patients are also advised to avoid head trauma and wear helmets during sports.

1.2.8 Cochlear Aperture Abnormalities

The **cochlear aperture** (CA), **cochlear fossette**, or **bony cochlear nerve canal** transmits the cochlear nerve from the cochlea to IAC. This can be visualized in the midmodiolar view as well as coronal sections on HRCT (Fig. 1.2a).

The cochlear aperture is considered hypoplastic (Fig. 1.12a) if the width is less than 1.4 mm [21]. The CA is considered to be aplastic when the canal is completely replaced by bone or there is no canal on midmodiolar view (Fig. 1.12b).

CA aplasia is typically accompanied by cochlear nerve aplasia. CN may be hypoplastic or aplastic when CA is hypoplastic. CA hypoplasia and aplasia can also be observed in a normal cochlea.

CA abnormalities may be accompanied by a narrow IAC on HRCT. IAC is considered narrow if the width of the midpoint of the IAC is smaller than 2.5 mm (Fig. 1.12c). Narrow IAC can accompany other malformations or with a normal cochlea. In cases of narrow IAC, MRI should be obtained to demonstrate if CN is normal, aplastic, or hypoplastic. Axial and sagittal oblique high T2-weighted (i.e., CISS, FIESTA, etc.)

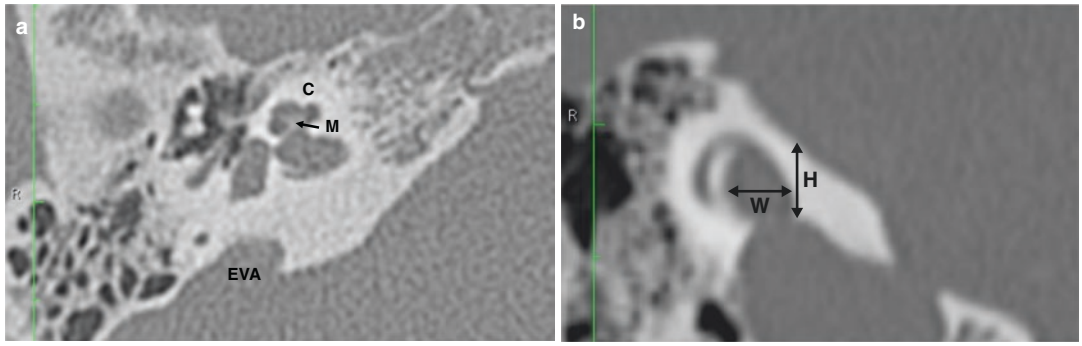


Fig. 1.11 Enlarged vestibular aqueduct. (**a**) Enlarged vestibular aqueduct (EVA) with normal cochlea (C) where modiolus (M) is normal. (**b**) Height (H) and width (W) of the vestibular aqueduct on coronal section

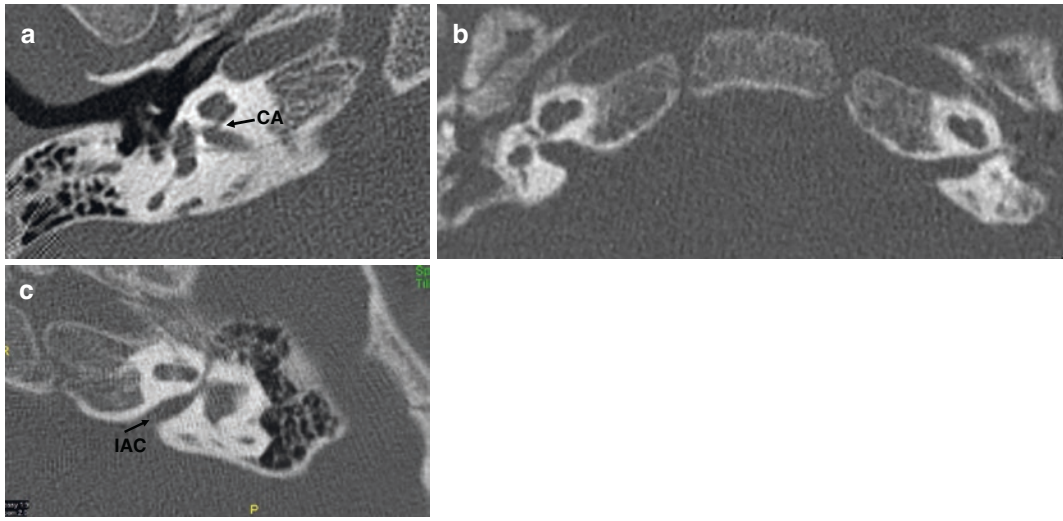


Fig. 1.12 Cochlear aperture (CA). (**a**) Hypoplastic, (**b**) aplastic, (**c**) narrow internal auditory canal (IAC)

images are necessary for this purpose. On sagittal oblique MR sections, four distinct nerves can be visualized in the IAC. In CN aplasia, no nerve can be identified in the anterior inferior part of the IAC. Please refer to the next section for a more detailed discussion on cochlear nerve abnormalities.

Audiological Findings

Severe to profound SNHL is usually present. As the cochlea is normal, otoacoustic emissions (OAE) may be present and the child may pass newborn hearing screening if automated auditory brainstem response (ABR) is not obtained. Their hearing loss is typically discovered later on in childhood based on the family's concerns of lack of sound awareness and language development. If the newborn screening protocol involves OAE and automated ABR, this malformation can be diagnosed during infancy. Diagnostic audiological evaluation will reveal profound hearing loss.

Management

Hearing aids usually do not provide sufficient amplification in patients with CA hypoplasia and aplasia. In patients with bilateral hypoplastic CA with hypoplastic cochlear nerve, hearing aid trial is necessary. If this does not provide adequate functional hearing, these patients usually become candidates for CI. The family should be counseled that if CI does not provide sufficient hearing in terms of auditory perception, contralateral ABI may be necessary to achieve improved audiologic and language outcomes.

In CA aplasia, ABI is indicated as first-line therapy.

1.3 Cochlear Nerve Abnormalities

The classification of cochleovestibular nerve is also important in the management of IEMs [1]. Sennaroglu L proposes the following classification for CN and CVN abnormalities in IEMs.

1. **Normal cochlear nerve (CN)**

   It is important to trace the CN until it enters the cochlea 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 Casselmann et al. [22] on parasagittal view the size of the CN is larger than the ipsilateral FN.

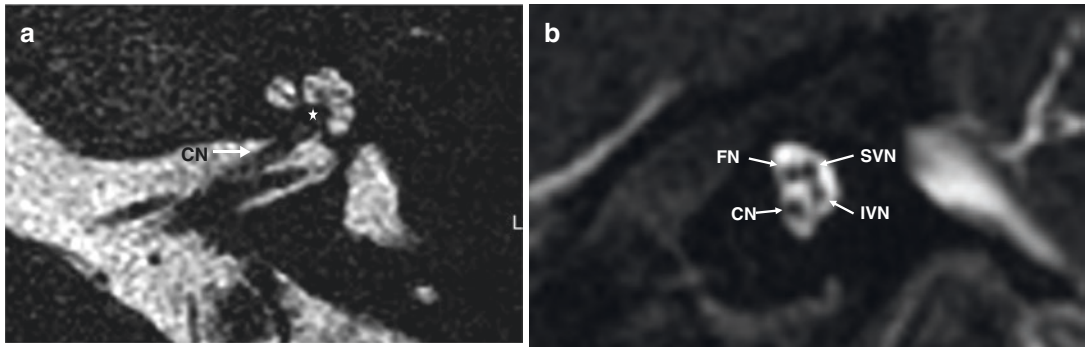


Fig. 1.13 Magnetic resonance imaging of cochlea and cochlear nerve. (**a**) Axial section (CN = cochlear nerve, white star = modiolus). (**b**) Parasagittal section perpendicular to the IAC (CN = cochlear nerve, FN = facial nerve, SVN = superior vestibular nerve, IVN = inferior vestibular nerve)

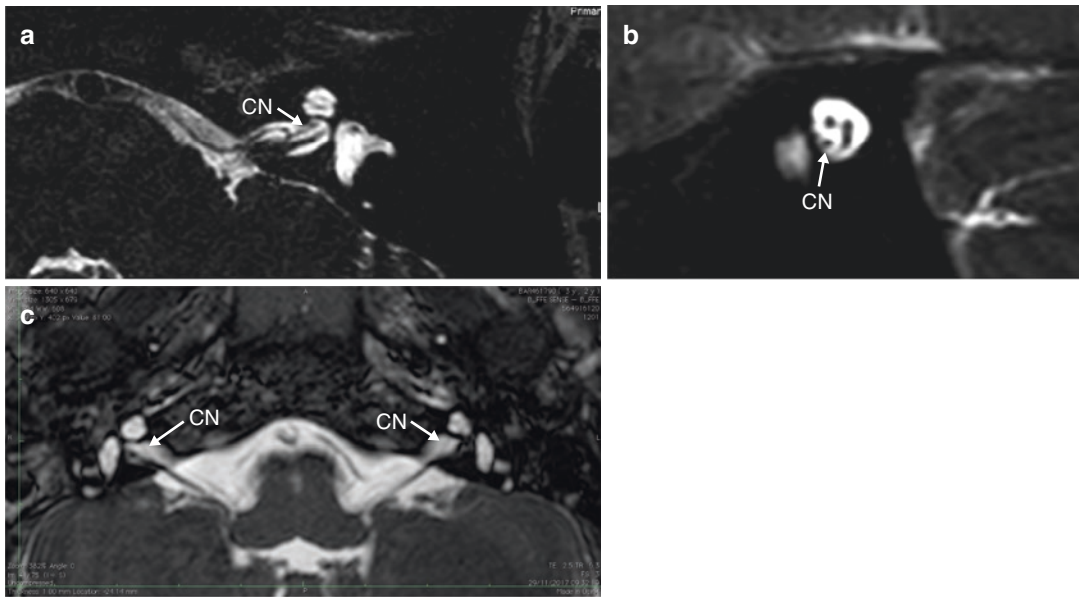


Fig. 1.14 Hypoplastic cochlear nerve (CN). (**a**) Separate CN but the size is less than ipsilateral normal facial nerve. (**b**) On direct parasagittal imaging CN appears to have a smaller diameter than ipsilateral FN. (**c**) Bilateral extremely thin and hardly visible CN

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). On direct parasagittal imaging CN appears to have a smaller diameter than ipsilateral FN (Fig. 1.14b).

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 extremely thin and hardly visible and on axial MRI it can be scarcely followed into the cochlea (<10% of the normal CN or ipsilateral FN) (Fig. 1.14c). These are the cases where a decision between CI and ABI has to be made.

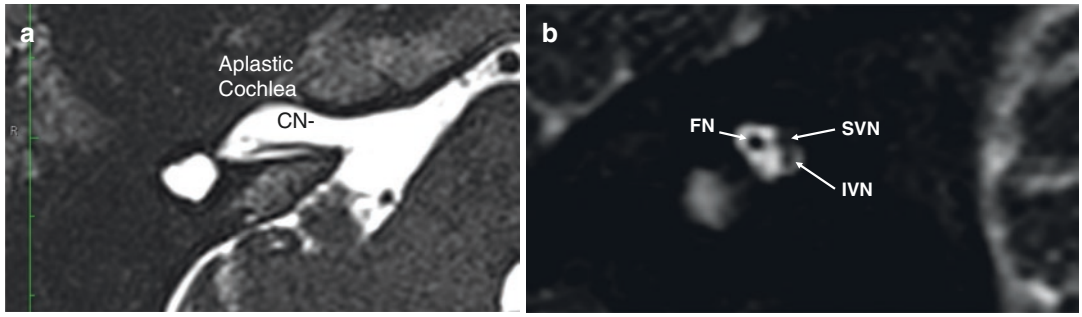


Fig. 1.15 Aplastic cochlear nerve. (**a**) Axial section. (**b**) Parasagittal section perpendicular to the IAC (FN = facial nerve, SVN = superior vestibular nerve, IVN = inferior vestibular nerve)

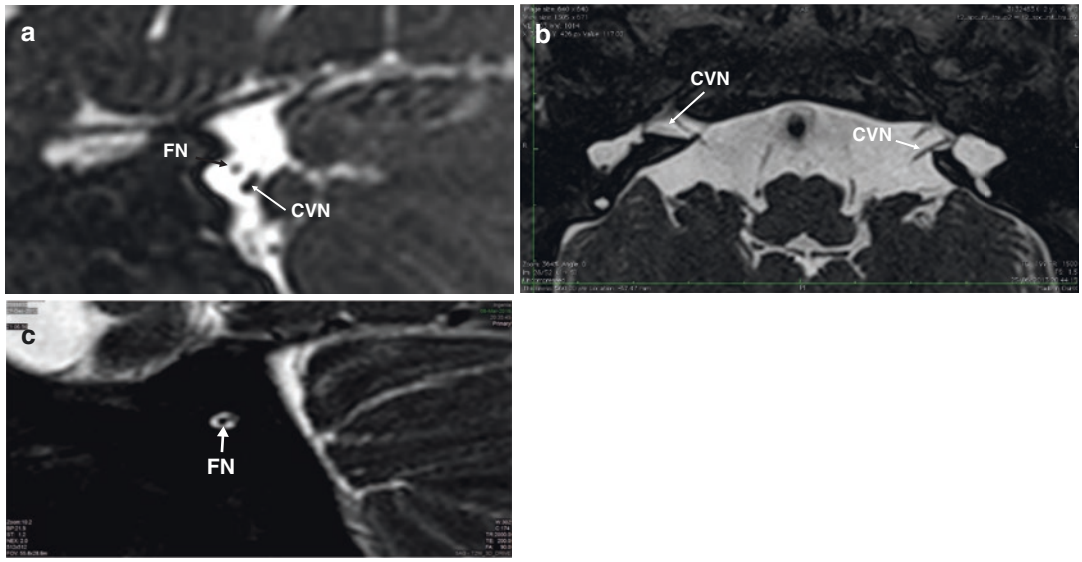


Fig. 1.16 Cochleovestibular nerve (CVN). (**a**) Normal where CVN is 1.5–2 times as much as the ipsilateral FN. (**b**) Right side hypoplastic, left side normal CVN. (**c**) Absent CVN in case of Michel deformity with absent IAC, CVN is also absent. Only FN can be identified (FN = Facial nerve, IAC = Internal auditory canal)

3. **Absent CN**

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

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 normal (Fig. 1.16a).

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.

6. **Absent CVN**

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

References

1. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
2. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. Laryngoscope. 1987;97(3 Pt 2 Suppl 40):2–14.
3. Mondini C. Minor works of Carlo Mondini: the anatomical section of a boy born deaf. Am J Otol. 1997;18(3):288–93.
4. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41.
5. Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27(5):615–23.
6. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41.
7. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
8. Sennaroglu L, Sennaroglu G, Ozgen B. Management of inner ear malformations. In: Sataloff RT, editor. Sataloff's comprehensive textbook of otolaryngology. JP Medical Publishers; 2015. p. 91–106.
9. Sennaroglu L, Ozkan HB, Aslan F. Impact of cochleovestibular malformations in treating children with hearing loss. In: Audiology and neuro-otology; 2013. p. 23–7.
10. Lemmerling MM, et al. Normal modiolus: CT appearance in patients with a large vestibular aqueduct. Radiology. 1997;204(1):213–9.
11. Ozgen B, et al. Complete labyrinthine aplasia: clinical and radiologic findings with review of the literature. AJNR Am J Neuroradiol. 2009;30(4):774–80.
12. Donaldson JA, Duckert LG, Lambert PM, Rubel EW, editors. Surgical anatomy of the temporal bone. 4th ed. New York, NY: Raven Press; 1992.
13. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
14. Sennaroglu L, et al. 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.
15. Sennaroglu L, et al. Cochlear hypoplasia type four with anteriorly displaced facial nerve canal. Otol Neurotol. 2016;37(10):e407–9.
16. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25(4):520–9; discussion 529.
17. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
18. Lo WW. What is a 'Mondini' and what difference does a name make? AJNR Am J Neuroradiol. 1999;20(8):1442–4.
19. Nance WE, et al. X-linked mixed deafness with congenital fixation of the stapedial footplate and perilymphatic gusher. Birth Defects Orig Artic Ser. 1971;07(4):64–9.
20. Phelps PD, et al. X-linked deafness, stapes gushers and a distinctive defect of the inner ear. Neuroradiology. 1991;33(4):326–30.
21. Wilkins A, et al. Frequent association of cochlear nerve canal stenosis with pediatric sensorineural hearing loss. Archiv Otolaryngol Head Neck Surg. 2012;138(4):383–8.
22. Casselman JW, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. Radiology. 1997;202(3):773–81.

Histopathologic Findings in Inner Ear Malformations

2

Rafael da Costa Monsanto, Sebahattin Cureoglu, Irem Gul Sancak, Levent Sennaroglu, and Michael Mauro Paparella

2.1 Introduction

The study of the ear structures has been a matter of great interest among anatomists, researchers, and physicians throughout the centuries [1]. Before the introduction of the first functional microscope in the 1660s, descriptions of the ear anatomy were limited to gross macroscopic studies of the external ear [1]. But the introduction of microscopy equipment and tissue staining techniques in the 1670s allowed the study of microscopic aspects of several diseases and malformations affecting the ear [1].

The first author to describe an inner ear abnormality was Carlo Mondini [2], in 1791, who reported the first gross anatomic features of a malformation which was later named by Siebenmann [3], in 1904, as "typus Mondini." [4] In his descriptions, Mondini [2] describes dilatation of the vestibule and vestibular aqueduct and several cochlear abnormalities, including incomplete interscalar septum, incomplete development of the modiolus, and hypoplasia of the upper turns, limiting the cochlea to one and a half turns [4]. Later, in 1838, Edward Cock [5] reported a wide communication between the cochlea and the vestibule in four human temporal bones, a deformity that is now known as "common cavity." In 1863, P. Michel [6] described another inner ear malformation characterized by bilateral absence of inner ear structures, which were later named after him (Michel aplasia). Other malformations affecting the membranous labyrinth have been described as

R. da Costa Monsanto
Department of Otolaryngology, Head & Neck Surgery, Universidade Federal de São Paulo/Escola Paulista de Medicina (UNIFESP/EPM), São Paulo, Brazil

Department of Otolaryngology, Head & Neck Surgery, University of Minnesota, Minneapolis, MN, USA

S. Cureoglu (✉)
Otopathology Laboratory, Department of Otolaryngology, Head & Neck Surgery, University of Minnesota, Minneapolis, MN, USA
e-mail: cureo003@umn.edu

I. G. Sancak
Department of Otolaryngology, Head & Neck Surgery, University of Minnesota, Minneapolis, MN, USA

Department of Veterinary Surgery, Ankara University Faculty of Veterinary Medicine, Ankara, Turkey

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

M. M. Paparella
Department of Otolaryngology, Head & Neck Surgery, University of Minnesota, Minneapolis, MN, USA

Paparella Ear Head & Neck Institute, Minneapolis, MN, USA
e-mail: papar001@umn.edu

© Springer Nature Switzerland AG 2022
L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_2

well, including complete membranous labyrinthine dysplasia [7], cochleosaccular dysplasia [8], and cochlear basal turn dysplasia [9]. Since membranous abnormalities are not seen in the available imaging tests, those are not included in most current classification systems of malformations [10].

For years, the term "Mondini dysplasia" has been used to describe virtually every type of congenital inner ear malformation detected by imaging tests [10]. However, further histopathologic and imaging have made possible the development of classification systems and to hypothesize potential pathophysiologic and embryogenic mechanisms involved in the genesis of each malformation [10, 11]. In addition, those new findings allowed the development of several hearing rehabilitation solutions for patients who have severe-to-profound hearing loss secondary to inner ear malformations [12–14]. Initially, these patients were excluded from the candidacy criteria for cochlear implantation, mostly due to concerns about potential complications and low expectations of a successful stimulation of the cochlear nerve [7, 12]; in addition, the auditory brainstem implants were not indicated for children under 12 years [15]. But technological advancements as well as the growing number of surgeons who successfully performed those implants in pediatric and adult patients with cochleovestibular malformations have led to gradual change to their indications [13, 14, 16]. Analysis of inner ears from donors who had cochlear malformations has a tremendous value to achieve a better understanding of the embryologic, pathophysiologic, morphologic, and developmental features associated with those malformations [1, 17].

2.2 Classification of Inner Ear Malformations

The introduction of high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) in the late 1980s, as well as further histopathologic analysis of human temporal bone specimens, resulted in detailed descriptions of new morphologic aspects and malformations that do not completely fit the initial descriptions of Mondini [2], Siebenmann [3], and Terrahe [17]. Moreover, new classifications became a necessity with the development of cochlear and brainstem implants—selection of potential candidates to those implants includes a thorough evaluation of the type and degree of malformations, considering their impact in the surgical decisions and potential prognosis of the rehabilitation [18].

Jackler et al. [10] were the first authors to describe a classification system for inner ear malformations based upon their radiologic appearance. The authors also described potential embryologic mechanisms (developmental arrest, aberrant development) involved in their genesis, and correlated the findings with the audiologic patterns and prognosis. Their classification has divided the bony malformations in 2 groups, one with absent or malformed cochlea and one with normal cochlea (Table 2.1). As a general rule, the authors observed a correlation between severity of the structural abnormalities and worse audiologic results [10].

Marangos [19], in 2002, developed a new classification method and highlighted some of the arrests in the embryogenic development that may cause each type of malformation. The authors also claim that some of the malformations may be considered as absolute contraindications to cochlear implant, due to potential complications and very low expectation of hearing rehabilitation. Marangos' [19] classification also includes other types of abnormalities not contemplated by Jackler et al. [10], such as long crista transversa, internal auditory canal tripartitus, and X-linked hearing loss (Table 2.1).

Sennaroglu and Saatci [11], in 2002, reported a classification method for cochlear malformations based on radiologic findings in 23 patients. The authors classified the abnormalities according to their locations in the inner ear and severity of the abnormalities in the inner and outer architecture. They also demonstrated that abnormalities in each labyrinthine compartment may occur in isolation or in association with malformations of other portions of the bony labyrinth (Table 2.1). With their findings, they hypothesized potential pathophysiologic mechanisms and embryogenic

Table 2.1 Different classifications of inner ear malformations proposed after introduction of high-resolution computed tomography

Author	Category		Characteristics
Jackler et al. [10]	Abnormal cochlea	1	Labyrinthine aplasia (Michel aplasia)
		2	Aplasia of cochlea, normal or deformed vestibule/semicircular canals
		3	Hypoplasia of cochlea, normal or deformed vestibule/semicircular canals
		4	Incomplete cochlea, normal or deformed vestibule/semicircular canals
		5	Common cavity
	Normal cochlea	1	Dysplasia of vestibule and lateral semicircular canal, normal vertical canals
		2	Enlarged vestibular aqueduct, normal or dilated vestibule, normal semicircular canals
Marangos [19]	Incomplete embryonic development	1	Labyrinthine aplasia (Michel aplasia)
		2	Common cavity (otocyst)
		3	Aplasia/hypoplasia of cochlea, normal vestibule/semicircular canals
		4	Normal cochlea, aplasia/hypoplasia of vestibule/semicircular canals
		5	Labyrinthine hypoplasia (cochlea and vestibule)
		6	Mondini dysplasia
	Aberrant embryonic development	1	Enlarged vestibular aqueduct
		2	Narrow internal auditory canal
		3	Long crista transversa
		4	Internal auditory canal tripartitus
		5	Incomplete cochleomeatal separation
	Isolated hereditary malformations	1	X-linked hearing loss
	Syndromes	2	Malformations associated with syndromes
Sennaroglu [11]	Cochlear malformations	1	Labyrinthine aplasia (Michel aplasia)
		2	Cochlear aplasia
		3	Common cavity
		4	Cochlear hypoplasia
		5	Incomplete partition type I
		6	Incomplete partition type II (Mondini dysplasia)
	Vestibular malformations		Labyrinthine aplasia, common cavity, absent vestibule, hypoplastic vestibule, dilated vestibule
	SCC malformations		Absent, hypoplastic, enlarged
	IAC malformations		Absent, narrow, enlarged
	Vestibular and cochlear aqueduct		Enlarged

SCC semicircular canal, *IAC* internal auditory canal

factors associated with the development of each abnormality [11, 20]. Later, in 2006, Sennaroglu et al. [12] modified their classification to accommodate the X-linked deafness as "incomplete partition type 3."

More recently, Jeong and Kim [21] suggested a new classification system, claiming that previous classifications have embryogenic implications but lack clinical application. Reviewing clinical, imaging, and surgical data of 59 patients with cochleovestibular malformations subjected to cochlear implantation, the authors suggested the following classification: (1) type A, normal cochlea and modiolus; (2) type B, malformed cochlea and partial modiolus; (3) type C, malformed cochlea and no modiolus.

2.3 Embryology of Inner Ear Malformations

After 3 weeks of development, the ectoderm on each side of the rhombencephalon thickens to form the otic placodes, which will later invaginate and form the otocysts [22]. Arrest in the development before formation of the otocyst leads to labyrinthine aplasia (Michel aplasia) [20]. The labyrinthine aplasia may occur either with presence or absence of the petrous bone and otic capsule [20]. If the arrest occurs in the initial stages of the otocyst formation, the resultant deformity is called "rudimentary otocyst." [23] At Week 5, the pars utriculovestibularis grows dorsocranially to later form the utricle, semicircular canals, and endolymphatic duct [22, 24]. The earlier formation of the posterior labyrinth explains why some cochlear malformations are not necessarily followed by abnormalities in the posterior labyrinth [20]. An arrest in development which occurs between the fourth and fifth week (before differentiation of the otocyst into cochlea and vestibule) will result in common cavity malformation. On the other hand, an insult affecting the inner ear within the fifth week (before development of the cochlear duct) will lead to an abnormality characterized by cochlear aplasia with hypoplastic or normal vestibular/endolymphatic structures [10, 20, 22].

During the sixth week, the cochlear duct starts to develop from the saccule, reaching the size of one turn at 7 weeks, 1.5 turns at 8 weeks, and the full 2.5 turns before the 11th week [10, 20, 22]. If there is any developmental arrest between sixth and 11th week, the cochlea will be hypoplastic with some degree of underdeveloped internal architecture depending on the time the insult happens [10, 20, 22]. After the cochlea reaches the full size of 2.5 turns, it will slowly continue to grow in caliber until it reaches adult size between the 22nd and 24th weeks. Thus, defects in the development occurring after the tenth week will result in a cochlea with normal or near-normal external dimensions [10, 20, 22].

After the cochlea reaches 2.5 turns around the tenth week, the mesenchyme surrounding the cochlear duct differentiates into cartilage, which will later become the perilymphatic cochlear ducts; in addition, epithelial cells of the cochlear duct differentiate into the spiral limbus and organ of Corti [10, 20, 22]. The development of the vestibulocochlear nerve occurs in parallel to the membranous labyrinth—before the eighth week, nerve fibers from the neuroblasts grow towards the labyrinth and vestibulocochlear nerve area at the brainstem [10, 20, 22]. Vestibulocochlear nerve is present by seventh week, while the hair cells in the basal turn of the cochlea receive nerve fibers around the tenth week [10, 20, 22]. The modiolus develops from the membranous bone within the cochlea, and bone deposition occurs exponentially around 20–21st week in the basal and middle turns of the cochlea and towards the apical turn by the 25th week. Thus, any cochlear developmental arrest after the tenth week will not affect the cochlear size, but may have a significant impact on the development of the internal architecture of the cochlea and vestibulocochlear nerve, comprising both incomplete partitions and membranous labyrinth anomalies [10, 20, 22].

2.4 Histopathologic Findings of Inner Ear Malformations and Their Clinical Implications

Histopathologic studies of human temporal bone from donors who had inner ear malformations provided (and will continue to provide) invaluable contributions to the current knowledge on several features of labyrinthine anomalies [1]. Throughout the years, several different types of inner ear malformations have been described, but adequate assessment of several of them was only made possible after introduction of high-resolution CT [10]. It is known today that over 20% of all cases of congenital sensorineural hearing loss are secondary to bony malformations, while the remaining 80% are probably membranous malformations located at the cellular level [11]. However, for many years, the term "Mondini deformity" has been used in such a way to encompass any type of bony malformation affecting the cochlea [10].

After the first demonstration of a cochlear implant in a patient with inner ear malformation by Mangabeira-Albernaz [16], in 1983, in addition to other following successful experiences [13, 14], the candidacy for cochlear implants has changed to accommodate patients with cochlear malformations, who were initially excluded from its indications [12]. The possibility for hearing restoration in this specific population brought the need for a broader and deeper understanding of the anatomic, morphologic and functional aspects of each specific type of inner ear malformation [10, 11]. In this regard, imaging and histopathologic studies already provided advances, demonstrating several different types of malformations and their respective morphologic features [25, 26]. The clinical implications of those new findings are clear, considering that some types of malformations are associated with neural and structural abnormalities that may either predict the results or even contraindicate cochlear implantation at all, as in cases of labyrinthine aplasia—those patients may benefit from brainstem auditory implants [12].

The classification systems proposed for inner ear malformations after the 1980s mostly comprised of bony malformations, considering that they can be identified in imaging tests [10]. However, descriptive histopathologic analysis of human temporal bones also demonstrated several malformations affecting only the membranous labyrinth as well [27, 28].

2.4.1 Malformations of the Membranous Labyrinth

In this group of malformations, the otic capsule and bony labyrinth are well developed and normal in size, but there is abnormal development of the internal architecture of the membranous labyrinth. The three main types of exclusive membranous malformations are:

1. Cochleosaccular dysplasia (Scheibe dysplasia): This type of malformation involves only the saccule and cochlea, while the utricle and semicircular canals are spared [29]. Several abnormalities secondary to this malformation were observed, such as degeneration of the stria vascularis and organ of Corti, loss of inner and outer hair cells, and collapsed Reissner's membrane; signs of hydrops may also be observed [28, 29]. However, there seem to be no significant decrease in the number of neuron cells. The saccular membrane is usually collapsed and its sensorial epithelium is deformed [29]. Some degree of atrophy of the cochlear and vestibular nerve have been reported as well [28]. This abnormality may occur as part of several syndromes (Down, keratitis-ichthyosis-deafness, Refsum, Waardenburg, Jervell and Lange, trisomy of 18), following viral infections (congenital rubella), or in isolation [28, 29].
2. Cochlear basal turn dysplasia (Alexander dysplasia): In his original description in 1904 [9], Alexander describes the abnormalities affecting the inner ear of a 38-year-old man who was deaf mute. The author describes normal *pars inferior*, while the cochlea had several abnormalities affecting the basal turn, including atrophy of the stria vascularis, defect of the interscalar septum from above the upper basal turn, loss of hair and pillar cells in the organ of Corti, and loss of spiral ganglion neurons. In the middle and apical turns, minor changes were observed, such as atrophy of the stria vascularis and spiral ligament and absence of helicotrema. However, no subsequent report of a malformation limited to the basal turn of the cochlea has been later described [7].
3. Complete membranous labyrinth aplasia (Siebenmann-Bing): The description of a complete dysplasia of the membranous labyrinth was first published by Siebenmann and Bing, in 1907 [10]. This type of malformation is exceedingly rare, with very few reports in the literature. The bony labyrinth has a normal development, and thus such abnormality cannot be seen in the currently available imaging tests [7]. It has also been suggested that it may be a severe variant of the Scheibe's dysplasia. An association with genetic syndromes such as Jervell and Lange, Usher, and oculo-auriculo-vertebral spectrum has been reported in the past [7].

2.4.2 Malformations of the Bony Labyrinth

In this group of malformations, abnormalities affect the otic capsule and internal/external cochlear and vestibular bony architecture, which may be accompanied by membranous abnormalities [10, 11, 20]. It comprises a wide range of abnormalities, ranging from complete labyrinthine aplasia (Michel aplasia) to near-normal inner ear. The most important bony malformations, as reported by Sennaroglu and Saatci [11], include labyrinthine aplasia, rudimentary otocyst, common cavity, cochlear aplasia, cochlear hypoplasia, and incomplete partition. Those malformations may be bilateral (more frequently) or unilateral, and the contralateral side may be normal or have a different type of malformation [11, 24]. In addition, they may occur in isolation, associated with middle ear or other organ anomalies, or in association with several syndromes, such as Wildervanck, Pendred, DiGeorge, and the trisomies [29]. The currently available histopathologic findings of human bone studies in the literature are listed below and in Table 2.2.

Labyrinthine aplasia and rudimentary otocyst comprise a group of malformations resulting from very early arrest in embryologic development, before the fifth week. To date, there are no histopathologic descriptions of those abnormalities in human temporal bones. Radiologic and clinical characteristics are described below:

2.4.2.1 Labyrinthine Aplasia (Michel aplasia)

It is characterized by the absence of both anterior and posterior labyrinth [6]. Petrous bone, as well as otic capsule, may be hypoplastic, aplastic, or normal [30]. There are no histopathologic human temporal bone descriptions of this malformation in the literature [20]. Patients with this abnormality have absence of response in all clinical audiologic tests and may benefit only from brainstem auditory implants [15, 31].

2.4.2.2 Rudimentary Otocyst

In the initial stages of embryologic development, an arrest in development may occur right after formation of the otocyst—such arrest results in a small, millimetric round or ovoid cavity, which may have parts of semicircular canals, called rudimentary otocyst. It is an earlier malformation as compared with the common cavity, which is larger in size and may have nerve connections through the internal auditory canal. Audiologic tests in patients with this abnormality are absent, and the only hearing rehabilitation option is brainstem auditory implant [15, 31]. Although (to the best of our knowledge) no human temporal bone description of this type of abnormality exists, studies in mice demonstrated the first maturational stages of the otocyst [32–34]. At Day 12 of embryologic development, areas of sensory structures can be identified [32–34], and at this stage, the otocyst is composed of pseudostratified columnar epitheliae surrounded by undifferentiated mesenchymal cells [32]. In some areas, there may be initial differentiation of those mesenchymal cells in chondrocytes [32, 33]. There are definite perilymphatic spaces, with respective projections resembling rudimentary semicircular canals; those canals sometimes have identifiable cristae, which usually end in a blind sac into the cartilaginous capsule [32]. No endolymphatic duct or sac is identified [32]. There may be formation of utriculosaccular spaces in a few cases, with or without sensorial macula [32]. In over 50% cases, there may be partial development of a cochlear duct, which does not coil and become a swollen sac [32–34]. Rudimentary cochlear sensory epithelia and tectorial membrane may also be present [32–34].

2.4.2.3 Common Cavity

The common cavity deformity, first described in 1838 by Cock [5, 7, 35], comprises a single, ovoid or round chamber as seen in imaging tests. It seems to occur secondary to an arrest in development around the fourth and fifth weeks [5]. It is important to differentiate a common cavity from a rudimentary otocyst due to the clinical implications. In both situations, a round or ovoid cavity is seen in the otic capsule, but in the rudimentary otocyst, the cavity is smaller in size and there is no internal auditory canal [5, 36]. In the common cavity, there is an internal auditory

Table 2.2 Most frequent histopathologic/imaging abnormalities seen in the different types of bony malformations

	Cochlea shape	Modiolus	ISS	CN	Vestibule	Semicircular canals	Associated abnormalities
Cochlear hypoplasia							
Type I	Smaller, bud-shaped	Aplasia/ hypoplasia	Aplastic	Aplasia/ hypoplasia	Aplasia/ hypoplasia/ dilated	Aplasia/ hypoplasia/ normal	Fixed footplate, aplastic oval window Aberrant course of facial nerve
Type II	Smaller, round	Hypoplasia	Aplasia/ hypoplasia	Hypoplasia/ normal	Hypoplasia/ dilated/normal	Hypoplasia/ dilated/normal	Fixed footplate, aplastic oval window Aberrant course of facial nerve
Type III	Smaller, normal	Shortened/ normal	Normal/ hypoplasia/ aplasia	Normal	Aplasia/ hypoplasia	Aplasia/ hypoplasia	Fixed footplate, aplastic oval window Aberrant course of facial nerve
Type IV	Smaller, hypoplastic upper turns	Shortened	Normal in the basal turn	Normal	Normal/dilated	Hypoplasia/ dilated/normal	Aberrant course of facial nerve
Incomplete partition							
Type I	Round or oval-shaped	Aplasia/ hypoplasia	Aplasia/ hypoplasia	Aplasia/ hypoplasia/ normal	Dilated	Aplasia/normal	Abnormal footplate
Type II	Normal	Hypoplasia	Normal, dislocated upwards	Normal	Dilated	Normal	Enlarged vestibular aqueduct, normal footplate
Type III	Normal	Absent	Absent	Normal	N/A; normal	N/A; normal	Medial displacement of vestibular aqueduct with varying degrees of enlargement; stapes fixation; aberrant course of facial nerve

ISS interscalar septum, *CN* cochlear nerve

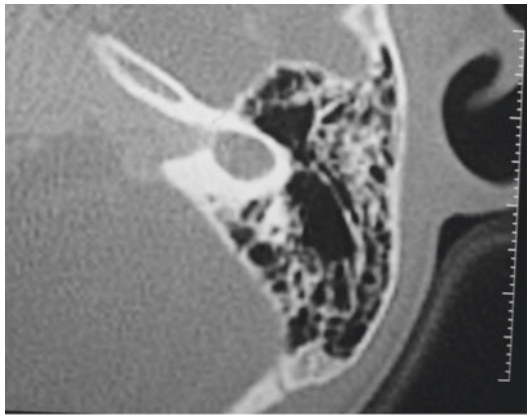


Fig. 2.1 Common cavity deformity. The cavity is bigger in size as compared with a rudimentary otocyst, and there is an internal auditory canal. (Source: Image kindly provided by Dr. Levent Sennaroglu, reproduced under his authorization)

canal (Fig. 2.1) and also neural spiral ganglion cells in the lateral wall, which allow cochlear implantation.

Patients with common cavity deformity usually have profound sensorineural hearing loss [30]. Considering the possibility of the presence of a viable cochlear nerve, cochlear implants may be indicated [5, 30]; nonetheless, it is imperative to perform an extensive audiologic assessment prior to surgery [5, 30]. In cases that a cochlear implant is indicated, the selected electrode should be short and expand against the external borders of the cochlea to reach the neural epithelium [5, 30, 36]; curved electrodes (designed to "hug" the modiolus) should be avoided [5, 37]. In cases of cochlear nerve aplasia or failure of cochlear implants, auditory brainstem implants may be offered [5, 15, 30].

2.4.2.4 Cochlear Aplasia

In this type of malformation, although the otic capsule is present and posterior labyrinth is formed, no cochlea is developed [10, 11, 20]; labyrinthine segment of the cochlear nerve is usually dislocated anteriorly. The arrest in development leading to this abnormality must occur before the development of the cochlear duct at fifth week. Frequently, the abnormality is bilateral and symmetric, suggesting a genetic origin. The vestibule and vestibular canals may be hypoplastic or dilated.

2.4.2.5 Cochlear Hypoplasia

Cochlear hypoplasia is defined as the presence of an identifiable cochlea with smaller external dimensions. The prevalence of cochlear hypoplasia accounts for 15–23.5% among all types of inner ear malformations [23, 38]. They may occur in isolation or associated with several syndromes, such as branchio-oto-renal syndrome, trisomy of 13, and triploidy [39]. The degree of structural abnormalities depends largely on the time and severity of arrest during the embryologic period [7, 10, 20]. The arrest in development leading to cochlear hypoplasia occurs between the sixth and eighth week, after the cochlear duct is formed but has not yet completed their development in caliber [20]. The only exception is cochlear hypoplasia type IV, in which the arrest seems to occur in a later developmental stage [10, 20].

There may be some coincidence in the morphologic appearance of cochlear hypoplasia, cochlear aplasia with enlarged vestibule and common cavity malformations [20]. The difference between those entities is that in the cochlear hypoplasia, the cochlea and the vestibule are distinct. Although morphologically similar, it is imperative to identify the type of malformation in the preoperative evaluation of patients with potential cochlear implant indication: patients with common cavity and cochlear hypoplasia may achieve good results with cochlear implants, but patients with cochlear aplasia only benefit from brainstem implants [20, 23].

When cochlear implants are indicated, there are some morphologic features that have to be assessed in the preoperative evaluation [18]. Patients who have bony cochlear nerve canal aplasia always have severe-to-profound hearing loss and do not benefit from cochlear implantation [37]. If the canal is hypoplastic, the hearing levels vary from mild (less frequently) to profound hearing loss [23, 40–42]. The presence of normal cochlear nerve canal also does not necessarily represent better hearing levels as compared with cochlear aperture hypoplasia,

considering the other structural and membranous abnormalities present in cochlear hypoplasia and also the status of the cochlear nerve fibers, which may be normal, hypoplastic, and aplastic in both conditions [20, 30, 37, 40]. Intraoperatively, considering the anatomic aberrations associated with the hypoplasia, identification of the round window niche/membrane through facial recess approach may be difficult; therefore, an additional transmeatal approach or a labyrinthotomy is needed in some cases [37, 43]. The course of the facial nerve may be abnormal as well [20, 23, 44]. It is recommended to use thin (0.8 mm) and short (<20 mm) electrodes, given the smaller cochlear size and fragility of its internal architecture [12, 30, 37].

Sennaroglu and Bajin [30] have categorized the group of malformations nominated as cochlear hypoplasia in four subgroups:

1. Cochlear hypoplasia type I: The cochlea is either oval- or round-shaped, and its internal architecture is severely deformed [11, 20, 23].
   (a) *Internal cochlear architecture:* The modiolus is absent or hypoplastic, with defective neural connections into the interior of the cochlea [11, 20, 23]. There is no interscalar septum or internal membranous structures [11, 20, 23, 30].
   (b) *Bony cochlear nerve canal/cochlear nerve:* The canal for the cochlear nerve may be aplastic (no cochlear nerve), or hypoplastic, with coincident cochlear nerve hypoplasia [11, 20, 23].
   (c) *Vestibular system:* The vestibular system may be hypodeveloped as well—there were reports of aplasia or hypoplasia of the vestibule and semicircular canals and dilatation of the utricle [20].
   (d) *Other associated malformations:* fixation of the footplate and aplastic oval window [11, 20].
   (e) *Clinical implications:* Most patients with cochlear hypoplasia type I have severe-to-profound sensorineural or mixed hearing loss [23]. Therefore, there are not many treatment options other than cochlear or auditory brainstem implants [30, 37]. In preoperative evaluation, the morphology of the bony cochlear nerve canal should be assessed carefully, considering that the cochlear implant may not provide good hearing outcomes if the canal is aplastic [30]. In cases of hypoplastic cochlear nerve and subtotal/total modiolar defect, it is recommended to first perform cochlear implants—although their functional results may be poorer than in normal patients, they are still better than brainstem implants, which should be performed in the contralateral side if cochlear implantation does not provide sufficient hearing restoration [30, 37]. If cochlear implants are indicated, short, straight electrodes are warranted for two reasons: (1) the neural epithelium is usually localized at the lateral cochlear wall, and thus the electrode should expand against these; and (2) if there is a communication between the cochlea and internal auditory canal, curved electrodes may be misplaced inside the meatus, increasing the risks for damaging the facial nerve [31, 37, 45].
2. Cochlear hypoplasia type II: The cochlea has an external shape that, although rounder, is more closely related to a normal cochlea as compared with cochlear hypoplasia type I.
   (a) *Internal architecture:* The internal architecture of the cochlea is also more developed in the cochlear hypoplasia type II as compared with type I [20]. The modiolus is always absent or hypoplastic (no development in the upper half of the cochlea) (Fig. 2.2) [20, 46]. The interscalar septum may be either hypoplastic or absent, giving the cochlea a rounder shape (Fig. 2.2) [20]. It has also been demonstrated the possibility of absence or loss in the population of spiral ganglion cells [20, 46–48]. Lateral wall in the basal turn is frequently normal in the basal turn, with varying degrees of hypoplasia/atrophy in the upper regions of the cochlea [20, 46].
   (b) *Bony cochlear nerve canal/cochlear nerve:* There were reports of complete absence of bony cochlear nerve canal and

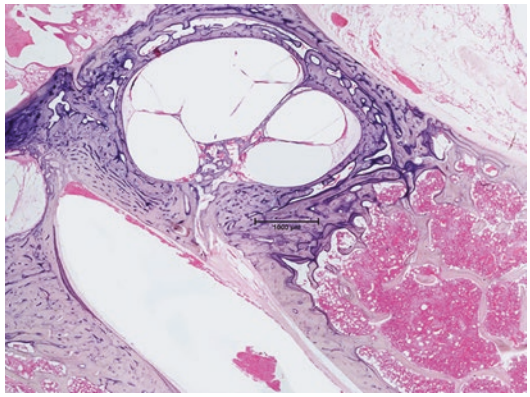


Fig. 2.2 A horizontal section of a human temporal bone with cochlear hypoplasia type II, harvested from a 5-year-old male patient (Hematoxylin and Eosin). The patient had congenital bilateral profound hearing loss, congenital heart problems, and complete cleft lip and palate. He also had a history of chronic otitis media bilaterally, as diagnosed by physical exam and type B tympanograms. Interestingly, he had a cochlear hypoplasia type III on the contralateral ear. Conditional audiometry revealed residual bone conduction thresholds at the frequencies of 250 and 500 Hz. In this section, it is possible to observe mucoid-purulent effusion, fibrous strands, and granulation tissue in the middle ear. The facial nerve is dehiscent in its tympanic segment. There is a clear abnormality in the footplate. The cochlea is hypoplastic, with a cystic apex; slight hydrops in the upper part of the scala media is observed. The modiolus is hypoplastic, absent in its upper portion. Interscalar septa are absent in the upper part of the cochlea as well. There are identifiable spiral ganglion cells in the Rosenthal's canal in the lower basal turn and in the lateral posterior wall of the cochlea. There is rudimentary organ of Corti in the basal turn of the cochlea, with adjacent normal stria vascularis and spiral ligament. The bony cochlear nerve canal is hypoplastic, with no cochlear nerve fibers within. Vestibule is hypoplastic, and semicircular canals are aplastic. Saccular wall is slightly dilated. Endolymphatic sac and duct are aplastic. (Source: Human temporal bone archive from the Otopathology Laboratory at the University of Minnesota (USA))

hypoplastic nerve, but they may also be normal (Fig. 2.2) [20].

(c) *Vestibular system:* The vestibule may be hypoplastic, normal, or minimally dilated; vestibular aqueduct is sometimes enlarged [20, 46].

(d) *Other associated malformations:* Aplasia of the oval window and stapes fixation are frequent features of cochlear hypoplasia type II [20].

(e) *Clinical implications:* The frequent abnormalities of the oval window and stapes may add a conductive component onto the sensorineural hearing loss, although pure sensorineural hearing loss is much more frequent than mixed hearing loss among patients with cochlear hypoplasia type II [23]. The majority of patients (over 80%) have severe-to-profound hearing loss, while moderate-to-severe hearing loss is less frequently observed [23, 30]. If functional hearing is observed, some patients may benefit from stapedectomy with postoperative hearing aids [23, 30]. In cases of severe-to-profound hearing loss, cochlear or auditory brainstem implants are warranted considering the presence or absence of aplasia of the bony cochlear nerve canal [31, 37, 45]. In cases with complete absence of the modiolus, surgeons must be aware of the possibility of intraoperative gusher and misplacement of the cochlear implant electrode inside of the internal auditory canal [49–51]. The high prevalence of stapes abnormalities secondary to this malformation may increase the risks of recurrent meningitis as well [36, 52, 53].

3. Cochlear hypoplasia type 3: In this type of hypoplasia, the external shape of the cochlea is very similar to normal, given the presence of interscalar septum [20]. In a case series published by Cinar et al. [23], this type of hypoplasia was the most frequent among all patients with cochlear hypoplasia. Although external architecture is near normal, the cochlea is limited to one and a half turns internally—this particular morphologic feature may have led authors in the past to classify patients with this abnormality as Mondini dysplasia [39]. Given that ossification of the cochlea begins only after the membranous labyrinth reaches full size, it is possible that, in this type of malformation, the cochlea is genetically predetermined to have a smaller size [20].

(a) *Internal architecture:* [20] The modiolus is usually well developed but shorter than in a normal cochlea (Fig. 2.3). Interscalar septum is always present. The anatomic configuration of the cochlear scalas, organ of Corti, and lateral wall is normal (Fig. 2.3)
(b) *Bony cochlear nerve canal/cochlear nerve:* [20] Although frequently normal, the bony cochlear nerve canal may be aplastic or hypoplastic.
(c) *Vestibular system:* [20] vestibule and semicircular canals are frequently hypoplastic.
(d) *Other associated malformations:* [20] oval window may be normal or aplastic, and stapes fixation is seen in some cases.
(e) *Clinical implications:* Given the near-normal internal cochlear architecture and potential abnormalities in the oval window, patients may present with virtually any type and degree of hearing loss [23, 30]; however, pure sensorineural, moderate-to-severe, and severe-to-profound hearing loss are the most frequent types of observed hearing loss among patients with cochlear hypoplasia type 3 [23]. Mixed hearing loss may be treated by stapedectomy and/or hearing aids, mild-to-moderate sensorineural hearing loss with hearing aid or middle ear implants and severe-to-profound hearing loss should receive cochlear or auditory brainstem implants [14, 30, 37, 54].

4. Cochlear hypoplasia type 4: Defined as normal basal turn, but hypoplastic middle and apical turn [20]. Descriptions of this type of abnormality are very recent [11, 20]. There is no current histopathologic description available for this type of cochlear hypoplasia [20]. Given the normal diameter of the basal turn, it

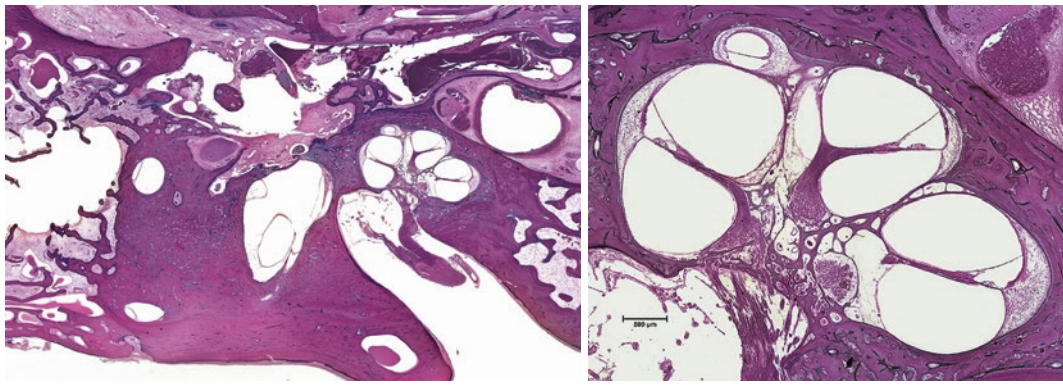


Fig. 2.3 A horizontal section of a human temporal bone with cochlear hypoplasia type III, harvested from a 50-year-old female patient (Hematoxylin and Eosin). The donor died of acute myeloid leukemia, renal failure, gastrointestinal bleeding, and sepsis. At physical examination prior to death, she had retracted tympanic membranes but no signs of inflammation. She complained of a mild hearing loss in the past, which got worse after she was started on ototoxic antibiotics to treat the infection in the moment of admission. Left side: In this panoramic view of the middle and inner ear, it is possible to observe mucoid-purulent effusion, fibrous strands, and granulation tissue in the middle ear and mastoid antrum. The facial nerve is partially dehiscent in its tympanic segment; footplate is normal. Vestibule, saccule, utricle, and semicircular canals are normal; endolymphatic duct and sac are also normal. Right side: A 4× magnification view of the cochlea, at the same level as the figure in the left. The cochlea is hypoplastic, with decreased number of turns as compared to a normal cochlea. The modiolus is normal (as per the size of the cochlea). Interscalar septa are present. Organ of Corti, basilar membrane, Reissner's membrane, and structures of the lateral wall are normal. There is no apparent decrease in the number of spiral ganglion cells in Rosenthal's canal; cochlear aperture for the cochlear nerve, internal auditory canal, and cochlear nerve are normal. (Source: Human temporal bone archive from the Otopathology Laboratory at the University of Minnesota (USA))

is possible that the insult leading to this type of malformation occurs between the tenth and 20th weeks of embryologic development, and the cochlea is probably genetically determined to have such morphology [20]. From imaging observations, it has been noted that the upper 1.5 turns of the cochlea are severely hypoplastic and dislocated to an anterior position [20, 44]. Among the available reports, none of the patients with this type of malformation had bony cochlear nerve canal aplasia. There may be fixation of the stapes and aplasia of oval window as well.

(a) Clinical implications: Considering the normal cochlear basal turn development, patients with his type of cochlear hypoplasia may have a mild hearing loss [23]. The frequency of moderate-to-severe hearing loss is also higher than in other types of cochlear hypoplasia. Those patients frequently have sensorineural or mixed hearing loss, but the presence of pure conductive hearing loss has also been demonstrated as well—for the other types of hypoplasia, there are no clinical descriptions of pure conductive hearing loss [23]. Treatment options are similar to what is described for cochlear hypoplasia type 3 [30].

2.4.2.6 Incomplete Partition Anomalies

In this type of malformation, the cochlea is well defined, with normal or near-normal size, and clearly distinct from the vestibule. According to Sennaroglu and Bajin [30], incomplete partitions account for 41% of all inner ear malformations. In their initial classification published in 2002, Sennaroglu and Saatci [11] initially reported that incomplete partition type I had more severe cochlear abnormalities as compared with cochlear hypoplasia and incomplete partition type II. However, those findings have been later questioned with the description of a wide range of potential types of hypoplasia, as previously mentioned [20]. In terms of development, cochlear hypoplasia seems to occur in an earlier developmental stage as compared with incomplete partition malformations, given that after the cochlea reaches its full size after the eighth week of development, it is not possible to have cochlear hypoplasia (except type IV) [20, 39].

The difference between the types of incomplete partitions is the degree of abnormalities affecting the internal cochlear architecture. Sennaroglu and Saatci [11] classified these abnormalities into three groups:

1. Incomplete partition type I: Externally, the shape of the cochlea is similar to the cochlear hypoplasia type II (round or oval shaped) (Fig. 2.4) [20, 55]. Considering its similarities with other malformations (such as common cavity, rudimentary otocyst, cochlear hypoplasia type I, and even Mondini dysplasia), some of the past clinical descriptions of the incomplete partition type I have been erroneously described under other type of malformation [37, 39]. The difference of the incomplete partition type I from those other abnormalities is that, in this type of malformation, it is possible to clearly identify the cochlea and vestibule (as opposed to common

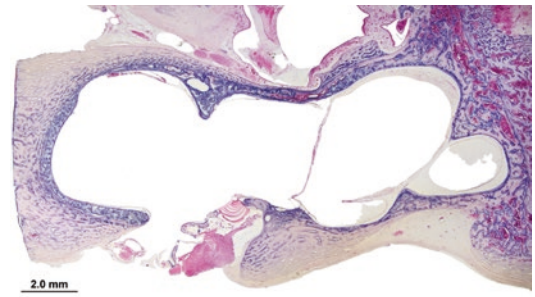


Fig. 2.4 A horizontal human temporal bone section from a donor who had incomplete partition type I malformation (Hematoxylin and Eosin). The size of the cochlea and vestibule is normal. However, there is no cochlear internal architecture, including modiolus and interscalar septa. In addition, there is a communication between the base of the cochlea and the internal auditory canal. The cochlea is separated from the vestibule by only a small thin membrane. (Source: Reproduced with permission of the "Massachusetts Eye and Ear Infirmary" and "Cochlear Implants International" journal)

cavity) and the external size of the cochlea is normal (as opposed to cochlear hypoplasia type II and rudimentary otocyst) [39]. Previous histopathologic descriptions of malformations which fit the characteristics of incomplete partition type I were nominated "empty cochlea," [56] "severe incomplete partition," [10] "severely dysmorphic cochlea," [57] and "pseudo-Mondini." [58]

(a) *Internal architecture:* The incomplete partition type I is characterized by the absence or subtotal development of the modiolus and absence of interscalar septum (Fig. 2.4). In some cases, there is no bony separation from the cochlear base and internal auditory canal. Lateral cochlear wall structures are hypoplastic or aplastic. The organ of Corti may or not be present—when present, it is rudimentary (Fig. 2.3) [5]. Those findings suggest an earlier developmental arrest than in incomplete partition type II [39].

(b) *Bony cochlear nerve canal/cochlear nerve:* There is usually a wide communication between the cochlea and internal auditory canal. The cochlear nerve may be aplastic (Fig. 2.4). There seems to be a distribution of spiral ganglion cells along the lateral wall of the cochlea, similarly to what is found in common cavity and cochlear hypoplasia type I deformities [5, 36].

(c) *Vestibular system:* Vestibule and semicircular canals are frequently grossly dilated (Fig. 2.5), but some cases of aplasia of the posterior semicircular canal have also been described [20]. Enlargement of vestibular aqueduct is very rare—in fact, imaging studies demonstrated that the vestibular aqueduct is not seen at all in some cases [59]. This may be explained by the fact that the vestibular aqueduct is the last structure of the posterior labyrinth to develop, suggesting a developmental arrest before this structure is completely formed [39]. The vestibule is separated from the cochlea by only a very thin membrane (Fig. 2.4), but in some cases a wide communication between the cochlea and vestibule can be seen [5, 20].

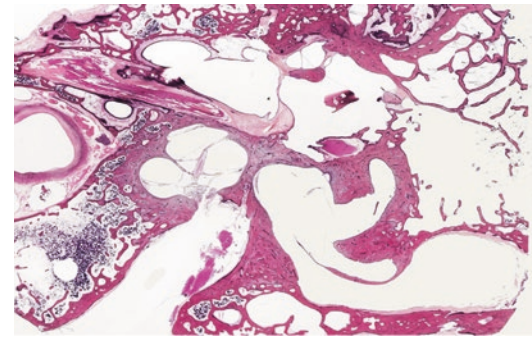


Fig. 2.5 A panoramic horizontal section of a right human temporal bone with bilateral incomplete partition type II, harvested from a 90-year-old female donor who died of pneumonia (Hematoxylin and Eosin). She had progressive hearing loss until she became completely deaf at the age of 6. Middle ear and mastoid are normal; cochlea is of normal size, flattened in the upper regions due to abnormal interscalar septum. Bony cochlear nerve aperture and internal auditory canal are normal. Vestibule is also normal, but lateral and posterior semicircular canals seem slightly dilated. The endolymphatic duct and sac are severely enlarged. There is a partial defect in the footplate, which is covered by membranous/fibrous tissue. (Source: Human temporal bone archive from the Otopathology Laboratory at the University of Minnesota (USA))

(d) *Vestibular system:* Vestibule and semicircular canals are frequently grossly dilated (Fig. 2.5), but some cases of aplasia of the posterior semicircular canal have also been described [20]. Enlargement of vestibular aqueduct is very rare—in fact, imaging studies demonstrated that the vestibular aqueduct is not seen at all in some cases [59]. This may be explained by the fact that the vestibular aqueduct is the last structure of the posterior labyrinth to develop, suggesting a developmental arrest before this structure is completely formed [39]. The vestibule is separated from the cochlea by only a very thin membrane (Fig. 2.4), but in some cases a wide communication between cochlea and vestibule can be seen [5, 20].

(e) *Other associated malformations:* Abnormalities in the stapes with areas of absent bone cover on the oval window have been described [20].

(f) *Clinical implications:* Most patients with incomplete partition type I have severe-to-profound hearing loss—it has been noted by Sennaroglu and Saatci [39] that none of their patients benefit from hearing aids. If a cochlear implant is indicated, considering that the modiolus is absent and the neuron cells are located in the marginal areas of the cochlea, straight electrodes should be used rather than ones designed to "hug" the cochlea, which may lead to poor results and displacement of the electrode into the internal auditory canal [30, 55, 60]. The surgery may be performed using the traditional transmastoid-facial recess approach, but the transmeatal approach may be used considering the possibility of aberrant facial nerve course [37, 55]. The surgeon must be prepared for profuse gusher due to the frequently observed large communication between the cochlea and the internal auditory canal, which should be sealed accordingly [30, 51, 55]. In cases of cochlear nerve aplasia, an auditory brainstem implant is indicated [15, 31]. Due to the anomalies in the footplate, recurrent meningitis is frequent among patients with incomplete partition type I [30, 55].

2. Incomplete partition type II (Mondini dysplasia): In this type of incomplete partition, the upper portion of the modiolus is absent [26]. This has been described by Mondini [2], in addition to the observations of enlarged vestibular aqueduct and dilated vestibule [20, 25, 26]. The association between enlarged vestibular aqueduct and partial modiolar defect has been demonstrated by several authors [27, 57, 61]. There are several histopathologic reports in the literature describing findings in human temporal bones with Mondini dysplasia, including absent vestibular labyrinth and vestibular nerve, enlarged vestibular aqueduct and sac, loss of both vestibular and cochlear hair cells, and atrophy of the stria vascularis [25, 26, 46, 62, 63]. However, it has to be considered that in the past most malformations (including common cavity, cochlear hypoplasia, and incomplete partition type I) were classified as Mondini's dysplasia [10].

(a) *Internal architecture:* The apex of the cochlea is frequently cystic and dilated, given the absence of the upper part of the modiolus and its adjacent interscalar septa (Figs. 2.5 and 2.6). The scala vestibuli in the basal turn of the cochlea may be dilated with its corresponded interscalar septum bulged upwards due to increased CSF pressure transmitted through an enlarged vestibular aqueduct (Figs. 2.5 and 2.6) [20, 64]. Some cases of bony defects in the base of the modiolus have been described as well [20, 27]. The internal architecture of the cochlea, especially in the basal turn, may be normal; however, different degrees of hair cell loss have been observed (Figs. 2.4 and 2.5) [26]. There were reports of spots of spiral ligament and stria vascularis atrophy as well, which were more pro-

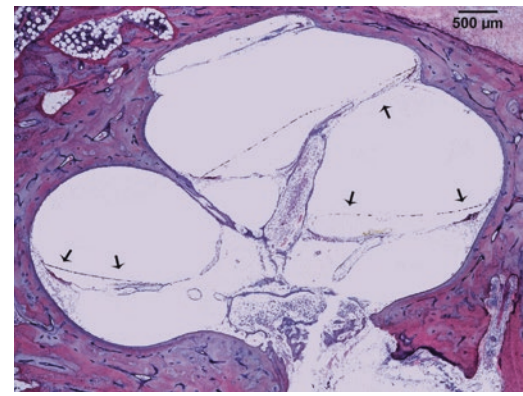


Fig. 2.6 A 4× cochlea-centered magnification of the human temporal bone section depicted in this figure (incomplete partition type II). The cochlea is of normal size, oddly shaped in the upper regions due to the abnormal interscalar septum. Scala vestibuli looks hydropic, especially in the upper basal turn, which reflects on the flattening of the Reissner's membrane (arrows pointing down) and the bulging of the interscalar septum upwards (arrows pointing up). The organ of Corti is absent in all cochlear turns, and there is atrophy of the stria vascularis and spiral ligament. The number of spiral ganglion cells is also decreased. (Source: Human temporal bone archive from the Otopathology Laboratory at the University of Minnesota (USA))

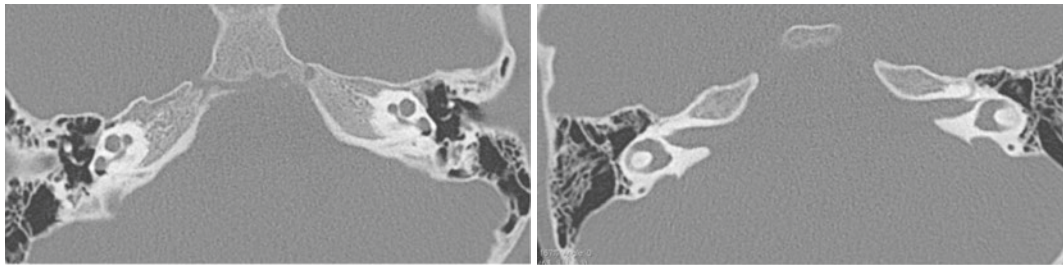


Fig. 2.7 A computed tomography scan of a female patient who had progressive bilateral mixed hearing loss, affecting the low frequencies. Figure in the left side: cochlea with cystic apex, Figure in the right side: Enlarged vestibular aqueduct

nounced in the upper cochlear turns (Figs. 2.5 and 2.6) [26].

(b) *Bony cochlear nerve canal/cochlear nerve:* Normal (Fig. 2.5).

(c) *Vestibular system:* Vestibule is frequently minimally dilated, and the vestibular aqueduct and sac are enlarged in virtually all cases (Figs. 2.5 and 2.7). The development of the vestibular aqueduct suggests that incomplete partition type II malformations occur in a later embryologic stage as compared with incomplete partition type I.

(d) *Other associated malformations:* The stapes footplate, oval window, and round window are usually unremarkable [20].

(e) *Clinical implications:* The degree of hearing loss resulting from the incomplete partition type II varies greatly—Özbal Batuk et al. [59] reported that those patients may have normal hearing or virtually any degree of conductive, mixed, or sensorineural hearing loss. However, in their casuistic, most patients (>75%) had severe-to-profound hearing loss. It is usually observed that patients with incomplete partition type II may have progressive hearing loss, possibly due to further sensorial and neural degeneration caused by the transmission of CSF pressure to the cochlea through the enlarged vestibular aqueduct and sac. Furthermore, it has been observed a positive relationship between the size of the vestibular aqueduct and presence of progressive hearing loss. At young age, patients may have near-normal or normal hearing, but as hearing degeneration progresses they eventually benefit from hearing aids. Most patients progress to (or are born with) severe-to-profound hearing loss, and thus become candidates for cochlear implants. Given the normal-sized cochlea and the normal development of the basal part of the modiolus, any type of electrode may be used. Due to the possible modiolar defects and the increased CSF pressure, either gusher, oozing, or pulsation is frequently observed intraoperatively. The cochlear nerve is always present, and therefore auditory brainstem implants are not indicated [37]. In comparison with patients with incomplete partition type I, patients with incomplete partition type II do have better functional hearing outcomes [60, 65], possibly due to the lesser architectural abnormalities in the cochlear neural structures [60].

3. Incomplete partition type III (X-linked deafness): To date, there are no histopathologic analysis of specimens with this type of malformation in the literature [20]. It is considered to be the least frequent form of inner ear malformation, accounting for 2% of all cases [30]. de Kok et al. [66] have mapped the gene responsible for the characteristic abnormalities to the Xq21 region by linkage analysis, more specifically the POU3F4 gene, located at the Xql3-q22 interval. High-resolution computed tomography scans demonstrate that

the otic capsule around the cochlea is very thin, following the outline of the membranous labyrinth. The interscalar septum is present, but cochlear base and modiolus are completely absent. The lateral end of the internal auditory canal is dilated, bulbous, and directly connected to the vestibule and cochlea, while in other cases there is a very thin separation [52, 67]. Such connection between internal auditory canal and the basal turn of the cochlea may transmit pressure from the internal auditory canal to the perilymphatic spaces, increasing the risks of gusher during surgical procedures; however, spontaneous fistulae have never been reported [20, 30]. The vestibular aqueduct is frequently abnormal, with varying degrees of dilatation [30, 68]. Cochlear nerve and cochlear nerve canal are normal. Stapes fixation is also frequent [52].

(a) *Clinical implications:* Most patients with incomplete partition type 3 have profound mixed or sensorineural hearing loss [30, 52] and also impaired vestibular function, as demonstrated by caloric and head impulse testing [52]. There are three possible explanations for the conductive component in the hearing loss: (1) stapes fixation [52, 66, 69]; (2) increase in the perilymphatic pressure due to the communication between cochlea and internal auditory canal and consequent pressure towards the footplate [70]; and (3) third-window phenomenon [71]. Given the increased pressure in the perilymphatic compartments, it is not recommended to perform stapedectomy in those patients due to the possibility of gusher and difficulties in placing the prosthesis [30, 69]. A loss in residual hearing secondary to this procedure could also be a potential complication [30, 69]. Thus, in patients with severe-to-profound hearing loss, a cochlear implant is indicated [49, 50]. Intraoperatively, the surgeon must be prepared to profuse gusher, which occurs in virtually all cases, which should be fully controlled during the procedure [69]. The literature suggests several different techniques to prevent continuous CSF leak: (1) using cochlear electrode with silicon or "cork type" stoppers to be placed in the site of cochleostomy [69]; (2) sealing around the cochleostomy site with temporalis muscle and fascia grafts [50]; (3) sealing cochleostomy site with muscle and fascia grafts followed by obliteration of the Eustachian tube and middle ear cleft [72]; and (4) subtotal petrosectomy and blind end closure of the external ear canal. In addition, considering the absence of the modiolus and separation from the cochlea and IAC, the electrode may be displaced into the internal auditory canal [69]. Therefore, ideally, the electrode to be selected should be short and have full rings or contact surfaces on both sides [69]. Pre-curved electrodes or other designs aimed to "hug" the cochlea should be avoided, due to increased risks of displacement into the internal auditory canal and damage to the facial nerve [69]. Intraoperative imaging tests must be used to ensure correct electrode positioning [50, 69]. Postoperatively, the patients should be followed periodically due to the increased risks of meningitis or intracranial infection [53].

2.4.2.7 Enlarged Vestibular Aqueduct

The first large study published demonstrating enlargement of the vestibular aqueduct was published by Valvassori and Clemis [73], in 1978. The authors demonstrated, using polytomography, presence of dilatation (>1.5 mm) of the vestibular aqueduct. It is known today that large vestibular aqueducts occur in combination with incomplete partition type II deformities [20]. However, in their series, Valvassori and Clemis found that the enlarged aqueduct was the only abnormal feature in 40% of their cases [73]. An enlarged vestibular aqueduct syndrome may lead to a variety of clinical presentations, from normal hearing to profound hearing loss [27, 65]. The hearing loss may be conductive at the beginning

(affecting preferentially the low frequencies, probably due to a third window effect) [29], but over time sensorineural hearing loss progresses, potentially leading to profound hearing loss [29]. It may occur in isolation, association with incomplete partition type II (more frequently) [20], or in association with Pendred syndrome [29]. Treatment of the hearing loss secondary to enlarged vestibular aqueducts should be performed in accordance with the degree of hearing loss (hearing aids or cochlear implants) [65].

2.4.2.8 Cochlear Aperture Abnormalities

The bony cochlear nerve canal, which allows passage of the cochlear nerve from the IAC into the cochlea, may be hypoplastic/narrow (<1.4 mm) or aplastic (canal closed by bone) [30]. They may be followed or not by narrowing of the internal auditory canal [30]. If the internal auditory canal or the bony cochlear nerve canal are hypoplastic, it is necessary to perform T2-weighted magnetic resonance imaging in axial and sagittal oblique planes to evaluate whether cochlear nerve is normal, hypoplastic, or absent [37]. Patients with absent or hypoplastic bony cochlear nerve aperture may have severe-to-profound hearing loss and may be treated with cochlear implants (cochlear nerve present) or auditory brainstem implants (absent cochlear nerve) [37].

2.5 Conclusions

Histopathologic studies of human temporal bones from donors who had malformations, in addition to improvements in the imaging tests, have brought new insights and developments in the current knowledge of inner ear malformations. However, there are still gaps to be filled regarding several anatomic and functional features associated with those abnormalities. Nonetheless, recently, information provided by those histopathologic and imaging studies allowed clinical advances in the treatment of the hearing loss associated with several of those malformations. To ensure continuous improvement in the rehabilitation of those patients, future histopathologic and imaging studies may shed additional light in the hidden aspects of the inner ear malformations.

Acknowledgements This project was funded by the NIH grant number: 1 UG3 NS107688-01, the International Hearing Foundation; the 5M Lions International; and the Starkey Foundation. The authors have no other funding, financial relationships, or conflicts of interest to disclose.

References

1. Monsanto RC, Pauna HF, Paparella MM, Cureoglu S. Otopathology in the United States: history, current situation, and future perspectives. Otol Neurotol. 2018;39(9):1210–4. https://doi.org/10.1097/MAO.0000000000001942.
2. Mondini C. Anatomica hati sectio. De Bononiensi Scientarum et Artium Instituto atque Academia Commentarii. Anat Hati Sect. 1791;7(28):419.
3. Siebenmann F. Grundzüge der Anatomie und Pathogenese der Taubstummheit. Wiesbaden: J.F. Bergmann; 1904.
4. Illum P. The Mondini type of cochlear malformation: a survey of the literature. Arch Otolaryngol. 1972;96(4):305–11. https://doi.org/10.1001/archotol.1972.00770090481002.
5. Khan AM, Levine SR, Nadol JB. The widely patent cochleovestibular communication of Edward Cock is a distinct inner ear malformation: implications for cochlear implantation. Ann Otol Rhinol Laryngol. 2006;115(8):595–606. https://doi.org/10.1177/000348940611500805.
6. Michel P. Mémoire sur les anomalies congénitales de l'oreille interne. Gaz Méd Strasbg. 1863;23:55–8.
7. Jackler RK. Chapter 152: Congenital malformations of the inner ear. In: Otolaryngology-head and neck surgery. 2nd ed. Mosby-Yearbook: Chicago, IL; 1993.
8. Scheibe A. Ein Fall von Taubstummheit mit Acusticusatrophie und Bildungsanomalien im häutigen Labyrinth beiderseits. Z Ohrenheilkd. 1892;22:11–24.
9. Alexander G. Zur Pathologie und pathologischen Anatomie der kongenitalen Taubheit. Arch Für Ohrenheilkd. 1904;61(3):183–219. https://doi.org/10.1007/BF01808972.
10. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. Laryngoscope. 1987;97(3 Pt 2 Suppl 40):2–14.
11. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41. https://doi.org/10.1097/00005537-200212000-00019.

12. Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27(5):615–23. https://doi.org/10.1097/01.mao.0000224090.94882.b4.
13. Silverstein H, Smouha E, Morgan N. Multichannel cochlear implantation in a patient with bilateral Mondini deformities. Am J Otol. 1988;9(6):451–5.
14. Tucci DL, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations. Arch Otolaryngol Head Neck Surg. 1995;121(8):833–8.
15. Monsanto RC, Bittencourt AG, Neto NJB, et al. Auditory brainstem implants in children: results based on a review of the literature. Int J Adv Otol. 2014;10(2):284–90.
16. Mangabeira-Albernaz PL. The Mondini dysplasia--from early diagnosis to cochlear implant. Acta Otolaryngol. 1983;95(5–6):627–31.
17. Terrahe K. Missbildungen des Innen- und Mittelohres als Folge der Thalidomidembryopathie: Ergebnisse von Röntgenschichtuntersuchungen. Fortschr Röntgenstr. 1965;102:14.
18. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(1 Pt 2 Suppl 106):1–26. https://doi.org/10.1097/00005537-200501001-00001.
19. Marangos N. Dysplasien des Innenohres und inneren Gehörganges. HNO. 2002;50:866–81.
20. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20. https://doi.org/10.1179/1754762815Y.0000000016.
21. Jeong S-W, Kim L-S. A new classification of cochleovestibular malformations and implications for predicting speech perception ability after cochlear implantation. Audiol Neurootol. 2015;20(2):90–101. https://doi.org/10.1159/000365584.
22. Rodriguez K, Shah RK, Kenna M. Anomalies of the middle and inner ear. Otolaryngol Clin N Am. 2007;40(1):81–96. https://doi.org/10.1016/j.otc.2006.10.006, vi.
23. Cinar BC, Batuk MO, Tahir E, Sennaroglu G, Sennaroglu L. Audiologic and radiologic findings in cochlear hypoplasia. Auris Nasus Larynx. 2017;44(6):655–63. https://doi.org/10.1016/j.anl.2016.12.002.
24. Bartel-Friedrich S, Wulke C. Classification and diagnosis of ear malformations. GMS Curr Top Otorhinolaryngol Head Neck Surg. 2008;6:Doc05. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3199848/. Accessed 26 Nov 2018.
25. Kaya S, Hızlı Ö, Kaya FK, Monsanto RD, Paparella MM, Cureoglu S. Peripheral vestibular pathology in Mondini dysplasia. Laryngoscope. 2017;127(1):206–9. https://doi.org/10.1002/lary.25995.
26. Paparella MM. Mondini's deafness. A review of histopathology. Ann Otol Rhinol Laryngol Suppl. 1980;89(2 Pt 3):1–10.
27. Hirai S, Cureoglu S, Schachern PA, Hayashi H, Paparella MM, Harada T. Large vestibular aqueduct syndrome: a human temporal bone study. Laryngoscope. 2006;116(11):2007–11. https://doi.org/10.1097/01.mlg.0000237673.94781.0a.
28. Cureoglu S, Schachern PA, Paparella MM. Scheibe dysplasia. Otol Neurotol. 2003;24(1):125.
29. Merchant SN. Genetically determined and other developmental defects. In: Schuknecht's pathology of the ear. 3rd ed. New Heven, CT: People's Medical Publishing House; 2010.
30. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411. https://doi.org/10.4274/balkanmedj.2017.0367.
31. Sennaroglu L, Ziyal I. Auditory brainstem implantation. Auris Nasus Larynx. 2012;39(5):439–50. https://doi.org/10.1016/j.anl.2011.10.013.
32. Li CW. Congenital malformation of inner ear. Dev Neurosci. 1979;2(1):7–18. https://doi.org/10.1159/000112434.
33. Ruben RJ. Development and cell kinetics of the kreisler (kr-kr) mouse. Laryngoscope. 1973;83(9):1440–68. https://doi.org/10.1288/00005537-197309000-00006.
34. Van De Water TR. Effects of removal of the statoacoustic ganglion complex upon the growing otocyst. Ann Otol Rhinol Laryngol. 1976;85(6 Suppl 33 Pt 2):2–31. https://doi.org/10.1177/00034894760850S602.
35. Schmidt RS, Sismanis A. Common cavity deformity. Otol Neurotol. 2008;29(4):567. https://doi.org/10.1097/MAO.0b013e31815ae9ef.
36. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. J Laryngol Otol Suppl. 2000;25:1–14.
37. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41. https://doi.org/10.1002/cii.416.
38. Joshi VM, Navlekar SK, Kishore GR, Reddy KJ, Kumar ECV. CT and MR imaging of the inner ear and brain in children with congenital sensorineural hearing loss. Radiogr Rev. 2012;32(3):683–98. https://doi.org/10.1148/rg.323115073.
39. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25(4):520–9; discussion 529.
40. Freeman SR, Sennaroglu L. Management of cochlear nerve hypoplasia and aplasia. Adv Hear Rehabil. 2018;81:81–92. https://doi.org/10.1159/000485542.
41. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. Am J Neuroradiol. 2002;23(4):635–43.
42. Peng KA, Kuan EC, Hagan S, Wilkinson EP, Miller ME. Cochlear nerve aplasia and hypoplasia: predictors of cochlear implant success. Otolaryngol Neck Surg. 2017;157(3):392–400. https://doi.org/10.1177/0194599817718798.
43. Beltrame MA, Frau GN, Shanks M, Robinson P, Anderson I. Double posterior labyrinthotomy tech-

nique: results in three Med-El patients with common cavity. Otol Neurotol. 2005;26(2):177–82.
44. Sennaroğlu L, Bajin MD, Pamuk E, Tahir E. Cochlear hypoplasia type four with anteriorly displaced facial nerve canal. Otol Neurotol. 2016;37(10):e407–9. https://doi.org/10.1097/MAO.0000000000001220.
45. Tahir E, Bajin MD, Atay G, Mocan BÖ, Sennaroğlu L. Bony cochlear nerve canal and internal auditory canal measures predict cochlear nerve status. J Laryngol Otol. 2017;131(8):676–83. https://doi.org/10.1017/S0022215117001141.
46. Schuknecht HF. Mondini dysplasia; a clinical and pathological study. Ann Otol Rhinol Laryngol Suppl. 1980;89(1 Pt 2):1–23.
47. Otte J, Schunknecht HF, Kerr AG. Ganglion cell populations in normal and pathological human cochleae. Implications for cochlear implantation. Laryngoscope. 1978;88(8 Pt 1):1231–46. https://doi.org/10.1288/00005537-197808000-00004.
48. Pollak A, Felix H. Histopathological features of the spiral ganglion and cochlear nerve in temporal bones from three patients with profound hearing loss. Acta Otolaryngol Suppl. 1985;423:59–66.
49. Cosetti MK, Friedmann DR, Heman-Ackah SE, Perez R, Waltzman SB, Roland JT. Surgical techniques and outcomes of cochlear implantation in patients with radiographic findings consistent with X-linked deafness. Int J Pediatr Otorhinolaryngol. 2015;79(10):1689–93. https://doi.org/10.1016/j.ijporl.2015.07.027.
50. Saeed H, Powell HRF, Saeed SR. Cochlear implantation in X-linked deafness - how to manage the surgical challenges. Cochlear Implants Int. 2016;17(4):178–83. https://doi.org/10.1080/14670100.2016.1180018.
51. Cabbarzade C, Sennaroglu L, Süslü N. CSF gusher in cochlear implantation: the risk of missing CT evidence of a cochlear base defect in the presence of otherwise normal cochlear anatomy. Cochlear Implants Int. 2015;16(4):233–6. https://doi.org/10.1179/1754762813Y.0000000048.
52. Phelps PD, Reardon W, Pembrey M, Bellman S, Luxom L. X-linked deafness, stapes gushers and a distinctive defect of the inner ear. Neuroradiology. 1991;33(4):326–30.
53. Incesulu A, Adapinar B, Kecik C. Cochlear implantation in cases with incomplete partition type III (X-linked anomaly). Eur Arch Otorhinolaryngol. 2008;265(11):1425–30. https://doi.org/10.1007/s00405-008-0614-z.
54. Pulcherio JOB, Bittencourt AG, Burke PR, et al. Carina® and Esteem®: a systematic review of fully implantable hearing devices. PLoS One. 2014;9(10):e110636. https://doi.org/10.1371/journal.pone.0110636.
55. Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N. Cochlear implant in incomplete partition type I. Acta Otorhinolaryngol Ital. 2013;33(1):56–62.
56. Reisner K. Tomography in inner and middle ear malformations: value, limits, results. Radiology. 1969;92(1):11–20. https://doi.org/10.1148/92.1.11.
57. Davidson HC, Harnsberger HR, Lemmerling MM, et al. MR evaluation of vestibulocochlear anomalies associated with large endolymphatic duct and sac. AJNR Am J Neuroradiol. 1999;20(8):1435–41.
58. Phelps PD. Mondini and "pseudo Mondini". Clin Otolaryngol Allied Sci. 1990;15(2):99–101.
59. Özbal Batuk M, Çınar BÇ, Özgen B, Sennaroğlu G, Sennaroğlu L. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233–8. https://doi.org/10.5152/iao.2017.3030.
60. Kontorinis G, Goetz F, Giourgas A, Lenarz T, Lanfermann H, Giesemann AM. Radiological diagnosis of incomplete partition type I versus type II: significance for cochlear implantation. Eur Radiol. 2012;22(3):525–32. https://doi.org/10.1007/s00330-011-2301-5.
61. Lemmerling MM, Mancuso AA, Antonelli PJ, Kubilis PS. Normal modiolus: CT appearance in patients with a large vestibular aqueduct. Radiology. 1997;204(1):213–9. https://doi.org/10.1148/radiology.204.1.9205250.
62. Zheng Y, Schachern PA, Cureoglu S, Mutlu C, Dijalilian H, Paparella MM. The shortened cochlea: its classification and histopathologic features. Int J Pediatr Otorhinolaryngol. 2002;63(1):29–39.
63. Sampaio ALL, Cureoglu S, Schachern PA, Kusunoki T, Paparella MM, Oliveira CACP. Massive endolymphatic sac and vestibular aqueduct in Mondini dysplasia. Arch Otolaryngol Head Neck Surg. 2004;130(5):678–80. https://doi.org/10.1001/archotol.130.5.678.
64. Holden PK, Linthicum FH. Mondini dysplasia of the bony and membranous labyrinth. Otol Neurotol. 2005;26(1):133.
65. Sefein I, Younes A, Omara A, Hamada S, Sami A, El Rouby I. Outcome of cochlear implantation in children with enlarged vestibular aqueduct (EVA) and Mondini dysplasia (incomplete partition type II). J Med Sci Res. 2018;1(1):17. https://doi.org/10.4103/JMISR.JMISR_7_18.
66. de Kok YJ, van der Maarel SM, Bitner-Glindzicz M, et al. Association between X-linked mixed deafness and mutations in the POU domain gene POU3F4. Science. 1995;267(5198):685–8.
67. Cremers CW, Hombergen GC, Scaf JJ, Huygen PL, Volkers WS, Pinckers AJ. X-linked progressive mixed deafness with perilymphatic gusher during stapes surgery. Arch Otolaryngol. 1985;111(4):249–54.
68. Talbot JM, Wilson DF. Computed tomographic diagnosis of X-linked congenital mixed deafness, fixation of the stapedial footplate, and perilymphatic gusher. Am J Otol. 1994;15(2):177–82.
69. Sennaroğlu L, Bajin MD. Incomplete partition type III: a rare and difficult cochlear implant surgical indication. Auris Nasus Larynx. 2018;45(1):26–32. https://doi.org/10.1016/j.anl.2017.02.006.
70. Tang A, Parnes LS. X-linked progressive mixed hearing loss: computed tomography findings. Ann Otol

Rhinol Laryngol. 1994;103(8 Pt 1):655–7. https://doi.org/10.1177/000348949410300814.
71. Snik AF, Hombergen GC, Mylanus EA, Cremers CW. Air-bone gap in patients with X-linked stapes gusher syndrome. Am J Otol. 1995;16(2):241–6.
72. Wootten CT, Backous DD, Haynes DS. Management of cerebrospinal fluid leakage from cochleostomy during cochlear implant surgery. Laryngoscope. 2006;116(11):2055–9. https://doi.org/10.1097/01.mlg.0000240286.43289.87.
73. Valvassori GE, Clemis JD. The large vestibular aqueduct syndrome. Laryngoscope. 1978;88(5):723–8.

Pathophysiology of IEMs

3

Levent Sennaroglu

Special Features

1. Membranous labyrinth development determines the final shape of the inner ear
2. Otic capsule layers have different development mechanisms; inner endosteum develops separately from outer enchondral and periosteal layers
3. Pressure transfer is responsible for IP-II

It is difficult to find a single mechanism which explains all inner ear malformations (IEM). It is possible that different mechanisms may play a role in certain anomalies. In some IEMs there may be a combination of certain factors. In most cases diminished vascular supply appears to be the most important factor for the development of IEMs.

3.1 Embryology

According to Gulya, otic placode, which is a plaque-like thickening of the surface ectoderm, can be seen at the end of third week [1] (Fig. 3.1a). Within a few days it forms auditory pit by invagination into the underlying mesenchyme (Fig. 3.1b). The endolymphatic appendage appears at this stage, much earlier than semicircular and cochlear ducts. Expansion of the auditory pit and fusion of overlying tissue create the otocyst (otic vesicle), separated from the surface (Fig. 3.1c). Mesenchymal tissue around the otocyst differentiates and forms the future otic capsule (bony labyrinth). By the fourth week, two flanges (the future semicircular ducts) arise from the otocyst. Development then involves elongation of the otocyst and the appearance of three deepening folds (I, II, and III), which demarcate the utricle with its three semicircular ducts, the endolymphatic duct and sac, and the saccule with its cochlear duct.

Gulya [2] explained cochlear development in detail. In the 6-week embryo, the cochlear duct forms as a tubular diverticulum from the saccular portion of the otic vesicle. This ventral projection coils with medial growth, completing one turn by the sixth week and its entire two and one-half turns by the eighth week (Fig. 3.2a–d). Meanwhile, the internal architecture (modiolus and interscalar septa) develops parallel to the development of the scala. Development of the membranous labyrinth starts from the basal turn and advances to the apex within 2 weeks. Having completed its requisite two and one-half turns by the 8- to 10-week stage, further growth of the cochlear duct occurs in caliber only (first basal turn, then, middle and apical turns expand to reach full size) and is essentially completed by midterm. By the 20th week, the membranous labyrinth is of maximum size and is housed

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_3

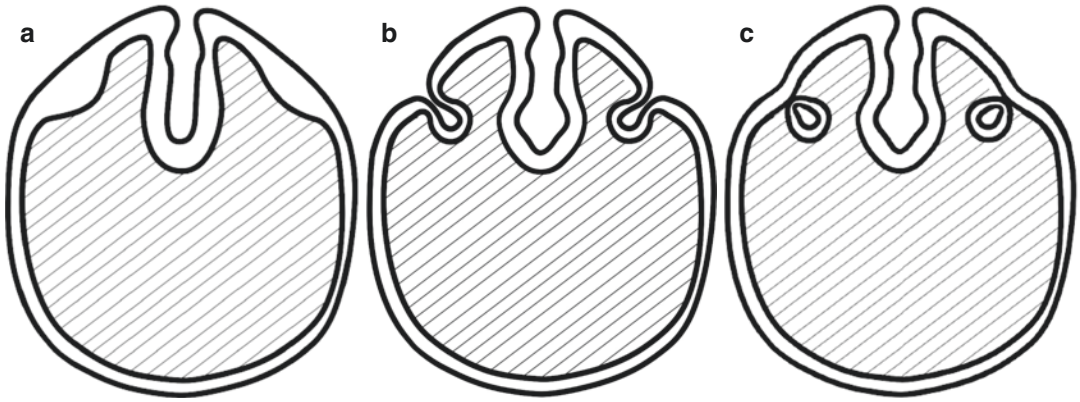


Fig. 3.1 Initial development of the inner ear. (**a**) Otic placode, (**b**) auditory pit, (**c**) otocyst

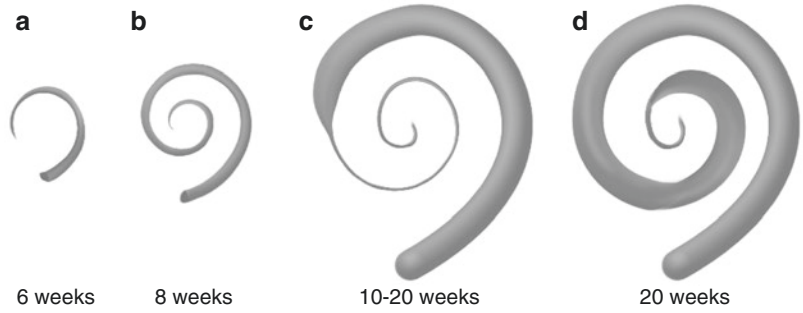


Fig. 3.2 Embryological development of cochlea. (**a**) 6 weeks, (**b**) 8 weeks, (**c**) 10–20 weeks, (**d**) 20 weeks

within a bony capsule. By 25 weeks, the inner ear displays an essentially adult configuration.

As can be seen after the cochlea finishes its coiling, turns reach full size and stop further growth. When the growth stops, ossification starts. In other words, the membranous labyrinth completes its development and the otic capsule ossification takes place. **Therefore, the shape of the inner ear is directly related to the membranous labyrinth development and the final shape of the membranous labyrinth most probably determines the type of IEM**.

Otic capsule surrounding inner ear has three layers. If embryology of the inner ear is investigated, it is noted that out of the three layers of the otic capsule, the endosteum (inner periosteal layer) has a vascular supply coming from the IAC, whereas the middle enchondral and outer periosteal layers have their vascular supply from the middle ear mucosa [3]. According to Donaldson [4], blood vessels from the IAC supply the developing modiolus, the walls of the scala, the osseous spiral lamina, and the partition between cochlear turns.

The membranous labyrinth and endosteum have vascular supply from the IAC. It looks possible that the development of endosteum and later on outer two layers of the otic capsule is parallel to membranous labyrinth development. Developmental abnormalities of the membranous labyrinth may be the reason for certain abnormalities such as complete labyrinthine aplasia, rudimentary otocyst, common cavity, and cochlear hypoplasia. Endosteum which has a vascular supply from the IAC develops parallel to the membranous labyrinth. When it completes its development, ossification of the outer two layers takes place and the final shape of the inner ear is achieved. According to the timing and location of developmental arrest (vascular supply from the IAC or middle ear), we can have different IEMs. In addition, pressure transfer plays a role in IP-II.

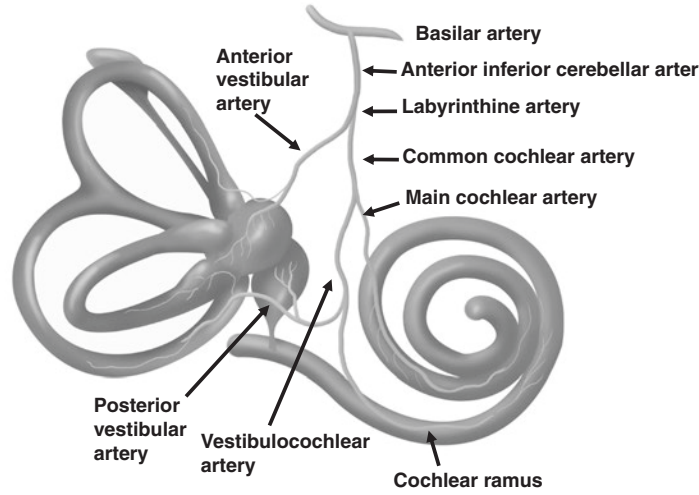


Fig. 3.3 Vascular supply of the inner ear. (Modified from Vascular anatomy, Chapter 7, page 216, in Gulya, ed. Gulya and Schuknecht's Anatomy of the Temporal Bone with Surgical Implications. Third edition ed. 2007, Informa Healthcare USA: New York)

As it will be seen, endosteum development is separate from outer two layers. If one pathology affects the development of endosteum, we can have different anomalies than pathologies affecting outer two layers. This is the main mechanism used to explain the pathophysiology of IEMs.

Vascular supply from the IAC appears to be the main cause of developmental arrest (Fig. 3.3). Disruption of main vessels probably causes more severe abnormalities. Terminal branches, however, may cause less severe abnormalities.

Complete labyrinthine aplasia (CLA), rudimentary otocyst (RO), common cavity (CC), cochlear aplasia (CA), and incomplete partition type III have no histopathological specimens. Therefore, pathophysiology is explained using information from radiological images. As a result, it is not possible to comment on the layers of the otic capsule. The remaining IEMs have histopathological specimens which were used together with radiological information to explain possible mechanisms.

Three different mechanisms are responsible for IEMs (Table 3.1).

1. **Membranous labyrinth developmental abnormality (MLDA)**. If the membranous labyrinth develops normally, this will result in normal shape of the cochlea. If there is MLDA, it is not possible to reach the stage to have the shape of normal cochlea. CLA, RO, CC, CA, and cochlear hypoplasia (CH) are all examples of MLDA where developmental arrest of the membranous labyrinth determines the final shape of the cochlea. In incomplete partitions (IP) the outline of the cochlea is normal. Therefore, MLDA is not responsible for IP anomalies.
2. **Otic capsule abnormalities**: Endosteum develops separately with a different mechanism than enchondral and outer periosteal layers. If there is endosteal abnormality, stapes footplate may be defective; IP-I and CH-II may be the result of endosteum development abnormality. In IP-III, on the other hand, outer two layers of the otic capsule are missing, with very thick endosteal layer.
3. **Pressure transfer** into cochlea: IP-II

3.2 Pathophysiology of Individual Inner Ear Malformations

3.2.1 Complete Labyrinthine Aplasia

Complete Labyrinthine Aplasia (CLA) is the developmental arrest before the formation of the otocyst and the membranous labyrinth. As there is no postmortem specimen with CLA neither in Massachusetts Eye and Ear Infirmary (MEEI)

Table 3.1 Mechanisms responsible for the development of inner ear malformations. MLDA: Membranous labyrinth developmental abnormality, CLA = Complete labyrinthine aplasia, RO = rudimentary otocyst, CC = common cavity, CA = cochlear aplasia, IP = incomplete partition CH = cochlear hypoplasia

	Time of insult	Mechanism	Membranous developmental arrest	Endosteal layer	Enchondral and outer periosteal layers	Pressure
CLA and hypoplastic petrous bone	First 2 weeks	MLDA + outer 2 layers defective + petrous bone defect	+	+	+	
CLA and otic capsule +		MLDA	+			
CLA w/o otic capsule		MLDA + outer 2 layers	+		+	
Rudimentary otocyst	Third week	MLDA	+			
CC	Fourth week	MLDA	+			
CA	Fifth week	MLDA	+			
CH-I	Sixth to eighth week	MLDA	+	+		
CH-II	Sixth to eighth week	MLDA + vascular defect from IAC	+	+		
CH-III	Sixth to eighth week	MLDA	+			
CH-IV	Tenth to 20th week	MLDA	+			
IP-I		Endosteal		+		
IP-II		Genetic EVA + pressure transmission into cochlea				+
IP-III		Enchondral and outer periosteal layers of otic capsule			+	

and the University of Minnesota (UOM), radiology is used to explain pathophysiology. Based on radiology, three different types of CLA are present. In CLA subtypes, otic capsule may be normally developed or is completely absent. This suggest that the development of the membranous labyrinth and the otic capsule may be independent from each other. Even if the membranous labyrinth is completely absent, it is possible to have the otic capsule present or absent. Therefore, there must be separate mechanisms for the development of the membranous labyrinth and otic capsule. This is most probably due to the fact that the outer two layers of the otic capsule have their vascular supply from the middle ear, while endosteal layer together with membranous labyrinth receives its vascular supply from the IAC [4]. According to Donaldson [4], the inner endosteum receives its vascular supply from the IAC, and the enchondral and outer periosteal layers get their vascular supply from the middle ear mucosa; if both vascular supply pathways are damaged, it may result in CLA without otic capsule formation; or if only the vascular supply from the IAC is damaged, it may result in CLA with otic capsule formation.

As there is complete absence of membranous labyrinth in all three subgroups, it can be speculated that vascular supply coming from the IAC is absent since the beginning of embryological

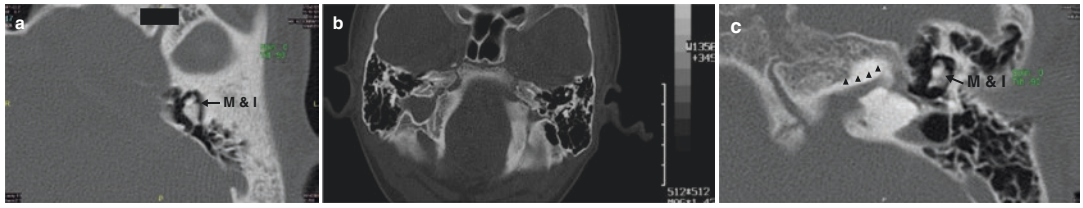


Fig. 3.4 Complete labyrinthine aplasia. (**a**) With aplasia/hypoplasia of the petrous bone, (**b**) absent otic capsule, (**c**) with normal otic capsule (▲ = course of the labyrinthine segment of the facial nerve). M&I = malleus and incus

development. Subgroups are the result of presence or absence of otic capsule.

1. **CLA with aplastic/hypoplastic petrous bone:** This is the most severe subgroup of CLA which is accompanied by aplasia/hypoplasia of the petrous bone (Fig. 3.4a). In addition to severe vascular deficiency coming from the IAC and middle ear, the petrous bone is severely defective. It is difficult to know the reason but probably greater vessels related to the development of temporal bone are involved in addition to the ones coming from the IAC and middle ear.
2. **CLA without otic capsule:** In this subgroup, formation of the petrous bone is normal, but the otic capsule is completely absent. As there is no membranous labyrinth formation, vascular supply from the IAC must be absent during the initial phase of inner ear development. Thus in this subgroup of CLA, the vascular supply from the IAC and middle ear appears to be severely damaged, resulting in the absence of all three layers of the otic capsule (Fig. 3.4b).
3. **CLA with otic capsule:** These are cases of CLA where the formation of the petrous bone and the otic capsule is normal (Fig. 3.4c). It is probably due to complete absence of vascular supply from the IAC. It may be speculated that vascular supply coming from the middle ear is normal. This is the only subgroup of CLA where facial canal can be traced in the temporal bone. Only in this group of CLA with otic capsule development, the facial canal occupies its normal location. This may show us that otic capsule formation is essential for the facial canal to obtain its normal position.

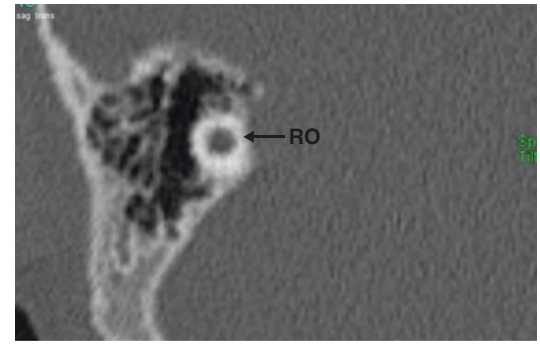


Fig. 3.5 Rudimentary otocyst (RO)

3.2.2 Rudimentary Otocyst

Rudimentary otocyst (RO) is a relatively newly described anomaly without any histopathological specimen [5]. Between the third and fourth week, the inner ear is in the form of an otocyst (otic vesicle). The insult probably occurs at the beginning of the formation of the otocyst, resulting in RO deformity. There is no IAC between RO and brainstem. However, when the images are examined, otic capsule formation around the RO appears to be normal (Fig. 3.5). This shows that it is most probably the result of membranous labyrinth developmental arrest at the otocyst stage. Membranous labyrinth development comes to an end in the stage of otocyst and otic capsule development follows this outline resulting in rudimentary otocyst. It may be due to damage of the main arterial supply from the IAC around the third week.

3.2.3 Common Cavity

There was no specimen in the MEEI or UOM collections with a common cavity (CC); and thus it is difficult to comment on the layers of the otic capsule and membranous labyrinth. When the images in the paper by Graham et al. [6] are investigated carefully, pathology may be more correctly classified as an IP-I instead of a CC. Because of this, the pathophysiology is explained on the basis of the radiologic findings.

From our clinical experience a CC contains cochlear and vestibular neural elements. Although it is impossible to determine the amount of cochlear and vestibular neural tissue, radiological findings demonstrate a round or ovoid structure, with an IAC opening into the center (Fig. 3.6a). This represents development arrest just before there is a clear differentiation into cochlea and vestibule (probably around fourth to fifth week), placing CC between rudimentary otocyst (RO) and cochlear aplasia. As both cochlea and vestibule development are arrested, pathology may be due to labyrinthine artery pathology around the fourth week. It must be differentiated from cases of **RO** and **cochlear aplasia with a dilated** vestibule (CADV). **RO** is an earlier anomaly without an IAC, where performing an ABI surgery is the only way to restore hearing. In cases of CC, if there is a well-developed cochleovestibular nerve, the patient may benefit from CI; otherwise ABI is the only treatment option for hearing restoration. **CADV** is a more developed anomaly, where the vestibule forms as a completely separate structure, located in its normal position, which is the posterolateral part of the IAC. As the cochlea is absent, an enlarged vestibule can be misdiagnosed as CC.

At the time of insult, CC is only a few millimeters in size, as a developed otocyst. CC may have small or large dimensions: usually, a CC with a diameter of 1–3 cm is encountered. This shows that its capacity to differentiate into cochlea and vestibule may terminate but it can still enlarge; so a CC larger than an initial otocyst may be encountered.

What Is the Reason for a Small or Large CC? According to explanations by Sennaroglu L [3], in embryological terms, ossification starts when the membranous labyrinth reaches its maximum size; final size of the CC may be related to the time when the membranous labyrinth terminates its development. In a RO, development of the membranous labyrinth stops at around the third week, and ossification results in a millimetric structure without an IAC. According to Gulya [2], the CVN arises from cells of the anteromedial aspect of the otic placode. During the fourth week of gestation, these cells migrate between the epithelium of the otic vesicle and its basement membrane; they then penetrate the basement membrane through minute defects to reach the region in which the VIIIth nerve ganglion forms. If the arrest is at the end of the fourth week, after the unification with the IAC, there is a CVN and a **common cavity** occurs; it seems likely that the developing membranous laby-

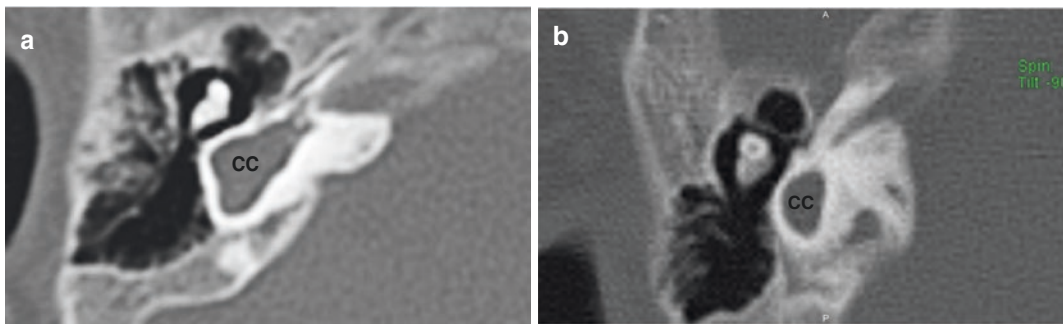


Fig. 3.6 Common cavity. (**a**) Large common cavity (CC); internal auditory canal opening into the center of ovoid structure. (**b**) Small common cavity

rinth is severely damaged because of the insult, without differentiating into a separate cochlea and vestibule and ossification continues from this stage on, resulting in a small-sized CC (Fig. 3.6b). If only the differentiation capacity is destroyed but the membranous labyrinth is still capable of enlarging, it will enlarge until the second and third layers of the otic capsule become mature and prevent further enlargement. This will result in a large CC (Fig. 3.6a). It is possible that during the early stages, the second and third layers of the otic capsule have not yet ossified, and that there is, therefore, no resistance to the expansion of the membranous labyrinth, which may continue to enlarge. The shape is ovoid or round because ossification forms following the shape of the otocyst at the time of developmental arrest.

The size of the IAC does not appear to be correlated with the size of CC. The IAC may be narrow or enlarged in a large CC. Therefore, CSF pressure does not appear to be correlated with the size of the CC.

3.2.4 Cochlear Aplasia

Computerized tomography shows bony otic capsule development in cochlear aplasia; usually otic capsule formation fills that particular space left for the cochlea, and the labyrinthine segment of the facial nerve is anteriorly dislocated. It is possible that this bone consists of enchondral and outer periosteal layers.

After the development of the otic vesicle at the end of the fourth week, the membranous labyrinth develops in three areas: the cochlea, the vestibule, and the endolymphatic duct. Cochlear aplasia is the absence of the cochlear duct, where vestibular and endolymphatic structures may develop normally (CANV) or result in dilated vestibule (CADV) (Fig. 3.7a, b). The insult most probably inhibits membranous labyrinth formation but outer two layers of the otic capsule develop normally. The time of the insult must be around the fifth week. (It is possible that genetic abnormality may be present at the beginning of inner ear development.) Otic capsule development is always normal, and the facial nerve is anteriorly displaced into the usual location of the cochlea.

It has been observed that CANV cases are bilateral and almost always symmetric, with similar features repeating in a similar way in different patients. It is very unlikely that an external cause would destroy only the cochlear bud completely, leaving the vestibular development normal. Therefore, there is a strong possibility that the origin in CANV is genetic. CADV, on the other hand, is usually asymmetric suggesting that it may be genetic or environmental [7].

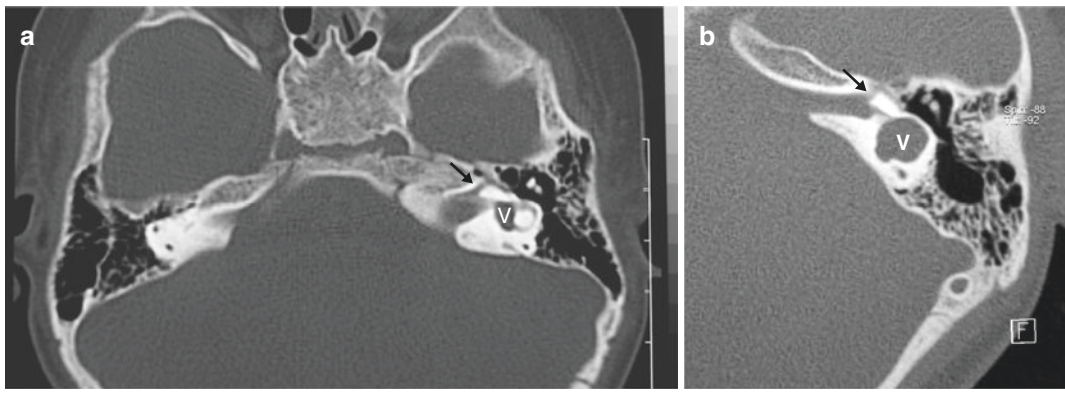


Fig. 3.7 Cochlear aplasia. (**a**) With normal vestibule, (**b**) with dilated vestibule (V = vestibule, black arrow = labyrinthine segment anteriorly dislocated to the usual location of the cochlea)

3.2.5 Cochlear Hypoplasia

In this group of IEMs, the cochlea and vestibule can be identified separately but external dimensions of the cochlea are less than those of a normal cochlea. As the cochlea and vestibule can be positively identified, separate from each other, cochlear hypoplasia (CH) group is more differentiated than common cavity. Therefore, developmental arrest must be later than sixth week and before 20th week.

There are four different subgroups, all of which have smaller external dimensions:

1. CH-I: bud type with absent internal architecture (modiolus and interscalar septa), and with or without a thin bony partition between the cochlea and the IAC (Fig. 3.8a). This is the most severe form of cochlear hypoplasia.
2. CH-II: a cystic cochlea with smaller external dimensions, with partial modiolar development (Fig. 3.8b).
3. CH-III: cochlea with smaller external dimensions, and normal internal architecture (Fig. 3.8c).
4. CH-IV: cochlea with smaller external dimensions, normal basal turn, however, with hypoplastic middle and apical turns (Fig. 3.8d).

It is possible to explain the pathophysiology with genetics and decreased vascular supply. Genetics determines the length of the cochlear duct. Cochlear duct completes one turn by sixth week, and 2.5 turns by eighth week (Fig. 3.2a–d). Further growth is by caliber only where first basal, then middle and apical turns reach adult size by midterm. By 20th week, the membranous labyrinth is maximum in size and it is housed within a bony capsule. By 25th week, the inner ear displays an adult configuration.

If development of the cochlear duct stops earlier than eighth week, we have CH-III. If, in addition, vascular deficiency is present, it may result

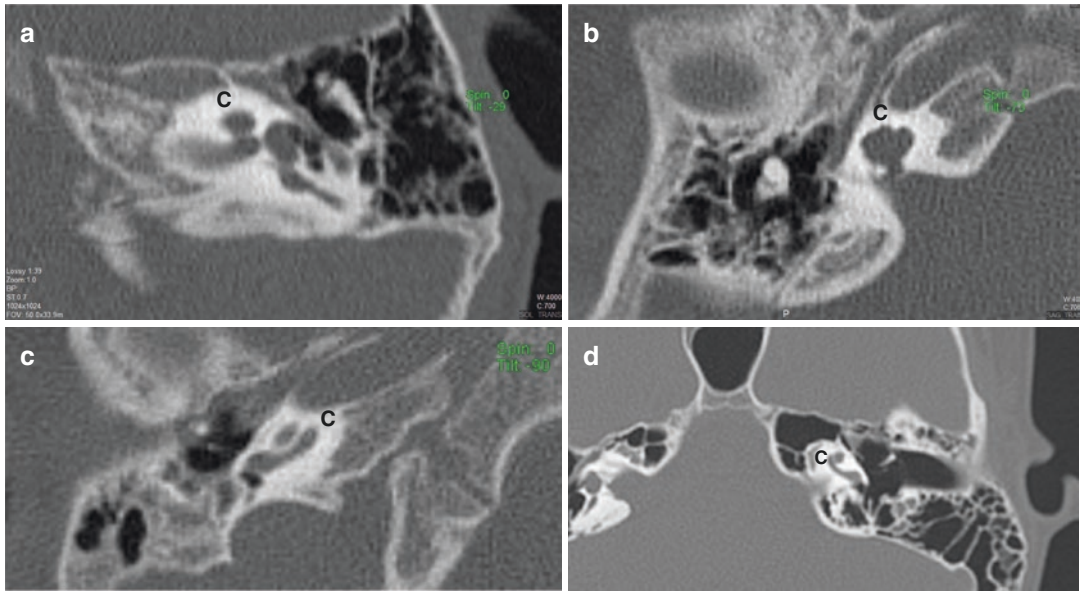


Fig. 3.8 Cochlear hypoplasia (CH): (**a**) CH-I (bud type) with absent internal architecture (modiolus and interscalar septa), and without a thin bony partition between the cochlea and the internal auditory canal, (**b**) CH-II (cystic type) with defective internal architecture (modiolus and interscalar septa), (**c**) CH-III with smaller dimensions but normal internal architecture (modiolus and interscalar septa), (**d**) CH-IV with normal basal turn but small middle and apical turns

in CH-I or CH-II. If development stops between tenth and 20th week, CH-IV may form.

It is very difficult to find a common pathophysiological explanation for the whole group of CH. This is because in some malformations, more than one factor may be responsible for the developmental anomaly. Before discussing the pathophysiology of the cochlear hypoplasia, it is appropriate to evaluate the embryology of the inner ear.

According to Gulya [2], in the 6-week embryo, the cochlear duct forms as a tubular diverticulum from the saccular portion of the otic vesicle. Cochlear duct starts coiling with medial growth, completing one turn by the sixth week and its entire two and one-half turns by the eighth week (Fig. 3.2a–d). Meanwhile, modiolus and ISS develop parallel to the development of the scala. Development of the membranous labyrinth starts from the basal turn and advances to the apex within 2 weeks. Having completed its requisite two and one-half turns by the 8- to 10-week stage, further growth of the cochlear duct occurs in caliber only (first basal turn, then, middle and apical turns expand to reach full size) and is essentially completed by midterm. By the 20th week, the membranous labyrinth is of maximum size and is housed within a bony capsule. By 25 weeks, the inner ear displays an essentially adult configuration.

It is apparent from the embryology that once the membranous labyrinth makes two and one-half turns at the eighth week, it is not possible to have cochlear hypoplasia, except CH-III and CH-IV. Therefore, developmental arrest in cases of cochlear hypoplasia must occur between the sixth and eighth week. This theory is valid for CH-I, CH-II, and CH-III. Ossification proceeds on this underdeveloped membranous labyrinth. CH-IV shows a normal-sized basal turn with hypoplastic middle and apical turns. The insult must happen during the tenth to 20th week after the basal turn reaches its full size, and before the middle and apical turns develop to their final size.

Before ossification starts, the cochlear duct attains its final length, with fully developed internal architecture. If the final length of the cochlear duct is smaller than normal, this results in CH. Vascular supply from the IAC is vital for internal cochlear development. If the vascular supply from the IAC is normal, this results in normal internal architecture, forming either CH-III or CH-IV. If the vascular supply from the IAC is additionally defective, the internal architecture will be defective, resulting in CH-I or CH-II (MLDA ± endosteal defect).

Erixon [8] demonstrated that the length of the cochlea shows variation among normal individuals; the lower limit is, therefore, a good starting point to understand cochlear hypoplasia with normal internal architecture. Some types of hypoplastic cochlea (CH-III and CH-IV) have dimensions less than the inferior limit of normal cases. A cochlea consisting of a small basal turn and a small apical turn is regarded as hypoplastic, and this is usually accompanied with semicircular (SCC) abnormalities. If CH-III and CH-IV are examined, it can be seen that they are smaller versions of a normal cochlea. According to Donaldson [4] the otic capsule ossification does not start until the membranous labyrinth reaches full size. This means that CH-III and CH-IV are most probably genetically predetermined to have a small size and development of the membranous labyrinth stops at a point earlier than normal, resulting in a shorter membranous labyrinth. At the time of complete ossification, this results in a cochlea with small external dimensions and normal internal architecture.

Therefore, in CH-III, the developmental arrest in the membranous labyrinth most probably occurs between 6 and 8 weeks, resulting in a cochlea whose dimensions are smaller than normal, with normal internal architecture. As the internal architecture is normal, vascular supply from the IAC must be normal (Membranous labyrinth developmental abnormality).

In CH-IV there is a normal basal turn, but small middle and apical turns. Arrest in the membranous labyrinth must be between tenth and 20th week, after the basal turn reaches full size, but before the middle and apical turns enlarge to their normal dimensions (Membranous labyrinth developmental abnormality).

In CH-I and CH-II there is arrested development of the internal architecture in addition to a

small-sized cochlea. In CH-I there is an oval or round, bud-shaped cochlea, not resembling the normal external shape of the cochlea. This implies that cochlear duct length must have stopped earlier than normal. The fact that the architecture is severely deformed shows that modiolar and endosteal development is also abnormal. Most likely, these areas have a severely defective vascular supply from the IAC. The main cochlear artery must be defective, resulting in defective endosteal development with an absent modiolus and ISS. The result is an undifferentiated bony bud. CN is usually hypoplastic or aplastic (membranous labyrinth developmental abnormality and severe otic capsule abnormality (endosteal layer)).

CH-II is better developed than CH-I. The outline of CH-II resembles that of a normal cochlea. It is round or ovoid with a partial modiolar defect. The modiolar base is normal, showing that only the internal radiating arteriole from the main cochlear artery may be defective, while the cochlear ramus of the vestibulocochlear artery supplying the base of the modiolus is normal (Fig. 3.2) (membranous labyrinth developmental abnormality and mild otic capsule abnormality (endosteal layer)).

Benefit from CI

In CH-II cases there is better development compared to CH-I cases. The basal part of the modiolus is present, and theoretically these patients may have better hearing. Patients with CH-III and CH-IV have normal internal architecture development, so they have more chance to benefit from CI if they have a separate CN. In CH BCNC may be aplastic together with hypoplastic/aplastic CN. Kondo [9] reported a case of CH-II with obliterated BCNC and absent spiral ganglion cells. These cases may not benefit from CI.

Coexistent Pathologies

CH-II and IP-I have similar mechanisms responsible for their development. Both pathologies are similar because both have a cystic cochlea with similar clinical features. But only their dimensions are different: in CH-II, the external dimensions are less than in IP-I. Both anomalies have similar clinical features; footplate fistula, CSF gusher during cochleostomy. In MEEI collection, there was a specimen with CH-II which had a subtotal modiolar defect with an intact bony partition on one side with an IP-I on the contralateral side. As there are two different pathologies on each side, it is very unlikely that there is a genetic etiology for this patient. It appears that the two sides are affected at different levels after the insult. These different anomalies can be seen in the same patient and a case with IP-I on one side and CH-II on the contralateral side is reported in Chap. 23 (Case 23.7).

The clinical findings in CH-II and IP-I are similar. The author has operated on six CH-II cases for CI; there was a gusher in four of these patients. A spontaneous CSF fistula can be seen in CH-II, and he has operated on one case with a defect in the stapes footplate. These may be due to defective development of the endosteum as a result of decreased vascular supply.

The accompanying vestibular system may be hypoplastic as well, as was demonstrated in a histopathologic report by Sekhar [10]. This can be explained by the developmental arrest of the membranous vestibular labyrinth. If the development of the membranous labyrinth stops earlier than normal, a hypoplastic or rarely aplastic vestibular system may occur, because the otic capsule development will follow the underdeveloped membranous labyrinth. This is most probably genetic.

It is also common to have stapes fixation in CH. Majority of the stapes fixation cases are observed in CH-II and CH-III; and this can be explained by embryology: the stapes footplate is part of the otic capsule, and according to Donaldson [4], the base of the stapes is originally continuous with the otic capsule, and then it is segregated through a retrogressive process in the cartilage. The reorganized tissue will be annular ligament. A transcapsular channel (fissula ante fenestram) is formed as a result of invasion of the primitive cartilage by periotic tissue. If there is an arrest of the otic capsule development before the formation of the footplate, it is natural that the stapes will become fixed to the oval window.

The CH-II cases are accompanied by stapes fixation and a small modiolus. Because of the resolution of HRCT, the partial modiolar defect may not be diagnosed, but histopathological examination shows the defective modiolus in all cases. Because of the shorter cochlea they have SNHL, while the fixed footplate provides the conductive component. The author has performed stapedotomy in cases of CH with mixed hearing loss; postoperatively these cases benefit more from HA. Patients with CH who have profound SNHL are candidates for CI. CH with cochlear aperture aplasia necessitates an ABI.

The external outline of CH-II cases resembled that of a normal cochlea, but it was rounder; the absence or hypoplasia of the ISS gave the cochlea a rounder shape, and the presence of ISS provided the normal shape of the cochlea. It is possible to explain this due to vascular deficiency of the main cochlear artery, as the basal part of the modiolus is normal.

Etiology may be due to genetic and environmental factors [3]. If the etiology is genetic, the size is smaller than a normal cochlea, and the endosteum follows the development of the membranous labyrinth. The development of membranous labyrinth stops at a predetermined point caused by the genetic abnormality, and ossification proceeds as in a normal case. If the etiology is environmental the space for the cochlea is normally present; but as the membranous development unexpectedly terminates earlier, the excess space is filled with a thicker enchondral layer [3]. This shows us that the place for the cochlea is normally predetermined.

Incomplete Partition Anomalies

In incomplete partition (IP) anomalies, there is a clear differentiation between the cochlea and vestibule, and dimensions and external shape of the cochleovestibular system are similar to normal cases. Therefore, it can be assumed that membranous labyrinth development is normal. Different anomalies are the result of otic capsule anomalies or pressure transfer into cochlea via EVA.

3.2.6 Incomplete Partition Type I

This is the cochlea which has external dimensions similar to a normal cochlea, but with absent modiolus, and interscalar septa (ISS). It is accompanied by a grossly dilated vestibule (Fig. 3.9a).

In IP-I membranous labyrinth development is normal, but there is endosteal development abnormality. There are internal architecture abnormalities but outer two layers of the otic capsule are normal.

There were five specimens with IP-I cochlea in the temporal bone collection of MEEI. No specimen with IP-I anomaly was present in UOM collection. Histopathological findings in IP-I can be summarized as follows:

1. Otic capsule around the IP-I cochlea consisted of three layers, but endosteum (inner periosteal lining) was much thinner than a normal cochlea, and at some locations it was completely absent. However, the middle enchondral and outer periosteal layers appeared to have developed normally.
2. Modiolus and interscalar septa (ISS) were defective in all cases. Four specimens had **subtotal modiolar defects** where only the base of the modiolus was present forming a thin, bony partition between the cochlea and the IAC (Fig. 3.9b). Remaining specimen showed **complete absence of modiolus**, forming a wide communication between the IP-I cochlea and the IAC (Fig. 3.9c). All five specimens also displayed a wide connection between the cochlea and the vestibule.
3. ISS was absent together with hypoplasia or aplasia of modiolus resulting in severely deformed internal architecture of the cochlea. The absence of the ISS, together with a thin endosteum, gave the cochlea a rounder shape than is found in the normal cochlea with interscalar partitions.
4. Stapes footplate defect: This was present in three specimens. The bony footplate was par-

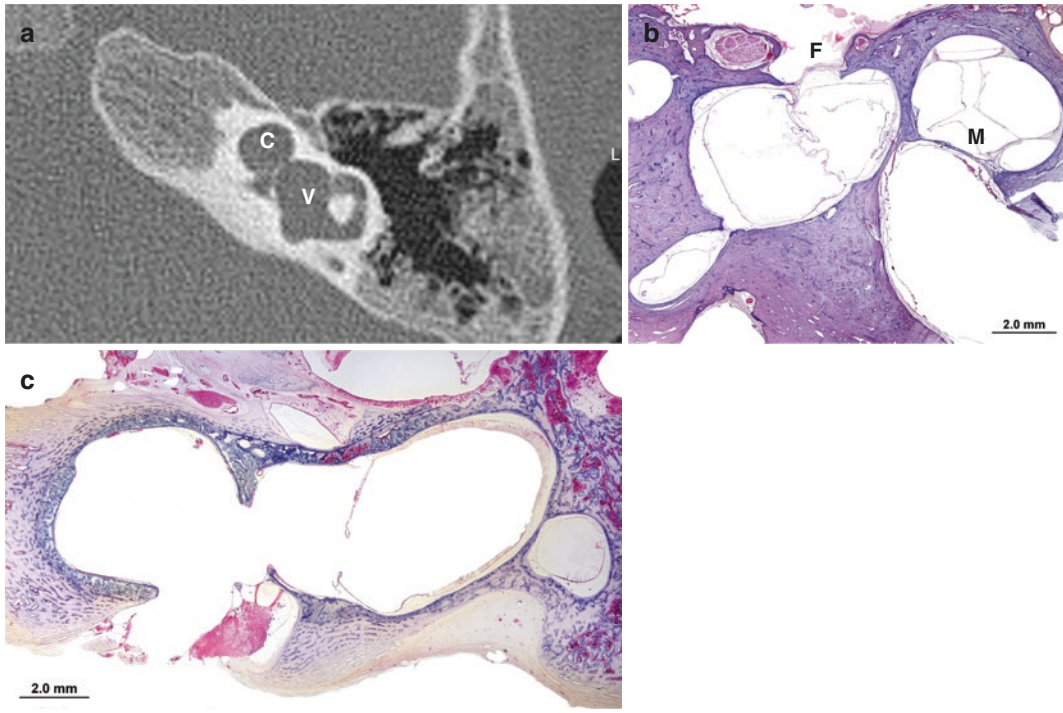


Fig. 3.9 Incomplete partition (IP) type I. (**a**) IP-I cochlea (C) with normal external dimensions but absent modiolus and interscalar septa. It is accompanied by a grossly dilated vestibule (V). (**b**) IP-I cochlea **subtotal modiolar defect** (type VI modiolar defect) where only the base of the modiolus (M) is present forming a thin, bony partition between the cochlea and the IAC. Footplate (F) is defective proving that footplate defect is not due to high CSF pressure but endosteal developmental abnormality (with permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary). (**c**) IP-I cochlea **total modiolar defect** (type VII modiolar defect) forming a wide communication between the IP-I cochlea and the IAC. (With permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary)

tially replaced either by a fibrotic or a thin membrane (Fig. 3.9b). In none of these three specimens, there was any connection of the cochleovestibular space with CSF-containing IAC, because the basal part of the modiolus was present.

5. All IP-I cochleae were accompanied by a dilated vestibule. The utricle and saccule were dilated in all specimens: three specimens had dilatation in all the SCCs. Two specimens had a dilated superior and lateral SCC, but the posterior SCC was aplastic bilaterally. As in the cochlear findings, the endosteum was defective all around the vestibular system, while the middle enchondral and outer periosteal layers were normal.
6. The basilar part of the modiolus was intact in four specimens, preventing any connection with the CSF-containing areas, and making it unlikely that pressure was the cause of footplate fistula or dilatation in the vestibular system.

Pathophysiology

The majority of the specimens had a subtotal modiolar defect, and only one showed complete absence of the modiolus. The ISS was absent in all cases. Three of the five cases with IP-I had a bony defect at the stapes footplate. The defect was covered with a thin membrane. One of the remaining patients had an aplastic oval window. Embryologically, the vestibular part of the stapes footplate is derived from the endosteum, and a

defective endosteum may be responsible for this defect.

As the shape of the cochlea is normal, it can be assumed that membranous labyrinth development is normal. There are three possible explanations for the observed pathologies:

1. **High CSF pressure inside the cochlea as a result of open communication with the IAC:** According to this hypothesis, observed changes and an occasional fistula at the oval window may be due to CSF filling the cochlea and exerting continuous pulsating pressure. On HRCT, it is seen that these patients have a defective fundus between the IAC and the cochlea. Although the fundus appears to be defective on the HRCT, not all patients have CSF leakage during cochleostomy. Out of the 50 patients with IP-I who received CI at Hacettepe University, 26 did not have CSF leakage (oozing or a gusher) during cochleostomy. This implies that although the fundus appears to be defective, there is a separation between the cochlea and the IAC, and CSF does not communicate with the cochlea in all cases. Histopathological support for the latter hypothesis was present in the specimens, because four of them had a thin, bony modiolar base separating the cochlea from the IAC; at today's level of radiological precision it may not be possible to determine this on HRCT or MRI. Therefore, the cochlea is not always filled with pulsating CSF. Based on these observations, the developmental anomaly of the internal architecture of the cochlea, as well as the footplate, could not be the result of high pulsating CSF pressure coming from the IAC and acting on these structures, because the thin bony partition separated the IP-I cochlea and IAC completely. In addition, the fundus defect in IP-III was larger and present in all cases, but no fistula was observed in the oval window. As a result, high CSF pressure cannot be held responsible for the observed changes.
2. **Timing of the insult:** Modiolar abnormalities resulting in a thin bone separating the IP-I cochlea and IAC may be explained by the timing of the insult. The base is the first part of the modiolus to develop. This is then followed by the middle and apical part of the modiolus. The subtotal modiolar defect can be explained by the developmental arrest taking place after the formation of the base: a thin bony layer is frequently formed before the insult. However, it is difficult to explain the associated abnormalities, such as the stapes footplate defect, ISS defect, and dilatation in the vestibular system, with this theory.
3. **Defective endosteum due to deficient vascular supply from the IAC:** Endosteal layer (innermost layer of the otic capsule) in all five patients with IP-I was thinner and defective all around the cochleovestibular space when compared to normal cochlea, while the middle enchondral and outer periosteal layers were normal.

If the embryology of the inner ear is investigated, it is noted that out of the three layers of the otic capsule, the endosteum (inner periosteal layer) has a vascular supply coming from the IAC, whereas the middle enchondral and outer periosteal layers have their vascular supply from the middle ear mucosa. According to Donaldson [4], blood vessels from the IAC supply the developing modiolus, the walls of the scala, the osseous spiral lamina, and the partition between cochlear turns. If Fig. 3.9b, c, which is a characteristic example of IP-I, is examined, it can easily be noticed that all these structures are absent. Therefore, it looks quite possible that histopathological changes in IP-I may be a result of defective blood supply from the IAC. As the vestibular surface of the stapes footplate is also derived from the endosteum, defective footplate development may be the result of abnormal endosteal development due to defective vascular supply from the IAC. During development of the fetus, vascular channels are being formed circumferentially on the footplate, through the growth of the endosteal bone around blood vessels that are already present. Therefore, reduced vascular supply may cause a defective footplate.

Three of the specimens had a thin, but intact modiolar base, and a stapes footplate defect at the

same time. Intact modiolar base prevents CSF from inside cochleovestibular space. Therefore, it prevents high CSF pressure acting on the footplate to create a fistula. These rare patients demonstrate that high CSF pressure cannot be held responsible for bony defects at the stapes footplate. The author has operated on nine cases of IP-I with spontaneous CSF fistula and found that there was a cystic structure present at the stapes footplate. Once it was punctured, CSF came in from this defect. This was repaired by inserting a piece of fascia through the defect into the vestibule, in a dumbbell shape. It is possible that in IP-I cochlea, where the modiolus is completely absent, high CSF pulsations acting on the thin membrane at the footplate may easily produce an oval window fistula. If there is a bony separation between the IAC and the cochlea, the footplate defect may not be noticed at all during the patient's lifetime. One case had recurrence of meningitis and she was operated with subtotal petrosectomy (see Case 23.4 in Chap. 23 Incomplete Partition Type I).

None of the nine spontaneous CSF fistulas due to inner ear malformation operated on by the author had a fistula at the round window. All of the reported spontaneous CSF fistula cases in IP-I in the literature are located at the oval window [11–14]. This also shows that this is observed in cases with a defective footplate. In IP-III there is a larger defect in the fundus, a high pulsating CSF pressure in the cochlea, and always a severe gusher upon cochleostomy; but a spontaneous CSF fistula has never been encountered. The reason may be that the endosteum, which is deficient in IP-I (probably due to defective vascular supply coming from the IAC), is properly formed in IP-III. In IP-III, it looks possible that the endosteum is well developed (and may even be hyperplastic as well), causing no fistula at the stapes footplate and preventing spontaneous CSF fistula formation.

Stapes footplate defect is most probably present at birth. High CSF pressure can cause a fistula through the defective footplate, or otitis media during childhood may result in recurrent meningitis. All cases that have been operated on by the author were children. No adult patient has been operated on so far: it appears that it is not a progressive disease, and that it must be present during childhood. If it had been the result of high pressure only, it would have been possible to see this clinical entity at all ages. This shows us that high pressure is not necessary all the time for the development of the oval window fistula. The defective development is most probably a result of a deficient endosteum present at birth, but high CSF pressure may produce a fistula in this already defective area.

As the external dimensions of the cochlea are similar to those of a normal cochlea, it can be assumed that the vascular supply from the middle ear is normal, and hence the outer two layers of the otic capsule develop normally. This is most probably the factor preventing abnormal dilatation of the cochlea. The pathology is at the fundus, modiolus, and interscalar septa. The vascular supply of the modiolus and endosteum comes from the arteries of the IAC. In IP-I the etiology appears to be the defective vascular supply coming from the IAC.

This causes different stages of developmental pathology in the modiolus and cribriform area. It is possible to observe a very thin layer of bone between the IAC and IP-I cochlea (a subtotal modiolar defect). As it is very thin, it cannot be detected with present-day radiological modalities such as HRCT and MRI. This is the reason why there is no oozing and no gusher in patients where the base of the cochlea appears to be defective—because the ¼ lower part of the modiolus may be present. As mentioned before almost half of the IP-I cases did not have a CSF leakage during cochleostomy. The reason for not having any CSF leakage must be the thin layer of basal modiolus, which cannot be demonstrated with HRCT and MRI. It is possible to explain this finding through developmental embryology. According to Gulya [2], the ¾ superior part of the modiolus receives its vascular supply from the internal radiating arteriole of the main cochlear artery. The ¼ basal part of the modiolus receives the vascular supply from the cochlear ramus of the vestibulocochlear artery. In cases with a thin layer of bone at the fundus, the former artery must be damaged while the latter is intact. In cases where the modiolus is completely absent,

with a wide connection between the cochlea and the IAC, both arteries are affected.

All these features are valid for CH-II as well. The only difference is the size of the cochlea, which is less in CH-II when compared to IP-I. The occurrence of a gusher, a spontaneous CSF fistula at the footplate, and all other clinical findings, can be observed here as well. One important clinical difference is the fact that in CH-II shorter electrodes (around 20 mm in length) should be used, whereas in IP-I, electrodes with a length of 25 mm can be used. If longer electrodes are used in cochlear hypoplasia, they will make more turns in the cochlea, and this may cause electrode displacement into the IAC.

Dilatation in the vestibule: The accompanying vestibular system showed dilatation which was present in the vestibule as well as the SCCs. This dilatation occurs only internally where external dimensions are normal. As indicated before, the outer two layers of the otic capsule, receiving vascular supply from the middle ear, are normal. Dilatation in the vestibular system is most probably due to a defective endosteum as well. Once the endosteum is defective, the factor limiting the enlargement of the membranous labyrinth may not be effective, resulting in a dilated vestibule and SCCs. The outer two layers of the otic capsule appear to be normally developed all around the vestibular system. This may be the factor limiting abnormal external enlargement of the vestibular system. These two layers have their vascular supply from the middle ear mucosa, and hence of different origin than the IAC. In all cases there was a wide connection between the cochlea and vestibule, as indicated before by Khan [15].

3.2.7 Incomplete Partition Type II

In IP-II, the size of the cochlea is normal. Therefore, IP-II is not due to membranous labyrinth developmental anomaly. Endosteum and outer layers of otic capsule are normal.

In IP-II, the apical part of the modiolus is defective, giving rise to a cystic cochlear apex with normal external dimensions (Fig. 3.10a). This anomaly is accompanied by a minimally dilated vestibule and an enlarged vestibular aqueduct (EVA) (Fig. 3.10b).

There were four specimens in MEEI and two specimens in UOM with IP-II pathology: Histopathological findings in IP-II can be summarized as follows:

1. All four specimens had a cochlea with an enlarged scala vestibuli (SV). The interscalar septum (ISS) was pushed upwards in all four specimens (Fig. 3.10c). The way that the ISS is expanded upwards in all four specimens gives a strong impression that it is the result of high pressure inside the SV. In two specimens the scala media was normal, while in the other two it was considerably compressed. The scala tympani was normal in all four specimens; and all four specimens had a very large endolymphatic sac (EVS) and aqueduct (EVA).
2. Two specimens had partial modiolar defects where superior 2/4 of the modiolus was defective (Fig. 3.10d).
3. None of the six cases had a defective stapes footplate. All three layers of the otic capsule were completely normal around the cochlea and vestibule. All SCCs were normal.
4. The specimens were bilateral and symmetric, and each had an enlarged vestibular aqueduct (EVA). It was noted that the endolymphatic epithelium in the vestibule showed mild dilatation. The saccule showed slight dilatation in all specimens, while the utricle was slightly dilated in two cases and normal in the remaining two. Interestingly there was no dilatation in the scala media, which is the continuation of the endolymphatic system. The scala vestibuli (SV), however, showed dilatation in all four specimens, where the hydropic changes had resulted in expansion of the ISS upwards (Fig. 3.10c). Instead of at its normal origin, the ISS was attached to a higher point, indicating that the pressure increase in the SV was the causative factor. This was present in all four specimens in the same way, in the canal side of the cochlea. There was no defect in the modiolus, connecting the CSF-containing IAC and the scala vestibuli.

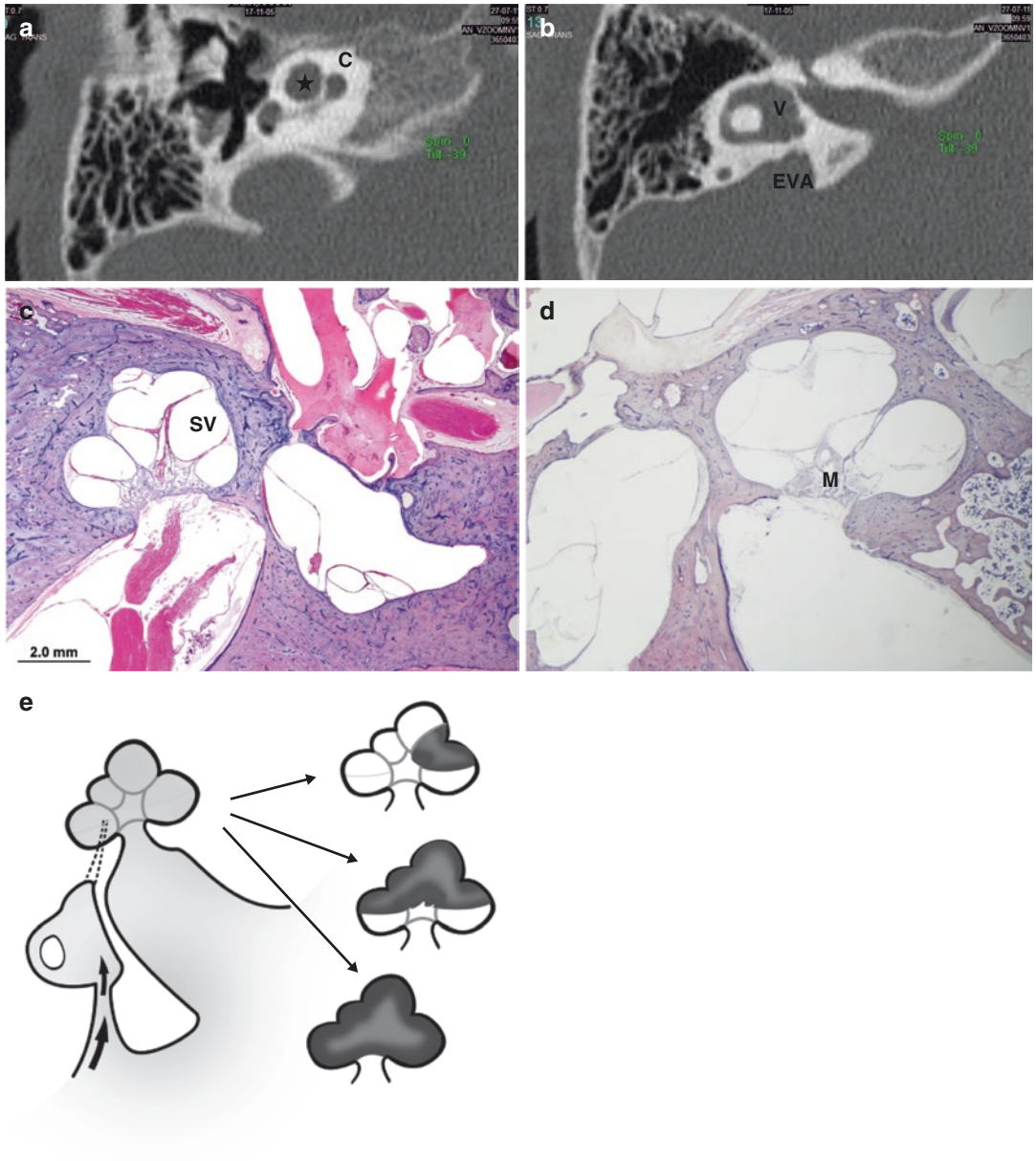


Fig. 3.10 Incomplete partition type II (IP-II). (**a**) IP-II cochlea (C) with normal external dimensions and cystic apex (star), (**b**) IP-II cochlea accompanied by a minimally dilated vestibule (V) and an enlarged vestibular aqueduct (EVA), (**c**) IP-II cochlea with scala vestibuli dilatation (with permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary), (**d**) IP-II cochlea with partial modiolar (M) defect (type V modiolar defect) (with permission of Department of Otolaryngology of University of Minnesota), (**e**) IP-II cochlea. Pressure transmission via enlarged vestibular aqueduct resulting in various modiolar defects

Scala communis was observed in all four specimens; this was always in the apical part. The scala tympani was normal in all the specimens without any dilatation. The scala media was completely normal bilaterally in one individual, and compressed bilaterally in the second one.

A similar case of scala vestibuli dilatation was reported histopathologically by Holden and

Linthicum [16], demonstrating the association of EVA and SV hydrops. According to the severity and timing of EVA, the pathology may stay at this stage, or cause destruction in the apex, resulting in the cystic cochlear apex originally described by Mondini and later called IP-II. During CI surgery in IP-II or EVA, high pulsation is almost always noted. This may result in the loss of the hair cells, causing progressive SNHL.

None of the six cases had a defective stapes footplate, and the endosteal layer was normal in all cases. This finding also demonstrates that a defective endosteum (inner periosteal lining) in IP-I may be responsible for footplate defects.

Pathophysiology

Observed pathology is SV dilatation (bilaterally in four specimens) and partial modiolar defects (bilateral in two specimens). EVA appears to be genetic and transmits the CSF pressure into the inner ear causing pathologies ranging from SV dilatation to various levels of modiolar defects. If it occurs early in life and the size of the defect is large, it is even possible to have complete modiolar defects resulting in gusher during CI surgery.

There are three possible theories to explain the pathophysiology of IP-II and EVA:

1. **Production of a large quantity of endolymph:** As the endolymphatic sac is responsible for endolymph production and is grossly enlarged, it is natural to blame endolymph overproduction for cochlear destruction. According to this theory, the production of an abnormal quantity of endolymph will cause dilatation in the endolymphatic compartment and allow transmission of high pressure into the cochlea, creating cochlear abnormalities. On the contrary, histopathological examination revealed the opposite. MEEI specimens showed either no dilatation or minimal dilatation in the saccule and utricle, which are part of the endolymphatic system [3]. In addition, ductus media is completely normal bilaterally in two specimens, and completely compressed bilaterally in the remaining two. This shows that it is very unlikely to explain cochlear morphological changes (expansion of SV and modiolar destruction) and minimal dilatation in the vestibule by increased endolymph volume or endolymphatic hydrops.
2. **Decreased cochlear vascular supply:** Decreased vascular supply may be the cause of developmental abnormality of the modiolus according to this theory. The cochlear artery which is a branch of labyrinthine artery provides the main supply of the modiolus and the endosteal layer. In IP-I both modiolus and endosteal layers were deformed, suggesting vascular origin. The scala media, tympani, and organ of Corti were completely normal. It is difficult to explain the expansion of SV on the basis of decreased vascular supply where the rest of the inner ear structures are completely normal. If there was a decrease in vascular supply, one would more probably expect an absent or thin endosteum, abnormalities of the scala, and a bony ISS and modiolus. It is, therefore, very difficult to explain the cochlear abnormalities in IP-II with the theory of unilateral decreased vascular supply.
3. **Transmission of high CSF pressure into the cochlea:** The cochlea, with its excellent bony otic capsule, is well protected from the high pulsating pressure of the CSF. Any bony opening in the otic capsule or fundus may allow the transmission of this pressure into the cochlea. This abnormally high and pulsating pressure has been observed almost on every occasion by surgeons when they perform a cochleostomy in cases of EVA (see Case 24.4 in Chap. 24). Even though there is no CSF leakage, pulsation is frequently observed. This may be the reason for the continuous pressure transmission into the inner ear, which results in vestibular dilatation and SV dilatation, modiolar defects, and progressive SNHL. A large sac with more endolymph production and/or high pressure may cause EVA.

There is variability among individuals. The EVA may have different patency in individuals: it may be enlarged, but if it is filled with fibrous tis-

sue, it may not allow pressure transmission into the cochlea. In such a case hearing loss may occur later in life. Depending on the transmission of the pressure into the cochlea and timing, this may result in variable defects in the cochlear apex, as shown in Fig. 3.10e.

During CI surgery in cases with IP-II, high pulsation is almost always noted (Video 15.1). This is the result of pressure transmission (without CSF itself) via the EVA into the cochlea. This may cause erosion of the modiolar apex, resulting in cystic apex (a superior or partial modiolar defect, third finding in IP-II). In patients with higher pressure transmission there may be more destruction, resulting in subtotal or total modiolus defects (Fig. 3.10e). CSF leakage during cochleostomy is not due to EVA. It is due to modiolar defects which are created as a result of pressure transmission by EVA. The author has operated on 93 cases with IP-II. Six of these had a severe gusher. This is due to the complete modiolar destruction enabling CSF to reach inside the cochlea, which produces a gusher during cochleostomy. Oozing is a less severe form of CSF leakage, and it is due to a partial modiolar defect incompletely connecting the cochlea and IAC. Oozing has occurred in 42 of the 93 cases with IP-II. If there is oozing or a gusher, most probably this comes via a defect in the modiolus. The remaining patients did not have any CSF leakage; only pulsation was present.

We still do not know the etiology of EVA. This usually occurs bilaterally and symmetrically in many patients. When the radiological information from patients with an enlarged vestibular aqueduct (EVA) and sac is combined with the information from the histopathological study, an enlarged endolymphatic sac (EES) appears to be the only genetic abnormality that is causing the other abnormalities by allowing transmission of high CSF pressure to reach inside the cochlea and vestibule. EVA may be an accompanying anomaly due to hypersecretion of endolymph by the abnormal sac. During fetal life, the pressure inside the endolymphatic canal may be higher than normal, because of the high volume of endolymph produced by the abnormal sac. This may cause enlargement in the surrounding VA, which is cartilaginous and soft at this stage. The EVA, in turn, transmits the high pulsating CSF pressure into the vestibule. Within the vestibular system, CSF pressure produces a mild dilatation in the walls of the vestibule (second finding in IP-II), with mild or no enlargement in the endolymphatic system. If there had been an enlarged (hydropic) endolymphatic system, a high volume of endolymph might have been the cause of cochlear destruction. However, no hydropic changes were observed in the endolymphatic space. Depending on the severity and timing of the insult, the pathology may stay at this stage and cause EVA only, or it may be transmitted into the cochlea, causing a spectrum of anomalies ranging from SV dilatation, scala communis, superior (cystic apex), partial, subtotal, and in some cases complete modiolar defects. In the cochlea, the SV adjacent to the IAC is enlarged. In two specimens the scala media was normal, while in the other two it was considerably compressed. This shows that pressure transfer is not via the ductus reuniens, which would have caused enlargement of the scala media. The result is hydrops in the SV. The scala tympani was normal in all four specimens. The high pressure in the SV causes bulging of the ISS upwards. This is a constant finding in all four cases, showing that cochlear pathology may be the result of high pressure in the SV and that it happened during the developmental phase. In the present cases, this must have occurred during the fetal development period when it was still cartilaginous; otherwise high pressure would have caused fracture of the osseous spiral lamina. If there is higher pressure, it is natural to expect more destruction at the upper, and possibly the lower part of the modiolus. This is the reason for the CSF oozing and gusher sometimes observed during CI surgery.

Histopathological support for this hypothesis is present in the literature. Hirai [17] reported that the most frequently associated abnormality seen in EVA was modiolar deficiency, observed in 73% of the temporal bones they studied. Holden and Linthicum [16] showed hydropic changes in the SV where the ISS was pushed upwards, in a specimen with EVA. Sampaio [18] demonstrated a partial modiolar defect in a case of massive EVA and EES.

Pyle [19] summarized the theories into two groups; Valvassori and Clemis [20], Emmett [21], and Arcand [22] suggested an arrest in development occurring at 5 weeks of gestation. Okumura [23], however, proposed that EVA results from aberrant development of the aqueduct and sac later in fetal and postnatal life. He concluded that it is unlikely that EVA results from an arrest in development or failure of narrowing early in embryonic life.

Whether the sac continues to secrete an abnormal quantity of endolymph is not known, but once there is EVA, it continues to transmit pressure into the cochlea. During later stages of life, CSF pressure is still transmitted into the cochlea and may cause progressive or sudden sensorineural hearing loss with further damage to the cochlear hair cells.

Radiological support was recently reported by Sennaroglu L [24].

3.2.8 Incomplete Partition Type III

This is the cochlea with interscalar septa but absent modiolus. As the size and shape of the cochlea is normal, **membranous labyrinth development must be normal. Most probably the wall of the cochlea consists of only a thick endosteum and outer two layers of the otic capsule are absent.**

As we have no histopathological specimen with a cochlea having IP-III malformation, information obtained from the radiology of the temporal bone is used to explain pathophysiology.

From the radiological evaluation of the cases with X-linked deafness, it can easily be seen that cochlea has a very specific appearance; as soon as one examines a CT of an IP-III cochlea the malformation is evident. Modiolus is absent, but interscalar septa is present. Otic capsule around the membranous labyrinth appears to be thinner in IP-III when compared to that in a normal cochlea (Fig. 3.11a). If a specimen of a normal cochlea is examined under light microscopy, the inner endosteal layer of the otic capsule follows the outline of the membranous labyrinth (Fig. 3.11b). The middle enchondral and outer periosteal layers increase the thickness of the otic capsule, without following the contour of the membranous labyrinth. With today's precision, it is not possible to observe the normal thin layer of

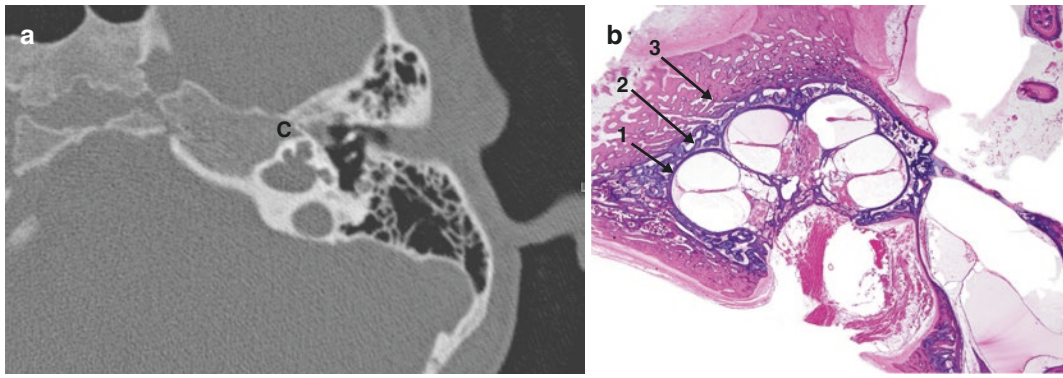


Fig. 3.11 Incomplete partition type III (IP-III). (**a**) IP-III cochlea with interscalar septa and absent modiolus. Please note the thinner otic capsule probably consisting of only a thicker endosteal layer, (**b**) normal cochlea: the inner endosteal layer (1) of the otic capsule follows the outline of the membranous labyrinth. The middle enchondral (2) and outer periosteal (3) layers increase the thickness of the otic capsule, without following the contour of the membranous labyrinth. (With permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary)

the endosteum on HRCT. However, HRCT demonstrates that in IP-III, the otic capsule around the cochlea is thinner and follows the outline of the membranous labyrinth as if it is formed only by a thick endosteal layer. Instead of the usual three layers, probably the second and third layers are either absent or very thin. The otic capsule most probably consists of a thickened inner endosteal layer without enchondral and outer periosteal layers.

This can be explained by embryological development of the otic capsule. According to Donaldson [4], vascular supply of the cochlea comes from two different sources: the outer periosteal and middle enchondral layers get the vascular supply from the middle ear mucosa. The innermost endosteal lining and modiolar areas get their vascularization from the IAC. When the images are carefully examined, it is seen that these patients have an intact inner endosteal layer and interscalar septum. This shows that there is an absence of either the middle enchondral layer alone or the enchondral and outer periosteal layers together (most probably due to reduced or absent vascular supply from the middle ear). As a result, the pathophysiology in X-linked deafness appears to be abnormal vascular supply from the middle ear mucosa as a result of a genetic abnormality.

Absence of outer two layers resulting in a thinner otic capsule has other distinctive features which are specific for IP-III. In normal anatomical conditions the cochlea is located at the anterolateral part of the IAC. Vestibule occupies the posterolateral part of the IAC. In IP-III, it looks as if the cochlea is directly located at the lateral end of the IAC, almost in a straight line. This is a unique finding not present in any of the other anomalies. However, this is a misinterpretation, because of the absence of enchondral and outer periosteal layers at the base of the cochlea. In a normal cochlea, the middle enchondral layer constitutes the major part of the cochlear base, with a contribution from the inner periosteal layer. Therefore, the missing bony layer or layers give a false impression that the cochlea is situated directly lateral to the IAC.

The labyrinthine segment of the facial nerve normally courses around the basal part of the cochlea, but in this anomaly it is always located above the cochlea. The labyrinthine segment of the facial nerve is not in the normal position, because missing layers of the otic capsule cause the labyrinthine segment move superiorly above the cochlea. The outer layers of the otic capsule, therefore, play an important role in the normal positioning of the labyrinthine segment of the facial nerve. From this aspect, this is similar to CLA without the otic capsule.

The cochlear base and modiolus are completely absent. The cochlear base consists of two layers: the endosteum and the middle enchondral layer. The middle enchondral layer provides the bulk to the cochlear base. If this layer is absent, the endosteal layer may not be sufficient to provide a thick base where the modiolus will have its support. Therefore, the absence of layers 2 and 3 results in a defective cochlear base and absent modiolus, in spite of normal vascularization from the IAC to the modiolus. It is quite possible that, if the base is defective, the modiolus cannot form the attachment points and develop appropriately.

3.2.9 Enlarged Vestibular Aqueduct

Enlarged Vestibular Aqueduct (EVA) appears to be responsible for the changes observed in IP-II. High CSF pressure transmitted into the inner ear via EVA may cause different modiolar defects resulting in IP-II. The mildest deformity observed in the cochlea is dilatation of scala vestibuli, and the greatest deformity is the complete modiolar absence. It is also possible that patency of the EVA is minimal or EVA occurs after the development of the inner ear and no pathology is observed in the cochlea. If the cochlea is completely normal on imaging, it is appropriate to use the term EVA.

In the past EVA was diagnosed more often. However, with the development of better scanners it has been possible to detect minor cochlear

changes with the present-day HRCT and MRI. MRI particularly can demonstrate even scala vestibuli dilatation. Therefore, many of the EVA cases have IP-II deformity in the cochlea rather than being EVA only.

It is appropriate to accept EVA as a separate IEM group because the cochlea and vestibule are normal on imaging in spite of EVA. However, audiological findings are closely related to IP-II cases.

3.2.10 Cochlear Aperture Abnormalities

The exact mechanism causing cochlear aperture abnormalities remains unknown. Normal development of the inner ear can be affected by a number of factors in various stages of the development. It was speculated that a developmental insult of the otic capsule may inhibit normal production of neural growth factor. This may cause neuronal degeneration and prevent normal growth of the developing CN. Majority of patients with profound SNHL are thought to have an abnormality involving the membranous labyrinth [25]. This may inhibit the normal trophic effects of nerve growth factor, resulting in a hypoplastic CN and CA [26]. Similar to developing IAC, which requires the presence of a normal cochlear nerve as a stimulus to obtain normal adult dimensions, it is likely that the cochlear aperture also requires a similar neural stimulus for its normal development [26, 27]. Lack of an adequate stimulus because of a hypoplastic cochlear nerve may prevent the cochlear aperture from reaching its normal diameter.

References

1. Gulya AJ. Developmental anatomy of the temporal bone and skull base. In: Shambaugh-Glasscock surgery of the ear. 5th ed. Hamilton, ON: BC Decker; 2003. p. 4–33.
2. Gulya AJ, Schuknecht HF, editors. Anatomy of the temporal bone with surgical implications. 3rd ed. New York, NY: Informa Healthcare USA; 2007.
3. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
4. Donaldson JA, Duckert LG, Rubel EW, editors. Surgical anatomy of the temporal bone. 4th ed. New York, NY: Raven Press; 1992.
5. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
6. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. J Laryngol Otol Suppl. 2000;25:1–14.
7. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41.
8. Erixon E, et al. Variational anatomy of the human cochlea: implications for cochlear implantation. Otol Neurotol. 2009;30(1):14–22.
9. Kondo K, et al. Temporal bone histopathologic findings in a case of interstitial deletion of the long arm of chromosome 2 [del(2) (q31q33)]. Int J Pediatr Otorhinolaryngol. 1999;48(1):31–7.
10. Sekhar HK, Sachs M. Mondini defect in association with multiple congenital anomalies. Laryngoscope. 1976;86(1):117–25.
11. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
12. Shetty PG, et al. Cerebrospinal fluid otorhinorrhea in patients with defects through the lamina cribrosa of the internal auditory canal. AJNR Am J Neuroradiol. 1997;18(3):478–81.
13. da Cruz MJ, Ahmed SM, Moffat DA. An alternative method for dealing with cerebrospinal fluid fistulae in inner ear deformities. Am J Otol. 1998;19(3):288–91.
14. Syal R, Tyagi I, Goyal A. Cerebrospinal fluid otorhinorrhea due to cochlear dysplasias. Int J Pediatr Otorhinolaryngol. 2005;69(7):983–8.
15. Khan AM, Levine SR, Nadol JB Jr. The widely patent cochleovestibular communication of Edward Cock is a distinct inner ear malformation: implications for cochlear implantation. Ann Otol Rhinol Laryngol. 2006;115(8):595–606.
16. Holden PK, Linthicum FH Jr. Mondini dysplasia of the bony and membranous labyrinth. Otol Neurotol. 2005;26(1):133.
17. Hirai S, et al. Large vestibular aqueduct syndrome: a human temporal bone study. Laryngoscope. 2006;116(11):2007–11.
18. Sampaio AL, et al. Massive endolymphatic sac and vestibular aqueduct in Mondini dysplasia. Arch Otolaryngol Head Neck Surg. 2004;130(5):678–80.
19. Pyle GM. Embryological development and large vestibular aqueduct syndrome. Laryngoscope. 2000;110(11):1837–42.
20. Valvassori GE, Clemis JD. The large vestibular aqueduct syndrome. Laryngoscope. 1978;88(5):723–8.

21. Emmett JR. The large vestibular aqueduct syndrome. Am J Otol. 1985;6(5):387–415.
22. Arcand P, et al. The large vestibular aqueduct syndrome and sensorineural hearing loss in the pediatric population. J Otolaryngol. 1991;20(4):247–50.
23. Okumura T, et al. Sensorineural hearing loss in patients with large vestibular aqueduct. Laryngoscope. 1995;105(3 Pt 1):289–93; discussion 293–4.
24. Sennaroglu L. Another evidence for pressure transfer mechanism in incomplete partition two anomaly via enlarged vestibular aqueduct. Cochlear Implants Int. 2018;19(6):355–7.
25. Fatterpekar GM, et al. Hypoplasia of the bony canal for the cochlear nerve in patients with congenital sensorineural hearing loss: initial observations. Radiology. 2000;215(1):243–6.
26. Casselman JW, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. Radiology. 1997;202(3):773–81.
27. Glastonbury CM, et al. Imaging findings of cochlear nerve deficiency. AJNR Am J Neuroradiol. 2002;23(4):635–43.

4 Genetic Causes of Sensorineural Hearing Loss Associated with Inner Ear Malformations

Wenxin Liu, Emre Ocak, Guney Bademci, and Mustafa Tekin

4.1 Introduction

Hearing loss (HL) is the most common sensory disorder in humans. It is estimated that 1.6 in every 1000 infants in the U.S. are born with sensorineural hearing loss (SNHL) [1]. One mechanism of congenital SNHL is developmental anomalies affecting the inner ear. Inner ear malformations (IEMs), detected by a computerized scan (CT) or magnetic resonance imaging (MRI) study, can be found in up to one-third of children with SNHL [2]. The presence of IEMs as well as the specific malformation detected may then have an impact on treatment options. For instance, if the cochlear nerve or the inner ear is absent in its entirety, placement of a cochlear implant would not be an effective treatment for the patient, although it is generally an effective treatment option for patients presenting with other forms of IEMs.

Despite studies suggesting over half of profound deafness has genetic causes [3], the genetic basis of IEMs still remains largely to be discovered. Understanding the genetic underpinnings of SNHL associated with IEMs is important in the diagnosis and timely management of patients presenting with SNHL. Not only would it help diagnose family members who may present with a milder form of IEMs and assist impacted individuals with family planning, but its diagnosis may also be an early indication of developmental abnormalities in other organ systems that would not have manifested until later in life. Both these implications of understanding the genetic basis of IEMs have the potential to greatly impact how a patient may be managed in a clinical setting.

W. Liu
Genomic Medicine MD/MS Program, University of Miami Miller School of Medicine, Miami, FL, USA
e-mail: wxl276@med.miami.edu

E. Ocak
Department of Otolaryngology, University of Miami Miller School of Medicine, Miami, FL, USA

Department of Otolaryngology, Ankara University Faculty of Medicine, Ankara, Turkey

G. Bademci
John P. Hussman Institute for Human Genomics, University of Miami Miller School of Medicine, Miami, FL, USA

Dr. John T. Macdonald Foundation Department of Human Genetics, University of Miami Miller School of Medicine, Miami, FL, USA
e-mail: g.bademci@med.miami.edu

M. Tekin (✉)
Department of Otolaryngology, University of Miami Miller School of Medicine, Miami, FL, USA

John P. Hussman Institute for Human Genomics, University of Miami Miller School of Medicine, Miami, FL, USA

Dr. John T. Macdonald Foundation Department of Human Genetics, University of Miami Miller School of Medicine, Miami, FL, USA
e-mail: mtekin@med.miami.edu

© Springer Nature Switzerland AG 2022
L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_4

This review thus aims to summarize current understanding of the genetic etiology for IEMs, specifically its clinical presentation, the genes involved, and the roles they play in embryology.

4.2 A Brief Molecular Embryology of the Inner Ear

The embryological base of the inner ear originates from a thickening of the ectoderm known as the otic placode (OP), which then invaginates to form the otic vesicle. The OP, in turn, arises from a primitive embryological region that surrounds the neural plate known as preplacodal region (PPR).

4.2.1 Preplacodal Region

Inner ear development begins in the PPR. Studies in animal models have shown that the development of the PPR in the ectoderm of the developing embryo is determined mainly by the interaction of bone morphogenic protein (Bmp), Wnt and their antagonists, and proteins in the fibroblast growth factor (FGF) pathway [4] (Fig. 4.1a). The anterior-posterior differentiation of the PPR is further modulated by the mutual repression of transcription factors Gbx2 and Otx2, with Gbx2 playing an especially large role in the development of the OP [5] (Fig. 4.1b).

The transcriptional co-activator Eya1 and homeobox gene *Six1* function in the early differentiation and maintenance of neurons [6] and are thought to be regulated in turn by Foxi1 [7]. Eya1 and Six1 together with Dach form a regulatory network that leads to transcriptional activation, cell proliferation, and organogenesis of the inner ear [8].

4.2.2 Development of Otic Vesicle

The PPR develops into the OP, which invaginates to form the otic vesicle. The FGF pathway continues to be important in the induction of the OP and signaling of this factor has been shown to induce the expression of zebrafish otic genes such as *pax8*, *pax2a*, *fgf24*, and *sox3* throughout the PPR, which is important in setting the pattern of the otic vesicle [9]. This transformation is believed to be, at least in part, regulated by the *Hoxa1* gene [10]. The FGF pathway is also influenced by foxi1 and gata3 during otic development, with foxi1 shown to inhibit, and gata3 shown to promote its signaling [11].

In a study investigating the RhoA activity for apical constriction in inner ear placode invagination in a chick model, investigators showed that invagination of OP to form the otic vesicle occurs via the activation of myosin-II not only through FGF signaling, but also through the RhoA-ROCK pathway [12].

After the development of the otic vesicle, the otocyst then gives rise to the mature inner ear structures: the vestibular system in the dorsal plane and the auditory system in the ventral plane.

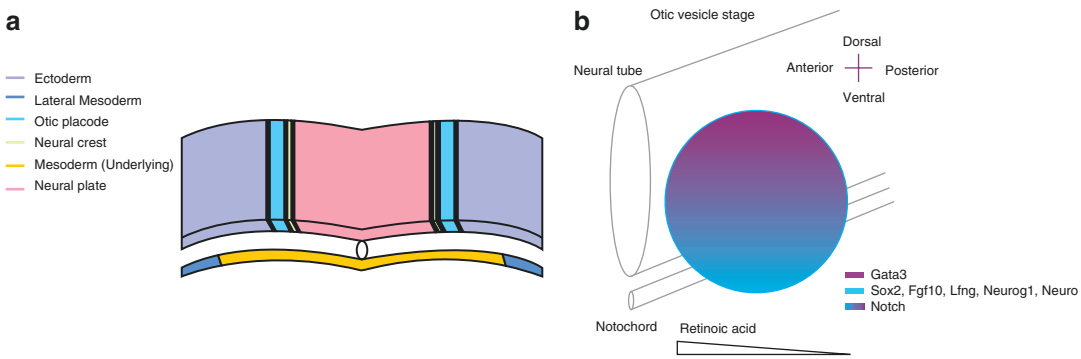


Fig. 4.1 (**a**) The anterior-posterior differentiation of the preplacodal region at ~22 days. (**b**) Differentiation of the otic placode and relevant factors

4.2.3 Molecules and Factors in the Neurogenesis of the Inner Ear

After the otic vesicle stage, many factors play important roles in the further development of neural inner ear structures. Previous studies indicate that high levels of Sox2 protein inhibit sensory cell development in the inner ear [13]. Thus in order for neuronal precursors to commit to a neuronal fate, Fgf10 signals the expression of Ngn1 and Neurod1, which act to inhibit the activity of Sox2 [13, 14] (Fig. 4.1b). Notch signaling then plays a role in specifying the sensory domains within the OP by inducing the proliferation of undifferentiated pre-sensory cells, upregulating Sox2, and inhibiting Ngn1 [15] (Fig. 4.1b). Furthermore, tfap2a is believed to modulate the activity of FGF and notch through activating the inhibitor bmp7a, playing a key role in neural development as well [16]. Another important neural aspect of the inner ear, the formation of the inner radial bundle, is mediated by Eph/Ephrin signaling, a target of Pou3f4 transcription factor activity [17, 18].

4.2.4 Cochlea Formation

The cochlea develops in the ventral plane in the antero-posterior development axis after the otic vesicle stage. Several factors and molecules are involved in the development of the cochlea, such as *Jag1*, *Sox2*, and *Lfng* [19]. Deletion of $p27^{kip1}$ can cause changes in the cochlea, such as overproduction of hair cells, interestingly causing sensorineural hearing loss [20]. *Atoh1* is the earliest discovered factor expressed in the prosensory domain associated with sensory hair cells. In the early period it can be detected in the whole cochlea but over time the expression of *Atoh1* is restricted with hair cell progenitors. Numerous factors, which can up- or down-regulate *Atoh1*, have been described. Among them, *Sox2* is one of the most investigated molecules. Although being required for the expression, *Sox2* also down-regulates *Atoh1*. Other regulators of this transcription factor are the Id (inhibitor of differentiation) genes (*Id1*, *Id2*, and *Id3*). These genes are known to negatively regulate *Atoh1* [21]. Cochlear lumen formation begins at the base of the cochlea and proceeds towards the apex. This is partly controlled by fluid secretion in the vestibular labyrinth, which is then absorbed into the endolymphatic sac, a process mediated by *Slc26a4*-encoded channels [22].

4.2.5 Semicircular Canal Development

The vestibular system is located in the dorsal plane of the inner ear. The semicircular canals and their neural elements are derived from the two prominences of the otocyst: the horizontal and vertical canal pouches. The hindbrain is the source of ventral-dorsal axial signaling for the inner ear and Wnts from the dorsal hindbrain are important signals for semicircular canal development [23]. Dlx5 has been shown to be one of the downstream genes that respond to Wnt signaling. Previous studies demonstrate that the lack of Dlx5 can affect canal and crista formation [24]. Also required for appropriate semicircular canal formation are the molecule Hmx3 and the gene *Chd7*, which encodes a chromodomain-containing helicase protein, and is proposed to act as a selector gene that encodes transcription factors essential for semicircular canal genesis [25].

4.3 Syndromic Causes of Inner Ear Malformations in Humans

Inner ear malformations can be found alone or as part of a syndrome involving other systems. While numerous syndromes can be associated with IEMs in humans, we summarize the most frequently diagnosed ones with clinical findings and causative genes in Table 4.1.

Table 4.1 Selected syndromic causes of inner ear malformations

Syndrome	Clinical features	Gene involved
Bosley-Salih-Alorainy	Autosomal recessive syndrome with congenital deafness due to inner ear malformations associated with Duane retraction syndrome of the eye, autism, and brainstem abnormalities. Inner ear malformations range from labyrinthine aplasia to cochlear anomalies	*HOXA1*
Branchio-oculo-facial	Autosomal dominant condition characterized by branchial skin defects, ocular anomalies (i.e., cataracts), facial anomalies, malformed pinnae, and HL. Patients may have EVA, cochlear dysplasia, and poorly differentiated cochlear turns	*TFAP2A*
Branchio-oto-renal	Autosomal dominant syndrome leading to malformations of the outer, middle, or inner ears associated with HL, branchial fistulae and cysts, and renal abnormalities. Inner ear presentations of patients include dysplastic vestibule, cochlear hypoplasia with defective modiolus, EVA, hypoplastic semi-circular canals, and dilated internal auditory canals. Onset of hearing loss ranges from childhood to young adulthood	*EYA1*, *SIX1*
CHARGE	Autosomal dominant disorder. CHARGE stands for *C*oloboma, *H*eart defects, Choanal *A*tresia, *R*etarded growth and development, *G*enital abnormalities, and *E*ar anomalies	*CHD7*, *SEMA3E*
Kabuki	Autosomal dominant disorder characterized by minor skeletal anomalies, unique facial features, "fetal" fingertip pads, mild to moderate intellectual disability, and growth deficiencies. IEM presentations include cochlear aplasia, vestibular dysplasia, and EVA	*KMT2D*, *KDM6A*
LAMM	Autosomal recessive condition. LAMM stands for *L*abyrinthine *A*plasia, *M*icrotia, and *M*icrodontia, which are its characteristic presentations along with profound congenital SNHL. The cerebellopontine angle may show the bilateral absence of cochleovestibular nerve with otherwise normal cerebral and cerebellar structures	*FGF3*
Microtia, hearing impairment, and cleft palate	Autosomal recessive condition characterized by mixed symmetric severe to profound hearing loss, microtia, and partial cleft palate. Inner ear malformations reported include labyrinthine aplasia	*HOXA2*
Pendred	Autosomal recessive condition with congenital severe-to-profound SNHL associated with EVA and either abnormal perchlorate discharge test or goiter	*SLC26A4*
Waardenburg	Auditory-pigmentary syndrome characterized by pigmentary abnormalities of the hair, including a white forelock and premature graying; pigmentary changes of the iris, such as heterochromia irides and brilliant blue eyes; and congenital SNHL. All types (I–IV) are inherited in an autosomal dominant manner, while a subgroup of type IV is autosomal recessive	*PAX3*, *MITF*, *SOX10*, *EDN3*, *SNAI2*, *EDNRB* and *KITLG*

HL hearing loss, *SNHL* sensorineural hearing loss, *EVA* enlarged vestibular aqueduct

4.4 Non-syndromic Causes of Inner Ear Malformations

Non-syndromic deafness is a type of hearing impairment in which HL is the only clinical finding in the patient. Only a few gene mutations have thus far been discovered to cause non-syndromic IEMs and they are summarized below.

4.4.1 SLC26A4

In 1997, the gene responsible for Pendred syndrome was identified as *SLC26A4* [26], which encodes a transmembrane protein named pendrin. Subsequently, *SLC26A4* mutations were also discovered in individuals with autosomal recessive non-syndromic deafness associated with enlargement of the vestibular aqueduct

(EVA). More than 200 mutations have since been reported related to sporadic and familial forms of Pendred syndrome and non-syndromic SNHL with EVA, and autosomal recessive mutations in the *SLC26A4* gene is therefore currently one of the leading causes of non-syndromic SNHL. Although the number of mutant alleles of *SLC26A4* has been shown to correlate with the auditory and thyroid phenotypes, no connections between the type of mutation and thyroid phenotype have been reported [27].

Recent studies with molecular testing for *SLC26A4* mutations and radiologic imaging of temporal bones demonstrated that enlargement of the vestibular aqueduct (EVA) can be recognized as the most penetrant feature of Pendred syndrome [28]. EVA is the most common radiologic anomaly of the inner ear and is mostly identified in either of two different contexts, non-syndromic EVA or Pendred syndrome.

While variants in *FOXI1* and *KCNJ10* were reported to cause SNHL with EVA when the same person is heterozygous for an *SLC26A4* mutation (i.e., digenic inheritance), this observation has not yet been confirmed by subsequent reports.

4.4.2 POU3F4

Variants in the *POU3F4* gene are a major cause of X-linked deafness worldwide at locus DFN3. *POU3F4* is the first nuclear gene implicated in non-syndromic deafness. The type of hearing loss found may be SNHL or mixed associated with IP-III (incomplete partition type 3) and stapes fixation (DFN3) [29–32]. In addition to mutations located within the gene, copy number variants not involving the coding part of the gene have been reported: de Kok and colleagues identified a hot spot for microdeletions in patients with X-linked deafness 900 kb proximal to the DFN3 gene [33]. Given the *POU3F4*'s uniqueness as an X-linked cause of IEMs, if hearing loss in a patient is found to be X-linked and associated with an inner ear malformation, mutations in *POU3F4* gene must be part of the differential and evaluated.

As a clinical pearl, if *POU3F4* is identified as the causative gene for hearing loss in a patient, the surgeon should be on the alert for perilymphatic gusher (in fact "cerebrospinal fluid") during stapes surgery and avoid stapes surgery which may result in total hearing loss.

4.4.3 COCH

The *COCH* gene is located at 14q11.2-q13 and encodes a secretory protein called Cochlin [34]. The postulated pathogenetic mechanism of *COCH* gene-related hearing loss is the accumulation of acidophilic deposits in the area of the spiral osseous lamina, spiral ligament, and vestibular nerve channels [35]. Several reports indicate high probability of a link between mutations in the *COCH* gene and presentation of IEMs. Hildebrand et al. described a patient who presented with semicircular canal dehiscence (SSCD) associated with a mutation in the *COCH* gene [36]. Dodson et al. described a patient heterozygous for a mutation in the *COCH* gene and who showed an EVA upon CT imaging [37]. Finally, de Varebeke described nine patients with the same mutation in the *COCH* gene. On CT imaging, eight of them were found to have sclerotic lesions and/or narrowing of the semicircular canals, and in one patient, the posterior vestibule was also affected [38]. Based on these findings, *COCH* mutations are a possible autosomal dominant inherited cause of IEMs and are postulated to play a role, along with type II collagen bundles, in laying down the structure of the inner ear [39].

4.4.4 ROR1

ROR1 (receptor tyrosine kinase-like orphan receptor 1) is an integral transmembrane protein consisting of extracellular and intracellular conserved domains. A *ROR1* gene mutation was found to be the cause of congenital autosomal recessive non-syndromic SNHL and common cavity anomaly in one reported family with two children [40].

References

1. Mehl AL, Thomson V. The Colorado newborn hearing screening project, 1992-1999: on the threshold of effective population-based universal newborn hearing screening. Pediatrics. 2002;109(1):E7.
2. Bamiou DE, Phelps P, Sirimanna T. Temporal bone computed tomography findings in bilateral sensorineural hearing loss. Arch Dis Child. 2000;82(3):257–60.
3. Nance WE. The genetics of deafness. Ment Retard Dev Disabil Res Rev. 2003;9(2):109–19.
4. Litsiou A, Hanson S, Streit A. A balance of FGF, BMP and WNT signalling positions the future placode territory in the head. Development. 2005;132(18):4051–62.
5. Steventon B, Mayor R, Streit A. Mutual repression between Gbx2 and Otx2 in sensory placodes reveals a general mechanism for ectodermal patterning. Dev Biol. 2012;367(1):55–65.
6. Zou D, Silvius D, Fritzsch B, Xu PX. Eya1 and Six1 are essential for early steps of sensory neurogenesis in mammalian cranial placodes. Development. 2004;131(22):5561–72.
7. Ishihara T, Sato S, Ikeda K, Yajima H, Kawakami K. Multiple evolutionarily conserved enhancers control expression of Eya1. Dev Dyn. 2008;237(11):3142–56.
8. Li X, Oghi KA, Zhang J, Krones A, Bush KT, Glass CK, et al. Eya protein phosphatase activity regulates Six1-Dach-Eya transcriptional effects in mammalian organogenesis. Nature. 2003;426(6964):247–54.
9. Padanad MS, Bhat N, Guo B, Riley BB. Conditions that influence the response to Fgf during otic placode induction. Dev Biol. 2012;364(1):1–10.
10. Makki N, Capecchi MR. Identification of novel Hoxa1 downstream targets regulating hindbrain, neural crest and inner ear development. Dev Biol. 2011;357(2):295–304.
11. Yao D, Zhao F, Wu Y, Wang J, Dong W, Zhao J, et al. Dissecting the differentiation process of the preplacodal ectoderm in zebrafish. Dev Dyn. 2014;243(10):1338–51.
12. Sai X, Yonemura S, Ladher RK. Junctionally restricted RhoA activity is necessary for apical constriction during phase 2 inner ear placode invagination. Dev Biol. 2014;394(2):206–16.
13. Evsen L, Sugahara S, Uchikawa M, Kondoh H, Wu DK. Progression of neurogenesis in the inner ear requires inhibition of Sox2 transcription by neurogenin1 and neurod1. J Neurosci. 2013;33(9):3879–90.
14. Alsina B, Abello G, Ulloa E, Henrique D, Pujades C, Giraldez F. FGF signaling is required for determination of otic neuroblasts in the chick embryo. Dev Biol. 2004;267(1):119–34.
15. Jeon SJ, Fujioka M, Kim SC, Edge AS. Notch signaling alters sensory or neuronal cell fate specification of inner ear stem cells. J Neurosci. 2011;31(23):8351–8.
16. Kantarci H, Edlund RK, Groves AK, Riley BB. Tfap2a promotes specification and maturation of neurons in the inner ear through modulation of Bmp, Fgf and notch signaling. PLoS Genet. 2015;11(3):e1005037.
17. Coate TM, Raft S, Zhao X, Ryan AK, Crenshaw EB III, Kelley MW. Otic mesenchyme cells regulate spiral ganglion axon fasciculation through a Pou3f4/EphA4 signaling pathway. Neuron. 2012;73(1):49–63.
18. Raft S, Coate TM, Kelley MW, Crenshaw EB III, Wu DK. Pou3f4-mediated regulation of ephrin-b2 controls temporal bone development in the mouse. PLoS One. 2014;9(10):e109043.
19. Wu D, Kelley MW. Molecular mechanisms of inner ear development. Cold Spring Harb Perspect Biol. 2012;4(8):a008409.
20. Chen P, Segil N. p27Kip1 links cell proliferation to morphogenesis in the developing organ of Corti. Development. 1999;126:1581–90.
21. Jones JM, Montcouquiol M, Dabdoub A, Woods C, Kelley MW. Inhibitors of differentiation and DNA binding (Ids) regulate Math1 and hair cell formation during the development of the organ of Corti. J Neurosci. 2006;26:550–8.
22. Kim HM, Wangemann P. Failure of fluid absorption in the endolymphatic sac initiates cochlear enlargement that leads to deafness in mice lacking pendrin expression. PLoS One. 2010;5(11):e14041.
23. Riccomagno MM, Takada S, Epstein DJ. Wnt-dependent regulation of inner ear morphogenesis is balanced by the opposing and supporting roles of Shh. Genes Dev. 2005;19:1612–23.
24. Acampora D, Merlo GR, Paleari L, Zerega B, Postiglione MP, Mantero S, Bober E, Barbieri O, Simeone A, Levi G. Craniofacial, vestibular and bone defects in mice lacking the distal-less-related gene Dlx5. Development. 1999;126:3795–809.
25. Hurd EA, Micucci JA, Reamer EN, Martin DM. Delayed fusion and altered gene expression contribute to semicircular canal defects in Chd7 deficient mice. Mech Dev. 2012;129(9-12):308–23.
26. Everett L, Glaser B, Beck J, Idol J, Buchs A, Heyman M, Adawi F, Hazani E, Nassir E, Baxevanis A, Sheffield V, Green E. Pendred syndrome is caused by mutations in a putative sulphate transporter gene (PDS). Nat Genet. 1997;17:411–22.
27. Ito T, et al. SLC26A4 genotypes and phenotypes associated with enlargement of the vestibular aqueduct. Cell Physiol Biochem. 2011;28(3):545.
28. Phelps PD, Coffey RA, Trembath RC, Luxon LM, Grossman AB, Britton KE, Kendall-Taylor P, Graham JM, Cadge BC, Stephens SG, Pembrey ME, Reardon W. Radiological malformations of the ear in Pendred syndrome. Clin Radiol. 1998;53:268–73.
29. Gong WX, Gong RZ, Zhao B. HRCT and MRI findings in X-linked non-syndromic deafness patients with a POU3F4 mutation. Int J Pediatr Otorhinolaryngol. 2014;78(10):1756–62.
30. Choi BY, An YH, Park JH, Jang JH, Chung HC, Kim AR, et al. Audiological and surgical evidence for the presence of a third window effect for the conduc-

tive hearing loss in DFNX2 deafness irrespective of types of mutations. Eur Arch Otorhinolaryngol. 2013;270(12):3057–62.
31. Anger GJ, Crocker S, McKenzie K, Brown KK, Morton CC, Harrison K, et al. X-linked deafness-2 (DFNX2) phenotype associated with a paracentric inversion upstream of POU3F4. Am J Audiol. 2014;23(1):1–6.
32. de Kok YJM, van der Maarel SM, Bitner-Glindzicz M, Huber I, Monaco AP, Malcolm S. Association between X-linked mixed deafness and mutations in the POU domain gene POU3F4. Science. 1995;267:685–8.
33. de Kok YJM, Vossenaar ER, Cremers CWRJ, Dahl N, Laporte J, Hu LJ. Identification of a hot spot for microdeletions in patients with X-linked deafness (DFN3) 900 b proximal to the DFN3 gene POU3F4. Hum Mol Genet. 1996;5:1229–35.
34. Parzefall T, Frohne A, Koenighofer M, et al. Identification of a rare COCH mutation by whole-exome Sequencing. Wien Klin Wochenschr. 2017;130:299. https://doi.org/10.1007/s00508-017-1230-y.
35. Robertson NG, Lu L, Heller S, Merchant SN, Eavey RD, et al. Mutations in a novel cochlear gene cause DFNA9, a human nonsyndromic deafness with vestibular dysfunction. Nat Genet. 1998;20:299–303.
36. Hildebrand MS, Tack D, Deluca A, Hur IA, Van Rybroek JM, McMordie SJ, et al. Mutation in the COCH gene is associated with superior semicircular canal dehiscence. Am J Med Genet A. 2009;149A(2):280–5.
37. Dodson KM, Georgolios A, Barr N, Nguyen B, Sismanis A, Arnos KS, et al. Etiology of unilateral hearing loss in a national hereditary deafness repository. Am J Otolaryngol. 2012;33(5):590–4.
38. de Varebeke SP, Termote B, Van Camp G, Govaerts PJ, Schepers S, Cox T, et al. Focal sclerosis of semicircular canals with severe DFNA9 hearing impairment caused by a P51S COCH-mutation: is there a link? Otol Neurotol. 2014;35(6):1077–86.
39. Shindo S, Ikezono T, Ishizaki M, Sekiguchi S, Mizuta K, Li L, et al. Spatiotemporal expression of cochlin in the inner ear of rats during postnatal development. Neurosci Lett. 2008;444(2):148–52.
40. Diaz-Horta O, Abad C, Sennaroglu L, Foster J, DeSmidt A, Bademci G, et al. ROR1 is essential for proper innervation of auditory hair cells and hearing in humans and mice. Proc Natl Acad Sci U S A. 2016;113(21):5993.

5 Preoperative Otolaryngology Examination

Munir Demir Bajin

Special Features

1. Patients with inner ear malformations may apply with recurrent meningitis which may be a fatal situation.
2. Spontaneous footplate fistula may be seen in incomplete partition I and cochlear hypoplasia II.
3. Progressive hearing loss with sudden hearing loss attacks may be due to incomplete partition type II (IP-II).

Preoperative evaluation of patients with inner ear malformations (IEM) usually represents audiological assessment such as behavioral testing, auditory brainstem response testing, otoacoustic emission testing, educational testing, and radiological testing such as computed tomography and magnetic resonance imaging. However, in this chapter we would like to focus on otolaryngological evaluation and emphasize certain clinical points which must be addressed prior to surgery.

5.1 History Taking

Characteristics of hearing loss and recurrent meningitis are very important to think about inner ear malformations. The patient may have unilateral or bilateral hearing loss. Unilateral hearing loss at birth is a strong indication for radiological evaluation for unilateral IEM.

Family may give a history of progressive hearing loss. There may be situations where the patient passes neonatal hearing screening tests at birth but then experiences progressive hearing loss with sudden hearing loss attacks. This is suggestive of incomplete partition type II (IP-II).

History may reveal rhinorrhea after head trauma. This is a situation seen in IP-I or cochlear hypoplasia type II (CH-II) where there is a stapes footplate fistula. There may be a history of recurrent meningitis. This presents an ENT emergency.

5.2 ENT Examination

Otolaryngological examination starts with a full physical examination. Otoscopy is particularly important. Otitis media with effusion (OME) should be treated prior to surgery particularly for cochlear implantation. Persistent effusion after adequate medical therapy necessitates ventilation tube placement. Cochlear implantation in the presence of effusion complicates surgery by obscuring the anatomical structures both by

M. D. Bajin (✉)
Department of Otolaryngology, Hacettepe University Medical Faculty, Ankara, Turkey
e-mail: dbajin@hacettepe.edu.tr

© Springer Nature Switzerland AG 2022
L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_5

hypertrophic mucosa and bleeding. Tube placement should be done at least 3 months prior to cochlear implantation. Implantation can be done with the tubes in place. If the patient has a subtype of IEMs with a risk of gusher it is advisable to remove the tubes and allow sufficient time for the perforation to heal before CI surgery.

Another very important point is to differentiate effusion from cerebrospinal fluid in the middle ear. IEMs like IP-I and CH-II are prone to stapes footplate fistulas which may lead to CSF leakage in the middle ear and subsequent meningitis. Cases of recurrent meningitis and persistent effusion behind tympanic membrane are suggestive of IP-I or CH-II. OME is a mucosal disease and tympanic membrane shows increased vascularity. This is very useful to differentiate the patients with CSF in the middle ear where there is no increased vascularity of the tympanic membrane. It is very important to keep in mind that patients with unilateral effusion or CH-II and IP-I might have CSF in the middle ear. In these patients, stapes footplate fistula is the point of CSF leakage and must be closed either preoperatively or intraoperatively.

These patients may also present with rhinorrhea and they may be operated by neurosurgery for suspected anterior skull base defects without success. Patients with rhinorrhea and hearing loss must be investigated for IEMs and stapes footplate fistulas.

Patients may have visible signs such as unilateral cup ear or hypoplastic auricle. Sometimes unilateral facial paralysis may accompany cochlear hypoplasia cases.

IEMs may be unilateral where contralateral ear and hearing are normal, making the diagnosis more challenging. This delay may cause recurrent meningitis leading to ossification within the ipsilateral labyrinth as well as ossification of the contralateral normal hearing ear. Therefore, patient may lose the option of cochlear implant [1]. Therefore, it is an ENT emergency to notice and repair stapes footplate fistulas particularly in IP-I and CH-II.

A significant number of the patients suffer from comorbidities. These are really important for anesthesiology both for radiological evaluation and surgery. Each comorbidity must be consulted with the relevant specialty. For example, a patient with CHARGE syndrome should be consulted with the department of pediatrics, ophthalmology, neurology, cardiology, orthopedics, and pediatric surgery.

Genetic consultation is also important both for etiology and family consulting.

5.3 Radiological Evaluation

A patient with fluctuating or progressive SNHL needs a radiological evaluation. If the patients benefit from hearing aids and a CI is not indicated, only MRI is sufficient for the diagnosis of IP-II. This is for the concern of avoiding radiation to the child. If the patient has profound SNHL and a CI is planned, temporal CT and MRI are necessary. This is usually done at 6 months of age and if available, cone beam CT is advantageous to minimize radiation.

5.4 Management

In case of total hearing loss there is a possibility of a diagnosis of complete labyrinthine aplasia, rudimentary otocyst, cochlear aplasia, or cochlear nerve aplasia which are definite indications for an ABI [1]. In such a situation, **radiological evaluation should be done as early as possible, not later than 6 months of age** [2].

If an IP-II is diagnosed the patient is advised to wear helmets during sports and avoid contact sports because head trauma may cause further hearing loss in IP-II. In addition, the family is informed that the patient may have attacks of sudden hearing loss. **In such a case they need to apply to a hospital immediately for steroid treatment to recover hearing**. Aim is to provide natural hearing and delay cochlear implantation as long as possible.

Patients with stapes footplate fistula need immediate middle ear exploration to check the footplate for a fistula. This is an ENT emergency to avoid further meningitis. Pneumococcal vaccination is mandatory but if the fistula is not

repaired vaccination will not provide protection against meningitis.

5.5 Conclusion

Patients with IEMs require special attention throughout the management. A meticulous preoperative evaluation prevents further problems and complications in an already complicated patient group.

References

1. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
2. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.

6 Preoperative Audiological Evaluation

Gonca Sennaroglu and Merve Ozbal Batuk

Special Features

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

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

6.1 Preoperative Evaluation Process

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

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

Supplementary Information The online version of this chapter (https://doi.org/10.1007/978-3-030-83674-0_6) contains supplementary material, which is available to authorized users.

G. Sennaroglu · M. Ozbal Batuk (✉)
Department of Audiology, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_6

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

6.2 Audiological Evaluation

6.2.1 History of Hearing Loss

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

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

6.2.2 Behavioral Testing

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

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

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

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

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

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

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

cal characteristics of incomplete partition malformations have been reported in a prior study [6].

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

6.2.2.1 Behavioral Observation Audiometry

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

6.2.2.2 Visual Response Audiometry

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

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

6.2.2.3 Conditioned Play Audiometry

The intention behind the play audiometry procedure is to pair the auditory stimulus with an interesting game, such as overlapping blocks, throwing a cube into a box, or inserting rings onto bars. Children with IEMs are usually diagnosed during the first year of life. Due to this early diagnosis, play audiometry is only rarely used in the preoperative audiological evaluation

of IEMs. For children with progressive HL or sudden HL (i.e., EVA, IP-II), the HL tends to be diagnosed later, and the decision concerning implantation made at an older age, when compared to other IEMs.

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

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

6.3 Electrophysiological Measurements

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

6.3.1 Electroacoustic Immitancemetry

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

6.3.2 Otoacoustic Emission Testing

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

The second consensus meeting on the management of IEM and decision-making between cochlear implantation (CI) and auditory brainstem implantation (ABI) highlighted the importance of the preoperative audiological evaluation. Sennaroğlu et al. [8] stated that the use of OAE

with the CM responses in auditory brainstem response testing might provide an indication of cochlear nerve aplasia/hypoplasia. The clinician should thus be careful when interpreting the audiological results.

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

6.3.3 Auditory Brainstem Response Testing

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

6.3.4 Electrical Auditory Brainstem Response Testing

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

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

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

Based on our clinical experience, eABR is not always the most efficient tool for predicting the functionality of the cochlear nerve in patients with IEMs. The decision-making process should, therefore, consist of a preoperative audiological evaluation, the radiological findings, and the intraoperative eABR results. Despite good

responses being observed in both behavioral tests and daily life, in some cases it is not possible to observe any waveform in the eABR testing. Hence, a correct decision can only be made in consultation with the implant team and after the available options have been discussed with the family.

6.4 Follow-Up with Hearing Aids

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

6.5 Case Studies

Case 1: ZÇ, A Two-Year-Old Female

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

Case 2: AMİ, A Three-Year-Old Female

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

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

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

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

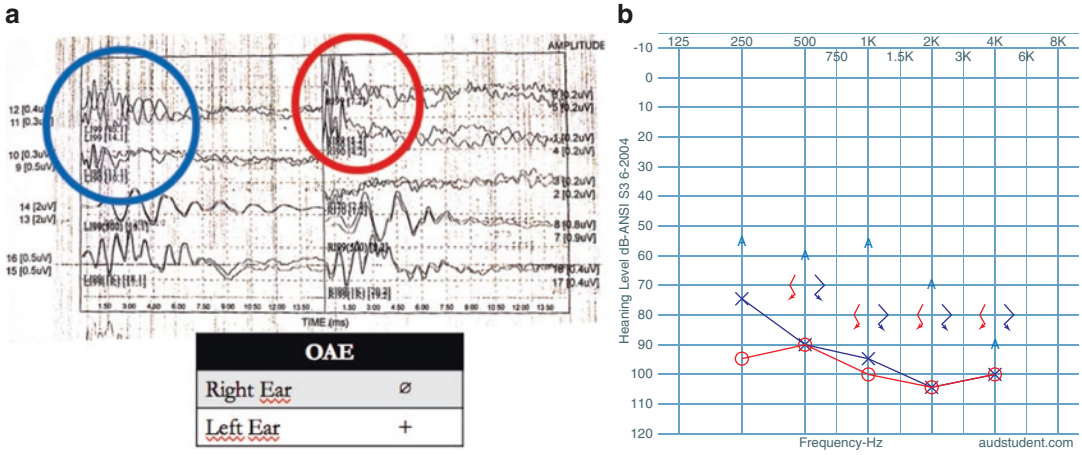


Fig. 6.1 Case 1: (**a**) Auditory brainstem response (ABR) showing cochlear microphonics without any response at the level of 99 dBnHL on both ears and otoacoustic emission (OAE) test results. (**b**) Audiogram with insert earphones presenting responses on both ears and thresholds with hearing aids revealing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

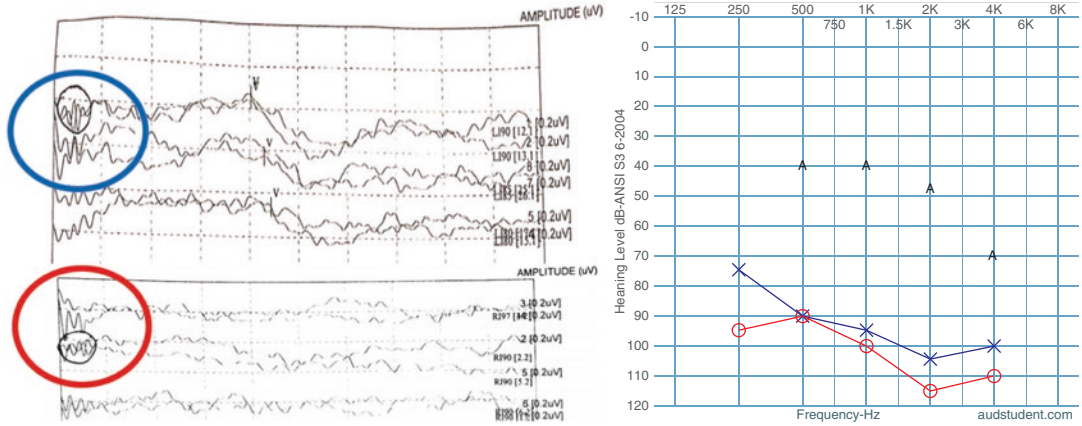


Fig. 6.2 Case 2: ABR test result showing bilateral cochlear microphonics with repeatable wave V on the left ear and the audiogram with insert earphones indicating good responses on both ears

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

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

Case 4: NO, A Nine-Year-Old Female

She passed the NHS in both ears. She had the risk factor of prematurity. During the first audiological evaluation, which was performed at the age of 6 months, the ABR results showed bilateral CM responses with positive OAE in both ears, in which the amplitude of the CM responses was wider on the right side (Fig. 6.4a). These findings

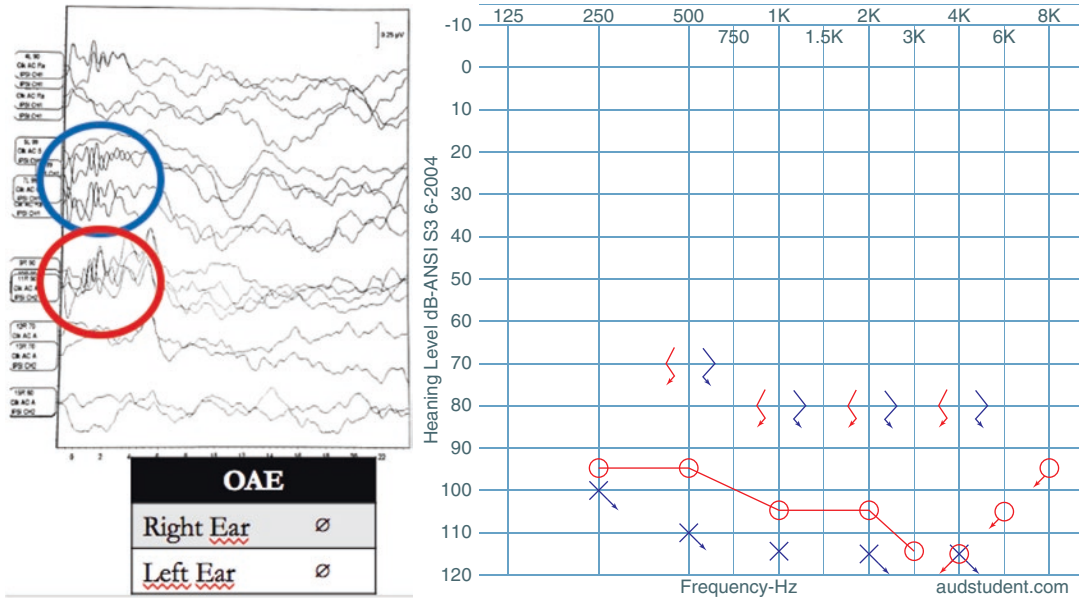


Fig. 6.3 Case 3: ABR indicating cochlear microphonics bilaterally with negative OAE and audiogram with insert earphones indicating good responses at the level of 95–110 dB from low to high frequencies on the right ear and no response on the left ear

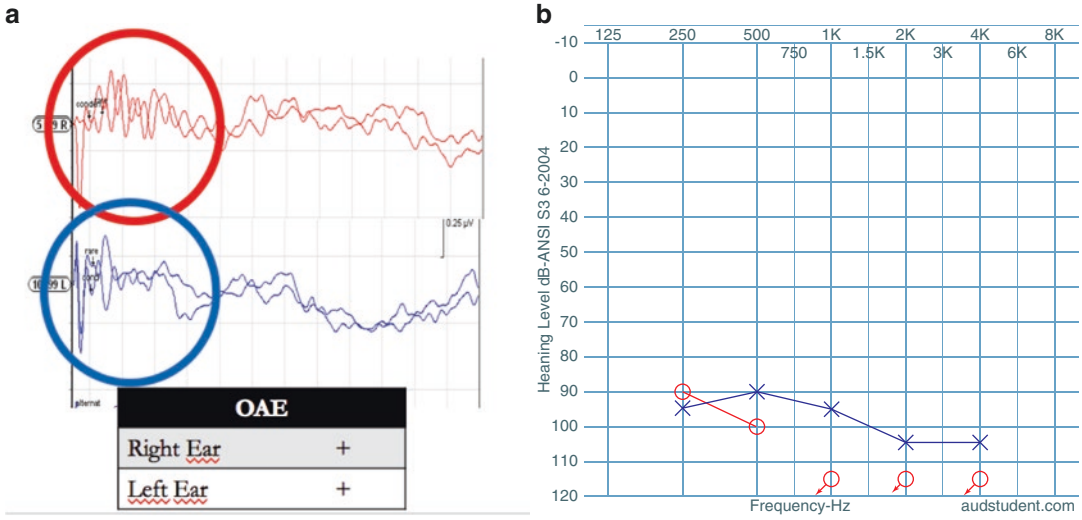


Fig. 6.4 Case 4: (**a**) ABR showing bilateral cochlear microphonics without any repeatable waveform, in which amplitude of the cochlear microphonics was wider on the right side. OAE test result revealed bilateral positive responses. (**b**) Audiogram with insert earphones indicating good responses on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

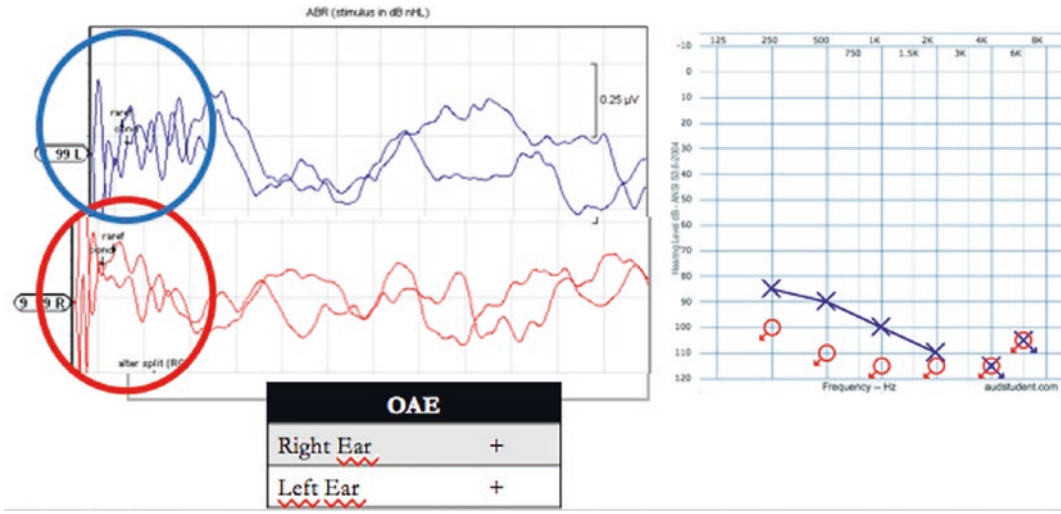


Fig. 6.5 Case 5: ABR presenting bilateral cochlear microphonics with positive OAE responses test result and audiogram with insert earphones indicating good responses on the left ear in the frequency range of 250–2000 Hz despite no response to the sound on the right ear

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

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

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

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

Case 6: ŞO, A Two-Year-Old Female

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

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

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

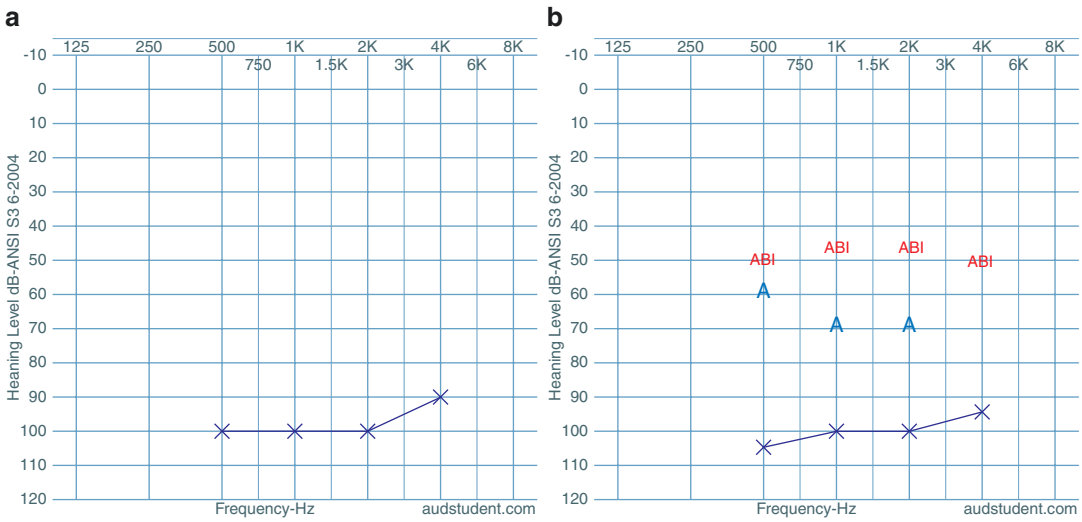


Fig. 6.6 Case 6: (**a**) Hearing thresholds of the left side with insert earphones indicating behavioral responses on the left ear when she was at the age of 9 months. (**b**) Hearing thresholds with left hearing aid (A) presenting responses in 0.5–2 kHz and right auditory brainstem implant (ABI) showing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

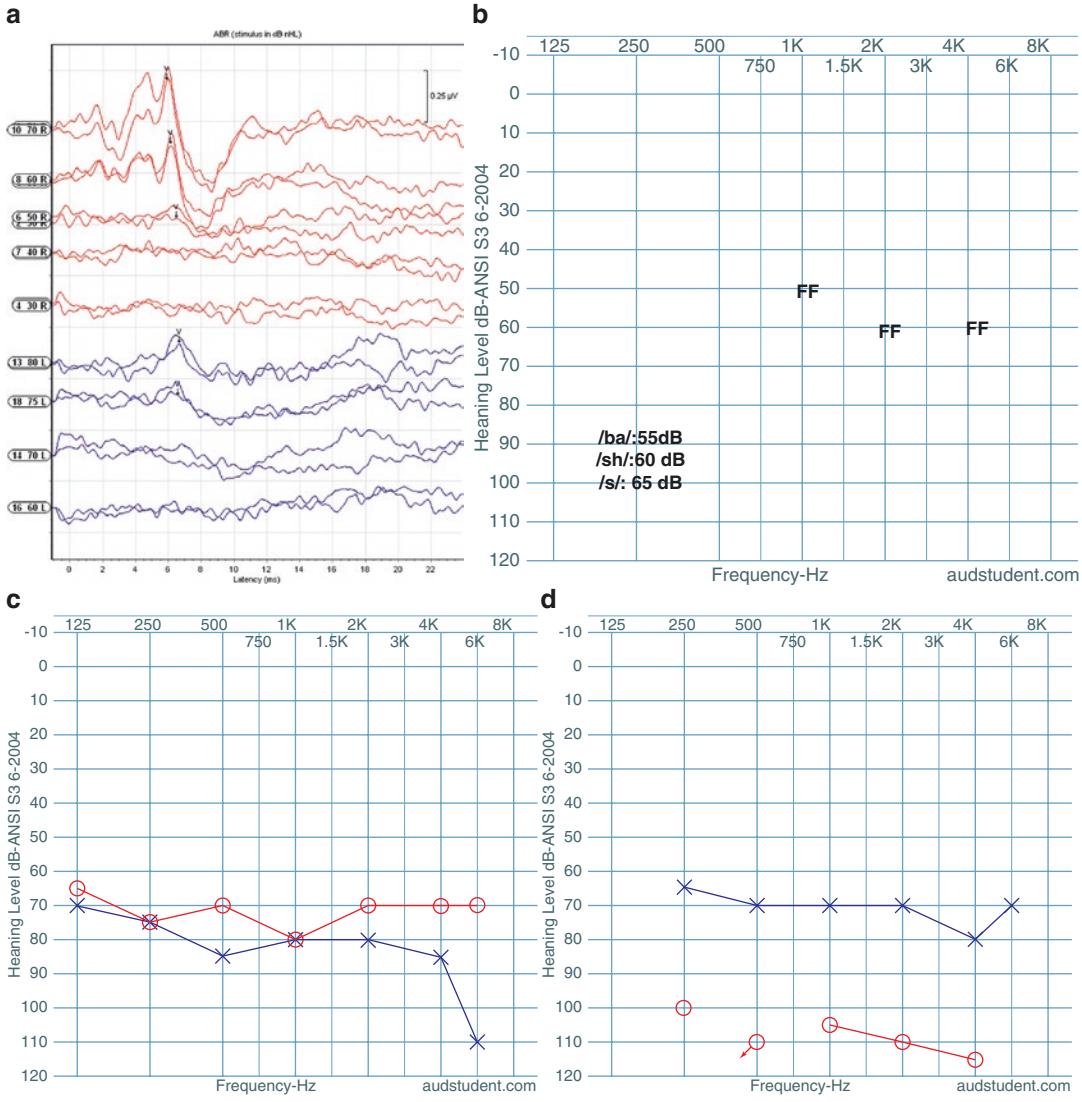


Fig. 6.7 Case 7: (**a**) First ABR test results when he was 2 years old presenting asymmetric hearing loss in which wave V was observed at the level of 50 dBnHL on the right ear and 75 dBnHL on the left ear with click stimulus. (**b**) First audiogram in free field showing hearing thresholds (FF: Free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

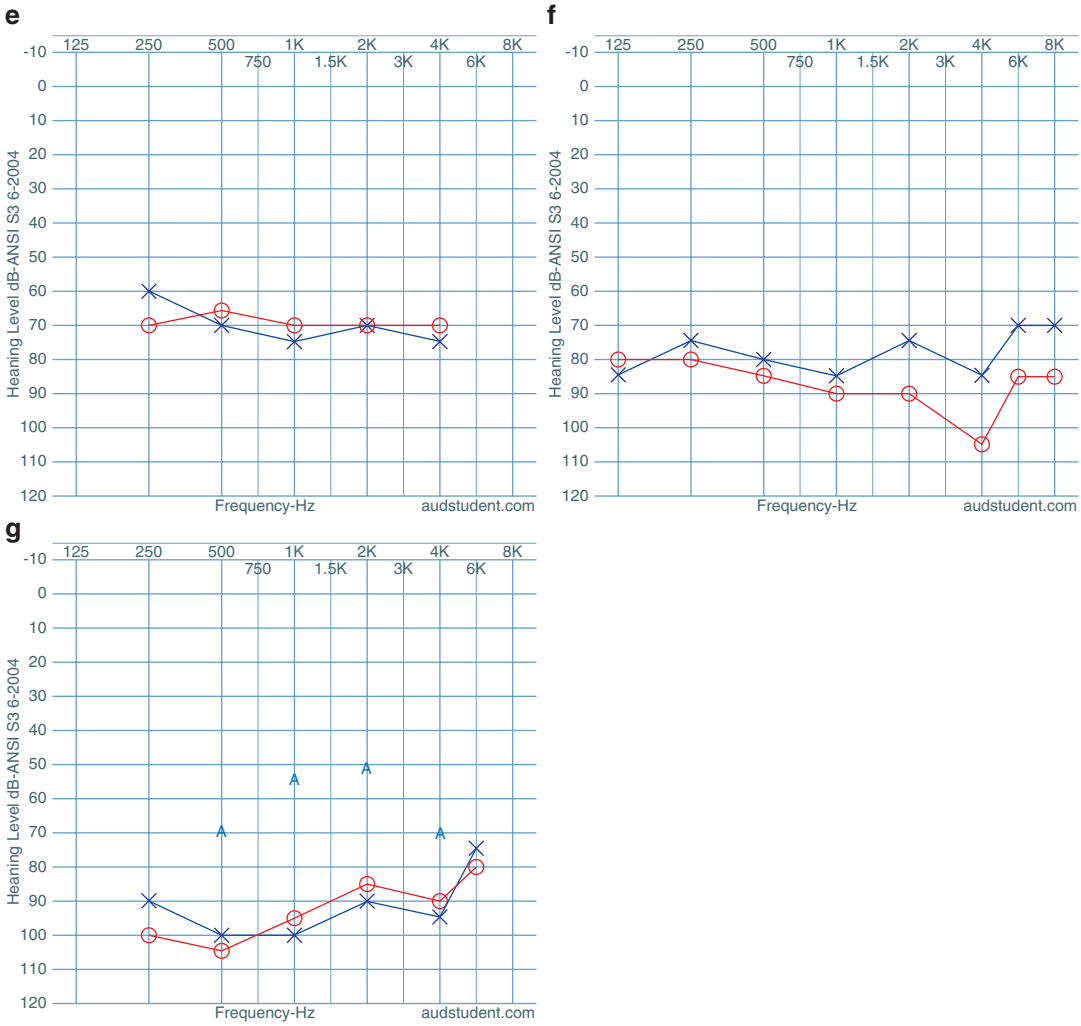


Fig. 6.7 (continued)

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

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

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

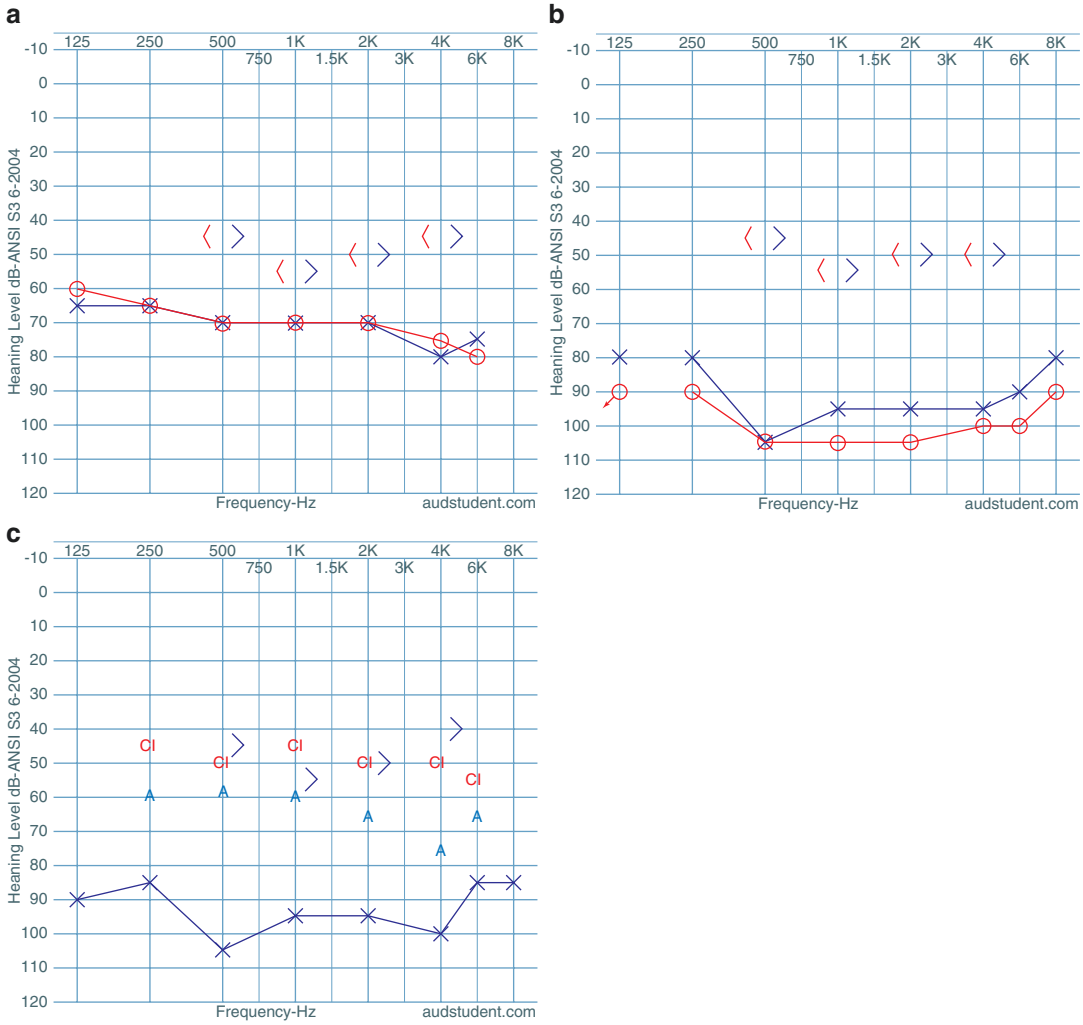


Fig. 6.8 Case 8: (**a**) First audiogram with insert earphones showing bilateral severe mixed-type hearing loss. (**b**) Follow-up audiogram requiring cochlear implantation due to the bilateral profound mixed-type hearing loss. (**c**) Hearing thresholds with right cochlear implant (CI) and left hearing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

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

6.6 Take-Home Message

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

Table 6.1 Signs suggesting cochlear malformation for audiologists

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

6.7 Putting the Pieces Together

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

References

1. Kimura Y, Masuda T, Kaga K. Vestibular function and gross motor development in 195 children with congenital hearing loss—Assessment of inner ear malformations. Otol Neurotol. 2018;39(2):196–205.
2. Van HT, et al. Functional magnetic resonance imaging may avoid misdiagnosis of cochleovestibular nerve aplasia in congenital deafness. Otol Neurotol. 2000;21(5):663–70.
3. Lim C-H, et al. Bony cochlear nerve canal stenosis in pediatric unilateral sensorineural hearing loss. Int J Pediatr Otorhinolaryngol. 2018;106:72–4.
4. Freeman SR, Sennaroglu L. Management of Cochlear nerve hypoplasia and aplasia. In: Advances in hearing rehabilitation. Basel: Karger; 2018. p. 81–92.
5. Weiss AD, et al. Efficacy of earphones for 12-to 24-month-old children during visual reinforcement audiometry. Int J Audiol. 2016;55(4):248–53.
6. Batuk MÖ, et al. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233.
7. Shaw P, Nikolopoulos T. The effect of initial stimulus type for visual reinforcement audiometry. Int J Audiol. 2004;43(4):193–7.
8. Sennaroğlu L, et al. 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.
9. James AL, Dixon PR, Harrison RV. Cochlear nerve aplasia with detectable Olivocochlear efferent function: a distinct presentation of auditory neuropathy Spectrum disorder. Audiol Neurotol. 2018;23(1):39–47.
10. Yamazaki H, et al. Usefulness of MRI and EABR testing for predicting CI outcomes immediately after cochlear implantation in cases with cochlear nerve deficiency. Otol Neurotol. 2015;36(6):977–84.
11. Cinar BC, et al. 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.
12. Ehrmann-Müller D, et al. Outcomes after cochlear implant provision in children with cochlear nerve hypoplasia or aplasia. Int J Pediatr Otorhinolaryngol. 2018;112:132–40.

7 Preoperative Radiological Evaluation

Burce Ozgen

7.1 Introduction

Cross-sectional imaging has become an indispensable tool in the preoperative assessment of cochlear implant (CI) and auditory brainstem implant (ABI) patients.

In infants and young children, computerized tomography (CT) and/or magnetic resonance (MR) imaging examinations of the inner ear are routinely performed to identify a potential etiology for hearing loss, to define the anatomy of the temporal bone and the auditory pathways, and for surgical planning [1]. In the setting of preoperative imaging, the evaluation of the cochlea and cochlear nerve determines the eligibility of the patient for the cochlear versus auditory brainstem implantation. Additionally, the imaging of the posterior fossa as well as supratentorial structures is crucial for appropriate preoperative assessment of a CI/ABI candidate.

7.2 Imaging Modalities

The ideal initial imaging modality for the evaluation of children with newly diagnosed SNHL is currently a topic of debate. Historically, CT has been the study of choice, but there is an increased use of MR imaging due to concerns regarding the ionizing radiation exposure in small children who are more radiosensitive [2]. However dual-technique imaging (with CT and MR imaging) was found to identify a larger number of abnormalities in preimplant candidates than either technique alone [3]. CT and MR imaging are mostly complementary in the preoperative work-up, each with its own strengths and weaknesses [1, 4–9].

7.3 CT Imaging

CT enables accurate anatomical surgical planning, visualizing the bony structures of the ear and anatomical variants that may influence surgery such as facial nerve course or mastoid pneumatization [10]. Multidetector CT (MDCT) can be performed in a relatively short time without a need for sedation and at a lower cost.

CT of the temporal bone is able to delineate the detailed anatomy of the inner ear, but can also help to demonstrate anatomical variants that may influence surgery. The two currently available and recommended CT scanners for the imaging of the temporal bone are multidetector CT (MDCT) and cone-beam CT (CBCT).

7.3.1 MDCT

MDCT is the most commonly used and widely available method to evaluate the temporal bone

B. Ozgen (✉)
Department of Radiology, University of Illinois at Chicago, Chicago, IL, USA
e-mail: bozgen2@uic.edu

© Springer Nature Switzerland AG 2022
L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_7

with CT. The image acquisition is performed in the axial plane; however, isotropic voxels allow reformatted images with high resolution in any additional plane. Scans have to extend from the top of the petrous apex to the mastoid tip in the axial direction with reformatted coronal images from the anterior tip of the petrous apex to the posterior margin of the mastoid. The imaging parameters are scanner specific but the collimation is usually chosen to be 0.5–0.625 mm and has to be less than 1 mm. Images should always be processed with a bone algorithm and viewed with a window width of 4000 HU and a window level of 200–500 HU [11].

7.3.2 CBCT

Although MDCT is used worldwide, CBCT using flat panel detector technology is slowly taking over for detailed evaluation of the small temporal bone structures [11]. The CBCT uses a rotating gantry and a cone-shaped X-ray beam that generates 3D volumetric dataset [11]. Newer cone-beam techniques offer higher resolution (0.15 mm thickness) but at lower radiation doses compared to traditional MDCT, making it valuable in the pediatric patient group. This technique is however more sensitive to motion as the acquisition usually lasts for 40 s and anesthesia may be required for small children.

For either technique, the CT of the temporal bone for the assessment of SNHL is routinely done without intravenous contrast.

7.4 MR Imaging

One of the main advantages of the MR imaging is the lack of ionizing radiation. However, due to longer exam times (of at least 20 min) the patient cooperation is paramount and anesthesia is usually required for small children. MR imaging has a higher soft tissue contrast compared to CT and is critical in the assessment of the cochlear nerve and also for a detailed evaluation of the auditory pathway [6, 8, 12–14]. Advances in MR imaging technology, including high-field-strength magnets, improved coil technology, and new sequence designs, allow increasingly more detailed imaging of the inner ear [15–18].

MR imaging should be performed with a 3.0 Tesla scanner, whenever possible, as higher field strength improves the signal-to-noise ratio (SNR) and increases the spatial resolution [17]. MR imaging for the evaluation of an implant candidate should include high-resolution heavily T2-weighted (T2W) sequence for a detailed evaluation of the membranous labyrinth but especially for the assessment of the cochlear nerve.

These sequences can be achieved with both gradient-echo (GRE) and fast spin-echo (FSE) T2-weighted techniques but the choice of which sequence to prefer is a heavily debated and published topic [19]. The most commonly used and widely available sequences include constructive interference into steady state (CISS), fast imaging employing steady-state acquisition (FIESTA), driven equilibrium radio frequency reset pulse (DRIVE), 3D true-fast imaging with steady-state precession (FISP), 3D T2 FSE, or 3D T2 FSE with fast recovery (FRFSE) depending on the scanner vendor. A high resolution of these sequences should be obtained despite thin slice thickness (of less than 1 mm) with appropriate increased scanning time. Those high T2 weighted images (with a spatial resolution approaching 0.4 mm) enable detailed evaluation of very small cochlear structures such as the interscalar septum and lamina spiralis but more importantly allow for rigorous assessment of the neural structures. For the accurate assessment of the cochlear nerve sagittal oblique images are required and although reformatted images can be obtained in the sagittal oblique plane from the axial dataset, bilateral direct sagittal oblique images, perpendicular to the IACs, with the same heavily T2-weighted sequence should always be acquired as the direct sagittal oblique images have a better resolution than reformatted images [20]. The T2-weighted imaging of the entire brain is also required to assess the auditory pathway [21, 22].

Intravenous contrast is not routinely administered in children assessed for SNHL unless there is a clinical concern for underlying neoplasm or infectious/inflammatory cause of hearing loss.

7.5 Imaging Evaluation

7.5.1 CT Evaluation

The imaging evaluation of a child with congenital SNHL primarily focuses on the detection of possible inner ear anomaly. The imaging appearances of different types of inner ear anomalies are further detailed in respective chapters.

Probably the most important function of the CT evaluation is to detect contraindications for the cochlear implantation including cochlear aplasia but also aplasia of the cochlear nerve canal/cochlear aperture (Fig. 7.1).

The atresia of the cochlear aperture is a strong indicator of underlying cochlear nerve anomaly [23, 24]. Tahir et al. reported that all 21 cases with cochlear aperture atresia in their series had accompanying cochlear nerve deficiency (either aplasia or hypoplasia) [23]. The dimension of a patent cochlear aperture thus needs to be assessed, as its diameter is a marker of the cochlear nerve status [23, 25]. The cochlear aperture is considered stenotic when it is narrower than 1.4 mm (Fig. 7.2) [7, 26–29]. It is critical to realize that the aperture can be stenotic in the presence of a normal-appearing and normal-sized cochlea; thus, a normal cochlear shape does not always indicate normal cochlear nerve structure and further imaging with MR is required to assess the cochlear nerve status [23]. The internal auditory canal size is also crucial for the preoperative assessment.

The IAC is considered stenotic when the diameter at its midpoint is smaller than 2 mm [30]. The IAC stenosis or atresia may easily be demonstrated by CT and the finding of a narrow or aplastic IAC again should raise concern for a deficiency of the cochlear nerve [24, 31]. However, the IAC morphology is an unreliable surrogate marker of CN integrity and as reported by Adunka et al., a normal IAC diameter can be

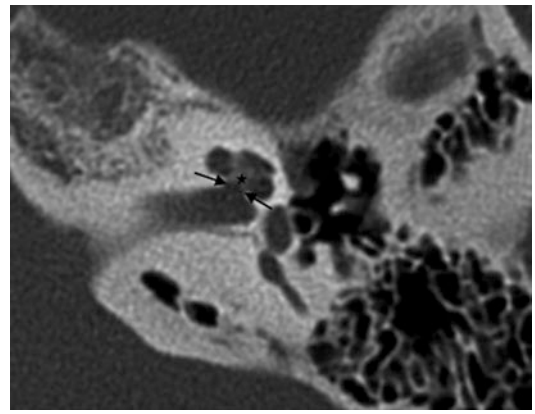


Fig. 7.1 CT anatomy of the cochlea. Axial temporal bone CT image demonstrates normal appearance of the cochlear aperture (delineated by the arrows). Note the normal appearance of the Modiolus (star) at the base of the cochlea

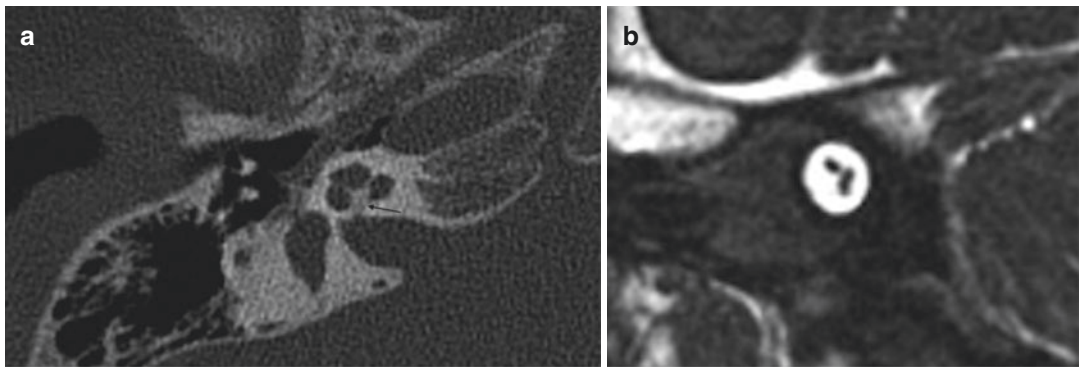


Fig. 7.2 Axial temporal bone CT (**a**) and sagittal oblique 3D-DRIVE image (**b**) of a patient with bilateral congenital severe SNHL. The CT image of the right ear reveals atresia of the right cochlear aperture (arrow) with aplasia of the cochlear nerve on the corresponding MR image (**b**)

seen in up to half of cochlear nerve aplasia patients [23, 32].

During the imaging evaluation, the cochlea should be carefully assessed not only for possible malformations as the type and severity of the cochlear anomaly will determine the type of the implant used, but also for possible presence of labyrinthine ossificans (Fig. 7.3). Furthermore, CT may demonstrate anomalies of the bony labyrinth such as Paget and otosclerosis (Fig. 7.4) that could increase the incidence of postimplant complications such as facial nerve irritation.

Preoperative assessment should also detect entities that may increase the degree of surgical difficulty such as mastoid sclerosis, abnormal sigmoid sinus position, high riding jugular bulb, aberrant carotid artery, and especially dehiscent or aberrant facial nerve. The size of the round window should also be assessed as there might be congenital or acquired stenosis of the round window [33]. Additionally, the thickness of the bone at the potential pedestal placement and large emissary veins in that location should also be noticed and conveyed to the referring physician.

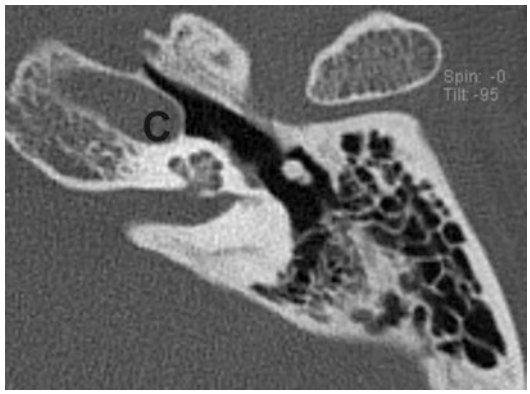


Fig. 7.3 Axial temporal bone CT revealing mineralization within the cochlea consistent with labyrinthine ossificans (*c* carotid canal)

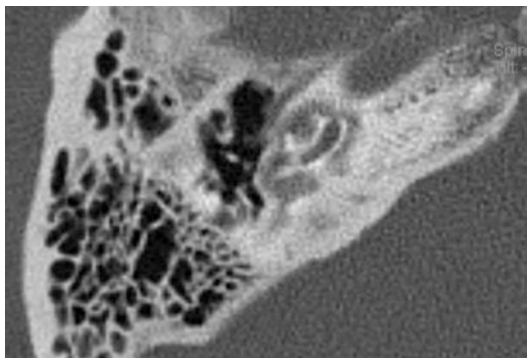


Fig. 7.4 Axial temporal bone CT revealing pericochlear lucency consistent with **extensive** retrofenestral otosclerosis

7.5.2 MR Imaging Evaluation

MRI with the heavily T2-weighted gradient-echo sequence allows a high spatial resolution that allows detailed visualization of the cochlear structures (Fig. 7.5a), allowing accurate depiction of the modiolus, lamina spiralis, and interscalar septum [34, 35].

Radiological assessment with MR not only gives detailed information regarding the inner ear structures but it is also essential for the assessment of the cochlear nerve. The evaluation of the cochleovestibular nerve and especially of its cochlear branch is of extreme importance prior to cochlear implantation. In a normal-sized IAC the diagnosis of cochlear nerve aplasia is relatively straightforward with dedicated sagittal oblique high-resolution images (Fig. 7.5b, c) [36]. However, in a very stenotic IAC, the diagnosis may be difficult because of the inability to separate the nerves [30, 32]. Again the visualized inner ear may be normal or have subtle abnormality despite severe deficiency of the cochlear nerve [12, 36]. Differentiation between hypoplasia and a normal size of the cochlear nerve can also be challenging and requires the highest possible resolution [12]. There is not a well-defined consensus regarding the definition of cochlear nerve hypoplasia. Li et al. defined cochlear nerve hypoplasia as a cochlear nerve with a diameter smaller than that of the facial nerve, seen on the oblique sagittal images. Similarly Glastonbury designated the cochlear nerve as small when it appeared decreased in size compared with the other nerves of the IAC [12]. It is critical to recognize that there might be

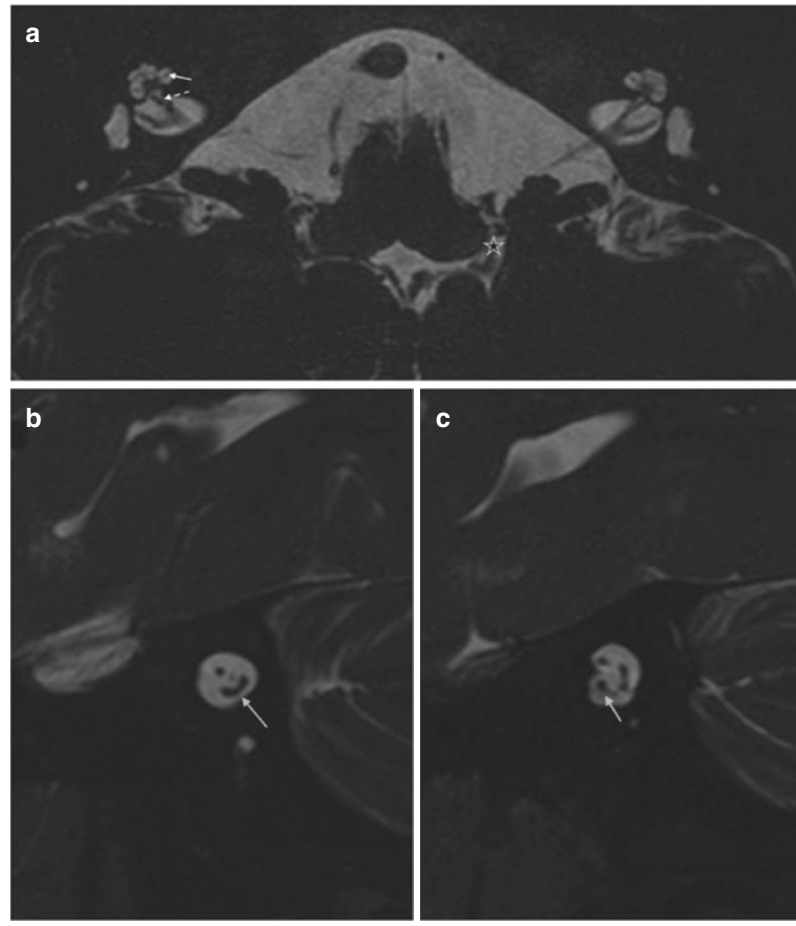


Fig. 7.5 Anatomy of the cochlea and cochlear nerve by high-resolution MR. Heavy T2-weighted driven equilibrium (DRIVE) images in axial (**a**) and sagittal oblique (**b** and **c**) planes. The cochlear turns with internal spiral lamina (arrow) are visible with this high T2-weighted axial image (**a**). The cochlear nerve (dotted arrow) is seen at the fundus of the IAC (**a**). With sagittal oblique imaging, the vestibulocochlear nerve (arrow) is seen as a crescent-shaped structure at the medial aspect of the IAC (**b**); however more laterally the cochlear nerve (arrow) can be seen separately from the inferior and superior vestibular nerves (**c**)

occasional discrepancy between the imaging and audiological findings regarding the presence/functionality of the cochlear nerve [37, 38]. Several studies have shown that subsets of patients with cochlear nerve aplasia have positive audiological responses and might derive benefit from cochlear implantation [37–39]. Anatomical connections between the cochlear nerve and other branches of the vestibulocochlear complex that are below the resolution of the current MR imaging might be responsible for this radiological-audiological inconsistency [40]. Imaging with ultra-high field magnets with DTI fiber tractography might solve this problem in the future [18, 41].

In every patient who is a candidate for a CI or ABI placement, the imaging of the brainstem and supratentorial brain structures with MRI is crucial to verify the integrity of the auditory pathways up to the temporal cortex but also to determine possible underlying congenital or acquired malformations that might hinder post-implant rehabilitation [42, 43].

MR imaging is better in delineating details of the brain anatomy but it is somewhat limited in the brainstem [44]. The difficulties in assessing the brainstem by using MR imaging arise not only from the small size of various brainstem structures but also from the fact that those anatomical components do not exhibit enough contrast to enable their individual identification [45]. Therefore, when relaxation-based MR image contrast is used, despite high resolution, conspicuity of those structures such as cranial nerve nuclei cannot be achieved in clinical field strengths [46]. Nevertheless, the bulge of the medulla into the lateral recess of the fourth ventricle and to the foramen of Luschka caused by

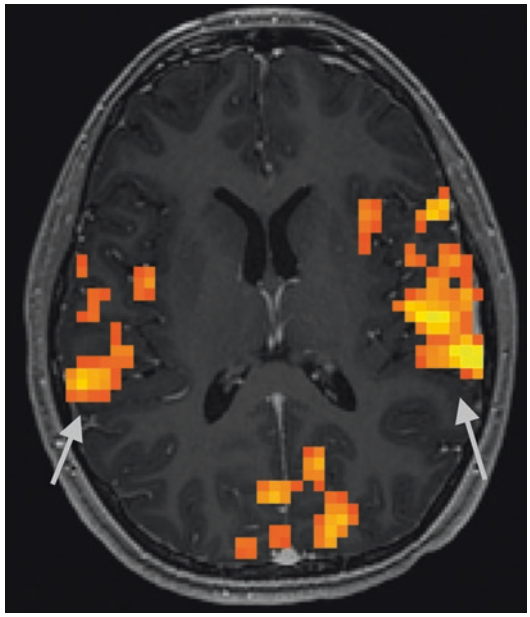


Fig. 7.6 Axial image from a BOLD fMRI study demonstrating bilateral activation of the auditory cortex (arrows). (Courtesy of Dr. Keith Thulborn)

the cochlear nuclear complex can be easily identified by MR imaging [47]. There is significant variability of the anatomy of the lateral recess in children with congenital deafness due to abnormalities of embryonic and fetal development [48].

It has been previously reported that congenital developmental abnormalities of the brain is more common in patients with auditory neuropathy spectrum disorder [1, 22, 48]. In patients with bilateral cochlear nerve deficiency hindbrain anomalies, such as pontine hypoplasia, were reported to be the most common abnormal intracranial finding [48]. Additionally there might be evidence of central pathologies such as chronic changes of hypoxic-ischemic injury, kernicterus, and chronic changes of congenital CNS infections [49, 50]. White matter lesions are also common findings in the preimplant imaging of the CI/ABI candidates [22, 51]. These lesions are nonspecific but more diffuse and prominent parenchymal changes were found to represent negative prognostic factors for speech and language development [21, 50, 51]. It is therefore critical to make a comprehensive evaluation of the brainstem and cerebrum in each CI and ABI candidate.

Similar to the cochlear nuclear complex, the ascending fibers of the auditory pathway are not visible with normal visual inspection of routine MR sequences. The auditory radiation can only be demonstrated with dedicated fiber tracking obtained from diffusion tensor imaging [52]. With new developing technologies, MRI also has the potential to study the anatomical and functional organization of the auditory cortex through voxel-based morphometry and functional MRI (fMRI) (Fig. 7.6) [53]. DTI metrics, such as fractional anisotropy, may prove important in selecting patients and predicting outcomes after the implantation [54].

References

1. Joshi VM, Navlekar SK, Kishore GR, Reddy KJ, Kumar ECV. CT and MR imaging of the inner ear and brain in children with congenital sensorineural hearing loss. Radiographics. 2012;32(3):683–98. https://doi.org/10.1148/rg.323115073.
2. Brenner DJ, Hall EJ. Computed tomography—an increasing source of radiation exposure. N Engl J Med. 2007;357(22):2277–84. https://doi.org/10.1056/NEJMra072149.
3. Trimble K, Blaser S, James AL, Papsin BC. Computed tomography and/or magnetic resonance imaging before pediatric cochlear implantation? Developing an investigative strategy. Otol Neurotol. 2007;28(3):317–24. https://doi.org/10.1097/01.mao.0000253285.40995.91.
4. Lo WW. Imaging of cochlear and auditory brain stem implantation. AJNR Am J Neuroradiol. 1998;19(6):1147–54.
5. Marsot-Dupuch K, Meyer B. Cochlear implant assessment: imaging issues. Eur J Radiol. 2001;40(2):119–32.
6. Sennaroglu L, Saatci I, Aralasmak A, Gursel B, Turan E. Magnetic resonance imaging versus computed tomography in pre-operative evaluation of cochlear implant candidates with congenital hearing loss. J Laryngol Otol. 2002;116(10):804–10. https://doi.org/10.1258/00222150260293619.
7. Miyasaka M, Nosaka S, Morimoto N, Taiji H, Masaki H. CT and MR imaging for pediatric cochlear implantation: emphasis on the relationship between the cochlear nerve canal and the cochlear nerve. Pediatr Radiol. 2010;40(9):1509–16. https://doi.org/10.1007/s00247-010-1609-7.

8. Connor SE. Contemporary imaging of auditory implants. Clin Radiol. 2017;73(1):19–34. https://doi.org/10.1016/j.crad.2017.03.002.
9. Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. Otol Neurotol. 2005;26(5):976–82.
10. Henderson E, Wilkins A, Huang L, Kenna M, Gopen Q. Histopathologic investigation of the dimensions of the cochlear nerve canal in normal temporal bones. Int J Pediatr Otorhinolaryngol. 2011;75(4):464–7. https://doi.org/10.1016/j.ijporl.2010.11.024.
11. Lemmerling M, de De Foer B. Temporal bone imaging. Berlin, Heidelberg: Springer; 2014.
12. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. AJNR Am J Neuroradiol. 2002;23(4):635–43.
13. Russo EE, Manolidis S, Morriss MC. Cochlear nerve size evaluation in children with sensorineural hearing loss by high-resolution magnetic resonance imaging. Am J Otolaryngol. 2006;27(3):166–72. https://doi.org/10.1016/j.amjoto.2005.09.007.
14. Young NM, Rojas C, Deng J, Burrowes D, Ryan M. Magnetic resonance imaging of Cochlear implant recipients. Otol Neurotol. 2016;37(6):665–71. https://doi.org/10.1097/MAO.0000000000001053.
15. van Egmond SL, Visser F, Pameijer FA, Grolman W. Ex vivo and in vivo imaging of the inner ear at 7 tesla MRI. Otol Neurotol. 2014;35(4):725–9. https://doi.org/10.1097/MAO.0000000000000276.
16. van Egmond SL, Visser F, Pameijer FA, Grolman W. In vivo imaging of the inner ear at 7T MRI: image evaluation and comparison with 3T. Otol Neurotol. 2015;36(4):687–93. https://doi.org/10.1097/MAO.0000000000000621.
17. Schulze M, Reimann K, Seeger A, Klose U, Ernemann U, Hauser TK. Improvement in imaging common temporal bone pathologies at 3 T MRI: small structures benefit from a small field of view. Clin Radiol. 2017;72(3):267e261–12. https://doi.org/10.1016/j.crad.2016.11.019.
18. Thylur DS, Jacobs RE, Go JL, Toga AW, Niparko JK. Ultra-high-field magnetic resonance imaging of the human inner ear at 11.7 tesla. Otol Neurotol. 2017;38(1):133–8. https://doi.org/10.1097/MAO.0000000000001242.
19. Glastonbury C. The vestibulocochlear nerve, with an emphasis on the normal and diseased internal auditory canal and cerebellopontine angle. In: Imaging of the temporal bone. New York, NY: Thieme Medical; 2009. p. 480–558.
20. Noij KS, Remenschneider AK, Kozin ED, Puram S, Herrmann B, Cohen M, Cunnane MB, Lee DJ. Direct parasagittal magnetic resonance imaging of the internal auditory canal to determine cochlear or auditory brainstem implant candidacy in children. Laryngoscope. 2015;125(10):2382–5. https://doi.org/10.1002/lary.25228.
21. Moon IJ, Kim EY, Park GY, Jang MS, Kim JH, Lee J, Chung WH, Cho YS, Hong SH. The clinical significance of preoperative brain magnetic resonance imaging in pediatric cochlear implant recipients. Audiol Neurootol. 2012;17(6):373–80. https://doi.org/10.1159/000341818.
22. Lapointe A, Viamonte C, Morriss MC, Manolidis S. Central nervous system findings by magnetic resonance in children with profound sensorineural hearing loss. Int J Pediatr Otorhinolaryngol. 2006;70(5):863–8. https://doi.org/10.1016/j.ijporl.2005.09.022.
23. Tahir E, Bajin MD, Atay G, Mocan BO, Sennaroglu L. Bony cochlear nerve canal and internal auditory canal measures predict cochlear nerve status. J Laryngol Otol. 2017;131(8):676–83. https://doi.org/10.1017/S0022215117001141.
24. Li Y, Yang J, Liu J, Wu H. Restudy of malformations of the internal auditory meatus, cochlear nerve canal and cochlear nerve. Eur Arch Otorhinolaryngol. 2015;272(7):1587–96. https://doi.org/10.1007/s00405-014-2951-4.
25. Fatterpekar GM, Mukherji SK, Alley J, Lin Y, Castillo M. Hypoplasia of the bony canal for the cochlear nerve in patients with congenital sensorineural hearing loss: initial observations. Radiology. 2000;215(1):243–6. https://doi.org/10.1148/radiology.215.1.r00ap36243.
26. D'Arco F, Talenti G, Lakshmanan R, Stephenson K, Siddiqui A, Carney O. Do measurements of inner ear structures help in the diagnosis of inner ear malformations? A review of literature. Otol Neurotol. 2017;38(10):e384–92. https://doi.org/10.1097/mao.0000000000001604.
27. Lan M-Y, Shiao J-Y, Ho C-Y, Hung H-C. Measurements of normal inner ear on computed tomography in children with congenital sensorineural hearing loss. Eur Arch Otorhinolaryngol. 2009;266(9):1361–4. https://doi.org/10.1007/s00405-009-0923-x.
28. Stjernholm C, Muren C. Dimensions of the cochlear nerve canal: a radioanatomic investigation. Acta Otolaryngol. 2002;122(1):43–8.
29. Yi JS, Lim HW, Kang BC, Park S-Y, Park HJ, Lee K-S. Proportion of bony cochlear nerve canal anomalies in unilateral sensorineural hearing loss in children. Int J Pediatr Otorhinolaryngol. 2013;77(4):530–3. https://doi.org/10.1016/j.ijporl.2012.12.031.
30. Romo LV, Casselman JW, Robson CD. Temporal bone: congenital anomalies. In: Som PM, Curtin HD, editors. Head and neck imaging. 5th ed. St. Louis, MO: Elsevier Health Sciences; 2011. p. 1097–165.
31. Shelton C, Luxford WM, Tonokawa LL, Lo WW, House WF. The narrow internal auditory canal in children: a contraindication to cochlear implants. Otolaryngol Head Neck Surg. 1989;100(3):227–31. https://doi.org/10.1177/019459988910000310.
32. Adunka OF, Roush PA, Teagle HFB, Brown CJ, Zdanski CJ, Jewells V, Buchman CA. Internal auditory canal morphology in children with Cochlear nerve deficiency. Otol Neurotol. 2006;27(6):793–801. https://doi.org/10.1097/01.mao.0000227895.34915.94.

33. Monsanto RC, Sennaroglu L, Uchiyama M, Sancak IG, Paparella MM, Cureoglu S. Histopathology of inner ear malformations: potential pitfalls for Cochlear implantation. Otol Neurotol. 2019;40(8):e839–46. https://doi.org/10.1097/mao.0000000000002356.
34. Reinshagen KL, Curtin HD, Quesnel AM, Juliano AF. Measurement for detection of incomplete partition type II anomalies on MR imaging. Am J Neuroradiol. 2017;38(10):2003–7. https://doi.org/10.3174/ajnr.A5335.
35. Davidson HC, Harnsberger HR, Lemmerling MM, Mancuso AA, White DK, Tong KA, Dahlen RT, Shelton C. MR evaluation of vestibulocochlear anomalies associated with large endolymphatic duct and sac. Am J Neuroradiol. 1999;20(8):1435–41.
36. 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. https://doi.org/10.1148/radiology.202.3.9051033.
37. Peng KA, Kuan EC, Hagan S, Wilkinson EP, Miller ME. Cochlear nerve aplasia and hypoplasia: predictors of Cochlear implant success. Otolaryngol Head Neck Surg. 2017;157(3):392–400. https://doi.org/10.1177/0194599817718798.
38. Young NM, Kim FM, Ryan ME, Tournis E, Yaras S. Pediatric cochlear implantation of children with eighth nerve deficiency. Int J Pediatr Otorhinolaryngol. 2012;76(10):1442–8. https://doi.org/10.1016/j.ijporl.2012.06.019.
39. Acker T, Mathur NN, Savy L, Graham JM. Is there a functioning vestibulocochlear nerve? Cochlear implantation in a child with symmetrical auditory findings but asymmetric imaging. Int J Pediatr Otorhinolaryngol. 2001;57(2):171–6. https://doi.org/10.1016/S0165-5876(00)00458-4.
40. Ozdogmus O, Sezen O, Kubilay U, Saka E, Duman U, San T, Cavdar S. Connections between the facial, vestibular and cochlear nerve bundles within the internal auditory canal. J Anat. 2004;205(1):65–75. https://doi.org/10.1111/j.0021-8782.2004.00313.x.
41. Vos SB, Haakma W, Versnel H, Froeling M, Speleman L, Dik P, Viergever MA, Leemans A, Grolman W. Diffusion tensor imaging of the auditory nerve in patients with long-term single-sided deafness. Hear Res. 2015;323:1–8. https://doi.org/10.1016/j.heares.2015.01.010.
42. Sennaroglu L, Ziyal I. Auditory brainstem implantation. Auris Nasus Larynx. 2012;39(5):439–50. https://doi.org/10.1016/j.anl.2011.10.013.
43. Colletti G, Mandala M, Colletti L, Colletti V. Nervus intermedius guides auditory brainstem implant surgery in children with Cochlear nerve deficiency. Otolaryngol Head Neck Surg. 2016;154(2):335–42. https://doi.org/10.1177/0194599815615858.
44. Sclocco R, Beissner F, Bianciardi M, Polimeni JR, Napadow V. Challenges and opportunities for brainstem neuroimaging with ultrahigh field MRI. Neuroimage. 2018;168:412–26. https://doi.org/10.1016/j.neuroimage.2017.02.052.
45. Beissner F. Functional MRI of the brainstem: common problems and their solutions. Clin Neuroradiol. 2015;25(Suppl 2):251–7. https://doi.org/10.1007/s00062-015-0404-0.
46. Lambert C, Lutti A, Helms G, Frackowiak R, Ashburner J. Multiparametric brainstem segmentation using a modified multivariate mixture of Gaussians. Neuroimage Clin. 2013;2:684–94. https://doi.org/10.1016/j.nicl.2013.04.017.
47. Gebarski SS, Tucci DL, Telian SA. The cochlear nuclear complex: MR location and abnormalities. AJNR Am J Neuroradiol. 1993;14(6):1311–8.
48. Huang BY, Roche JP, Buchman CA, Castillo M. Brain stem and inner ear abnormalities in children with auditory neuropathy spectrum disorder and cochlear nerve deficiency. AJNR Am J Neuroradiol. 2010;31(10):1972–9. https://doi.org/10.3174/ajnr.A2178.
49. Jallu AS, Jehangir M, Ul Hamid W, Pampori RA. Imaging evaluation of pediatric sensorineural hearing loss in potential candidates for Cochlear implantation. Indian J Otolaryngol Head Neck Surg. 2015;67(4):341–6. https://doi.org/10.1007/s12070-015-0819-6.
50. Xu XQ, Wu FY, Hu H, Su GY, Shen J. Incidence of brain abnormalities detected on preoperative brain MR imaging and their effect on the outcome of Cochlear implantation in children with sensorineural hearing loss. Int J Biomed Imaging. 2015;2015:275786. https://doi.org/10.1155/2015/275786.
51. Hong P, Jurkowski ZC, Carvalho DS. Preoperative cerebral magnetic resonance imaging and white matter changes in pediatric cochlear implant recipients. Int J Pediatr Otorhinolaryngol. 2010;74(6):658–60. https://doi.org/10.1016/j.ijporl.2010.03.014.
52. Javad F, Warren JD, Micallef C, Thornton JS, Golay X, Yousry T, Mancini L. Auditory tracts identified with combined fMRI and diffusion tractography. Neuroimage. 2014;84:562–74. https://doi.org/10.1016/j.neuroimage.2013.09.007.
53. Semenza C, Cavinato M, Rigon J, Battel I, Meneghello F, Venneri A. Persistent cortical deafness: a voxel-based morphometry and tractography study. Neuropsychology. 2012;26(6):675–83. https://doi.org/10.1037/a0029688.
54. Huang L, Zheng W, Wu C, Wei X, Wu X, Wang Y, Zheng H. Diffusion tensor imaging of the auditory neural pathway for clinical outcome of Cochlear implantation in pediatric congenital sensorineural hearing loss patients. PLoS One. 2015;10(10):e0140643. https://doi.org/10.1371/journal.pone.0140643.

8 Preoperative Speech and Language Evaluation

Esra Yücel, Filiz Aslan, and Burcu Özkan

The performance of cochlear implanted children varies widely across subjects. That variation in both speech perception/production and in communication skills depends on physiologic and environmental factors. Inner ear malformations (IEM) are found in approximately %20 of children with congenital sensorineural hearing loss and are among the primary causes of congenital deafness [1].

Children with IEMs may confront different developmental issues. Kaga [2, 3] reported three main difficulties specifically common in this group; their language development is poor as they do not get enough auditory benefit with hearing aids and cochlear implantation is therefore indicated. Due to poor and variable auditory skills it is difficult to predict the outcome of cochlear implantation. Depending on the type of IEM and auditory nerve, especially balance issues are noteworthy. Balance problems usually result in delayed fine and gross motor development which is crucial for independent learning and sensory integration tasks. Therefore, especially for children with IEMs, more extensive evaluation process should be planned.

Decision-making for a cochlear implantation in a child with IEM requires careful consideration and thorough counseling. The success of an interdisciplinary approach depends upon collaboration among an experienced team that includes the family as equal partners in the decision-making process.

The process consists of detailed medical and audiological evaluation which should also be supported by an evaluation of rehabilitative audiologist, physiotherapist/ergo therapist, other interventionists, educators, and always the parents. During the evaluation process the following questions should be addressed:

- Which technology will convey the clearest and audible auditory signal that enables the child's auditory learning, helps developing speech and communication skills, conveys proficiency in academic circumstances and psychosocial competence?
- Are there qualified and experienced interventionists who will actively play role in meeting child's listening, communication, speech-language, and learning needs?
- Are there any aspects of the child that will require consideration of different forms of communication?
- If the child needs different forms of communication excluding auditory verbal form, is it possible to address the effects of these alternative learning strategies on expected outcomes of cochlear implantation?
- What are the safety issues that should be considered to minimize any potential risk for the cochlear implant surgery, mapping, and habilitation?

E. Yücel (✉) · F. Aslan · B. Özkan
Department of Audiology, Hacettepe University Faculty of Health Sciences, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_8

In order to give answers to the questions listed above, cochlear implant team members should approach the child and family with a holistic approach. Traditionally, during a preoperative speech-language and communication evaluation vocabulary, receptive and expressive language development and speech production should be examined in terms of quantity and quality. These findings identify areas that need improvement and also laying out the current state of the child. Also serve as comparative data for the measurement of benefit and progress.

The speech-language and communication outcomes with any degree of IEMs are characterized by a wide spectrum in functional listening skills and linguistic competency. For that reason, for any child from this group the communication assessment should involve functional listening evaluation in different circumstances. This evaluation conveys valuable data about speech perception skills such as using auditory or visual cues in maintaining language learning and communication.

8.1 Comprehensive Evaluation

There are two main objectives in the evaluation process:

1. To draw a map of child's communication form and communicative weaknesses and strengths.
2. To monitor the child's general developmental progress and effectiveness of the rehabilitation-mapping process.

During the initial evaluation the most appropriate assessment tools and method for ongoing evaluation process should be determined. It is also crucial to integrate the medical-audiological findings and developmental history of the child and also expectations of the parents. In general, the initial assessment takes about 2 h with breaks accompanied by play, arts, and crafts or reading which also provides valuable observation opportunity about child's social interaction and behavior, cognitive skills, and parental involvement. It is also useful to inform parents to bring some favorite materials and toys from home as familiar objects always will enable the child to feel himself safe and secure during the activities.

Findings from an initial speech-language evaluation, and general development, and functional listening evaluation are used to identify the baseline in several skill domains as following; Ying [4]

- Social and emotional: Getting along with people and caring for personal needs.
- Language/communication: Hearing, understanding, and using language.
- Cognitive (learning, thinking, problem-solving)/fine motor-adaptive: Eye–hand coordination, manipulation of small objects, and problem-solving.
- Gross motor: Sitting, walking, jumping, and overall large muscle movement.
- Phonemic awareness: Detection, discrimination, and identification skills of vowels and consonants as they occur in isolation or in different positions within words.
- Word recognition: Open-set word identification under unilateral and binaural listening conditions in quite if possible in noise.
- Vocabulary: Understanding or expressive use of real words or sound associations representing real words (e.g., hop hop for ball).
- Language comprehension/receptive language: Understanding or/and contingent response to spoken language, including pointing to pictures, manipulating the objects or verbally responding.
- Expressive language: Both verbal and nonverbal communication attempts.
- Speech production: Phonemic repertoire, articulation of isolated sounds, repeated and named in words, word combinations, prosody, and intonation.
- Pragmatic functioning: Spoken language used for communication such as labeling, commenting, directing, and questioning.

Also, through formal and informal evaluation detailed information should be questioned such

as age of identification, communication modality, usage of hearing aids (age at start using, regularity, willingness for each ear), cognitive skills (attention, memory), parental factors (participation, being aware of child's needs and strengths, expectation, participation in the provision of the rich stimuli which is required for child's integrated developmental path).

Table 8.1 Assessment tools for comprehensive evaluation

Assessment tools	Age range	Areas of assessment	Reference
Language			
Test of Early Language Development 3: Turkish Version **(TELD-3)**	2–8 years	Syntax, morphologic, semantic	Güven and Topbaş [5]
Test of Language Development-4 Primary: Turkish **(TOLD-4)**	4–9 years	Syntax, morphologic, semantic	Topbaş and Güven [6]
Turkish Expressive and Receptive Language Test **(TIFALDI)**	2–12 years	Expressive and receptive language vocabulary sub-scales	Berument and Güven [7]
Manchester Spoken Language Development Scale **(MSLDS)** Levels	0–6 years	Expressive language	Wallis et al. [8]
Auditory perception			
Children Test **(CIAT)**	2–15 years	1. Detection of speech sounds 1.a) phoneme detection 1.b)MAIS/IT-MAIS 2. Perception of suprasegmentals 2.a) synthetic syllable discrimination 2.b) synthetic syllable identification 2.c) pattern perception (Turkish speech perception test) 3. Speech identification 3.a) word identification (Turkish speech perception test) 3.b) sentence identification 3.c) sentence identification for young children (Mr. potato head test) 4. Integration of visual and auditory input 5. Modified open-set speech recognition 6. Open-set speech recognition 6.a) Turkish sentence recognition test 6.b) comprehension of basic instructions	Yücel and Sennaroglu [9]
Categories of Auditory Performance **(CAP)**-II	Birth–6 years	To evaluate child's auditory abilities	Archbold et al. [10]
Speech intelligibility			
Ankara Articulation Test **(AAT)**	2–12 years	To assess overall of correctness of phonemes (e.g., place, manner, voicing)	Ege et al. [11]
Turkish Articulation and Phonology Test	2–8 years	To assess articulation and phonological development	Topbaş [12]
Speech Intelligibility Rating **(SIR)**	2–older	To understand the development of speech intelligibility	Allen et al. [13]
Developmental screening			
Denver Developmental Screening Test II: Turkish Version	Birth–7 years	Personal-social, fine motor-adaptive, language, gross motor	Anlar et al. [14]

(continued)

Table 8.1 (continued)

Assessment tools	Age range	Areas of assessment	Reference
Ankara Developmental Screening Inventory **(AGTE)**	Birth-6 years	Cognitive development, fine motor development, gross motor development, and social skills self-development	Savaşır et al. [15]
Functional auditory assessment			
Functional Auditory Performance Inventory **(FAPCI)**	Birth–5 years	To assess the communication skills of children with hearing loss	Clark et al. [16]
Meaningful Auditory Integration Scale **(IT/MAIS)**	Birth–6 years	To evaluate meaningful use of sound and everyday situations	Robbins et al. [17]
Sensory Integration Sensory Profile	3–10 years	To measure children's responses to commonly occurring sensory experiences	Dunn [18]

Tests and questionnaires which are commonly used for this group are summarized in Table 8.1.

8.2 Selection of Test Protocols

Children with hearing loss have distinctly differing diagnostic and habilitative requirements. It is very important to describe individualized needs of each child in order to select the most appropriate evaluation techniques and constitute the individualized habilitation program. It is also critical to select the age appropriate diagnostic tools and formulate the developmental expectations based on interpretations of the outcomes.

Children with IEMs may also suffer from a number of problems. First, language development based on auditory skills is poor when using hearing aids. Second, because of the poor hearing ability in these children, it is often encountered that they were not able to develop a formal language system that would be insusceptible for standard evaluation. Third, children with congenital deafness also tend to have balance disorders, resulting in delayed motor development that may complicate the application of standard tests in particular period and procedure. It is also controversial to conduct a speech-language evaluation and functional listening assessment for children who have only recently been diagnosed and had not developed any listening and/or attention skills with their partially intact auditory system. Usually these factors leave professionals to make available more than one assessment tool and procedure at the same time of evaluation. When the child with inner ear malformations continues to exhibit limited functional listening or oral language skills, the most appropriate evaluation may be parent-report inventories or criterion-referenced assessment tools; keeping in mind that is critical to include some tool to assess the nature and consistency of the child's auditory and communicative demands [19].

The most effective method is to observe the child's reactions to different noisemakers in various frequency ranges by exposing the child in several opportunities in a natural setting. By providing controlled exposure to the multi-sensorial stimuli in a natural environment child's attention, memory, learning and problem-solving skills should be observed. This information is required for prediction of the developmental process of the child and the alternative habilitation techniques after cochlear implant surgery. For children who have hypoplastic nerve and had cochlear implantation it is crucial to observe their ongoing developmental rate on every higher auditory skills such as temporal and spectral analysis, ordering, loudness, and intensity analysis in order to decide the ABI need for contralateral ear.

References

1. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34:397–411.
2. Kaga K. Vestibular compensation in infants and children with congenital and acquired vestibular

loss in both ears. Int J Pediatr Otorhinolaryngol. 1999;49:215–24.
3. Kaga K, Suzuki J, Marsh RR. Influence of labyrinthine hypoactivity on gross motor development of infants. Ann N Y Acad Sci. 1981;374:412–20.
4. Ying E. Speech/language/auditory management of infants and children with hearing loss. In: Madell JR, Flexer C, editors. Pediatric audiology: diagnosis, technology, and management. 2nd ed. New York: Thieme Medical; 2014.
5. Güven S, Topbaş S. Adaptation of the test of early language development-(TELD-3) into turkish: reliability and validity study. Int J Early Childhood Spec Educ. 2014;6(2):151–76.
6. Topbaş S, Güven O. TODİL: Türkçe Okulçağı Dil Gelişim Testi (TOLD-P: 4-Turkish Version). In: TODİL Projesi: Anadili Türkçe Olan Tek-Dilli ve İki Dilli Okul Öncesi ve İlköğretim Çağı Çocuklarında (2: 0-9: 0) Özgül Dil Bozukluklarını Ölçme ve Değerlendirme Çalışması; 2013. p. 71–90.
7. Berument SK, Güven AG. Türkçe İfade Edici ve Alıcı Dil (TİFALDİ) Testi: I. alıcı dil kelime alt testi standardizasyon ve güvenilirlik geçerlik çalışması. Türk Psikiyatri Dergisi. 2013;24(3):192–201.
8. Wallis A, Sadadcharam M, Fullwood C, Henderson L, Bruce IA, Freeman S. Validation of the manchester speech and language development scale. Under journal review. 2017.
9. Yucel E, Sennaroglu G. Çocuklar için işitsel algı testi (ÇİAT). İstanbul: Advanced Bionics; 2011.
10. Archbold S, Lutman ME, Marshall DH. Categories of auditory performance. Ann Otol Rhinol Laryngol. 1995;166:312–4.
11. Ege P, Acarlar F, Turan F. Ankara Artikülasyon Testi (AAT). Ankara: Ankara Üniversitesi, Bilimsel Araştırmalar Projesi Yayını; 2004.
12. Topbaş SS. Türkçe sesletim sesbilgisi testi. Milli Eğitim Bakanlığı. Ankara: TDK Yayınları; 2005.
13. Allen MC, Nikolopoulos TP, O'donoghue GM. Speech intelligibility in children after cochlear implantation. Am J Otol. 1998;19(6):742–6.
14. Anlar B, Bayoğlu BU, Yalaz K. Denver II developmental screening test "adaptation and standardization to Turkish children". Ankara: Developmental Child Neurology Association; 2009.
15. Savasir I, Sezgin N, Erol N. Ankara development inventory handbook. Turkey: Turkish Psychological Association; 1998.
16. Clark JH, Aggarwal P, Wang NY, Robinson R, Niparko JK, Lin FR. Measuring communicative performance with the FAPCI instrument: preliminary results from normal hearing and cochlear implanted children. Int J Pediatr Otorhinolaryngol. 2011;75(4):549–53.
17. Robbins AM, Renshaw JJ, Berry SW. Evaluating meaningful auditory integration in profoundly hearing-impaired children. Am J Otol. 1991;12:144–50.
18. Dunn W. Sensory profile: users manual. San Antonio, TX: Psychological Corp; 1999.
19. Anderson K, Arnoldi KA. Building skills for success in the fast-paced classroom. Hillsboro, OR: Butte; 2011.

9 Preoperative Neurosurgical Evaluation of Children Undergoing Auditory Brainstem Implantation

Burcak Bilginer and İbrahim Ziyal

Special Features

1. Families are informed about the important risks of auditory brainstem implantation.
2. Preoperative imaging may show certain contraindications to retrosigmoid approach.

Indication for auditory brainstem implantation (ABI) is given according to the radiological and audiological findings. Role of pediatric neurosurgeon in inner ear malformations (IEM) is in the surgery of ABI. As this is an intracranial operation, it involves important risks. As neurosurgeon we have the key role in the surgery, which includes evaluation of the possible risks before surgery and informing the parents about the risk of surgery.

Surgical complications are damage to the brainstem, vessels, and cranial nerves in the area [1]. Cranial nerves are facial, vestibulocochlear, glossopharyngeal, vagus, and accessories nerves. The parents are informed about the possible complications related to the damage to any of these nerves. Wound infection or an infection at the surgical field and CSF leak can also be seen as important postoperative complications. Until September 2018, we have performed 128 ABI surgeries in our center and we have noticed that with experience it is rare to have any of the nerve damage. There is an increased risk of surgical complications in revision surgery which is usually done in cases of device failure. Fibrosis in the surgical field increases not only the damage to the nerves but to the brainstem as well. If there is CSF leak at postoperative period, external lumber drainage systems can be used to solve the problem, but dural repair can rarely be needed. Surgical field infection is a rare but an important problem which can result in removing the device.

Imaging may show special features which may complicate a surgery or even provide a contraindication. Large emissary veins sometimes can cause problems. Because of the patients age, bleeding from these vessels requires blood transfusion. If such a condition has been detected preoperatively, operation side should be changed or translabyrinthine approach can be preferred. One such case is presented in Fig. 9.1. One-year male patient had unsuccessful cochlear implantation in a case of cochlear aplasia with vestibular dilatation on the right side and was referred for ABI on the left side. His computerized tomography revealed a large emissary vein in the area of retrosigmoid approach (Fig. 9.1). A dam-

B. Bilginer (✉)
Department of Neurosurgery, Hacettepe University Medical Faculty, Ankara, Turkey
e-mail: burcak@tr.net

İ. Ziyal
Department of Neurosurgery, Marmara University Medical Faculty, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_9

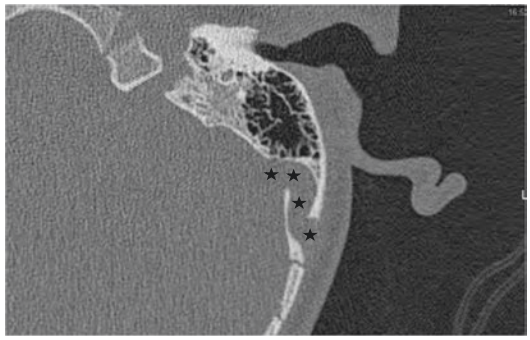


Fig. 9.1 Computerized tomography showing a large emissary vein (black stars) in the area of retrosigmoid approach on the left side

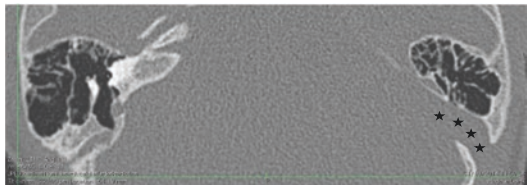


Fig. 9.2 A male patient with bilateral complete labyrinthine aplasia who had a large emissary vein on the left side (black stars)

age to this vessel may cause serious brain damage and therefore ABI was performed on the right side after removing the previously placed cochlear implant.

A similar male patient with bilateral complete labyrinthine aplasia was evaluated with similar condition. His CT revealed large emissary vein on the left side (Fig. 9.2) and ABI surgery was performed on the right side.

The aim is to choose the better side to provide hearing. Rarely previous shunt surgery in the surgical field may complicate the approach. It may be better to relocate the shunt before surgery.

Reference

1. Sennaroglu L, Ziyal I. Auditory brainstem implantation. Auris Nasus Larynx. 2012;39(5):439–50.

10 Genetic Evaluation in People with Sensorineural Hearing Loss

Emre Ocak, Guney Bademci, and Mustafa Tekin

10.1 Introduction

Hearing loss is the most common sensory disorder affecting approximately 300 million individuals worldwide, mostly diagnosed as sensorineural hearing loss (SNHL). SNHL can be either congenital or acquired and numerous etiologies can underlie this common disorder. SNHL is diagnosed in 1–3 of 1000 newborns [1]. Newborn hearing screening program has been globally accepted after the suggestion of American Speech-Language-Hearing Association (ASHA) in 1990. By means of this program, millions of children who have hearing loss at birth have been diagnosed and treated accurately in a timely manner. Conversely, there is also a considerable amount of adult population who have SNHL.

10.2 Principles of General Evaluation

Once a person is diagnosed with SNHL, a comprehensive evaluation is needed to identify its etiology. At this point, multiple disciplines including otorhinolaryngology, audiology, radiology, and genetics collaborate. Genetic factors are regarded as one of the leading causes of SNHL in developed countries, since other causes are generally prevented by vaccines or antibiotics. Therefore appropriate assessment of these people requires a genetic evaluation. In this manner, a 3-generation family history should be obtained addressing ancestral background, family members with congenital or later onset hearing loss as well as with relevant phenotypes affecting other systems and presence of parental consanguinity. It is a good practice to obtain audiograms from first degree relatives even hearing loss is not verbally reported [2].

10.3 Genetic Etiology in Sensorineural Hearing Loss

As mentioned previously, many factors can cause SNHL. More than 50% of infants diagnosed with congenital SNHL have genetic causes. Among this population, approximately 70% are related to genetic factors, which are not associated with clinical findings of a defined syndrome (non-

E. Ocak
Department of Otolaryngology, Faculty of Medicine, Ankara University, Ankara, Turkey

G. Bademci · M. Tekin (✉)
John P. Hussman Institute for Human Genomics, Miller School of Medicine, University of Miami, Miami, FL, USA
e-mail: g.bademci@med.miami.edu; mtekin@med.miami.edu

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_10

Table 10.1 Common syndromic causes of sensorineural hearing loss

Syndrome	Inheritance pattern	Clinical features	Genes
Alport	X-linked, AR, AD	Glomerulonephritis, lens abnormalities	*COL4A3, COL4A4, COL4A5*
Usher	AR	Retinitis pigmentosa	*ADGRV1, CDH23, CLRN1, HARS, MYO7A, PCDH15, SANS, USH1C, USH1E, USH2A, WHRN*
Jervell and Lange-Nielsen	AR	Long QT interval, cardiac arrhythmia	*KCNE1, KCNQ1*
Waardenburg	AD	Dystopia canthorum, heterochromia, pigmentary abnormalities of skin and hair	*PAX3, MITF, SNAI2, EDNRB, EDN, SOX10, KITLG*
Pendred	AR	Thyroid goiter, enlarged vestibular aqueduct	*SLC26A4*
Noonan	AD	Heart defects, short stature	*BRAF, KRAS, LZTR1, NRAS, PTPN11, RAF1, RIT1, SOS1, SOS2*
Branchio-Oto-renal	AD	Renal anomalies, middle/external ear anomalies, branchial fistulae/cysts	*EYA1, SIX1*
CHARGE	AD	Coloboma, heart defects, choanal atresia, retardation in growth and development, genital abnormalities	*CHD7*

AR autosomal recessive, *AD* autosomal dominant

syndromic congenital SNHL). The remaining 30% are associated with at least one additional of these syndromic causes (syndromic congenital SNHL) [3]. More than 400 forms of syndromic causes which are related to SNHL have been defined (www.omim.org).

Genetics of non-syndromic SNHL is heterogeneous with over 100 loci have already been identified. Among individuals with SNHL, autosomal recessive (AR) form is more frequent, accounting approximately 80% of cases (over 90% in countries with high rate of consanguineous marriage) and is typically congenital or prelingual-onset. The most common cause of non-syndromic SNHL in many populations is variants in the *GJB2* gene. Variants in this gene account for up to 50% of the individuals with non-syndromic AR SNHL in white populations of Europe and the USA. Mitochondrial DNA-related or X-linked forms of inheritance are rare compared to autosomal recessive and dominant forms [3].

Among many forms of syndromic causes for congenital or prelingual-onset SNHL, some are more common such as Pendred, Usher, Waardenburg, and Branchio-Oto-Renal syndromes. The responsible gene variants for several syndromes have been defined for which genetic testing is available. The clinical features and responsible genes for some common syndromic causes of SNHL are summarized in Table 10.1. Certain inner ear malformations are commonly associated with particular gene mutations; for instance, individuals who have *SLC26A4* and *POU3F4* mutations typically present with IP-II and IP-III cochlear malformations, respectively. Figure 10.1 shows three families with inner ear malformations due to mutations in the *SLC26A4*, *POU3F4*, and *EYA1* genes.

10.4 Comprehensive Genetic Testing

Mapping of the human genome and recent advances in DNA sequencing technology have created an extremely wide research area for numerous human disorders. Considering the extreme genetic heterogeneity of SNHL, comprehensive genetic testing via a set of genes (i.e., a gene panel) or whole exome/genome sequencing utilizing next-generation DNA sequencing makes a difference for the etiological evaluation of all individuals with SNHL [4]. It ideally should include all recognized genes for SNHL with both

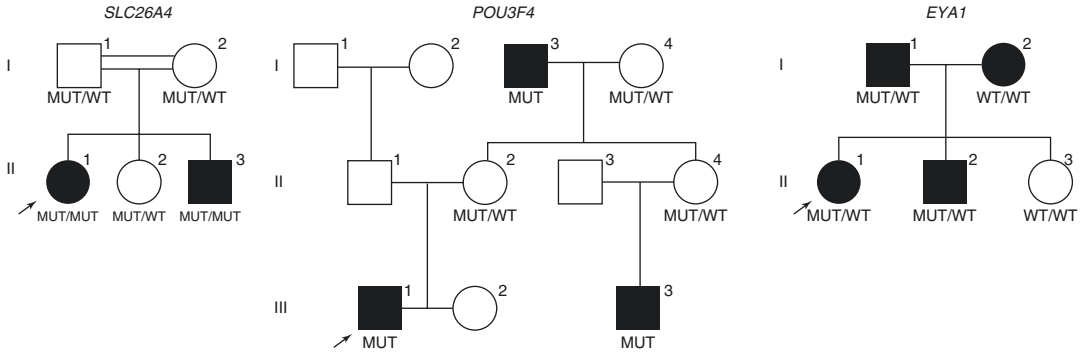


Fig. 10.1 Three families with mutations in *SLC26A4*, *POU3F4*, and *EYA1* genes. Left pedigree (autosomal recessive inheritance): two siblings with SNHL associated with IP-II cochlear anomaly are homozygous for a *SLC26A4* mutation. Unaffected sister and both parents are heterozygous for the same mutation. Double line between parents indicates parental consanguinity; Middle pedigree (X-linked inheritance): three males diagnosed with mixed hearing loss and IP-III cochlear anomaly are hemizygous for a *POU3F4* mutation. Individuals I:4, II:2, and II:4 (all females) are heterozygous for the same mutation without clinical presentation; Right pedigree (autosomal dominant inheritance): two siblings and their father have branchio-oto-renal syndrome and are heterozygous for the same *EYA1* mutation. Unaffected sister and the mother with non-syndromic hearing loss do not have the mutation. *Open circle* unaffected female, *open square* unaffected male, *closed circle or square* affected individual, *arrow* proband, *Mut* mutation, *WT* wild type

syndromic and non-syndromic forms and be performed in all cases when history and physical examination do not reveal a clear-cut environmental etiology. Comprehensive genetic testing may reveal the underlying genetic etiology even in late-onset hearing loss and aid in diagnosing syndromes even before additional symptoms appear [5].

The clinician should keep in mind that a genetic cause cannot be entirely excluded although clinical evaluation of a patient is performed with respect to the above-mentioned factors. Even when clinical findings are suggestive of a syndrome, there are often multiple genes to cause the clinically suspected syndrome. Therefore, in most cases, a next-generation sequencing gene panel or exome/genome sequencing offers a superior diagnostic yield compared to single gene testing [4].

There has been a dramatic decrease in the cost of the next-generation sequencing in the past years. It is practical, fast, accurate, and widely accepted by physicians.

Many of the recognized forms of syndromic hearing loss were reported in small number of families and sometimes in a single family. Syndromic phenotype might potentially involve any system, making a thorough review of systems and physical examination mandatory during genetic evaluation of hearing loss. Special emphasis should be placed on the clinical evaluation of the following systems:

- Visual anomalies: A full ophthalmological evaluation should be performed in every individual with SNHL as eye abnormalities involving all segments are seen in a number of syndromes. Usher and Stickler syndromes affecting the eye are among the most common forms of syndromic hearing loss with congenital/prelingual and postlingual SNHL, respectively.
- Endocrine anomalies, especially goiter and signs of hypothyroidism should be carefully evaluated, as Pendred syndrome is the most common form of syndromic hearing loss.
- Central and peripheral nervous system and general developmental history: Developmental delays and nervous system abnormalities are common in children with many syndromes as well as those with chromosomal abnormalities including copy number variants. Delay in gross motor development in an infant with severe/profound SNHL should prompt an investigation for Usher syndrome type 1 that is associated with vestibular impairment.

– Craniofacial morphology (e.g., palpebral fissures, auricules, nasal morphology, hair, sutures, etc.)
– Integumentary changes (e.g., as skin tags): especially hypo- and hyperpigmented macules should be carefully evaluated with a Wood's lamp. Waardenburg syndrome is one of the most common forms of syndromic hearing loss.
– Syncope attacks and epilepsy: individuals with Jervell and Lange-Nielsen syndrome might be misdiagnosed as having epilepsy.

Laboratory investigation

• With the recent advances in genomic sequencing technology, it is now feasible to screen mutations in most, if not all, known genes for SNHL. These tests yield underlying etiology in 20–60% of the cases depending on ethnicity, family structure, and clinical presentation. If a DNA variant known to cause only non-syndromic SNHL is identified through genetic testing, there is no need to perform costly and/or invasive clinical and laboratory investigations to rule out syndromic findings.
– Following tests should be considered based on clinical needs:

  Imaging studies (computerized tomography, magnetic resonance imaging) of the temporal bone for inner ear anomalies.

  Renal functions and morphology.

  Cardiac evaluation and EKG.

10.5 Conclusion

The etiological evaluation of individuals with SNHL requires a teamwork. As genetic factors play an important role in SNHL, appropriate and early assessment of these individuals is important to evaluate potential additional health problems, in some cases to predict the prognosis of hearing loss and to empower families to make informed decisions during planning of their families.

References

1. Korver AM, Smith RJ, Van Camp G, et al. Congenital hearing loss. Nat Rev Dis Primers. 2017;12(3):16094.
2. ACMG. Genetics evaluation guidelines for the etiologic diagnosis of congenital hearing loss. Genet Med. 2002;4(3):162–71.
3. Bademci G, Foster J II, Mahdieh N, et al. Comprehensive analysis via exome sequencing uncovers genetic etiology in autosomal recessive nonsyndromic deafness in a large multiethnic cohort. Genet Med. 2016;18(4):364–71.
4. Yan D, Tekin D, Bademci G, et al. Spectrum of DNA variants for non-syndromic deafness in a large cohort from multiple continents. Hum Genet. 2016;135(8):953–61.
5. Tekin D, Tutar E, Akay HO, et al. Comprehensive genetic testing can save lives in hereditary hearing loss. Clin Genet. 2015;87(2):190–1.

Treatment Alternatives in Inner Ear Malformations

11

Levent Sennaroglu

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.

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_11].

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_11

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

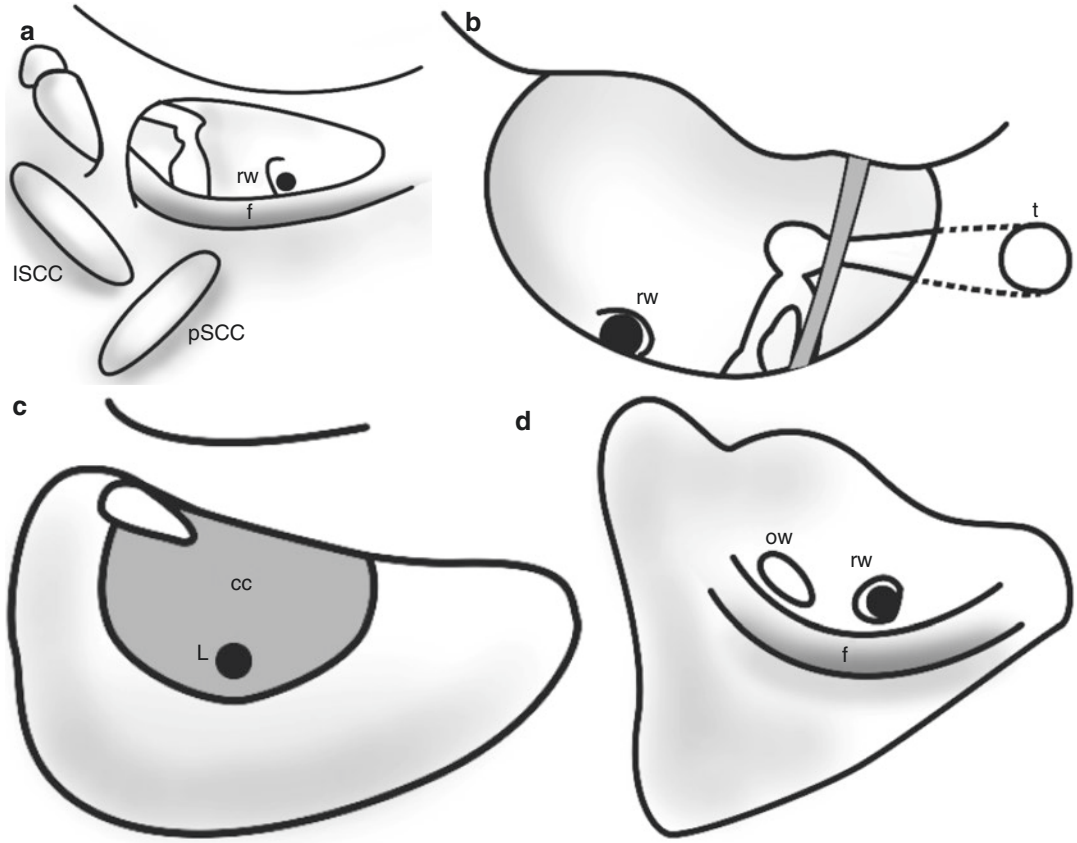


Fig. 11.1 Surgical treatment options: (**a**) Facial recess approach, (**b**) Transcanal approach, (**c**) Transmastoid labyrinthotomy approach, (**d**) Subtotal petrosectomy. *rw* Round Window; *f* Facial Nerve; *lSCC* Lateral Semicircular Canal; *pSCC* Posterior Semicircular canal; t tunnel for the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

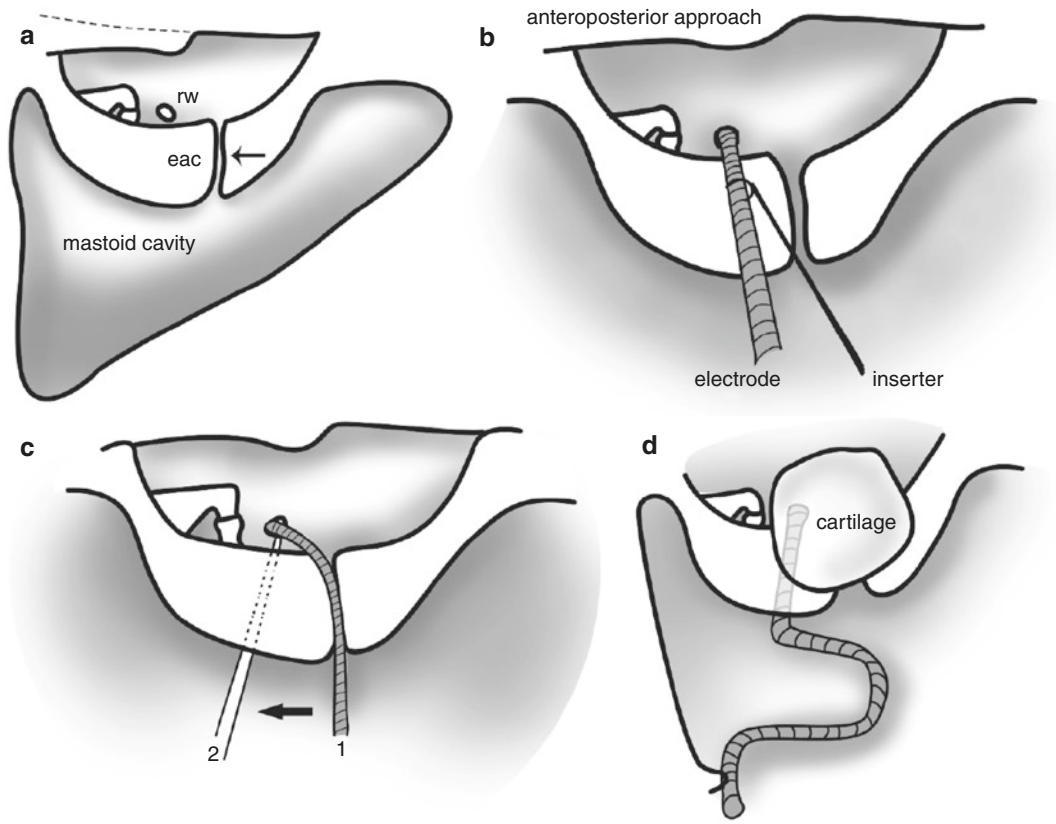


Fig. 11.2 Split ear canal technique. (**a**) A cut was produced in the bony ear canal with a tiny diamond bur. (**b**) Electrode insertion through the ear canal, (**c**) Transfer of the electrode through the opening into the mastoid cavity. (**d**) The cut was covered with a thin cartilage. (Modified 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

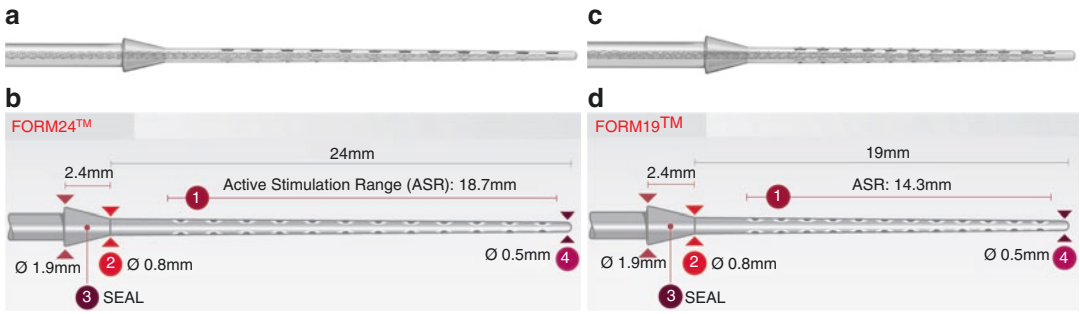


Fig. 11.3 FORM electrode series. (**a** and **b**) FORM 24 electrode. (**c** and **d**) FORM 19 electrode. (With permission from Med-El company)

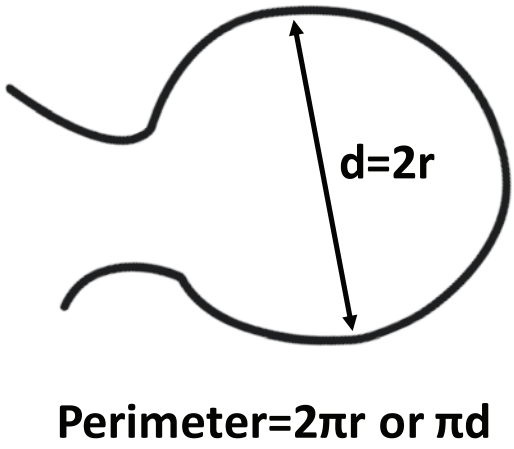


Fig. 11.4 Measuring the diameter of common cavity and using the formula perimeter = $2\pi r$ (or $d\pi$) gives the length of the electrode that makes one full turn in the cavity

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).

References

1. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
2. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
3. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
4. Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. Otol Neurotol. 2002;23(1):39–42; discussion 42-3.
5. House WF. The neurotology saga: a personal perspective, foreward. In: Jackler RK, Brackmann DE, editors. Neurotology. St. Louis: Mosby; 1994. p. xxi.
6. McElveen JT Jr, et al. Cochlear implantation in common cavity malformations using a transmastoid

labyrinthotomy approach. Laryngoscope. 1997;107(8):1032–6.
7. Beltrame MA, et al. Common cavity and custom-made electrodes: speech perception and audiological performance of children with common cavity implanted with a custom-made MED-EL electrode. Int J Pediatr Otorhinolaryngol. 2013;77(8):1237–43.
8. Kiratzidis T. 'Veria operation': cochlear implantation without a mastoidectomy and a posterior tympanotomy. A new surgical technique. Adv Otorhinolaryngol. 2000;57:127–30.
9. Kronenberg J, Migirov L, Dagan T. Suprameatal approach: new surgical approach for cochlear implantation. J Laryngol Otol. 2001;115(4):283–5.
10. Weber BP, et al. Pediatric cochlear implantation in cochlear malformations. Am J Otol. 1998;19(6):747–53.
11. Molter DW, Pate BR Jr, McElveen JT Jr. Cochlear implantation in the congenitally malformed ear. Otolaryngol Head Neck Surg. 1993;108(2):174–7.
12. Beltrame MA, et al. Double posterior labyrinthotomy technique: results in three med-El patients with common cavity. Otol Neurotol. 2005;26(2):177–82.
13. El-Kashlan HK, Arts HA, Telian SA. Cochlear implantation in chronic suppurative otitis media. Otol Neurotol. 2002;23(1):53–5.
14. Kim LS, et al. Cochlear implantation in children with inner ear malformations. Ann Otol Rhinol Laryngol. 2006;115(3):205–14.
15. Sennaroglu L, Atay G, Bajin MD. A new cochlear implant electrode with a "cork"-type stopper for inner ear malformations. Auris Nasus Larynx. 2014;41(4):331–6.
16. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25(4):520–9; discussion 529.
17. Slattery WH 3rd, Luxford WM. Cochlear implantation in the congenital malformed cochlea. Laryngoscope. 1995;105(11):1184–7.
18. Sennaroglu L, Ziyal I. Auditory brainstem implantation. Auris Nasus Larynx. 2012;39(5):439–50.
19. Sennaroglu LS, Sennaroglu G, Atay G. Auditory brainstem implantation in children. Curr Otorhinolaryngol Rep. 2013;1:80–91.
20. Bento RF, et al. Retrolabyrinthine approach for surgical placement of auditory brainstem implants in children. Acta Otolaryngol. 2012;132(5):462–6.

CSF Fistula and Meningitis

12

Levent Sennaroglu and Munir Demir Bajin

Special Features

1. Oval window is usually the site for spontaneous CSF fistula.
2. Cochleostomy can also be the source of CSF leakage.
3. Mostly seen in IP-I, to a lesser extent in CH-I and CH-II.
4. Stapes footplate defect is most probably due to endosteum developmental anomaly as a result of defective vascular supply from IAC.
5. Recurrent meningitis is common.
6. Vaccination against pneumococcus is very important but not sufficient.
7. Proper sealing of the leakage area with fascia in a dumbbell fashion is mandatory.
8. Subtotal petrosectomy may be necessary in some cases.
9. Stapes footplate must be examined for fistula during CI surgery in every IP-I and CH-II cases.
10. Surgeon should not leave operation theater without fully stopping the leakage.

Supplementary Information The online version of this chapter (https://doi.org/10.1007/978-3-030-83674-0_12) contains supplementary material, which is available to authorized users.

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

M. D. Bajin (✉)
Department of Otolaryngology, Hacettepe University Medical Faculty, Ankara, Turkey
e-mail: dbajin@hacettepe.edu.tr

12.1 Introduction

Today, inner ear malformations (IEM) are among the most important causes of recurrent meningitis with otogenic origin. In addition, cochlear implantation (CI) in this particular group of patients can also cause recurrent meningitis. Cerebrospinal fluid fistula (CSF) in IEMs is associated with considerable morbidity and mortality, usually presenting a diagnostic challenge to the otologist. It may easily lead to recurrent meningitis unless, there is a surgical intervention to repair the fistula. Majority of the CSF fistulae are located at the stapes footplate [1–6]. Rarely one may encounter CSF fistula at the cochleostomy site [7]. **As a surgical principle, surgeon should not leave operation theater without completely stopping the leakage.** Therefore, diagnosis and treatment of CSF leaks are very important to prevent meningitis in this special patient group.

There are two groups of patients: spontaneous CSF leakage in IEMs and CSF leakage after CI surgery. Trauma can also produce CSF leaks in this special group of patients easier than in subjects with normal anatomy. Both groups present with recurrent meningitis. It is more difficult to diagnose the pathology in spontaneous CSF leaks. High suspicion is the key to diagnosis. Therefore, **in cases of recurrent meningitis it is mandatory to evaluate inner ear radiologically to rule out an IEM**.

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_12

12.2 Histopathology and Pathophysiology

In 2016 Sennaroglu [8] reported the histopathology and possible pathophysiology in inner ear malformations. The findings provided understanding of the etiology and changed the concept of "CSF pressure causing footplate defect." Findings suggested that defective endosteum is most probably the cause of IP-I. All five patients with IP-I pathology in Massachusetts Eye and Ear Infirmary (MEEI) had a very thin and defective endosteum (innermost layer of the otic capsule) all around the cochleovestibular space, while the middle enchondral and outer periosteal layers were normal.

Three of the five cases with IP-I had a defect involving the stapes footplate. The defect was covered with a thin membrane. Embryologically, the vestibular part of the stapes footplate is derived from the endosteum, and a defective endosteum may be responsible for this defect.

If the embryology of the inner ear is investigated, it is noted that three layers of otic capsule have different vascular supply [9]. While inner endosteum (inner periosteal layer) has a vascular supply coming from the IAC, middle enchondral and outer periosteal layers have their vascular supply from the middle ear mucosa. According to Donaldson [9], blood vessels from the IAC supply the developing modiolus, the walls of the scala, the osseous spiral lamina, and the partition between cochlear turns. Therefore, it looks quite possible that histopathological changes in IP-I may be a result of defective vascular supply from the IAC blood vessels. As the vestibular surface of the stapes footplate is also derived from the endosteum, defective footplate development may be the result of abnormal endosteal development due to vascular arrest coming from the IAC. During development of the fetus, vascular channels are being formed circumferentially on the footplate, through the growth of the endosteal bone around blood vessels that are already present. Therefore, reduced vascular supply may cause a defective footplate.

In MEEI temporal bone collection, there were three IP-I specimens with a thin, but intact modiolar base, and a stapes defect at the same time (Fig. 12.1). Therefore, it is possible to have stapes footplate defect in cases with no CSF filling the cochlea. This is a very important finding demonstrating that high CSF pressure cannot be held responsible for the defective development at the stapes footplate. The author has operated on 11 cases with spontaneous CSF fistula and found that majority of the cases had a cystic structure present at the stapes footplate. Once it was punctured, CSF gushed out from this defect. This was repaired by introducing a piece of fascia through the defect into the vestibule, in a dumbbell fashion. Sometimes a bony defect with CSF leakage was also encountered. It is possible that in IP-I cochlea, where the modiolus is completely absent, high CSF pulsations acting on the thin membrane at the footplate may easily produce an oval window fistula. If there is a bony separation between the IAC and the cochlea, the footplate defect may not be noticed at all during the patient's lifetime. Stapes footplate fistula can only be noticed if there is a middle ear infection which may cause meningitis when the infection passes through the thin membrane at the oval window into CSF filling the cochlea.

None of the 11 spontaneous CSF fistulas due to IEMs operated on by the first author had a fis-

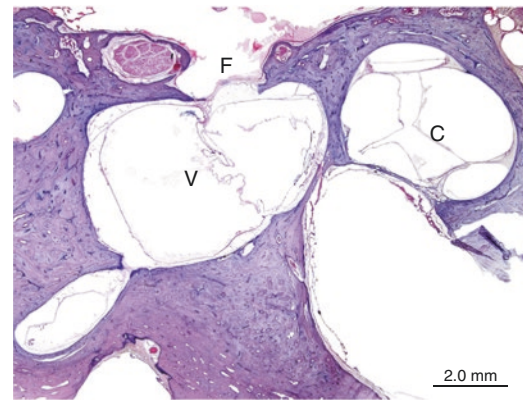


Fig. 12.1 Histopathological specimen showing stapes footplate where the normal bony footplate is replaced completely by thin fibrotic membrane (F) with intact modiolar base in a patient with IP-I specimen. As the cochlea is completely separated from internal auditory canal high cerebrospinal fluid pressure cannot be held responsible for the defective footplate (with permission of Massachusetts Eye and Ear Infirmary)

tula at the round window. All of the reported spontaneous CSF fistula cases in IP-I in the literature are located at the oval window [3, 5, 6, 10]. This also shows that this is observed in cases with a defective footplate.

In IP-III, which is observed in X-linked deafness, there is a larger defect in the fundus, a high pulsating CSF pressure in the cochlea. All 11 IP-III cases operated in our department had a severe gusher upon cochleostomy, but a spontaneous CSF fistula has never been encountered. The reason may be that the endosteum, which is deficient in IP-I (probably due to defective vascular supply coming from the IAC), is properly formed in IP-III. In IP-III, it looks possible that the endosteum is well developed. There is no histopathological specimen with IP-III but from HRCT it looks as if there is a thick endosteal layer in IP-III, while outer two layers of the otic capsule are missing. This causes stapes fixation without any fistula at the stapes footplate. Therefore, spontaneous CSF fistula formation has not been observed by our group or reported in the literature in IP-III. In other words, high CSF pressure cannot be held responsible for spontaneous footplate fistulas.

Another question is whether the footplate defect is present at birth or develops in time. The defect is most probably present at birth. High CSF pressure can cause a fistula through the defective footplate, or otitis media during childhood may result in recurrent meningitis. All cases that have been operated on in our department are children. No adult patient has been operated on so far: it appears that it is not a progressive disease, and that it must be present during childhood. If it had been the result of high pressure only, it would have been possible to see this clinical entity at all ages. This shows us that high pressure is not necessary all the time for the development of the oval window fistula. The defective development is most probably a result of a deficient periosteum present at birth, but high CSF pressure may produce a fistula in this already defective area. The prevalence of otitis media decreases considerably in adults. Therefore, adult patients with IP-I have very little chance to develop otitis media and meningitis.

All these features are valid for CH-I and CH-II as well. The only difference is the size of the cochlea; dimensions of the cochlea are less in CH-I and CH-II compared to IP-I. The occurrence of a gusher, a spontaneous CSF fistula at the footplate, and all other clinical findings can be observed in CH-II as well. The author has seen a spontaneous CSF fistula in two and a gusher during CI surgery in four CH-II cases and one spontaneous CSF fistula in CH-I.

12.3 Literature Review

In 1997 Hoffman et al. [11] reviewed the literature, along with the results of 200 institutions performing cochlear implants (which were queried by questionnaire) about IEMs. Out of 50 cases none had meningitis. In spite of this report, at present, IEMs are one of the most important causes of recurrent meningitis. This is the reason for devoting a chapter on this topic.

Phelps et al. [3] reported that meningitis occurred in 40% of the patients with severe cochlear dysplasia. They observed that patients with wider basal turn, possibly incomplete partition type I (IP-I), are more prone to risk of CSF fistula, whereas none of the patients with normal basal turn and enlarged vestibular aqueduct (possibly IP-II cases) had meningitis. Most probably their patients with "wider basal turn" are IP-I cases, where absence of interscalar septa and modiolus caused their basal turn look enlarged. This was an important observation indicating that stapes footplate defect was more often encountered in IP-I cases. However, the reason for recurrent meningitis is the absence of cribriform plate and modiolus resulting in a wide defect between the IAC and the cystic cochlea.

In 2010 Sennaroglu [1] reported in his review of IEMs that a wide defect in the cribriform plate and modiolus may cause the CSF to come adjacent to the medial surface of the oval and round window. Continuous CSF pressure may cause erosion, bony defect, and fistula at the stapes footplate. At that time CSF pressure was held responsible for bony erosion at the footplate. Recently, In 2016 Sennaroglu [8] reported the

histopathology and possible pathophysiology in IEMs and came to the conclusion that footplate defect is the result of endosteal developmental anomaly seen in IP-I and CH-II. If there is CSF filling cochlea, a middle ear infection causes meningitis when pathogens pass through the cystic or membranous footplate into inner ear.

In the literature, majority of the spontaneous CSF fistulas are reported to be located in the oval window. Phelps et al. [3] reported one patient with meningitis whose exploration of the middle ear revealed ballooning of the mucosa over stapes footplate and removal of the stapes produced a CSF gusher. Histopathological examination of another patient who had died of meningitis revealed a defect in the stapes footplate. Similarly Syal et al. [10] reported four cases where the site of leak was in and around oval window in all cases.

Shetty et al. [6] described two patients who had Mondini malformation in whom CT cisternography showed a CSF fistula at the lamina cribrosa (lateral wall of the internal auditory canal). They had CSF rhinorrhea in the presence of an intact tympanic membrane. Contrast material was seen in the middle ear cavity, having leaked through a defect in the lamina cribrosa and from there to an enlarged vestibule through the oval window. The cochlea was represented by a sac like diverticulum from the vestibule without apical turns. During surgery, they discovered a defect at the posterior aspect of the oval window, where the CSF was leaking.

It is possible to have multiple leak sides. Da Cruz et al. [5] reported a case with recurrent meningitis due to oval window fistula. After repairing the fistula with vein and fat graft, rhinorrhea continued and meningitis recurred. CT images in their report resembled a common cavity deformity. In their investigation, CT cisternography demonstrated two fistula sites: one at the oval window and the other in the tegmen. During revision surgery, two fistula were discovered: one fistula at the oval window and a second fistula at the tegmen. This necessitated a combined middle fossa and transmastoid approach. They made an important contribution that it is very important to look for another site of CSF fistula in recurrent cases.

CSF fistula may be seen in common cavity malformation as well. Mylanus et al. [4] reported recurrent meningitis in a patient with common cavity. Similar to other cases middle ear exploration revealed a fistula at the oval window which was sealed with temporalis fascia.

Although rarer than oval window, sometimes the leak site can be the cochleostomy. Page and Eby [7] reported a case of meningitis after minor head trauma developing 2 years after cochlear implantation in a child with Mondini malformation. This is the first case of meningitis after CI surgery in IEMs. The images in their report resemble IP-I malformation. CSF leakage was located at the cochleostomy around the electrode, and this was sealed with a temporalis fascia and muscle plug.

Contralateral side may also be the source of CSF leak and cause for meningitis in an implanted patient. Bluestone [12] stated that sometimes the implant may not be associated with the pathogenesis of post-implantation meningitis. Suzuki et al. [13] reported the temporal bone histopathological findings of a 6-year-old boy who had a CI surgery at another institution and was admitted with acute, fulminating pneumococcal meningitis, and died. His temporal bones were removed and the histopathology revealed bilateral Mondini malformations that had been implanted in his left ear. Interestingly, he had otitis media and labyrinthitis in the contralateral, right, non-operated ear that had spread to his meninges. The cochlear implant was not involved in the pathogenesis of the meningitis but the underlying inner ear anomaly and otitis media were. The left side with the cochlear implant was completely normal with no infection in the middle ear and no disruption in the footplate, annular ligament and the round window membrane. However, the non-operated right side had normal footplate and annular ligament but he had inflammatory necrosis of the round window membrane with many leucocytes in the scala tympani adjacent to the tympanic membrane. The suppurative labyrinthitis was attributable to the spread of middle ear infection through the round window membrane. The further spread of infection into intracranial CSF space was through the defective modiolus.

Findings in this patient demonstrated that round window may rarely be involved in transmission of infection as well.

12.4 Radiology

Radiology is the most important tool to diagnose preoperatively the CSF leak and the predisposing anatomic factors. HRCT is superior to MRI in demonstrating the bony defects or fractures. While MRI demonstrates the fluid characteristics better than HRCT. Shetty et al. [6] stressed the importance of CT in the evaluation of patients with CSF fistula. The presence of IEMs, a defect in the lamina cribrosa, or a bone fracture on HRCT may be the etiology of CSF gusher. CT cisternography can show CSF fistula in patients with CSF otorhinorrhea and unilateral hearing loss. A noninvasive method for confirming this finding is a fast spin-echo T2-weighted MR sequence through the region [14]. Retrospectively, Shetty et al. [6] concluded that plain high-resolution CT study of the temporal bones coupled with coronal MR cisternography of this region with the use of a fast spin-echo T2-weighted sequence would have noninvasively shown the site of the CSF fistula in their patients CSF fistula. Syal et al. [10] also found MRI (using 3D FSE T2WI and 3D FIESTA sequences) a useful technique in the assessment of patients with CSF fistulae; it is noninvasive, offers excellent anatomical detail, and has no radiation risk. Da Cruz et al. [5] also recommended a plain high-resolution CT coronal temporal bone study with MR cisternography to show the defect and the leak noninvasively, particularly in patients with bilateral CSF otorhinorrhea associated with unilateral hearing loss.

Radiology is the most important tool to diagnose CSF fistula. **All cases of recurrent meningitis, particularly with sensorineural hearing loss should have HRCT and MRI of the temporal bone to rule out a possible IEM**. Radiology may demonstrate:

1. Presence and type of IEM: In our department stapes footplate fistulas were observed in IP-I (Fig. 12.2a), CH-I (Fig. 12.2b, c), and CH-II (Fig. 12.2d).

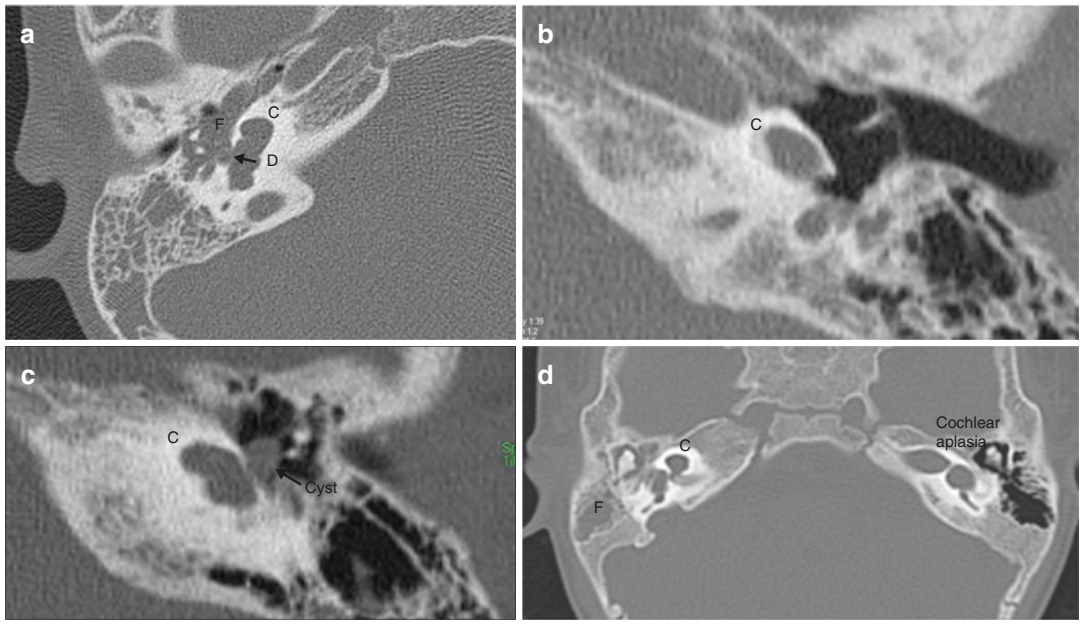


Fig. 12.2 Cases with stapes footplate fistula: (**a**) Incomplete partition type one cochlea (C), with defective footplate (D) and fluid (F) filling the middle ear, (**b** and **c**) cochlear hypoplasia type one (C) with cyst over the oval window, (**d**) cochlear hypoplasia type two (C) with fluid (F) filling middle ear and mastoid on the right side. Please note cochlear aplasia on the left side

2. Signs of oval window cyst: Opacity in the oval window area (Figs. 12.2c and 12.3).
3. Signs of CSF leak: Fluid filling middle ear and mastoid (Fig. 12.4a) and sometimes extending to nasopharynx via Eustachian tube (Fig. 12.4b). If this fluid has similar characteristic to the fluid filling the inner ear and IAC, it may be the sign of a CSF leak. This latter finding, if present, distinguishes OME from CSF leakage where both fluids demonstrate different characteristics on MRI.

To diagnose CSF leak in case of recurrent meningitis after CI surgery is more difficult. MRI cannot be performed. If there is no fluid in the middle ear and mastoid in the preoperative HRCT, and during operation, postoperative fluid signal on HRCT is an important sign indicating CSF leak (particularly in IP-I) (see **Case 3** below). One of our patients with oval window fistula presented like this. It may be necessary to have a CT cisternography in certain cases.

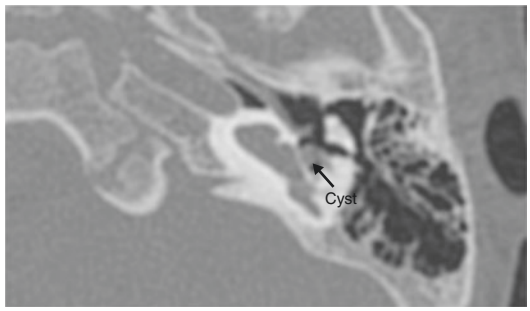


Fig. 12.3 Cystic opacity over the oval window area

12.5 Indications for Surgery

None of the patients who have been operated had a clear diagnosis of fistula before exploration. History of the patient is extremely important in the diagnosis and management of the patient with recurrent meningitis (see **Case 1** below). Therefore, suspicion of fistula in recurrent meningitis if there is one of the above pathologies is very important. If there is a suspicion of CSF leak it is advisable to go ahead with surgery rather than wait and see.

If one of the findings below is present there is possibility of a stapes footplate fistula necessitating middle ear exploration:

1. An opacity at the oval window area on HRCT in a patient with recurrent meningitis: oval window area has to be examined surgically.
2. Middle ear effusion in the above IEM types with recurrent meningitis.
3. Fluid in the middle ear which has similar characteristics on intensity with the inner ear fluids on MRI extending through Eustachian tube into nasopharynx.
4. Posttraumatic rhinorrhea or otorrhea in the above mentioned subtypes of IEM.

12.6 Treatment

SURGEON SHOULD ALWAYS KEEP IN MIND NOT TO LEAVE OPERATION THEATER WITHOUT FULLY CONTROLLING THE CSF LEAKAGE.

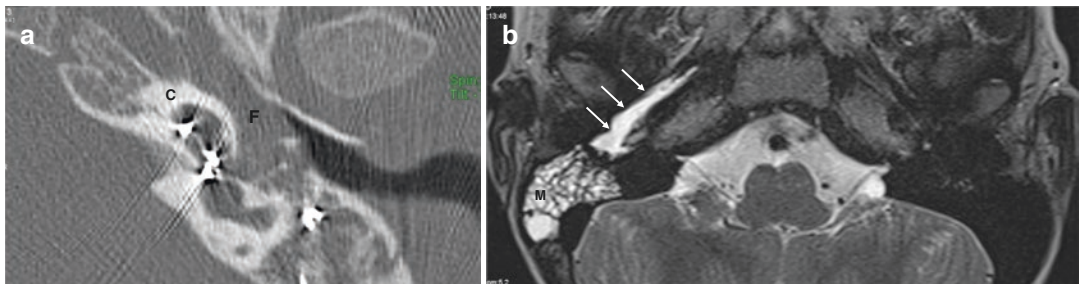


Fig. 12.4 (**a**) Incomplete partition type one cochlea (C) with implant and fluid (F) filling middle ear, (**b**) Fluid in the mastoid (M) and Eustachian tube (white arrows)

12.6.1 Vaccination

SNHL due to meningitis showed dramatic decrease after routine use of pneumococcal vaccination. However, IEMs still remain an important cause of recurrent meningitis. Although vaccination is more important in IEMs than normal population, appropriate management of the fistula site is priority. Vaccination only does not prevent meningitis in a case of IEM.

12.6.2 Middle Ear Exploration

Ear canal is narrower in children; exploration should be performed via endaural approach which allows enlargement of the ear canal. Atticotomy may be necessary to evaluate the ossicles. In general, more manipulation is necessary at the oval window than stapedotomy. Therefore, endaural approach provides larger exposure and is preferred over endomeatal approach in children. The latter approach has been used to manage footplate without damaging the electrode in an already implanted patient (see **Case 4** below).

There have been important changes in the management of these patients. In the past removal of the ossicles and obliteration of the vestibule was suggested in the management of these cases [1]. It has been observed that stapes is an important structure keeping the fascia in place against high pulsating CSF pressure. Now we believe that **every effort must be shown to keep the stapes in place**.

The fistula site is explored without removing any ossicles. Usually there is a cystic structure coming from the footplate (Fig. 12.5a). Without removing this cyst, it is impossible to evaluate the fistula. There is usually little serous fluid around. The fluid is removed with suction and facial nerve (FN) canal is evaluated. It is important to note any bony dehiscence at the tympanic segment at this stage because once the cyst is removed, there will be a gusher and during continuous CSF leakage it is not possible to evaluate the FN. During gusher it is possible to cause FN injury with surgical instruments if there is a bony dehiscence.

The cystic structure is then removed with cup forceps. As soon as it is removed there will be a CSF gusher from the footplate. It is important to avoid trauma to the facial nerve because clear fluid causes refraction and FN location may be misinterpreted. At this stage it is advisable to gently remove the fluid with suction and wait for 10–15 min until it decreases considerably and starts to pulsate.

At this stage the fistula site is inspected (Fig. 12.5b). Aim of surgery is to introduce a piece of fascia in a dumbbell fashion; one half of the fascia in the vestibule and the other half in the middle ear. If the defect is less than 0.7–0.8 mm, it is difficult to insert fascia through the opening and efficiently block the fistula. In that case it is advisable to use a 0.6 mm diamond burr to make a circular opening at the footplate (Fig. 12.5c). It is very important to keep the stapes intact at this stage. Then a fascia about 1 cm long and 2–3 mm wide is gently inserted into the defect (Fig. 12.5d). Ideally the fascia has to be placed in a dumbbell fashion in the defect (Fig. 12.5e).

There may be irregular defects or a defect at the anterior crus. It is advisable to enlarge the defect with a 0.6-mm diamond burr and apply the fascia as explained before.

The most difficult patients are the ones who will have a CI surgery at the same session where the stapes has a fistula. In two of them stapes dislocated during manipulation. Vestibule had to be obliterated. This carries the risk of filling up the space for electrode. It is advisable to insert the electrode and gently obliterate the vestibule (See **Case 5** below). However, the best treatment method is to keep the stapes in place and enlarge the fistula.

IF CI has to be applied as well, we start with transmastoid approach and open the facial recess. The facial recess is enlarged as much as possible because there will be prolonged surgical manipulation around the cochleostomy area. This approach is sufficient for electrode insertion. But it is insufficient for stapes footplate exploration. Therefore, additional transcanal exploration is necessary. The ear canal skin is elevated and then the footplate is managed as mentioned before.

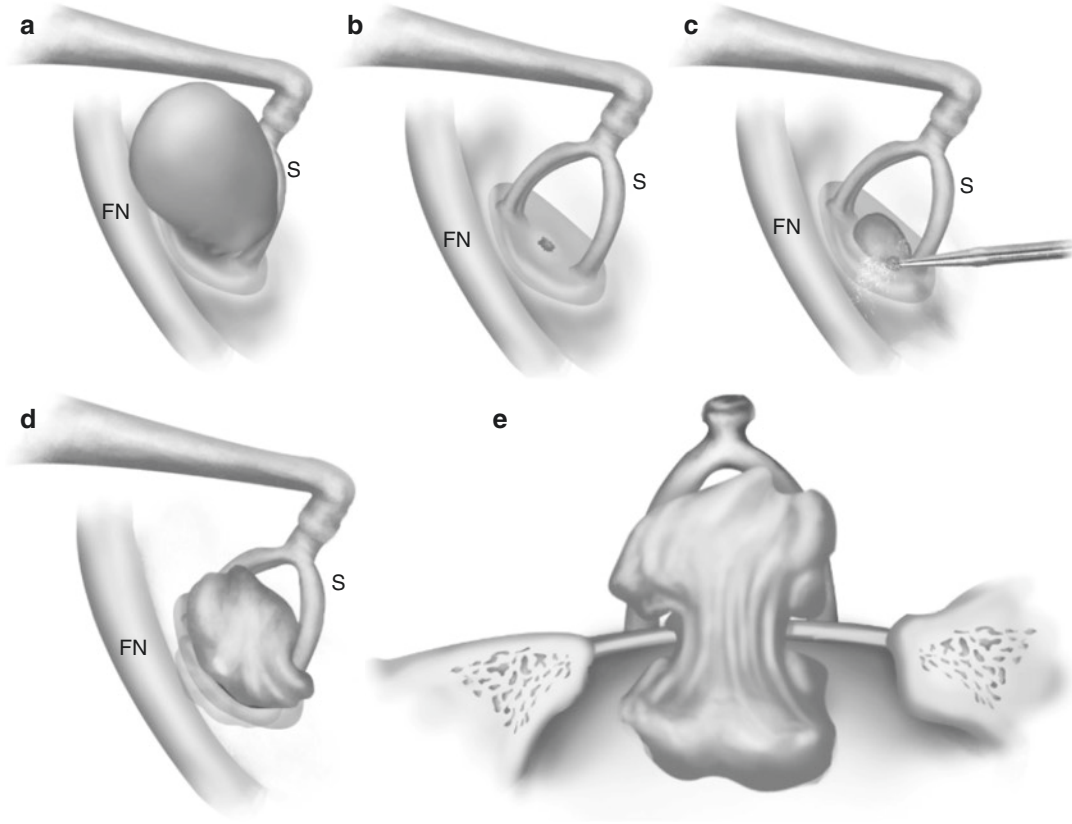


Fig. 12.5 Management of the stapes footplate fistula with a cyst: (**a**) A cystic structure coming from the footplate, (**b**) Footplate defect after cyst removal, (**c**) Enlargement of the fistula with diamond burr, (**d**) Placement of the fascia through the defect, while keeping the stapes in place to provide support to the fascia, (**e**) Fascia placed in dumbbell fashion through the defect

In situations where the stapes was removed during manipulation the vestibule is packed with fascia. It is advisable to block oval window by incus. Long process is removed by drilling and short process and body is inserted into the window with fascia between incus and the bony opening (Fig. 12.6a–d).

At times it is necessary to make prolonged manipulation at both windows. Keeping the fascia pieces in place against high pulsating CSF pressure is the most important part of the procedure. Therefore, postoperative CONTINUOUS LUMBAR DRAINAGE IS ALWAYS PART OF THE PROCEDURE.

In the literature, there are methods suggesting to remove the ossicles and to pack vestibule through the oval window with a layer of muscle or fascia followed by injection of fibrin glue [3, 4, 6, 10]. Another layer of muscle or fascia may be used on top. Syal et al. [10] placed intraoperative continuous lumbar drainage and lumbar drainage was continued postoperatively for 7 days in all four cases. Da Cruz [5] repaired the oval window fistula with vein and fat graft.

If a leakage is discovered at the cochleostomy site, this should be sealed as reported by Page and Eby [7]. Temporalis fascia was packed around the electrode array and into the vestibule until the leakage stopped. A piece of temporalis muscle was then placed in the middle ear space on top of the fascia. The wound was then closed, and a lumbar drain was inserted.

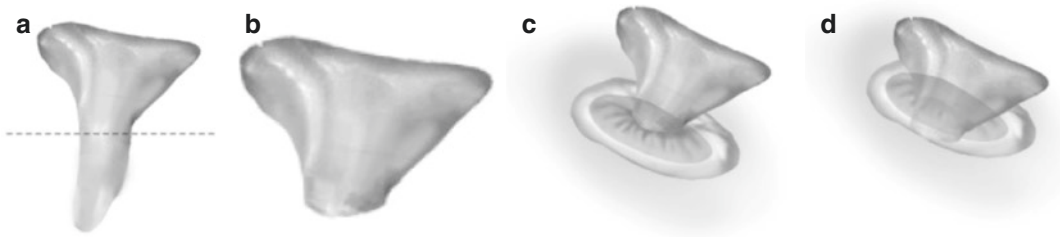


Fig. 12.6 (**a**–**d**) Stapes was removed during manipulation. Vestibule is obliterated with fascia and incus is placed into oval window defect after long process is removed

12.6.3 Subtotal Petrosectomy

Weber et al. [15] recommended subtotal petrosectomy with removal of the middle ear mucosa, closure of the Eustachian tube and ear canal in patients with recurrent meningitis.

Mylanus et al. [4] reported a patient with common cavity who had recurrent meningitis. Exploration of the middle ear revealed a fistula in the oval window which was sealed with temporal fascia. Upon recurrence of meningitis she had subtotal petrosectomy and a cochlear implantation through a labyrinthotomy, followed by a total obliteration with abdominal fat and closure of the external auditory canal. No CSF gusher was found and complete insertion was accomplished with an uncoiled, straight electrode array.

In their patient with multiple leakage sites Da Cruz et al. [5] used combined middle fossa and transmastoid approach. They repaired the oval window fistula with vein and fat graft. A soft tissue seal of temporalis muscle and fascia lata was inserted from above into the tegmen defect like a cork. Eustachian tube orifice was blocked and ear canal was closed as a blind sac.

We have performed this procedure in one patient who had recurrence of the stapes footplate fistula twice.

12.6.4 Continous Lumbar Drainage

This is a very important part of the management of the fistula. There is a strong pulsating CSF pressure acting on the reconstruction area. It tries to dislodge the fascia around the opening. In one case of footplate fistula, leakage was managed properly and no CLD was applied, there has been a recurrence of rhinorrhea 2 days after the surgery. Fistula site was re-explored and repaired once again and CLD was applied a week. No further leakage was noted.

Another patient with difficult surgery at the oval window and cochleostomy was managed properly in OR. At night CLD was removed accidentally and unfortunately rhinorrhea started the day after. He had 10 days of CLD and finally discharged without any leakage.

Our current management protocol is to employ CLD in all cases of gusher at the footplate or during cochleostomy even though the fistula is closed completely. As mentioned before, this diverts CSF to another area and lowers the CSF pressure acting on the fascia decreasing the chance of dislodging the fascia.

12.7 Clinical Experience

Eleven patients, who had spontaneous footplate fistula without any cochlear implant surgery, had the following subtypes of IEMs:

IP-I eight cases (Fig. 12.2a),
CH-I one case (Fig. 12.2b, c),
CH-II two cases (Fig. 12.2d).

Two of these patients had CI surgery in their contralateral ears and CI was not related to the fistula at all. Two of these cases received CI in the same setting during fistula closure (see **Case 5** below).

CSF leakage was seen after CI surgery in six cases: Two had been operated in our department: one had a leakage around electrode, other had stapes footplate fistula without any leakage around electrode (see **Case 3** below). Remaining four patients had been operated in other centers: two had CSF leakage around electrode, two had oval window fistula.

12.8 Site of Fistula

All 11 cases with spontaneous CSF fistula had their defect at the oval window. None of them had a spontaneous fistula at the round window. Literature findings support these findings. One case with accidental removal of CLD led to rhinorrhea. One case without CLD needed revision. One case with two recurrences had subtotal petrosectomy.

Six cases of recurrent meningitis were after CI surgery: three of these had fistula at the oval window and the remaining three had CSF leakage at the cochleostomy site. In cases with oval window fistula the footplate defect was repaired with fascia. In cases where the leak was from the cochleostomy site, area was closed with fascia. We had to change the modiolar hugging electrode in one patient with the FORM electrode.

In our department we had a patient who presented with recurrent meningitis due to spontaneous fistula on the contralateral side to CI surgery. She had bilateral IP-I defect and had been implanted 2 years prior to meningitis on the right side. She had two attacks of meningitis. Her CT demonstrated fluid on the left, contralateral middle ear, and mastoid. Exploration of the nonimplanted left ear revealed mucosal cyst coming from the stapes footplate. Perforation of the mucosa produced a CSF gusher which was controlled by packing fascia into the vestibule. She had two recurrences of CSF leak and the surgeon performed a subtotal petrosectomy in the end. This case demonstrates us that CI in IEMs may not always be the source of recurrent meningitis. This presents a diagnostic dilemma to the implant surgeon.

12.9 Cases

Case 1: EP 2y Old Male Patient, Operated July 2013

His sister was operated for ABI because of severe IEM. Her mother said she had a brother who had two attacks of meningitis. I asked her about his hearing and she said he was deaf on one side only. I told her that I want to examine him immediately. Next day he was brought from Istanbul to Ankara. His HRCT revealed an IP-I with fluid filling middle ear and mastoid on the right side. Left side had normal cochlea and well aerated middle ear and mastoid (Fig. 12.7). Next day he was operated via endaural approach and the footplate cyst was removed and defect was sealed with fascia. He had no further attack of rhinorrhea or meningitis (Video 12.1).

Case 2: NCB 3-Year-Old Female Patient Operated on 8.10.2013

She applied with recurrent meningitis. She had bilateral CH-II with fluid filling left middle ear and mastoid (Fig. 12.8). Left side was explored via endaural approach. There was a fistula at the stapes footplate with continuous CSF leakage. There was no cystic structure at the footplate.

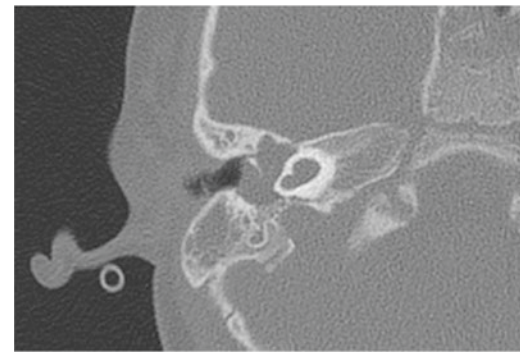


Fig. 12.7 Case 1. HRCT showing an IP-I with fluid filling middle ear and mastoid on the right side

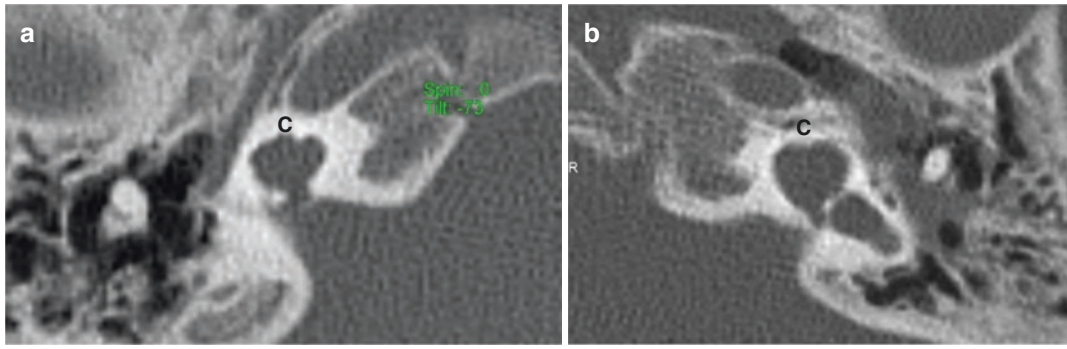


Fig. 12.8 Case 2. Bilateral CH-II with normal middle ear and mastoid on the right (**a**) and fluid filling middle ear and mastoid on the left side (**b**)

The ossicles were kept in place and the millimetric defect at the footplate was enlarged with a 1-mm diamond bur so that a piece of fascia can be passed through to stop the leakage. After the leakage was controlled tissue glue was used and a second layer of fascia was applied (Video 12.2).

Case 3: CK 6-Year Old Female Patient, Operated on 5.9.2005

In 2008 we were faced with recurrent meningitis in a child with bilateral IP-I who had received CI 4 years prior to meningitis attacks. She had bilateral IP-I deformity (Fig. 12.9a, b) with no fluid in the middle ear and mastoid. Right side was implanted in 2005. During cochleostomy there was gusher which was controlled completely and the electrode was fully inserted. Four years later she had meningitis. HRCT demonstrated normal electrode placement without any fluid on the right side (Fig. 12.9c). On the left side middle ear and mastoid were filled with fluid (Fig. 12.9d). Left side with fluid in the middle ear was explored via endaural approach. There was a cyst coming from the stapes fistula. The cyst was removed and this revealed a CSF leakage. Incus and stapes were kept in place and the defect was repaired with fascia. She had one more attack of meningitis and her surgeon performed subtotal petrosectomy on the left side.

Her HRCT enabled us to choose the correct side with CSF fistula. Her initial preoperative HRCT 4 years prior to meningitis attacks had normal ventilation in the middle ear. After meningitis CT demonstrated fluid in the middle ear and mastoid on the contralateral side to CI. Retrospective examination of the original CT demonstrates a soft tissue mass (cyst) on the left side (Fig. 12.9b).

Case 4: MP 1-Year-Old Female Patient, Operated on 14.6.2010

She had bilateral IP-I with defective fundus between cochlea and IAC (Fig. 12.10a). Right side was implanted in 2010. During surgery there was severe gusher. She was implanted with the initial prototype of FORM electrode. At the end of the surgery, there was no CSF leakage around the electrode. Six months later she applied with meningitis. HRCT demonstrated fluid filling middle ear and mastoid on the right side with cochlear implantation (Fig. 12.10b). Middle ear exploration was planned on the right side. Endomeatal approach was used. Electrode was functioning and the risk for damage to the electrode is avoided by using endomeatal approach. Middle ear was filled with CSF coming from a defect in the stapes footplate. No leakage was present at the cochleostomy area. Oval window fistula was repaired with a piece of fascia keeping the stapes in place (Video 12.3). She has not experienced any leakage or meningitis afterwards.

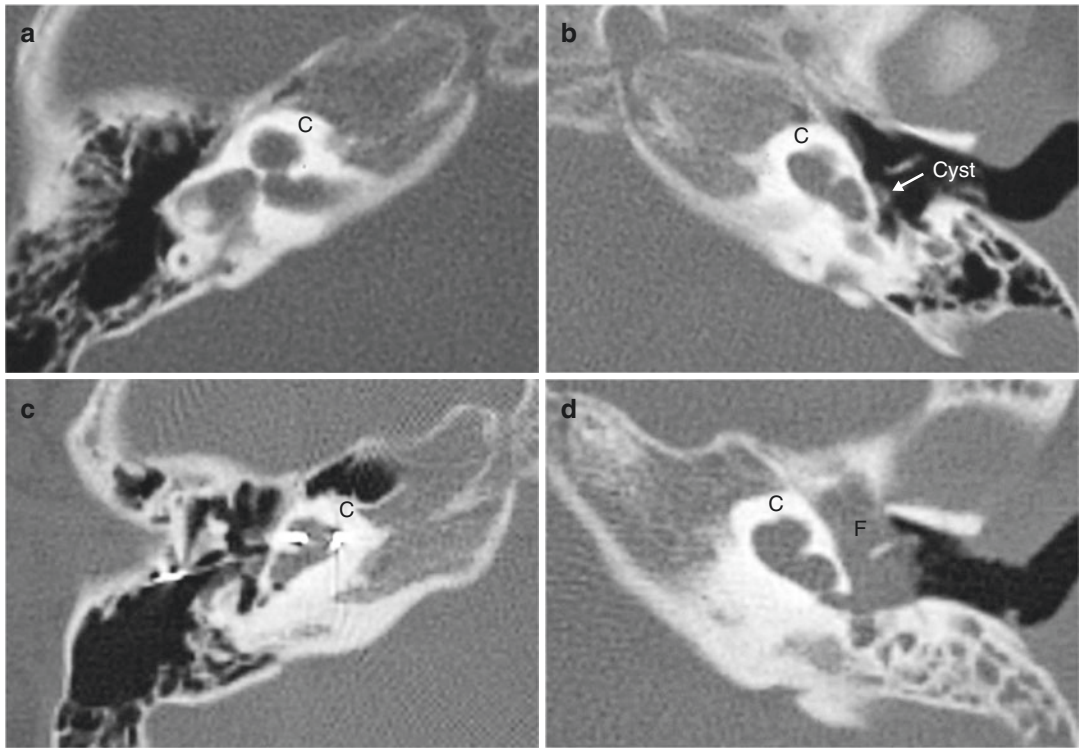


Fig. 12.9 Case 3 bilateral IP-I deformity (**a** = Right and **b** = Left) with no fluid in the middle ear and mastoid. Please note the soft tissue mass (cyst) on the left side which was noticed postoperatively. (**c**) HRCT showing normal electrode placement without any fluid on the right side, (**d**) Contralateral ear filled with fluid (*C* cochlea, *F* fluid)

Case 5: MD 1.5-Year-Old Male Patient Operated on 31.12.2012

He had posttraumatic rhinorrhea for 8 months prior to his visit. He had two times neurosurgical exploration of the anterior cranial fossa without any leakage point in another center. Main reason for application was deafness since birth. Unfortunately, his ears had not been evaluated before. Child was deaf on both sides since birth. HRCT demonstrated left side cochlear aplasia and right side CH-II with hypoplastic cochlear nerve. On the right side middle ear was filled with fluid.

CSF fistula reparation and cochlear implantation were planned at the same time. Postauricular approach was used. During mastoidectomy there was continuous CSF leakage coming from the antrum. Facial recess was opened. Leakage was coming from the stapes footplate. Initially footplate defect was explored via facial recess but it was impossible to evaluate and manage the fistula properly. Transmastoid approach was combined with transcanal approach where the footplate was controlled much better. Stapes had to be removed. Electrode was inserted via facial recess through cochleostomy. Defect was then closed with fascia and incus (Video 12.4).

He then received an ABI on the contralateral side with cochlear aplasia (Fig. 12.11).

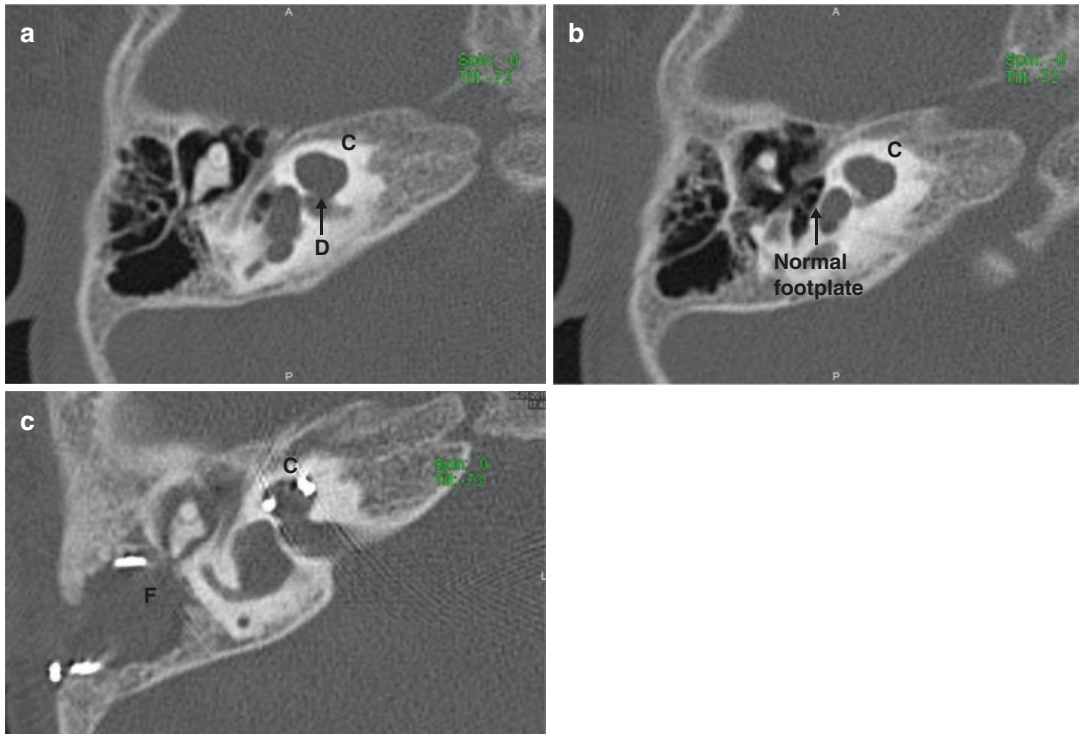


Fig. 12.10 Bilateral IP-I with defective fundus between cochlea and IAC. (**a**) Preoperative HRCT showing normal IP-I defect without footplate fistula or cyst, and normal ventilation, (**b**) Normal footplate, (**c**) HRCT showing normal electrode placement with fluid filling middle ear and mastoid (*C* cochlea, *F* Fluid)

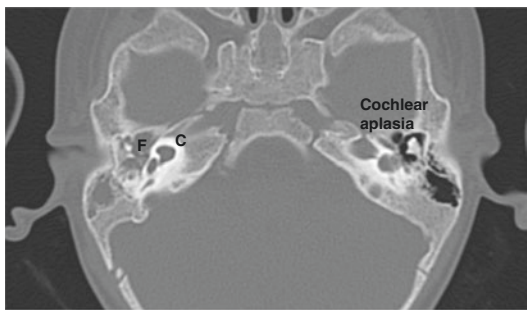


Fig. 12.11 HRCT showing left side cochlear aplasia and right side CH-II with hypoplastic cochlear nerve. On the right side, the middle ear was filled with fluid

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Luntz M, et al. Cochlear implants in children with congenital inner ear malformations. Arch Otolaryngol Head Neck Surg. 1997;123(9):974–7.
3. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
4. Mylanus EA, Rotteveel LJ, Leeuw RL. Congenital malformation of the inner ear and pediatric cochlear implantation. Otol Neurotol. 2004;25(3):308–17.
5. da Cruz MJ, Ahmed SM, Moffat DA. An alternative method for dealing with cerebrospinal fluid fistulae in inner ear deformities. Am J Otol. 1998;19(3):288–91.
6. Shetty PG, et al. Cerebrospinal fluid otorhinorrhea in patients with defects through the lamina cribrosa of the internal auditory canal. AJNR Am J Neuroradiol. 1997;18(3):478–81.
7. Page EL, Eby TL. Meningitis after cochlear implantation in Mondini malformation. Otolaryngol Head Neck Surg. 1997;116(1):104–6.
8. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
9. Donaldson JA, Rubel EW. Surgical anatomy of the temporal bone. 4th ed. New York: Raven Press; 1992.
10. Syal R, Tyagi I, Goyal A. Cerebrospinal fluid otorhinorrhea due to cochlear dysplasias. Int J Pediatr Otorhinolaryngol. 2005;69(7):983–8.
11. Hoffman RA, et al. Cochlear implantation in children with cochlear malformations. Am J Otol. 1997;18(2):184–7.

12. Bluestone CD. Cochlear malformations, meningitis, and cochlear implants: what have we learned? Otol Neurotol. 2003;24(2):349–50.
13. Suzuki C, et al. Histopathological features of a cochlear implant and otogenic meningitis in Mondini dysplasia. Arch Otolaryngol Head Neck Surg. 1998;124(4):462–6.
14. Eljamel MS, et al. MRI cisternography, and the localization of CSF fistulae. Br J Neurosurg. 1994;8(4):433–7.
15. Weber BP, et al. Pediatric cochlear implantation in cochlear malformations. Am J Otol. 1998;19(6):747–53.

Facial Nerve Abnormalities

13

Levent Sennaroglu and Emel Tahir

13.1 Introduction

Facial nerve (FN) is among the most important structures in the temporal bone. It is of particular importance to otologic surgeons because of its intricate route through the surgical field. FN provides a great challenge as its injury due to surgical trauma is the most devastating complication [1]. The intraoperative damage to FN can be caused by direct trauma or as a result of thermal energy from shaft of rotating burr [2].

FN can exhibit variations and anomalies in its usual route, which can be of significant surgical importance. The association between an anomalous FN route and congenital hearing loss is well known [3]. Tympanic segment of the FN located at the oval window (OW) can lead to congenital conductive hearing loss or can accompany OW and ossicular malformations; therefore, it is natural to encounter FN anomalies together with congenital middle ear anomalies.

Embryological development of the bony canal of the FN is closely related to the development of the structures of the inner ear. Since both FN and inner ear are closely related to the development of the otic capsule, FN abnormalities can accompany inner ear malformations (IEMs). Patients with IEMs are at higher risk of FN injury [4]. Hence a thorough knowledge of the intricate course of FN and its anatomic relation to the vital structures of the inner ear is essential to the surgeon who plans to operate in this area. CI surgeons should therefore always account for the possibility of an abnormal FN course to avoid iatrogenic FN damage [5].

13.2 Developmental Anatomy of the FN and Otic Capsule

It is necessary to briefly review the embryologic development of the otic capsule and FN to understand the relationship between them. The long, convoluted course of the FN through the temporal bone is thought to be dependent on the normal development of the bony structures derived from Reichert's cartilage. Thus, abnormal formation of related bony structures may lead FN to a different route or migration to its end organ (the muscles of facial expression).

The facial nerve canal develops from 2 separate structures: the primordial otic capsule and Reichert's cartilage from the second branchial arch. Reichert's cartilage provides cartilaginous sheath to the labyrinthine and tympanic segments of the FN [6]. At that time the otic capsule is entirely cartilaginous; Reichert's cartilage attaches to the otic capsule and provides the remaining cartilaginous sheath or the labyrin-

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

E. Tahir (✉)
Department of Otolaryngology, Ondokuz Mayıs University Medical Faculty, Samsun, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_13

thine and tympanic segments of the FN. Abnormal development of first and second branchial arch derivatives, including the bony wall of the facial nerve canal, the stapes, the styloid process, and/or external auditory canal, has been associated with an anomalous course of the tympanic and mastoid segments of the facial nerve, the nerve of the second branchial arch [7, 8]. Normally, the FN is located posterior to these structures which derive from Reichert's cartilage, the cartilage anlage of the second branchial arch [7]. The facial nerve canal develops from 2 separate structures: the primordial otic capsule and Reichert's cartilage from the second branchial arch. Reichert's cartilage provides cartilaginous sheath to the labyrinthine and tympanic segments of the FN [6].

Embryologically, FN develops from the facioacoustic primordium around third gestational week and lies at the anterior part of the otic vesicle. The vestibulocochlear nerve and facial nerve are seen as separate structures by the time the embryo is 14 mm in length. FN is well developed by the fifth week of gestation. At approximately the same period, the cochlear duct begins to develop and during its development it follows a convoluted course toward the facioacoustic primordium [9, 10]. The association of the geniculate to the vestibuloacoustic ganglion and the coiling of the cochlear duct explain the normal location of the geniculate ganglion, namely in the bony substance just above the basilar turn of the cochlea and between the superior semicircular canal and the cochlea proper [8, 11]. A developmental delay in the growth of the cochlea might permit the FN to dislocate. A normal sized and developed cochlea may behave in a manner similar to that of Reichert's cartilage derivatives, which prevent migration of the FN to an abnormal location. The FN canal has a periosteal layer forming 2 laminae, above and below the FN, both of which grow toward each other and enclose the nerve in a bony channel. Since facial canal encases the nerve, facial nerve promotes the development of facial canal. Depending on the developmental status of the temporal bone, distortion of FN may affect different segments of FN canal [12].

A sulcus develops around the FN at eighth week which ossifies at 26th week with partial closure of sulcus to form the facial canal [12]. At 10 weeks of gestation the facial nerve canal is a deep sulcus in the canalicular portion of the primordial otic capsule. At that time the otic capsule is entirely cartilaginous, Reichert's cartilage attaches to the otic capsule and provides the remaining cartilaginous sheath or the labyrinthine and tympanic segments of the FN. Therefore, it seems that the development of the facial canal is related to the facial nerve.

Instances of an anomalously coursing or dehiscent FN are rare. Henner was the first to describe such anomalies in 1960 [13]. Numerous theories have been developed to explain anomalous FN routes in the middle ear. One is that union between the otic capsule and the second branchial arch fails or occurs too late to prevent the FN from moving anteriorly. As normal development of the inner ear can be affected by a number of factors such as genetic mutations and teratogenic factors, FN and other structures in the ear can be a part of this pathological entity [1].

13.3 Imaging of the FN

Computed tomography (CT) has traditionally been the preferred modality for initial imaging workup of pediatric candidates for cochlear implantation, thus it is the first line study to detect any type of IEM. Temporal bone CT can detect deviations in the course and caliber of the intratemporal facial nerve, which can provide key information regarding IEMs.

CT of the petrous temporal bones enables detailed evaluation of the osseous anatomy of the inner and middle ear, as well as assessment of mastoid pneumatization and the degree of middle-ear aeration [14, 15]. Temporal bone CT is particularly useful in the evaluation of the caliber and the course of the IAC and bony FN canal in the temporal bone. In addition, CT has the advantage of demonstrating the relationship of the facial nerve canal to normal anatomic landmarks such as the ossicles which are not seen on MR [16].

However, there are some disadvantages to the use of CT. It exposes patients to ionizing radiation, which is of particular concern in the vulnerable pediatric population. Cone beam CT has less radiation compared to conventional CT and should be preferred if it is available. In addition, CT does not directly depict the cochlear nerve, only the bony channel that contains it [17].

Preoperative imaging workup for pediatric cochlear implantation varies among institutions, but the utility of MRI in patients with congenital sensorineural hearing loss is becoming increasingly appreciated. Some centers use MR imaging as the primary, and often only, modality to evaluate candidates for cochlear implantation, some others routinely perform both MR imaging and CT, and some still primarily rely on CT. However, it is important to recognize that CT does not demonstrate the presence of a normal cochlear nerve [15]. MRI of the internal auditory canal and inner ear enables direct visualization of the cochleovestibular nerve (CVN) and carries no radiation risk.

MRI also provides relatively detailed information about the fluid filled labyrinth and may demonstrate substantial inner ear malformations although its resolution is inferior to that of CT [17–19]. A major disadvantage of the use of MR imaging before cochlear implantation is its poor resolution in aerated or bony structures, it may not reliably identify bone landmarks and the degree of mastoid or middle-ear pneumatization. Course of the FN is not demonstrated in MRI. From the perspective of this chapter, although MRI is better to visualize neural structures, CT should be preferred in order to describe the IEM and FN canal relationship.

13.4 Facial Nerve Abnormalities

13.4.1 Background

Many abnormalities of the facial nerve canal in the temporal bone have been documented previously. The most common abnormality is accepted as congenital bony dehiscence of the FN canal that occurs in up to 55% of otherwise normal temporal bones, predominantly involving the tympanic portion (91%) [20]. With this high prevalence, it is more accurately described as a variation in normal anatomy [21].

Anomalies of the FN canal are usually seen in association with middle and IEMs [22, 23]. Anteromedial displacement of the labyrinthine segment of FN has been described in association with cochlear malformations, and an anteriorly displaced FN mastoid segment is often noted with congenital aural atresia [8]. An anomalous course of the tympanic segment has also been reported in association with oval window atresia [3, 24]. Bifurcation and trifurcation anomalies of FN are other entities described in the otolaryngology literature [4].

Few articles have been published regarding FN aberrations during CI. Raine et al. reported four cases of an anteriorly located vertical segment and one case of vertical segment bifurcation [25]. Fowler et al. found that anteromedial displacement of the vertical segment was the most common aberrant finding, followed by bifurcation of the vertical or horizontal segment. In their limited cohort composed of seven patients they demonstrated that cochlear hypoplasia was the most common IEM in aberrant FN cases. They also mentioned vestibular hypoplasia and FN abnormality relationship [26]. The relationship between the type of IEM and the FN abnormality is not well understood, making it difficult to estimate the risk of FN abnormalities before surgery. However, previous researchers have argued that some cochlear malformations definitely cause FN anomalies [27]. Hoffman et al. reported that IEMs occur in conjunction with anomalous FN anatomy in approximately 16% of cases [28]. Furthermore, in 2011, the author of this chapter indicated that the course of the FN may be altered in certain malformations such as IP-I, common cavity, cochlear hypoplasia, and semicircular canal malformation. Sennaroglu L. also stated that the vertical segment of the FN may be dislocated anteromedially toward the promontory and lies over the oval and round windows in patients with IEMs particularly cochlear hypoplasia cases. In addition, it was found that cochlear aplasia itself may cause anterior dislocation of

the labyrinthine segment of the FN [29]. In 2016, Jin published a paper about the classification of the labyrinthine segment of the FN and identified five types: anteromedial displacement at the beginning site, increase of the angle of the first genu, increase of length of the labyrinthine segment, and bifurcation [30].

As the relationship between the type of IEM and type of FN abnormality is not well defined in the literature, the authors of this chapter recently submitted a paper involving a detailed classification to clarify this association.

13.4.2 A New Classification of FN Abnormalities

13.4.2.1 Classification of Inner Ear Malformations

In order to understand the FN abnormalities, it is important to know the classification of IEMs. According to the most recent classification of IEMs proposed by the author of this chapter, there are eight groups: [31, 32]:

1. Complete labyrinthine aplasia (Michel deformity).
2. Rudimentary otocyst.
3. Cochlear aplasia.
4. Common cavity.
5. Cochlear hypoplasia: 4 subtypes.
6. Incomplete partition anomalies: 3 subtypes.
7. Enlarged vestibular aqueduct.
8. Cochlear aperture abnormalities.

Please refer to Chap. 1: Classification of IEMs for more details. In addition to cochlear morphology, vestibular structures also play a role in FN anomalies. Semicircular canal morphology can be evaluated in 4 groups as: normal, hypoplastic, aplastic, or dilated.

13.4.2.2 A New Classification of FN Abnormalities

FN canal abnormalities of different segments can be visualized on CT scans. FN has 4 different segments: meatal, labyrinthine, tympanic, and mastoid segments. Therefore, meatal, labyrinthine, tympanic, and mastoid segments should be investigated separately. This classification was introduced and published by the authors of this chapter in the year of 2020 [32]. This new classification is summarized in Table 13.1.

Meatal Segment

The meatal segment of the FN traverses in the IAC. Since IAC is a bony structure it can be evaluated and measured in CT. Normal development of the IAC is dependent on a normally developed CVN. Stenotic IAC may be the cause of CN deficiency. Meatal segment of FN is included in the classification of FN abnormalities because sometimes facial canal is the only canal instead of IAC or there may be a separate canal for FN which is completely separate than IAC. The classification of meatal segment is especially important in CI surgery for the evaluation of the cochlear nerve. The width of the IAC can be measured at its midportion on axial CT scans. Based on the current literature, IAC is considered stenotic when its midportion diameter is less than 2 mm [33]. This measurement could not be performed in cases such as complete labyrinthine aplasia (Michel deformity) and rudimentary otocyst due to the absence of the IAC. Types of meatal segment anomalies are classified as:

Type 1—Normal meatal segment (normal internal auditory canal): Facial nerve occupies the anterosuperior part of the IAC without a separate bony canal. IAC is accepted as normal when its midpoint diameter is ≥2 mm (Fig. 13.1a).

Type 2—Narrow meatal segment (narrow internal auditory canal): The midpoint diameter of the IAC is <2 mm. This might be accompanied by a hypoplastic or aplastic CN on MRI (Fig. 13.1b).

Type 3—Only facial canal: IAC consists of facial canal only, FN has its own canal in the absence of the IAC. The authors noticed that only facial canal is usually seen in severe IEMs such as Michel deformity and rudimentary otocyst (Fig. 13.1c).

Type 4—Separate facial canal/duplicated IAC: FN is separated from the CN by a bony canal. Canal for the meatal segment is separate from the IAC (Fig. 13.1d). This canal separates FN and cochlear nerve from each other. This type of anomaly was described in some case reports in the literature as "duplicated internal auditory canal" [34].

Table 13.1 The outline of the classification of facial nerve abnormalities

Meatal segment	Labyrinthine segment	Tympanic segment	Mastoid segment
Type 1: Normal IAC	*Type 1:* Normal	*Type 1:* Normal	*Type 1:* Normal facial recess
Type 2: Narrow IAC	*Type 2a/b/c:* Mild/moderate/severe anterior displacement	*Type 2:* Superior displacement	*Type 2:* Narrow facial recess
Type 3: Only FC	*Type 3:* Superior displacement	*Type 3:* At OW	*Type 3:* Unclassified
Type 4: Seperate FC/ duplicated IAC	*Type 4:* Straight labyrintine segment	*Type 4:* Inferior to OW	

FC facial canal, *IAC* internal auditory canal, *OW* oval window

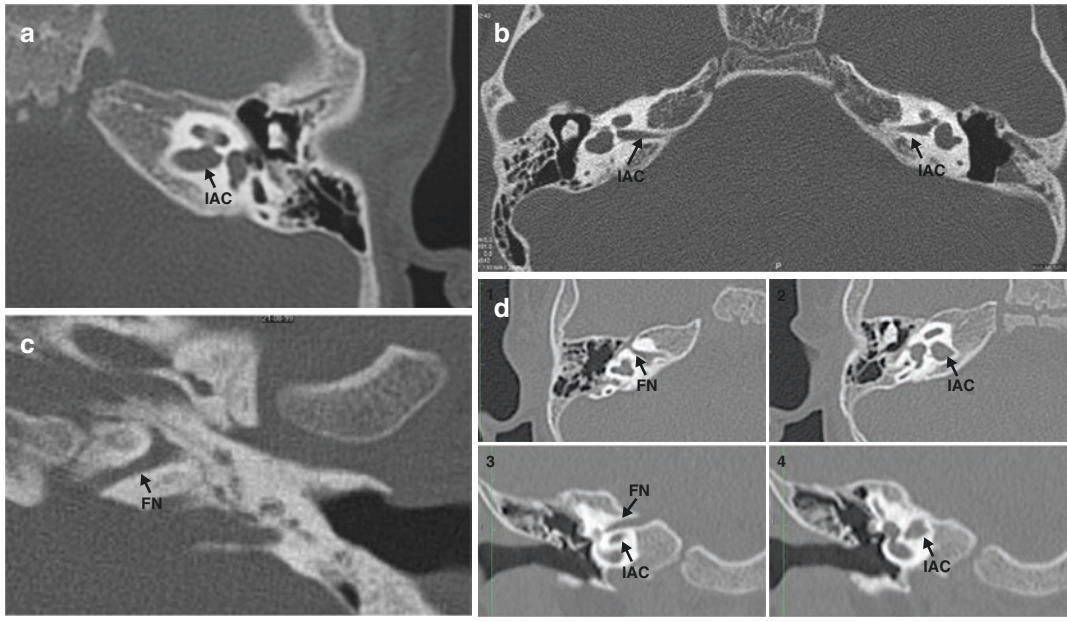


Fig. 13.1 (**a**) Normal meatal segment (normal internal auditory canal): Internal auditory canal (IAC) is normal when its midpoint diameter is ≥2 mm S. (**b**) Narrow meatal segment (Narrow IAC): Bilateral narrow IAC where the midpoint diameter is <2 mm (cochlear nerve may be hypoplastic or aplastic on MRI). (**c**) Only facial canal: IAC consists of facial canal (FN) only; FN has its own canal in the absence of the IAC. This is a Michel deformity. (**d**) Separate facial canal/duplicated IAC: FN is separated from the CN by a bony canal inside the IAC. This separate canal can be seen in axial (1) and coronal sections (3) and is located in the anterosuperior part of IAC. This was also reported as "duplicated internal auditory canal"

Labyrinthine Segment

Labyrinthine segment is situated between the anterior-superior part of the fundus of internal auditory canal and geniculate ganglion.

Abnormalities of the labyrinthine segment are classified as:

Type 1—Normal labyrinthine segment: Labyrinthine segment makes a gentle curve between the geniculate ganglion and the IAC around the basal turn. This segment is observed at the same section as basal turn of the cochlea (Fig. 13.2a).

Type 2—Displacement of the labyrinthine segment to the anterior part of the cochlea: This anterior displacement can be subdivided into 3 groups according to the degree of anterior course.

Type 2a—Mild anterior displacement: Cochlea is present and labyrinthine segment is located at the anterosuperior part of the cochlea instead of its usual posterolateral course (Fig. 13.2b).

Type 2b—Moderate anterior displacement: Labyrinthine segment is anteriorly dislocated, occupying the usual location of the cochlea, which is usually absent or hypoplastic (Fig. 13.2c).

Type 2c—Severe anterior displacement: Labyrinthine segment is further anteriorly displaced beyond the usual location of the cochlea (Fig. 13.2d).

Type 3—Superior displacement: In axial sections, labyrinthine segment appears superior to the basal turn. In other words, labyrinthine segment is situated above the cochlea (Fig. 13.2e).

Type 4—Straight labyrinthine segment: Instead of its usual curved course around the basal turn of the cochlea, labyrinthine segment courses from IAC to geniculate ganglion in a non-curved, straightway so that facial canal looks like a continuation of IAC (Fig. 13.2f).

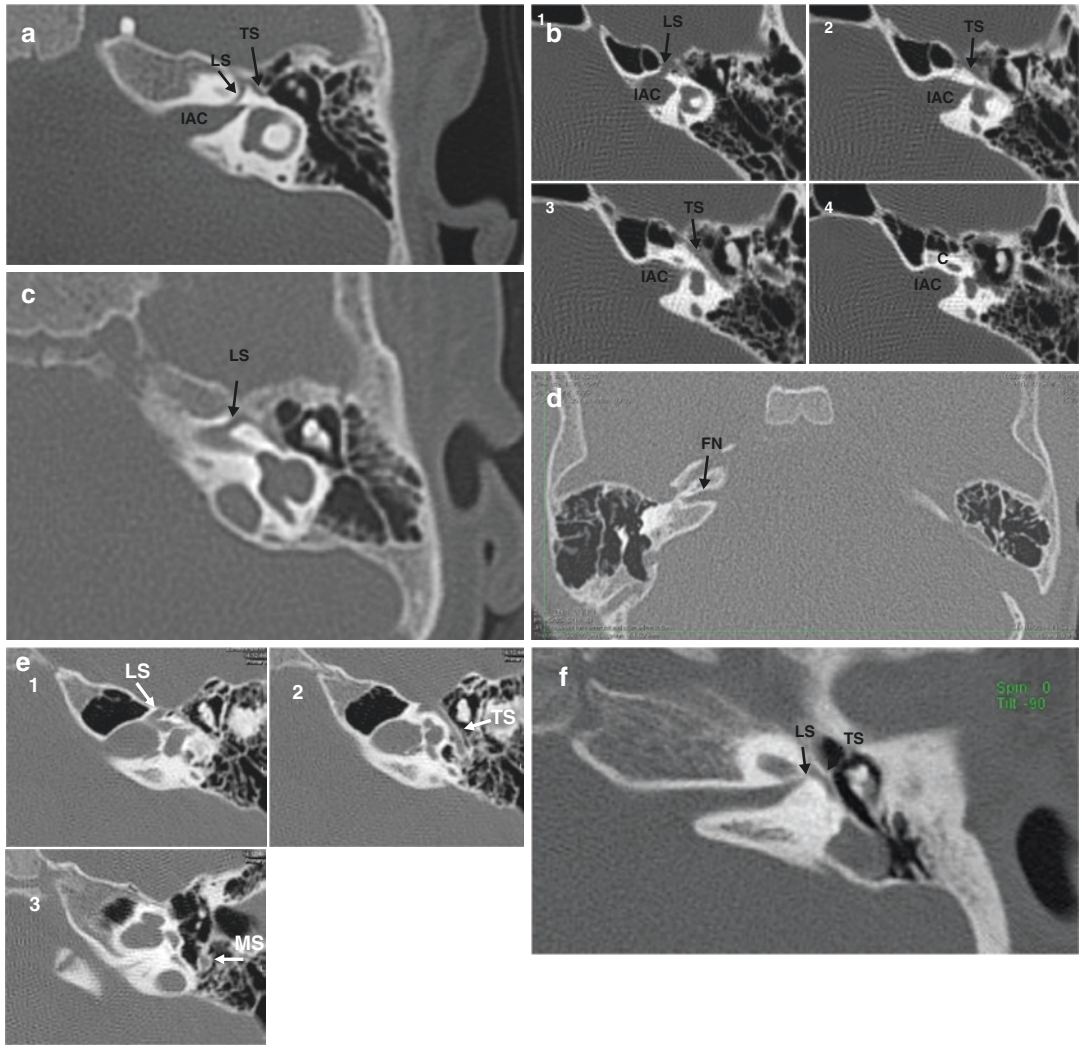


Fig. 13.2 (**a**) Normal labyrinthine segment (LS): LS makes a gentle curve between the geniculate ganglion and the IAC around the basal turn. This segment is observed at the same section as basal turn of the cochlea. Please note the tympanic segment (TS) of the facial nerve. (**b**) *Mild anterior displacement*: Cochlea is present and labyrinthine segment (LS) is located at the anterosuperior part of the cochlea instead of its usual posterolateral course. In the successive images from superior to inferior, first LS is visualized (1), then tympanic segment (TS) (2 and 3), and finally cochlea (4). Normally LS is visualized at the same section as the basal turn. (**c**) *Moderate anterior displacement*: A case of cochlear aplasia with dilated vestibule. Labyrinthine segment (LS) is anteriorly dislocated to the usual location of the cochlea, which is usually absent or hypoplastic. (**d**) *Severe anterior displacement*: A case of complete labyrinthine aplasia where labyrinthine segment (LS) is further anteriorly displaced beyond the usual location of the cochlea. (**e**) *Superior displacement*: In axial sections, from superior and inferior, labyrinthine segment (LS) appears superior to the basal turn (1), then tympanic segment (TS) is observed (2). In IP-III; cochlea is observed later (3) at the level of the section demonstrating the mastoid segment (MS) showing that labyrinthine segment is situated above the cochlea. (**f**) *Straight labyrinthine segment*: Instead of its usual curved course around the basal turn of the cochlea, labyrinthine segment (LS) courses from IAC to geniculate ganglion in a non-curved, straight line so that facial canal looks like a continuation of IAC. It then makes a sharp turn at the first genu forming the tympanic segment (TS)

Tympanic Segment

Tympanic segment was investigated on coronal sections and was evaluated according to its relationship with the oval window and lateral SCC as:

Type 1—Normal course of the tympanic segment: It is located superior and lateral to the oval window and stapes and inferior to lateral SCC (Fig. 13.3a).

Type 2—Superiorly displaced tympanic segment: Tympanic segment is located above the oval window, generally accompanying hypoplastic or aplastic lateral SCC (Fig. 13.3b).

Type 3—Tympanic segment at the oval window: Tympanic segment is located at the oval window at coronal sections (Fig. 13.3c).

Type 4—The tympanic segment inferior to oval window: Tympanic segment is located below the oval window, generally accompanying cochlear hypoplasia and abnormal SSCs (Fig. 13.3d).

Type 5—Unclassified: Because of the aplasia of the oval window, facial nerve cannot be classified in relation to oval window (in some situations such as Michel deformity and rudimentary otocyst).

Mastoid Segment

The anatomical borders of the facial recess (FR) are fossa incudis, chorda tympani, and facial nerve. FR is considered as narrow when the distance between the chorda tympani and vertical segment of the facial nerve is less than 2 mm [35]. Since the visualization of the chorda tympani is not always possible in CT sections, the distance between the external ear canal and FN can be used to determine FR in some cases. The width of the FR is especially important in cochlear implantation. Mastoid aeration is the principle factor that determines the distance between the external ear canal and FN, thus the width of the FR is not solely dependent on the

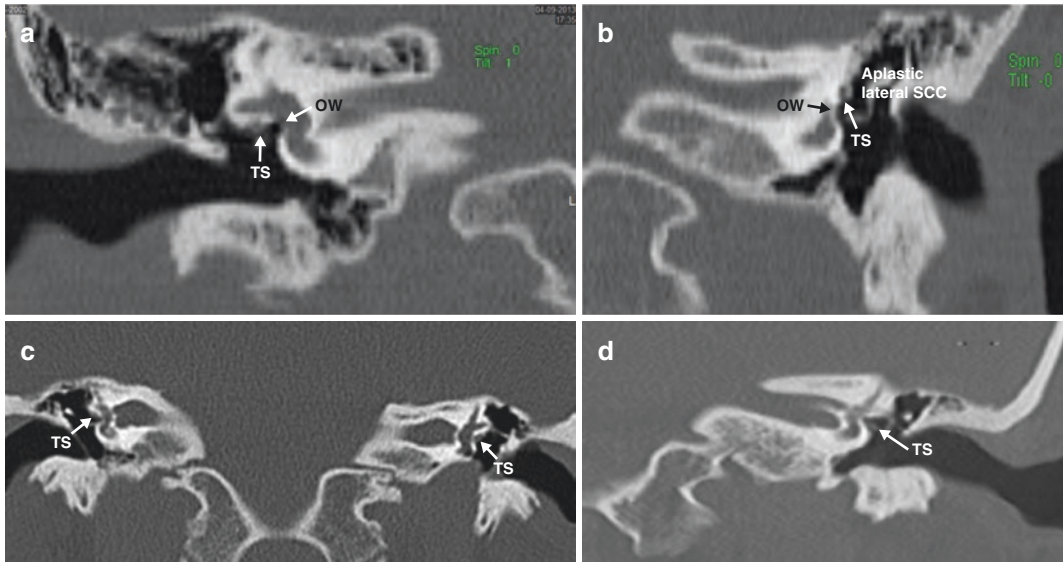


Fig. 13.3 (**a**) *Normal course of the tympanic segment*: Tympanic segment (TS) is located superior and lateral to the oval window (OW) and stapes and inferior to lateral SCC. (**b**) *Superiorly displaced tympanic segment*: Tympanic segment (TS) is located above the oval window (OW). Please note that lateral SCC is aplastic. (**c**) *Tympanic segment at the oval window*: On the left side tympanic segment (TS) is located at the oval window (OW) at coronal section. Please note normal location of the TS on the right side. (**d**) *Tympanic segment inferior to oval window*: Tympanic segment (TS) is located inferior to the oval window (OW), in a case of cochlear hypoplasia

development of otic capsule and Reichert's cartilage. Even though the width of the FR is not thoroughly an embryological process it was also inspected in all CT scans and classified as:

Type 1—Normal facial recess/normal mastoid segment: In the normal temporal bone if the space between the external auditory canal (EAC) and the FN is more than 2 mm, the width of the facial recess is accepted as normal (Fig. 13.4a).

Type 2—Narrow facial recess: The distance between the EAC and the mastoid portion of the FN is less than 2 mm which may result in diminished exposure for cochlear implantation via the posterior tympanotomy approach (Fig. 13.4b).

Type 3—Unclassified: The mastoid portion of the FN cannot be visualized and the course cannot be followed in cases of cochlear aplasia, Michel deformity, CC, or rudimentary otocyst since there are no accurate radiological landmarks.

13.4.3 Embryological Perspective from Inner Ear Malformations to Facial Nerve Abnormalities

13.4.3.1 Meatal Segment

Since the FN traverses inside the IAC, the meatal segment is part of the contents of IAC. In our classification system we categorized IAC width as: normal = 2–8 mm, narrow <2.0 mm, and enlarged >8 mm [31]. Narrow IAC is usually associated with CN and/or FN anomalies, whereas IAC wider than 8 mm is unlikely to be recognized as abnormal [36–38]. It is reported that IAC stenosis accounts for about 12% of all congenital temporal bone malformations and is associated with aplastic cochleovestibular nerve (CVN) [39].

Stenosis of the IAC is usually not related to FN anomalies and it is due to ipsilateral CN deficiency. There are two widely accepted hypothe-

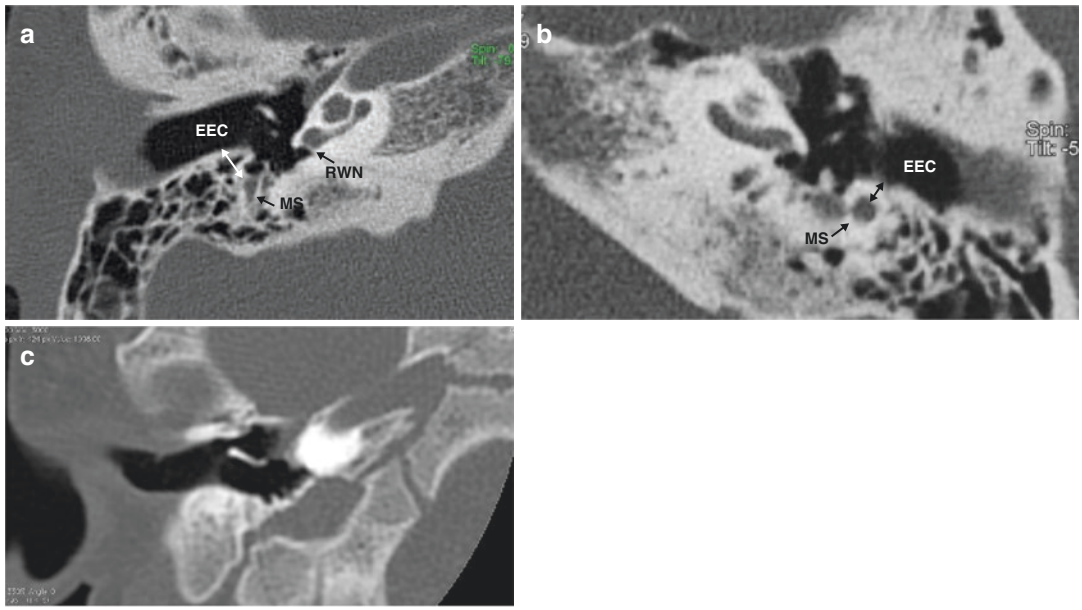


Fig. 13.4 (**a**) *Normal facial recess/normal mastoid segment*: The space between the external auditory canal (EAC) and the mastoid segment of FN (MS) (white arrow) is more than 2 mm; the width of the facial recess is accepted as normal so that a good exposure of the round window niche (RWN) can be expected during racial recess approach. (**b**) *Narrow facial recess*: The distance between the EAC and the mastoid segment (MS) of the FN is less than 1 mm. This resulted in diminished exposure for cochlear implantation via the posterior tympanotomy approach. (**c**) *Type 3. Unclassified Mastoid Segment*. A case of complete labyrinthine aplasia. Mastoid portion of the FN cannot be visualized and the course cannot be followed since there is no accurate radiological landmarks

ses for this association. The first hypothesis states that the embryonic cochlea and vestibule induce the growth of the CVN. The bony IAC develops around the facial and CVN by a process of chondrification and ossification of the enveloping mesoderm, in the eighth week of gestation. As a result, if there is aplasia or hypoplasia of the CVN, IAC becomes narrower. The second hypothesis emphasizes the stenotic canal as the primary defect, which inhibits the growth of the CVN and results in impaired transmission of neural growth factors to the cochlea [40, 41]. A narrow IAC (meatal segment) is often accompanied by a narrow cochlear aperture (CA) [42]. In CA abnormalities it might be expected that the IAC becomes narrow "type 2 meatal segment."

In Michel deformity there is no inner ear development and IAC. Meatal segment of the facial canal can be seen particularly in the presence of otic capsule. Rudimentary otocyst is a more severe anomaly than common cavity without an IAC. The common cavity contains cochlear and vestibular neural elements represented as a single cystic chamber with an IAC opening into the center. Cochlear aplasia with a dilated vestibule is a more developed anomaly with an IAC.

In other IEMs related to a developmental arrest in later stages, the development of the IAC is expected. In IP malformations only the internal architecture of the cochlea is defective and the IAC caliber is usually normal. In CH, cochlea is hypoplastic. Due to the diminished neural growth factor release from spiral ganglion, CN may be deficient which may cause a narrow IAC.

If separate bony canal is identified inside the IAC on a CT image, it is named as "duplicated IAC"/"separate facial canal" or "type 3 meatal segment." In that case FN is separated from the CN by a bony canal inside the IAC. This malformation is not necessarily associated with other IEMs of the temporal bone. In these cases, the facial nerve is usually described as running inside the anterosuperior canal, and these patients have normal facial nerve function.

The embryologic origin of duplicated IAC is uncertain, because FN never exists as separate bundles during its development. Formation of the FN begins early in gestation with the facioacoustic primordium separating into facial and acoustic components at the end of the fourth week. By the end of the fifth gestational week, the chorda tympani has differentiated from the distal facial nerve. By the eighth week, the orientation of FN within the temporal bone has been established, with the nerve's ultimate position and FN canal. An anomalous and precocious activation of one or more centers of ossification would lead to the formation of a complete or incomplete bony septum determining the presence of 2 two separate canals [42]. Kew et al. reported cases with unilateral duplicated IAC, with CVN aplasia and severe facial nerve hypoplasia [41].

Atresia of the IAC (only facial canal) is a rare malformation. It is theorized that IAC atresia is secondary to an agenesis of the CVN. Ozeki et al. noticed that in atresia of the IAC, the facial nerve takes a ventral and superior course, with its own canal starting at the point where the trigeminal nerve enters the Gasserian ganglion [43]. In their series, the CVN was missing in all of the affected ears. The FN was identified in all cases where function was normal. Interestingly, other cases of IAC atresia reported in the literature do not display facial nerve palsy [43, 44]. In early weeks of gestation, facial and CVN were represented as facioacoustic primordium. Later, they will separate and FN starts to migrate anteriorly to the facial musculature. It is speculated that if the CVN is absent, IAC shows no development and FN travels alone. This is seen mostly in severe IEMs such as Michel deformity and rudimentary otocyst.

To summarize the meatal segment anomalies:

- Meatal segment of the FN is part of the contents of IAC.
- A narrow or stenotic IAC (<2 mm in diameter) usually signifies a deficient CN.
- In severe IEMs such as Michel deformity where IAC is not developed, the FN traverses in its own canal "only facial canal."
- In some anomalies, there may be "separate facial canal" or "duplicated IAC."

13.4.3.2 Labyrintine Segment

Premature differentiation of the facial nerve during the embryonic development stage may be a reason for displacement of fallopian canal. Depending on the developmental status of the temporal bone, displacement of fallopian canal may affect different segments of fallopian canal. The labyrinthine segment of the facial nerve was shown to be displaced anteriorly by Raine et al., anteromedially by Hoffman et al., and anteroinferiorly by Huang et al. [25, 28, 45].

Labyrinthine segment is the mostly affected part of the FN by IEMs due to its anatomical border with cochlea. In severe IEMs such as Michel deformity, cochlear aplasia, common cavity, and rudimentary otocyst, it is very rare to find normal course of the FN. In those cases, FN traverses more directly such as straight labyrinthine segment and severe anterior displacement. This strengthens the theoretic assumption that cochlea is the most important structure for the proper localization of the LS.

Complete labyrinthine aplasia or Michel's deformity is caused by early arrest of differentiation of the otic placode during the third gestational week. This deformity is very rare and the diagnosis is made when the inner ear is completely absent on CT and MR. Common cavity malformations occur when development is arrested between the fourth and fifth week of gestation. Arrest of development during the fifth week (cochlear bud) results in cochlear aplasia, a rare malformation [33]. In such cases the FN traverses directly to its end organ, facial muscles, thus the maldevelopment of the cochlea might lead to more anteriorly directed FN course.

Romo and Curtin found that anteromedial migration of the labyrinthine segment of the facial nerve may occur in association with common cavity and cochlear hypoplasia cases [8]. Cochlear hypoplasia results when the normal development of the cochlear duct is impaired during the sixth week of gestation. The cochlea can be distinguished by its smaller dimensions. The vestibule and semicircular canals may be normal but are usually malformed. In CH-I, the cochlea is like a small bud arising from the IAC. CH-II is defined as small cochlea with defective modiolus and interscalar septa with normal external shape. In CH-III, the internal and external architecture is similar to that of a normal cochlea, but the dimensions are less and the number of turns is less (<2 turns). CH-IV has smaller external dimensions with normal basal turn but has hypoplastic middle and apical turn. Because of hypoplastic middle and apical turns FN is displaced in a very characteristic way to the anterior-superior part of the cochlea. This is a very characteristic sign for CH-IV [46]. In other words, middle and apical turns are also important for normal development of the labyrinthine segment. Therefore, labyrinthine segment of FN can demonstrate various abnormalities due to the hypoplastic cochlea. A hypoplastic cochlea leads the labyrinthine segment of FN to a more anterior location.

In incomplete partition anomalies the external size of the cochlea and labyrinth is normal but internal architecture is deformed [47]. Incomplete partition of the cochlea occurs when the development of the cochlea is arrested during the seventh week of gestation. IP-I is characterized by deformation of the internal architecture of cochlea, while the outer structure is normal together with normal cochlear dimensions. In IP-II anomaly, modiolar defect is seen together with minimally dilated vestibule and large vestibular aqueduct. IP-III is defined as the absence of the modiolus with the presence of interscalar septa. In all IP types the external dimensions of the cochlea (height and diameter) are normal. Since the cochlear dimensions are normal in IP-I and IP-II, the facial nerve has it normal curved labyrinthine course. All IP-III cases are characterized by superior displacement of the labyrinthine segment of the facial canal which appears superior to cochlea in axial images. This is a pathognomonic sign for IP-III. During its development the facial nerve is related to a sulcus in the otic capsule and after the completion of the endochondral ossification, the facial nerve is completely surrounded by its canal at 24th week of gestation. The author of this chapter revealed that IP-III has a thick endosteal layer, while the outer two layers (enchondral and outer periosteal) of the otic capsule are missing. Therefore, otic capsule in IP-III appears thinner

than normal because of the absence of outer layers. External dimensions are normal. Outer two layers of the otic capsule should have a critical role in the development of labyrinthine segment as it creates the difference between IP-III and other IP types. These two outer layers appear to prevent the superior displacement of the labyrinthine segment. It might be interpreted that the normal course of the labyrinthine segment is dependent on normal cochlear size as well as the normally enchondral ossification of the cochlea in the neighborhood of the facial canal.

To summarize the labyrinthine segment anomalies, we can speculate these theories:

- Normal sized cochlea is related to a normal labyrinthine segment.
- Defective outer periosteal layer and enchondral ossification as in IP-III cause superiorly displaced labyrinthine segment which is pathognomonic for IP-III.
- Aplastic or hypoplastic cochlea is associated with various degrees of anterior displacement.
- Severe IEMs such as Michel, rudimentary otocyst, and common cavity cause more prominent forms of anterior displacement.
- Hypoplastic middle and apical turns are the major factors that influence anterior-superior displacement which is very typical for CH-IV.

13.4.3.3 Tympanic Segment

Embryologically, the second branchial arch is deeply involved in guiding the course of the facial nerve, and many cases of abnormal facial nerve courses are associated with anomalies of the second branchial arch. The facial nerve, stapes, and otic capsule all develop from the second branchial arch derivates. A developmental problem about one of these structures may result in malformation of both the oval window and the stapes [21, 24]. Any displacement of the FN in the early stages of development may prevent normal stapes and oval window formation. Because of the association with FN anomalies or an aberrant course of FN, surgical intervention can be challenging. Preoperative identification of any anomaly in the tympanic segment in relation to OW is also important to avoid the risk of injury during middle ear surgery. It is previously reported as FN anomalies are found in 59% to 76% of patients with congenital round or oval window malformations. These include partial or complete overlay over the oval window or promontory, dehiscence or partial absence, and bifid facial nerves [48].

Between the sixth and eighth week the semicircular canals develop and the development is completed at 21st week of gestation. The superior semicircular canal develops first, the lateral semicircular canal last. A malformation of a semicircular canal, in association with a normal cochlea therefore most likely is due to an insult occurring between the seventh and 22nd week of gestation. Coronal high-resolution CT reconstruction of temporal bone is highly valuable in preoperative detection of the anomaly in the tympanic segment. At the level of the oval window, the tympanic segment of the facial canal is seen inferior to the lateral SCC in coronal CT sections.

In a recent paper of the authors of this chapter, almost all ears with normal lateral SCC dimensions had normal tympanic segment (99.7%), while the hypoplastic or aplastic lateral SCC was associated with superior or inferior displacement of the tympanic segment [32]. This finding might answer the question whether the lateral eminence of the lateral SCC affects the tympanic course of the FN. In superiorly displaced tympanic segments, vast majority of lateral SCCs were smaller than normal (hypoplastic or aplastic). Although the inferiorly displaced TS is relatively rare, it usually accompanies hypoplastic cochlea and SSCs. In all IP-III cases, FN has normal tympanic segment. In contrast to its key role in the location of labyrinthine segment, otic capsule has a lesser impact on the route of tympanic segment.

Coronal sections of CT allow diagnosis of an absent oval window or an abnormal facial nerve course before surgery. Preoperative evaluation of this segment is especially important in middle ear surgery [3].

As a result,

1. Lateral SCC affects the development of the tympanic segment.
2. When the lateral SCC is aplastic or hypoplastic the tympanic segment is found superior or inferior to oval window.
3. During embryological development, lateral eminence of the lateral SSC is responsible for normal location of the tympanic segment.
4. Inferior dislocation of the tympanic segment is very rare but it is usually seen in cochlear hypoplasia.
5. In ears with cochlear hypoplasia, hypoplasia of SSC is a common entity and this may result in displacement of TS.

13.4.3.4 Mastoid Segment

Air cells between the chorda tympani and the vertical part of the facial nerve are called the facial recess, which is located at the posterior tympanum wall. The anatomic boundary of the facial recess consists of the incudal fossa superiorly, facial nerve posteriorly, and chorda tympani laterally. The width of the facial recess changes with the development of the facial nerve canal and fibrous annulus during the intrauterine period. Changes in the facial recess may relate closely with the development of the facial nerve, tympanic cavity, mastoid air cells, and the position of the tympanic membrane. Zheng et al. [49] reported that the width of the facial recess in ears with congenital malformations was similar to that in the normal ear. The next consideration is preoperative assessment of the facial recess width. The distance between the chorda tympani nerve and the vertical part of the facial nerve less than 1 mm could be considered as a narrow facial recess, affecting the exposure of the round window, opening of the tympanic scala, and insertion of the electrodes, and causing injury of the chorda tympani [50]. However, it is difficult to show the chorda tympani in thin slice temporal bone CT scans. He et al. reported that the normal distance between the vertical facial nerve and the posterior wall of the external acoustic meatus can be used to determine facial recess width. If the space between the external auditory canal (EAC) and the FN is more than 2–3 mm, the width of the FR can be considered as normal [33, 51].

Telmessani et al. [52] classified the vertical segment of the FN (VSFN) in relation to lateral semicircular canals (LSSC). To assess the location of the VSFN in relation to the LSCC, two vertical lines were drawn, one adjacent to the most lateral aspect of the bony LSCC and another adjacent to the most lateral aspect of the fallopian canal of the VSFN. The location of the VSFN in relation to the LSCC was classified into three types: type I, medial to the LSCC; type II, in the same plane as the LSCC; and type III, lateral to LSCC. They observed some IEMs in VSFN abnormalities but there was no significant association between position of the VSFN and IEM [52]. In that classification they did not mention about cochlear hypoplasia and did not investigate the LSSC morphology as a separate entity. The authors of this chapter proposed a classification system regarding the LSSC canal, oval window, and tympanic segment of the FN [32].

As mentioned above, FN canal ossification from Reichert's cartilage is completed at fifth gestational week; however, the mastoid segment develops and completes its development postnatally, along with the growth and pneumatization of the mastoid bone. Surgically, the mastoid segment is of critical importance, as the facial recess is drilled out to expose the round window niche or promontorium for cochlear implantation.

The authors of this chapter evaluated if a particular IEM causes misplacement of the mastoid segment, creating difficulty during surgery via the facial recess approach. It was found that the frequency of narrow FR was higher in the CH and severe IEM group in contrary to Zheng et al. [49]. The mastoid segment of the FN should be considered a unique entity, as its location is dependent on the mastoid aeration [49]. However in severe IEMs it is more likely to encounter a narrow FR during cochlear CI.The surgeon should be careful during a posterior tympanotomy approach in severe IEMs.

FN may be misplaced over the oval and round window or promontory. In this situation it may be impossible to measure the width of the facial

recess. This situation is most often seen in cases with cochlear hypoplasia or severe IEMs. Therefore, in CH and severe malformations, due to an abnormal course of the tympanic and mastoid segments facial recess approach may be very difficult.

To summarize:

- The mastoid segment might not be directly affected by otic capsule development.
- Pneumatization of the mastoid segment continues after birth and continues until adulthood; therefore, changes in the route of the mastoid segment can be expected to occur afterwards.
- Narrow facial recess is not particularly associated with a specific sub-type IEM. However, FR becomes narrow in CH and severe IEM cases.
- In CH and severe IEMs, due to abnormal course of FN over oval, round windows and promontory facial recess approach may be difficult to use.

13.5 Clinical Significance of FN Abnormalities

When performing CI in syndromic patients, particularly those with craniofacial deformities or IEMs, surgeons should always consider the possibility of an aberrant course of the FN. Meticulous preoperative CT analysis regarding FN and ossicular anomalies and intraoperative nerve monitoring systems are mandatory to prevent FN trauma. When surgical access to the round window is restricted by an aberrant course of the FN, the surgeon should reassess the CT findings. In patients with cochleovestibular malformations, surgeons should always consider the possibility of an aberrant course or shape of the FN. This is most common in cases with cochlear hypoplasia. Through such systematic and cautious management, the majority of cases can be successfully implanted without major complications. Due to the aberrant course, dehiscence, or its proximity to the electrode array, FN anomalies can result in FN stimulation following device activation. Cushing et al. have argued that CI patients with inner ear malformations appear to be at an increasing risk of FN stimulation following device activation [53]. When FN stimulation occurs, the nearby electrodes may be switched off.

A narrow facial recess will adversely affect a surgeon's ability to expose the round window niche, open the scala tympani, and insert the electrode without injuring the chorda tympani. Axial sections of the temporal bone CT provide vital radiological data about the anatomical landmarks which are important for cochlear implantation surgery. If a narrow facial recess is detected preoperatively, alternative surgical approaches should be considered.

References

1. Aslan A, Goktan C, Okumus M et al. Morphometric analysis of anatomical relationships of the facial nerve for mastoid surgery. J Laryngol Otol. 2001;115(6):447–9.
2. Webb RL, Lehnhardt E, Clarj GM, et al. Surgical complications with the cochlear multiple channel intracochlear implant: experience at Hannover and Melbourne. Ann Otol Rhinol Laryngol. 1991;100(2):131–6.
3. Sennaroğlu L, Bajin MD, Atay G, Günaydın RÖ, Gönüldaş B, Batuk MÖ, Mocan BÖ, Sennaroğlu G. Oval window atresia: a novel surgical approach and pathognomonic radiological finding. Int J Pediatr Otorhinolaryngol. 2014;78(5):769–76.
4. Glastonbury CM, Fischbein NJ, Harnsberger HR, Dillon WP, Kertesz TR. Congenital bifurcation of the intratemporal facial nerve. AJNR Am J Neuroradiol. 2003;24(7):1334–7.
5. Song JJ, Park JH, Jang JH, Lee JH, Oh SH, Chang SO, Kim CS. Facial nerve aberrations encountered during cochlear implantation. Acta Otolaryngol. 2012;132(7):788–94.
6. Schuknecht HF. Pathology of the ear. Cambridge, MA: Harvard University Press; 1976. p. 332–3.
7. Streeter GL. On the development of the membranous labyrinth and the acoustic and facial nerves in the human embryo. Am J Anat. 1906;6:139–65.
8. Romo LV, Curtin HD. Anomalous facial nerve canal with cochlear malformations. AJNR Am J Neuroradiol. 2001;22(5):838–44.
9. Wysocki J, Skarzynski H. Distance between the cochlea and adjacent structures related to cochlear implant surgery. Surg Radiol Anat. 1998;20(4):267–71.
10. Anson BJ, Bast TH, Richany SF. Development of the second branchial arch (Reichert's cartilage),

facial canal and associated structures in man. Q Bull Northwest Univ Med Sch. 1956;30(3):235–49.
11. Curtin HD, Vignaud J, Bar D. Anomaly of the facial canal in a Mondini malformation with recurrent meningitis. Radiology. 1982;144(2):335–41.
12. Sataloff RT, Selber JC. Phylogeny and embryology of the facial nerve and related structures. Part II: Embryology. Ear Nose Throat J. 2003;82(10):764–6.
13. Henner R. Congenital middle ear malformations. AMA Arch Otolaryngol. 1960;71:454–8.
14. Rutherford KD, Lerer TS, Schoem SR, Valdez TA. Evaluation of pediatric sensorineural hearing loss: a survey of pediatric otolaryngologists. Ann Otol Rhinol Laryngol. 2011;20(10):674–81.
15. Casselman JW, Offeciers EF, De Foer B, Govaerts P, Kuhweide R, Somers T. CT and MR imaging of congenital abnormalities of the inner ear and internal auditory canal. Eur J Radiol. 2001;40(2):94–104.
16. Nager GT, Proctor B. The facial canal: normal anatomy, variations and anomalies. II. Anatomical variations and anomalies involving the facial canal. Ann Otol Rhinol Laryngol Suppl. 1982;97:45–61.
17. Adunka OF, Jewells V, Buchman CA. Value of com- puted tomography in the evaluation of children with cochlear nerve deficiency. Otol Neurotol. 2007;28(5):597–604.
18. Nauer CB, Rieke A, Zubler C, Candreia C, Arnold A, Senn P. Low-dose temporal bone CT in infants and young children: effective dose and image quality. AJNR Am J Neuroradiol. 2011;32(8):1375–80.
19. Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. Otol Neurotol. 2005;26(5):976–82.
20. Baxter A. Dehiscence of the fallopian canal. J Laryngol Otol. 1971;85:587–94.
21. Jahrsdoerfer RA. The facial nerve in congenital middle ear malformations. Laryngoscope. 1981;91(8):1217–25.
22. Marquet J. Congenital malformations and middle ear surgery. J R Soc Med. 1981;74(2):119–28.
23. Basek M. Anomalies of the facial nerve in normal temporal bones. Ann Otol Rhinol Laryngol. 1962;71:382–90.
24. Zeifer B, Sabini P, Sonne J. Congenital absence of the oval window: radiologic diagnosis and associated anomalies. AJNR Am J Neuroradiol. 2000;21(2):322–7.
25. Raine CH, Hussain SS, Khan S, Setia RN. Anomaly of the facial nerve *and* cochlear implantation. Ann Otol Rhinol Laryngol Suppl. 1995;166:430–1.
26. Fowler EP. Variations in the temporal bone course of the facial nerve. Laryngoscope. 1961;71:937–46.
27. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. J Laryngol Otol Suppl. 2000;25:1–14.
28. Hoffman RA, Downey LL, Waltzman SB, Cohen NL. Cochlear implantation in children with cochlear malformations. Am J Otol. 1997;18:184–7.
29. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
30. Jin A, Xu P, Qu F. Variations in the labyrinthine segment of facial nerve canal revealed by high-resolution computed tomography. Auris Nasus Larynx. 2018;45(2):261–4.
31. Sennaroğlu L, Bajin MD. Classification and current Management of Inner ear Malformations. Balkan Med J. 2017;34(5):397–411.
32. Sennaroğlu L, Tahir E. A novel classification: anomalous routes of the facial nerve in relation to inner ear malformations. Laryngoscope. 2020. Epub a head of print.
33. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1): 3–20.
34. Demir OI, Cakmakci H, Erdag TK, Men S. Narrow duplicated internal auditory canal: radiological findings and review of the literature. Pediatr Radiol. 2005;35(12):1220–3.
35. Kashio A, Sakamoto T, Karino S, Kakigi A, Iwasaki S, Yamasoba T. Predicting round window niche visibility via the facial recess using high-resolution computed tomography. Otol Neurotol. 2015;36(1):18–23.
36. Cho YS, Na DG, Jung JY, Hong SH. Narrow internal auditory canal syndrome: parasagittal reconstruction. J Laryngol Otol. 2000;114(5):392–4.
37. Sakashita T, Sando I. Postnatal development of the inter- nal auditory canal studied by computer-aided three-dimensional reconstruction and measurement. Ann Otol Rhinol Laryngol. 1995;104(6):469–75.
38. Ferreira T, Shayestehfar B, Lufkin R. Narrow, duplicated internal auditory canal. Neuroradiology. 2003;45(5):308–10.
39. Li Y, Yang J, Liu J, Wu H. Restudy of malformations of the internal auditory meatus, cochlear nerve canal and cochlear nerve. Eur Arch Otorhinolaryngol. 2015;272(7):1587–96.
40. Yates JA, Patel PC, Millman B, Gibson WS. Isolated congenital internal auditory canal atresia with normal facial nerve function. Int J Pediatr Otorhinolaryngol. 1997;41(1):1–8.
41. Kew TY, Abdullah A. Duplicate internal auditory canals with facial and vestibulocochlear nerve dysfunction. J Laryngol Otol. 2012;126(1):66–71.
42. Tahir E, Bajin MD, Atay G, Mocan BÖ, Sennaroğlu L. Bony cochlear nerve canal and internal auditory canal measures predict cochlear nerve status. J Laryngol Otol. 2017;131(8):676–83.
43. Ozeki M, Kato Z, Sasai H, Kubota K, Funato M, Orii K, Kaneko H, Fukao T, Kondo N. Congenital inner ear malformations without sensorineural hearing loss in children. Int J Pediatr Otorhinolaryngol. 2009;73(10):1484–7.
44. Everberg G, Ratjen E, Sorensen H. Unilateral atresia of the internal auditory meatus, confirmed by radiography. Br J Radiol. 1963;36:568–73.

45. Huang CC, Lin CY, Wu JL. Retrofacial approach of cochlear implantation in inner ear malformation with aberrant facial nerve: a case report. Auris Nasus Larynx. 2006;33:179–82.
46. Sennaroğlu L, Bajin MD, Pamuk E, Tahir E. Cochlear hypoplasia type four with anteriorly displaced facial nerve canal. Otol Neurotol. 2016;37(10):407–9.
47. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41.
48. Vincent R, Wegner I, Derks LS, Grolman W. Congenital oval or round window malformations in children: surgical findings and results in 17 cases. Laryngoscope. 2016;126(11):2552–8.
49. Zheng Y, Schachern PA, Djalilian HR, Paparella MM. Temporal bone histopathology related to cochlear implantation in congenital malformation of the bony cochlea. Otol Neurotol. 2002;23(2):181–6.
50. Wang L, Yang J, Jiang C, Zhang D. Cochlear implantation suergey in patients with narrow facial recess. Acta Otolaryngol. 2013;133(9):935–8.
51. Sennaroglu L, Tahir E. Cochlear implantation in a subject with a narrow facial recess: importance of preoperative radiological findings. Cochlear Implants Int. 2016;17(3):158–61.
52. Telmesani LM, Alrammah MK. Telmesani radiological classification of the location of the vertical segment of the facial nerve: impact on surgical approach in cochlear implant surgery. Otol Neurotol. 2017;38(9):335–8.
53. Cushing SL, Papsin BC, Strantzas S, Gordon KA. Facial nerve electromyography: a useful tool in detecting nonauditory side effects of cochlear implantation. J Otolaryngol Head Neck Surg. 2009;38:157–65.

Stapedotomy

14

Levent Sennaroglu, Munir Demir Bajin, and Betul Cicek Cinar

14.1 Introduction

Congenital ossicular chain abnormalities behind normal ear canal and tympanic membrane are much rarer when compared to conductive hearing loss (CHL) caused by acquired causes. They usually fail neonatal hearing screening tests. It can be diagnosed early if air and bone conduction ABR is done. They usually present as non-progressive hearing loss since birth. If there is additional sensorineural hearing loss component, they usually present with poor language development.

In congenital CHL stapes may be fixed alone or three ossicles may be involved in different degrees. They may all be fixed. Stapes fixation is the most important of all, because stapedotomy may improve hearing loss significantly. However, stapedotomy in pediatric population is controversial. In this group of patients there may be higher risk of sensorineural hearing loss (SNHL), facial nerve (FN) injury, and meningitis when compared to normal population. These are the main reasons for stapedotomy to be controversial in pediatric population.

In 2014, first author investigated the specimens with inner ear malformations in Massachusetts Eye and Ear Infirmary (MEEI) [1]. Interestingly 14 of 41 cases had oval window atresia or stapes footplate fixation. This brings the concept of performing stapedotomy in cases with IEMs. Here we present 11 cases of IEMs who underwent stapedotomy between 2008 and 2014.

14.2 Histopathology

Histologically stapes fixation and oval window anomalies can be encountered in IEMs. Sennaroglu L investigated 41 temporal bone specimens with IEMs in the MEEI [1]. Fourteen of the 41 cases with IEMs presented either with fixed stapes footplate (Fig. 14.1a) or oval window atresia (Fig. 14.1b). Twelve of these cases presented with cochlear hypoplasia (CH). As the oval window is part of the cochlea, fixation of the footplate or the oval window atresia can be expected in a hypoplastic cochlea. It was interesting to note that cochlear abnormalities, particularly cochlear hypoplasia, may cause CHL or mixed hearing loss by stapes fixation.

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_14].

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

M. D. Bajin (✉)
Department of Otolaryngology, Hacettepe University, Medical Faculty, Ankara, Turkey
e-mail: dbajin@hacettepe.edu.tr

B. C. Cinar
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_14

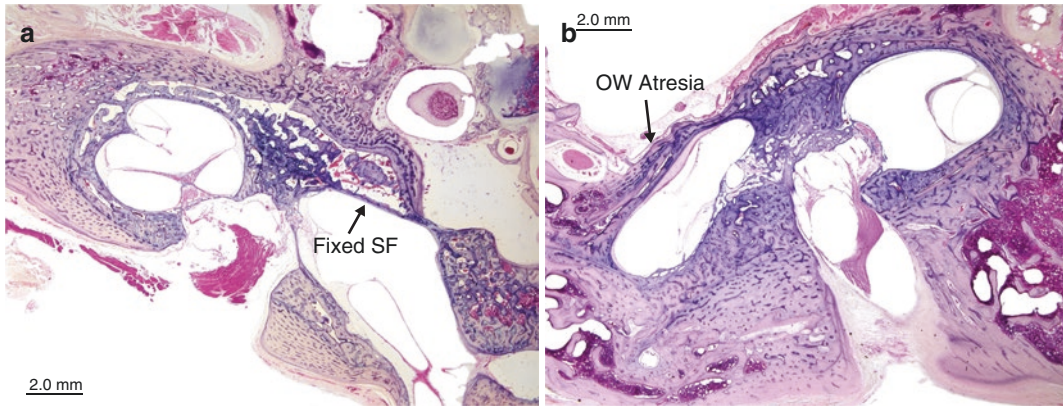


Fig. 14.1 Histopathological findings of oval window in cochlear hypoplasia: (**a**) stapes footplate (SF) fixation, (**b**) oval window (OW) atresia. (With permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary)

14.3 Literature Review

In his report on histopathology of IEMs, Sennaroglu L. mentioned that majority of stapes footplate fixation or oval window atresia are found in cochlear hypoplasia (CH) [1]. Out of three CH-I cochleae in MEEI, two had a fixed stapes footplate and one had an atretic oval window. In specimens with CH-II, oval window was atretic in one specimen, and the stapes footplate was fixed in three. There were five specimens with CH-III: oval window was normal in one specimen, fixed in one, and atretic in three. There was no specimen with CH-IV. These cases show that it is common to find stapes footplate fixation or oval window atresia in cochlear hypoplasia. It also implies that if there is conductive or mixed type hearing loss in CH stapedotomy may result in better hearing. The patient may have to use hearing aid after surgery depending on the bone conduction level and air-bone gap. If there is pure conductive loss, stapes surgery may result in near normal hearing.

According to Sennaroglu [1], in CH-III, the developmental arrest in the membranous labyrinth most probably occurs between 6 and 8 weeks, resulting in a cochlea whose dimensions are smaller than normal, with normal internal architecture. In CH-IV there is a normal basal turn but small middle and apical turns. Arrest in the membranous labyrinth must be between 10th and 20th weeks, after the basal turn reaches full size but before the middle and apical turns enlarge to their normal size.

Congenital stapes fixation can be explained by embryology: the stapes footplate is part of the otic capsule, and according to Donaldson et al. [2], the base of the stapes is originally continuous with the otic capsule. Then it is segregated through a retrogressive process in the cartilage. The reorganized tissue becomes the annular ligament. A transcapsular channel (fissula ante fenestram) is formed as a result of invasion of the primitive cartilage by periotic tissue. If there is an arrest of the otic capsule development before the formation of the footplate, it is natural that the stapes becomes fixed to the oval window; however, it is still difficult to explain stapes fixation in CH-IV with normal basal turn.

CH-I and CH-II cases also have stapes fixation in some cases. But due to the severe malformation and profound SNHL, they are candidates for implantation. CH-II cases are accompanied by a defective modiolus. Because of the resolution of present day HRCT, the partial modiolar defect may not be diagnosed, but histopathological examination shows the defective modiolus in all cases. Because of the shorter cochlea they have SNHL and the fixed footplate provides the conductive component. The author has performed stapedotomy in cases of CH with mixed hearing loss. Postoperatively, these cases benefit more from HA. Patients with CH who have profound sensorineural hearing loss (SNHL) are candidates for CI. CH with cochlear aperture aplasia necessitates an ABI.

14.4 Clinical Findings

Between 2008 and 2014, nine cases of cochlear hypoplasia underwent stapedotomy (two cases of vestibular dilatation are not included). They all applied with the complaint of hearing loss, which was non-progressive and present since birth. They present with a normal ear canal and tympanic membrane. Otitis media with effusion may be a coincidental finding but conductive hearing loss persists after the effusion is treated. In addition to stapes, other ossicles may also be fixed, the facial nerve may be misplaced at the oval window area, or the oval window may be atretic.

14.5 Radiological Findings

Radiology is very important in congenital conductive or mixed hearing loss. Radiology may demonstrate ossicular fixation to the attic wall.

Radiology also shows the type of IEMs. There are four groups of cochlear hypoplasia where external dimensions are smaller than normal cochlea with various internal architecture deformities (Chaps. 1 and 26 for more details):

1. CH-I: Cochlea with absent internal architecture (modiolus and ISS) with/without a thin bony partition between the cochlea and the IAC.
2. CH-II: External shape resembled a cochlea, but it was smaller and rounder than normal. Modiolus is defective resulting in a cystic cochlea.
3. CH-III: Small cochlea with normal internal architecture. The only difference from a normal cochlea was that the CH-III cochlea consisted of approximately one and a half turns.
4. CH-IV: Cochlea with normal basal turn, hypoplastic middle, and apical turn.

CH-I is like a bud without any internal architecture and outcome with stapes surgery is not expected to be good. In CH-II there may be a risk of gusher. CH-III (Fig. 14.2a, b) and CH-IV (Fig. 14.2c, d) are the best candidates for stapedotomy.

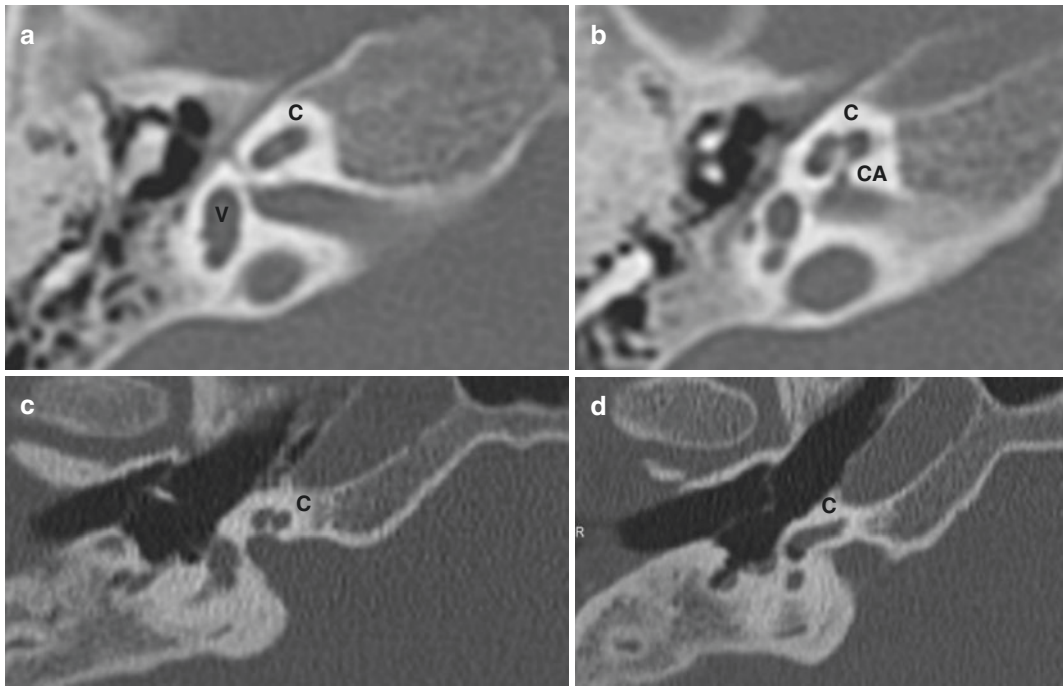


Fig. 14.2 (**a**, **b**) Cochlear hypoplasia type III. Cochlea (C) consists of hypoplastic basal and middle turns with hypoplastic vestibule (V). Note stenotic cochlear aperture (CA). (**c**, **d**) Cochlear hypoplasia type IV. Cochlea (C) consists of normal basal turn and hypoplastic middle and apical turns

Temporal CT shows the position of the facial nerve. Coronal sections in particular show the position of the facial nerve in relation to oval window. In the present series there are cases with facial nerve in more lateral position (Fig. 14.3a), at the OW (Fig. 14.3b) and inferior to the OW (Fig. 14.3c). The surgery becomes very challenging if the coronal section demonstrates the facial nerve at or inferior to the oval window on coronal section.

Another use of HRCT is to show the defect between the cochlea and internal auditory canal (IAC) which may cause CSF leakage. Case 14.1 who had CSF leakage did not have a defect between IAC and cochlea but presented with demineralization all around cochlea, which may cause CSF from subarachnoid space to reach cochlea and hence result in CSF leakage at the time of fenestration into the vestibule.

14.6 Audiological Findings

Audiological findings of these patients are given in Table 14.1. It is possible to diagnose pure conductive hearing loss (see Case 14.1) and mixed type hearing loss (see Case 14.2). Out of these nine ears, seven were pure conductive and two were mixed. Although both benefit from stapedotomy, there is a possibility for near normal hearing in cases of conductive hearing loss (see Case 14.1). In case of mixed hearing loss, aim of the surgery is to make the patient benefit more from hearing aids in the postoperative period (see Case 14.2).

All patients had cochlear hypoplasia (CH). More common presentation for CH is SNHL. Depending on thresholds, hearing aids or CI may be the method for habilitation. If there is no cochlear nerve ABI may be indicated as well.

14.7 Management

There are different treatment options in congenital ossicular fixation. Providing hearing aids is the earliest and most appropriate option, particularly in bilateral cases; however, in cases that present with severe ossicular pathology such as oval window atresia or ossicular discontinuity between the tympanic membrane and the oval

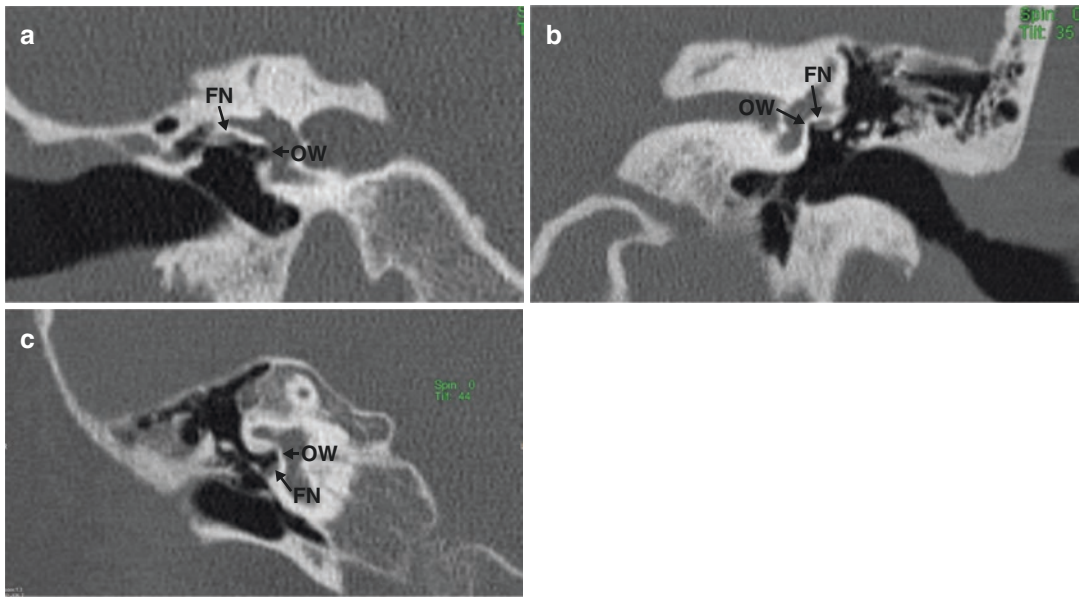


Fig. 14.3 Abnormalities of tympanic segment of facial nerve (FN) (coronal section) (**a**) FN in a more lateral position in relation to oval window (OW) and stapes and inferior to lateral semicircular canal (cochlear hypoplasia type IV), (**b**) FN at the OW, (**c**) FN inferior to oval window

window, hearing aids may not be sufficient or well tolerated because of the large air-bone gap (ABG). Stapedotomy is another option that is not universally accepted for children, because the procedure involves exposing the child's inner ear. Yet another option is bone anchored hearing devices, but for children, the surgical placement of a bone anchored device can be too early. Bone conduction devices with head band may be appropriate if the patient does not benefit from a hearing aid. As this is a treatment option for ear canal atresia cases, where the surgical reconstruction of the ear canal is usually not satisfactory, the families usually look for other treatment possibilities in ossicular pathologies. In such a situation, stapedotomy is one of the treatment options. In addition, other ossicles may become fixed, necessitating surgical options such as manubriostapedioplasty [3] or atticotomy.

14.8 Surgery

There are certain difficulties during the surgery of CHL in IEMs. These can be grouped under five headings:

1. **Facial Nerve Anomaly:** Facial nerve (FN) may have an abnormal location in congenital cases. In CH-IV the labyrinthine segment of the facial nerve (FN) is anterior-superior to the cochlea (patient #6 in Table 14.1). This is almost a pathognomonic finding. Therefore, tympanic segment may not be in the usual location. Two cases with CH-IV had stapedotomy and their tympanic segment was located more superior to its normal location relative to oval window (Fig. 14.3a). This did not cause any difficulty during stapedotomy. Her mother with identical abnormality underwent cochlear implantation where she needed subtotal petrosectomy to visualize round window because of mastoid segment anterior dislocation (see Case 26.3 Chap. 26). Most difficult situation is when tympanic segment of FN is located at the oval window (patient #1). Preoperatively, this situation can be seen on coronal sections of temporal bone CT passing through the oval window. This needs special technique for drilling into vestibule while the burr is very close to the dehiscent FN and incus has to be extended for piston placement [4]. FN can also be seen inferior to the OW

Table 14.1 Audiological findings of the operated patients

Patient	Age	Sex	Side	IEM	Op date	Operation findings	Preoperative			Postoperative		
							Bone	Air	ABG	Bone	Air	ABG
1-TU	16	M	Left	CH-III	2008	FN at OW, drill to make vestibulotomy, bone cement, stapedotomy	22	72	50	12	28	16
2-SA	11	F	Left	CH-IV	2012	Stapedotomy, oozing	10	54	44	10	45	35
3-HG	8	F	Left	CH-III	2012	OW atresia, drill to make vestibulotomy & stapedotomy	13	47	34	13	25	12
3-HG	10	F	Right	CH-III	2013	OW atresia, drill used to make vestibulotomy & stapedotomy	10	48	38	13	32	19
4-IH	4	M	Left	CH-III	2013	FN inferior to OW, OW atresia, drill to make vestibulotomy, gusher	10	59	49	13	28	15
5-HS	11	F	Left	CH-III	2013	All ossicles fixed, atticotomy, stapedotomy	37.5	85	47.5	30	61.25	31.25
6-DK	22	F	Right	CH-IV	2013	Stapedotomy	10	66	55	11	25	14
4-IH	5	M	Right	CH-III	2014	FN inferior to OW, OW atresia, drill used to make vestibulotomy, gusher	10	55	45	12	22	10
5-HS	13	F	Right	CH-III	2014	All ossicles fixed, atticotomy, stapedotomy	37.5	89	51.50	33.75	71.25	37.5

(patient #4 in Table 14.1). Surgically, these are the most challenging cases, requiring facial nerve monitoring and a stimulator.

2. **Oval Window Atresia:** Some cases do not have an oval window formation. Those cases can be classified as atretic and require drilling for making fenestra into vestibule. Using a laser is not advisable because the bone is thick, and a laser may cause excessive heating, resulting in thermal damage to FN. Correct location can be estimated by taking into account the position of the incus and if present, the remnants of stapes suprastructure. During drilling, it is necessary to avoid a tunnel, but carefully lower the thickness of the bone to open the vestibule at the final moment by taking into consideration the direction of the prosthesis from the incus to the fenestra. If this is not planned correctly, drilling after opening the fenestra may cause severe SNHL.
3. **Incus Abnormality**: In cases of otosclerosis incus is positioned more horizontally during surgical exploration. In cases of oval window atresia, incus is more obliquely positioned possibly because of the absence of the stapes, making stapes piston placement more difficult. There is a possibility of the piston sliding off the incus which is positioned more oblique than normal. It is advisable to use a few drops of cement to stabilize the piston and avoid its slipping off incus towards vestibule.
4. **Involvement of Other Ossicles**: This finding is likely to be unrelated to the IEMs but makes the surgery more challenging in this situation. An atticotomy is necessary removing all the bone immobilizing the ossicles. This was present in patient #5 in Table 14.1. It is advisable to use endaural approach in all these cases so that more manipulation can be done around malleus and incus if necessary.
5. **CSF Leakage:** This is the most serious complication of the surgery. If the leak is not controlled properly, it may lead to meningitis. It is mandatory to have the children vaccinated before the procedure. Nowadays, pneumococcal and haemophilus vaccination is routinely done in many countries.

CSF leakage can be expected in these cases. If a piston tightly fitting into the fenestra is used, inserting the fascia around the piston into the vestibule becomes difficult. Based on our experience, using a 0.6 mm drill and a 0.4 mm stapes piston is suggested so that fascia can be inserted sufficiently into the vestibule. Passing the piston shaft through the fascia and inserting them together allows the fascia to surround the piston all around (see Case 14.1).

Contraindications to surgery in congenital hearing loss: In patients with CH with CHL, stapedotomy can be suggested to the families, but it is difficult to think stapes footplate fixation in congenital mixed HL. The operation should never be done if HRCT demonstrates IP-II or IP-III (Fig. 14.4a–c).

14.9 Complications

This procedure carries the risk of injury to the facial nerve, sensorineural hearing loss, and meningitis. Patients without vaccination against pneumococcus and haemophilus influenza should not be operated.

14.10 Clinical Experience

Between January 2003 and September 2015, the first author performed stapedotomy in 355 cases that presented with normal tympanic membrane and ossicular pathologies. Fifty-one of these had congenital ossicular anomalies with a normal ear canal and tympanic membrane (ear canal atresia is not included). If the revision cases are excluded, 42 primary cases out of 355 presented with congenital fixation. Eleven of these cases had various inner ear malformations. Nine had cochlear pathologies and two had only vestibular anomalies.

Nine cases presented with cochlear hypoplasia (CH) (Table 14.1). As stapes is part of the cochlea, the surgical findings are investigated in particular from this perspective. They have been classified according to the recent classification system [5, 6].

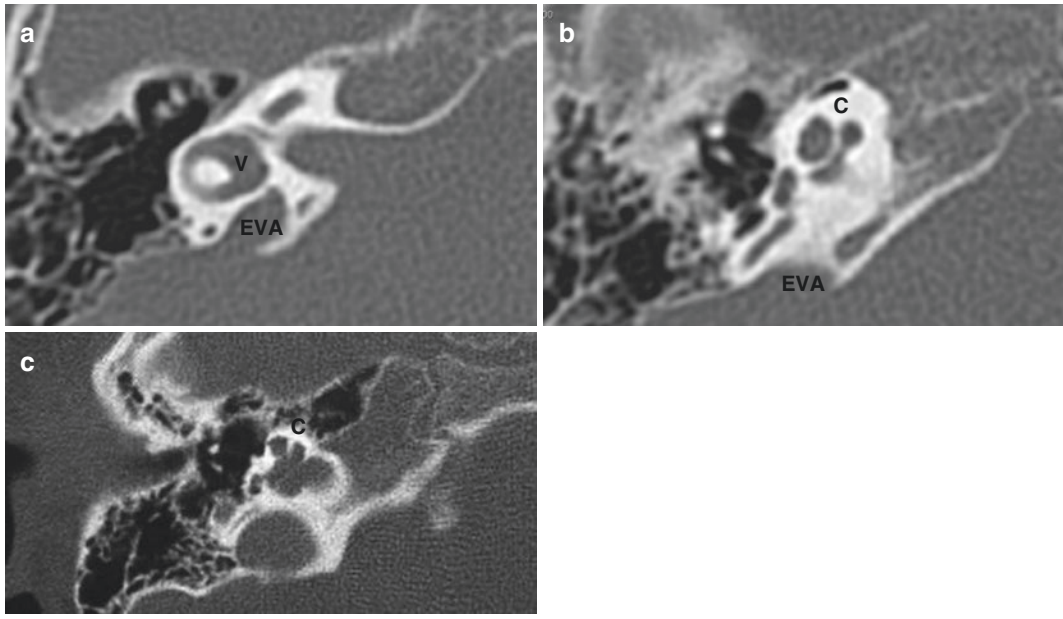


Fig. 14.4 Pathologies with mixed hearing loss where stapedotomy is contraindicated: (**a**, **b**) IP-II cochlea (C) with cystic apex, enlarged vestibular aqueduct (EVA), and minimally dilated vestibule (v). (**c**) IP-III cochlea (C) with interscalar septa and absent modiolus

The age of the patients was between 4 and 22 (Table 14.1). There were two males (three ears) and four females (six ears). There were seven cases of CH-III and two cases of CH-IV. Three cases had bilateral stapedotomy. Remaining three cases had unilateral surgery.

14.11 Cases

Case 14.1: IH (Patient #4) 4-Year-Old Male Patient

He applied with bilateral hearing loss since birth. He failed hearing screening on both sides. On HRCT of the temporal bone he had bilateral CH-III (Fig. 14.5a, b). Coronal sections showed that FN was located inferior to oval window on both sides (Fig. 14.5c, d). His preoperative audiological evaluation revealed bilateral moderately severe conductive hearing loss (Fig. 14.5e). His operation was the most difficult of the nine cases. Left side was operated on 20 September 2013 and right side on 25 November 2014. On both sides, the oval window was completely atretic without any footplate or annular ligament formation; however, round window was present bilaterally. Incus and malleus were present (Video 14.1). By taking into account the position of the incus, a vestibulotomy was created using 0.6 mm diamond burr. Cerebrospinal fluid (CSF) leakage occurred on both sides. A 0.4 mm stapes piston was passed through a piece of fascia (2 × 2 mm) and then placed between the incus and the fenestra. The fascia was inserted all around the piston into the vestibule in a dumb-bell fashion. CSF leak stopped completely. Incus was inclined medially on both sides, which made piston insertion difficult. A few drops of cement were used to fix the piston onto angled incus. The patient had already been immunized with Pneumococcal and Haemophilus influenza vaccination. Ear examination at 3 months' intervals revealed no fluid in the middle ear that would suggest CSF leakage.

After operation, his air conduction hearing thresholds showed improvement and ABG decreased to 20 dB from 50 dB. He had mild con-

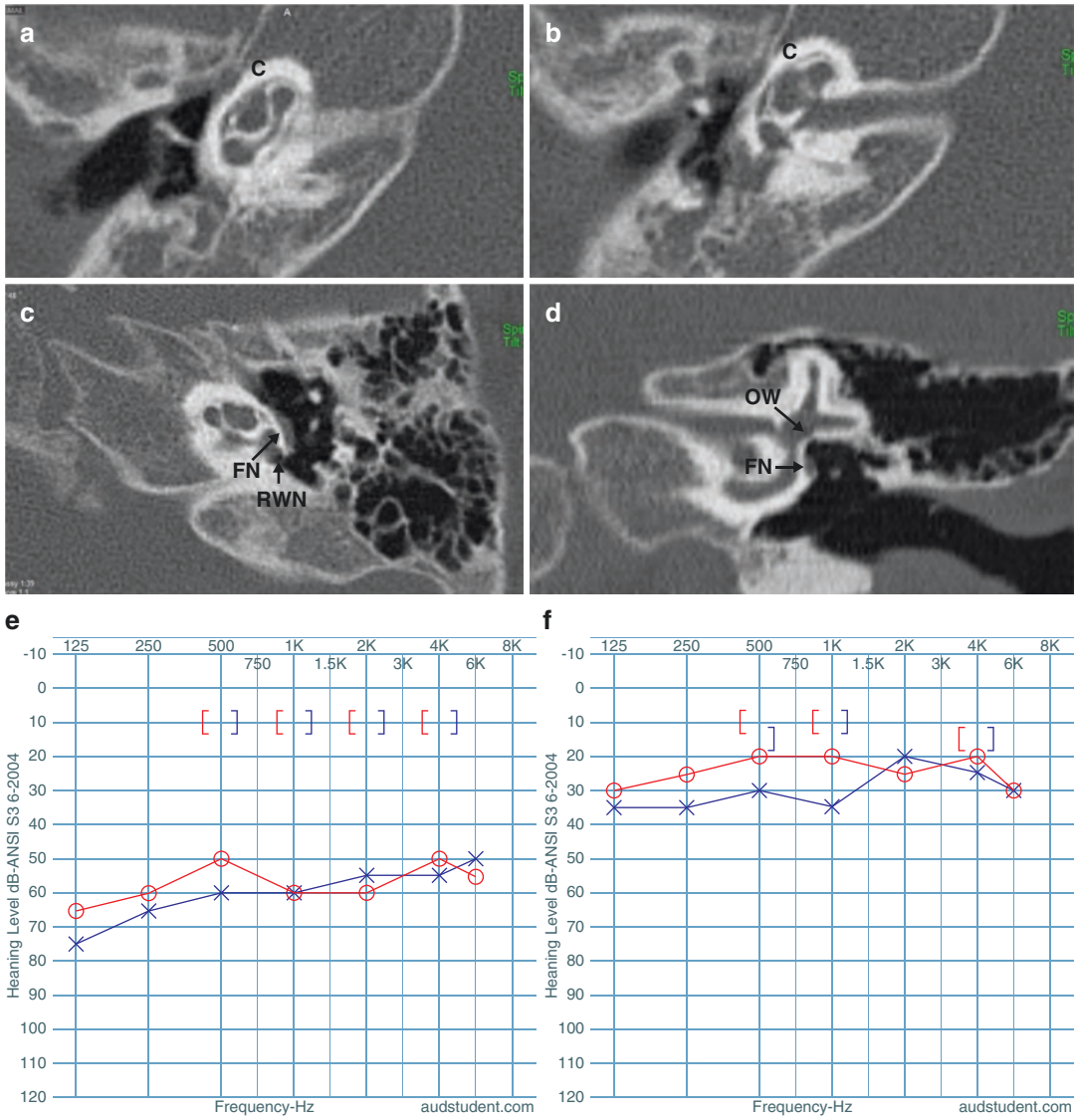


Fig. 14.5 Case 14.1. (**a**, **b**) Axial sections showing cochlear hypoplasia type III (C); note demineralization around cochlea. Modiolus appears to be normal. (**c**) Axial section showing tympanic segment of the facial nerve (FN) over the round window niche (RWN) on the promontory (left ear). (**d**) Coronal section showing FN below the oval window (OW). (**e**, **f**) Preoperative (**e**) and postoperative (**f**) hearing thresholds

ductive hearing loss and was able to hear without hearing aids. He developed excellent speech without hearing aid.

CSF leakage most probably came from bone demineralization around cochlea. Coronal section is very important to visualize FN in relation to oval window. This patient shows that it is possible to obtain hearing without hearing aids in pure conductive hearing loss.

Case 14.2: HS (Patient #5) 11-Year-Old Female Patient

She applied with bilateral severe mixed type hearing loss since birth. She had congenital cleft palate and developmental delay. She had been using hearing aids with very poor language development. CT revealed bilateral CH-III with extremely small middle and apical turns (Fig. 14.6a, b). Cochlear aperture was stenotic.

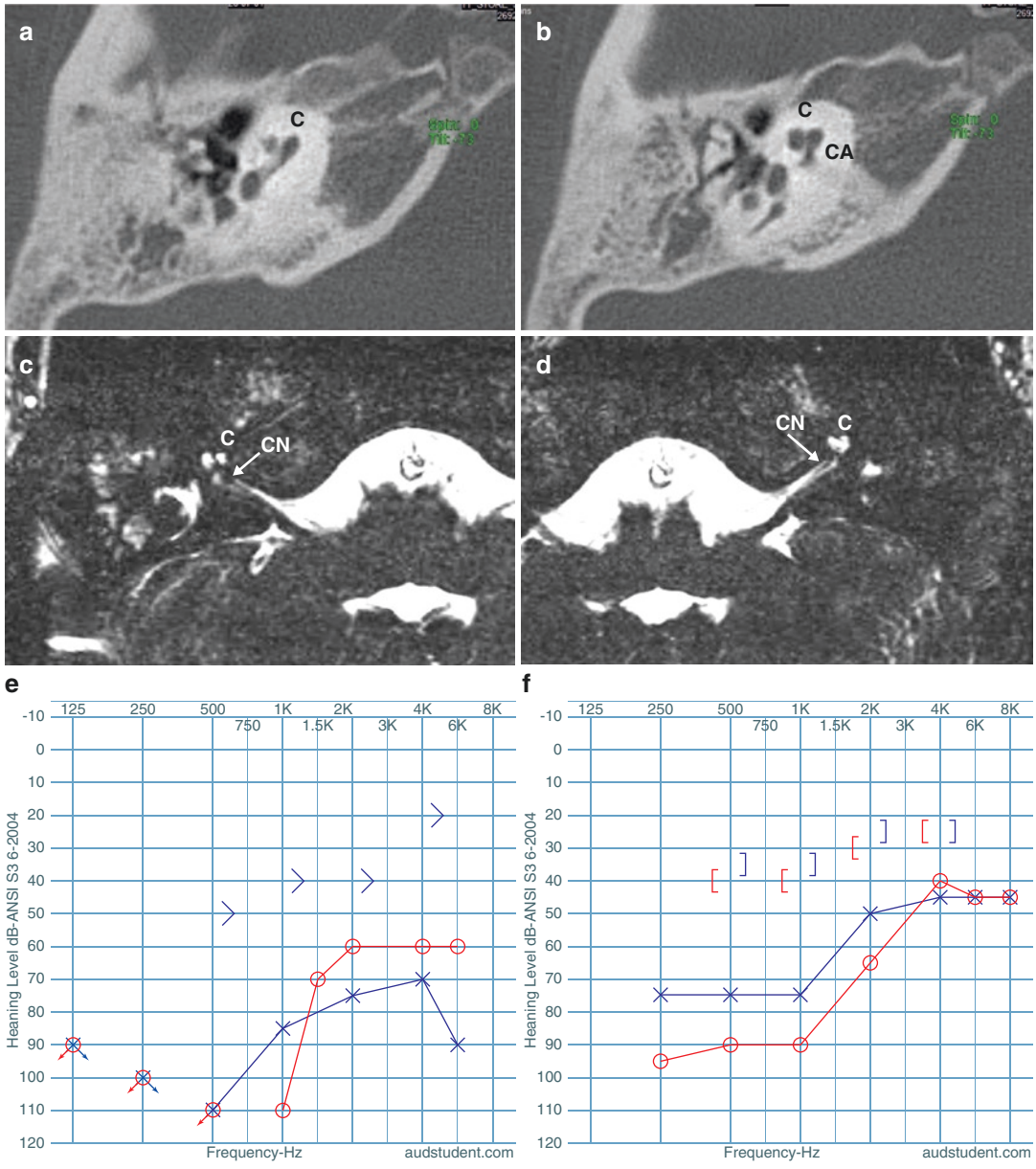


Fig. 14.6 Case 14.2. (**a**, **b**) Cochlear hypoplasia type III (C) consisting of hypoplastic basal turn and extremely small apical turn. Please note cochlear aperture stenosis. Axial sections showed cochlear nerve (CN) on right (**c**) and left (**d**) sides. (**e**, **f**) Preoperative (**e**) and postoperative (**f**) hearing thresholds

Cochlear nerve was present bilaterally (Fig. 14.6c, d). Due to developmental delay, testing was very difficult. Therefore, ears were tested at different sessions through insert earphones. First preoperative audiogram was done when she was 4 years old. She had rising audiogram and average air conduction thresholds (500–1000–2000–4000 Hz) were 85 dB for left and 89 dB for right ear. Average bone conduction threshold was 37.5 dB (masked bone conduction thresholds did not get). ABG was 47.5 dB for left and 51.50 for right ear (Fig. 14.6e).

On both sides she had all ossicles fixed. Left side was operated on 29 January 2011 and right

side was operated on 29 April 2014. In both operations, atticotomy and mobilization of the malleus and incus were performed before stapedotomy. A 0.6 mm stapes piston with 4.5 mm length was used on both sides. On the left side, the facial nerve was covering almost 75% of the footplate.

In her latest postoperative audiogram, air conduction hearing thresholds showed improvement. She had moderately severe mixed HL on the left ear and severe mixed HL on the right side with rising configuration. ABG also decreased from 47.5 to 31.25 for left ear and from 51.50 to 37.5 for right ear (Fig. 14.6f). She uses her hearing aids regularly and thresholds with hearing aids were between 35 and 20 dB. She made much better use of hearing aid after bilateral stapedotomy. Both her receptive and expressive language were significantly poorer than peers. Although she had better performance at closed-set tests (pattern perception and word identification), she performed poorly at open-set test (sentence recognition).

Case 14.3: (Patient #1) TU, 16-Year-Old Male Patient

His left ear was explored in 2008. Facial nerve was located at the oval window (Fig. 14.3b). Using a drill, a vestibulotomy was performed inferior to facial nerve. Using glass ionomer cement, incus was extended towards the opening in the vestibule, and a piston was attached between this extension and the vestibulotomy.

Surgery of Case 14.3 was difficult. We have to use a drill to make a fenestra. This may cause SNHL. Position of incus was over the FN. Therefore, cement was used to extend incus for attachment of the piston towards the vestibulotomy.

Preoperatively he had severe mixed type HL on the left and moderately severe mixed type HL on the right side with ABG of 50 and 41 dB (Fig. 14.7a). On the left side, there was a notch around 2000 Hz. He was operated on left side and his hearing thresholds were improved and ABG was decreased (Fig. 14.7b).

Case 14.4: HG 8-Year-Old Female Patient

She applied with bilateral moderate CHL. She had been using hearing aids until that time. In both ears, stapes was absent, and incus position was more oblique when compared to normal otosclerosis cases. Oval window was atretic bilaterally. It was not possible to make the fenestra using a perforator, necessitating the use of a 0.6 mm diamond drill to make the fenestra. Insertion of the hook of the stapes piston onto incus was more difficult when compared to nor-

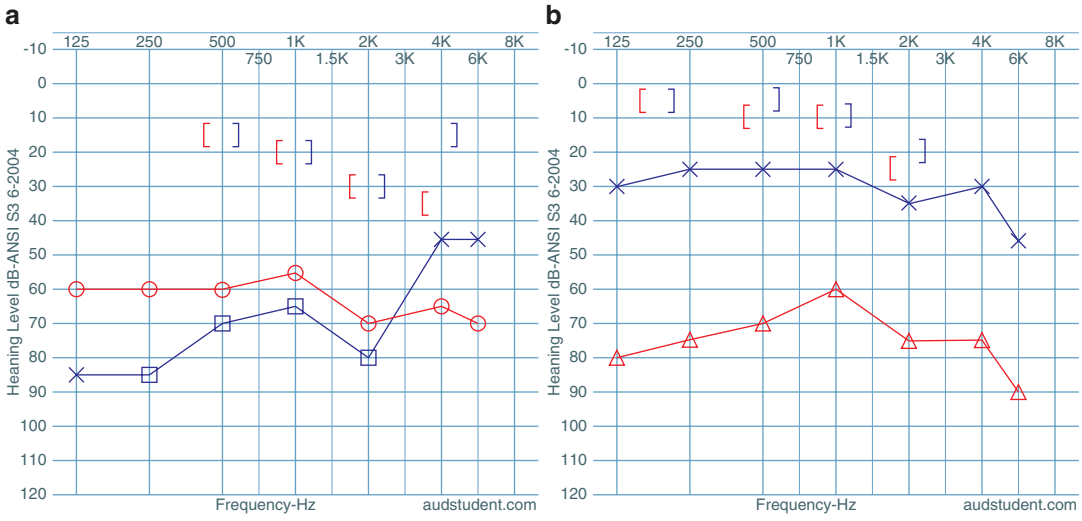


Fig. 14.7 Case 14.3 (**a**, **b**) Preoperative (**a**) and postoperative (**b**) hearing thresholds

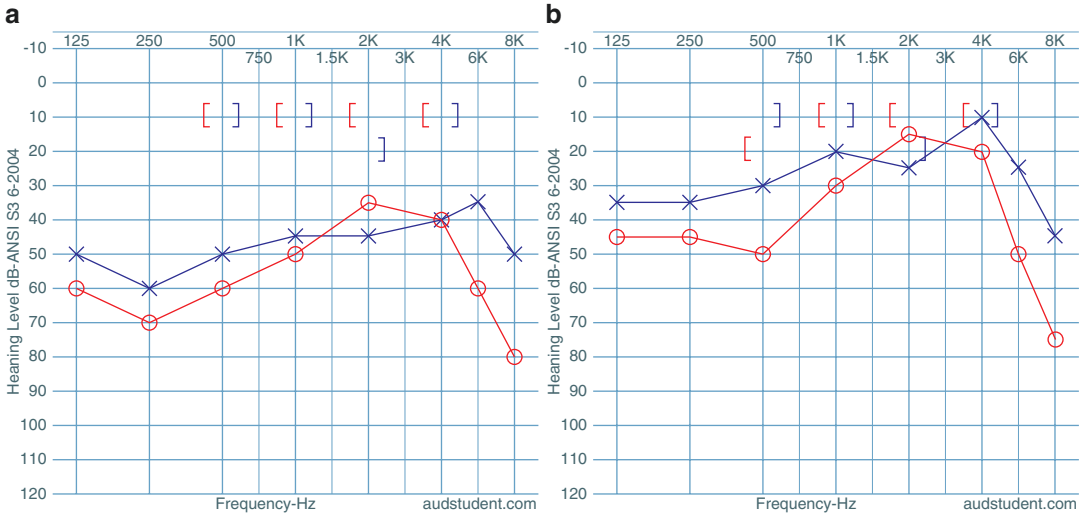


Fig. 14.8 Case 14.4 (**a**, **b**) Preoperative (**a**) and postoperative (**b**) hearing thresholds

mal stapedotomy, where the incus is more horizontal. Two drops of glass ionomer cement were used to fix the prosthesis to avoid sliding inferiorly.

Her preoperative audiogram showed bilateral moderate CHL with ABG of 34 dB on left ear and 38 dB on right ear (Fig. 14.8a). Although conductive component remains, postoperative audiogram showed bilateral improvement on air conduction thresholds. On left ear there was slight CHL with 12 dB ABG and on the right ear there was mild CHL with 19 dB ABG (Fig. 14.8b).

14.12 Outcome

Preoperative and postoperative hearing levels can be seen in Table 14.1. The average preoperative ABG was 46 dB and postoperative ABG was 21 dB.

In cases with pure CHL, it is possible to close the ABG and have near normal hearing without hearing aid. In children with mixed HL, the goal is to close the ABG as much as possible and allow the child to benefit more from the hearing aid. The latter should be made clear to the family when obtaining informed consent. In spite of tremendous improvement in all cases except patient #2 (Table 14.1), the results are not as good as stapedotomy in otosclerosis. This is due to the additional severe anatomical abnormalities.

14.13 Conclusion

Congenital stapes fixation and oval window abnormalities can be seen in inner ear malformations particularly accompanying cochlear hypoplasia. Stapedotomy is an acceptable treatment option as it provides sufficient hearing gain postoperatively. In mixed hearing loss the aim of the operation is to provide better benefit from hearing aid. The surgery may be complicated as a result of fixation of other ossicles, facial nerve abnormality, and CSF leakage. In the light of the findings of the present study stapedotomy has to be an option in conductive or mixed hearing loss in cochlear hypoplasia among other treatment alternatives.

References

1. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
2. Donaldson JA, Lambert PM, Duckert LG, Rubel EW. Surgical anatomy of the temporal bone. New York: Raven Press; 1992.

3. Sennaroglu L, et al. Manubrio-stapedioplasty: new surgical technique for malleus and incus fixation due to tympanosclerosis. J Laryngol Otol. 2015;129(6):587–90.
4. Sennaroglu L, et al. Oval window atresia: a novel surgical approach and pathognomonic radiological finding. Int J Pediatr Otorhinolaryngol. 2014;78(5):769–76.
5. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
6. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.

CSF Leakage and Its Management

15

Levent Sennaroglu and Munir Demir Bajin

Special Features

1. Gusher is the result of defect between IAC and cochlea; if not controlled completely, it may lead to recurrent meningitis.
2. Gusher occurs 100% in IP-III, 48% in IP-I, and 6.5% in IP-II.
3. Most important step is to control the point of leakage properly. Subtotal petrosectomy can be done once the leakage has been controlled.

15.1 Definition

Cerebrospinal fluid (CSF) leakage is common during CI surgery in inner ear malformations (IEM) while making the cochleostomy. This is a very critical situation which may lead to recurrent meningitis and death. In his review article Sennaroglu [1] highlighted the importance of proper control of CSF leakage during CI surgery in IEMs. The term "gusher" is generally used in the literature to describe the egress of profuse CSF upon making an opening into the inner ear. This can also be encountered during stapedotomy in patients with IEMs (particularly in IP-III). In the past, the term "perilymphatic fistula" was used to describe this situation. Today "perilymphatic fistula" is accepted as a misnomer for this situation. As pointed out by Janssens et al. [2], in cases of gusher, the fluid coming from the inner ear cannot be perilymph, as the inner ear contains only a few microliters of perilymph. The profuse fluid coming from the inner ear is CSF and is due to a defect between the malformed cochlea and internal auditory canal (IAC). Depending on the size of the defect, the magnitude of CSF leakage may vary.

15.2 Incidence

In 1986 Miyamoto et al. [3] made the first report of gusher during CI surgery in IEMs, which took place during cochleostomy in an ear with Mondini deformity. When the CT image in that paper is examined, the pathology appears to be a "common cavity" rather than "Mondini deformity." Later Silverstein et al. [4] reported the first multichannel cochlear implantation in a true Mondini deformity. CSF leakage in that patient can be described as "oozing."

After these initial case reports, in 1990 results of patient series started to appear in the literature. Frequency of gusher in IEMs shows great variation in these papers. In their questionnaire,

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_15].

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

M. D. Bajin (✉)
Department of Otolaryngology, Hacettepe University, Medical Faculty, Ankara, Turkey
e-mail: dbajin@hacettepe.edu.tr

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_15

Hoffman et al. [5] found that 40% of the patients with malformations had CSF gusher regardless of the type of malformation. Majority of the papers (Tucci et al. [6], Slattery et al. [7], Luntz et al. [8], Sennaroglu [1], Weber et al. [9], Eisenman et al. [10], Loundon et al. [11]) reported the incidence of gusher to be between 40 and 50% of their patients with IEMs.

In 2005 Papsin [12] made an extensive case review and reported that the gusher was present in 6.7% of 103 patients with malformations. He stated that it is very important to differentiate between "oozing" and "gusher." He stressed the point that the term gusher should be used only to describe the pulsatile egress of fluid that lasts for up to a minute and then subsides in order not to overestimate the incidence of gusher. He also reported that inclusion of minor leaks may lead to an overestimate of the number of CSF leaks and may help explain the huge range in incidence of leaks reported.

Wootten et al. [13] reported the lowest frequency of gusher, occurring in approximately 1% of patients undergoing cochlear implant surgery in IEMs. It was seen in equal incidence in children and adults in their series. Preoperative imaging had been predictive in only 50% of their cases.

Hoffman et al. [5] found that the incidence of gusher was almost similar regardless of the type of malformation: 50% of common cavity deformities, 40% of incomplete partition, hypoplasia and EVA cases had CSF gusher during the surgery. Loundon et al. [11] reported gusher according to the type of malformation. They noted that gusher was present in 50% of cases (9/18), with 11% having persistent leak (1/9), which resolved after 3 days with medical treatment. The type of malformation in those cases was an EVA in 33% of cases (3/9), a complex cochleovestibular malformation in 55% of cases (5/9), and a common cavity in 11% of case (1/9).

15.3 Types of CSF Leakage

As pointed out by Graham et al. [14] gusher of CSF is the result of an abnormal bony defect at the lateral end of the IAC. Normally, CSF in the subarachnoid space extends laterally into the IAC as far as the fundus, where it is separated from the perilymph by the bony plate of the lamina cribrosa. In some IEMs there is a defect at the lateral end of the IAC, allowing direct confluence of CSF and perilymph.

Three types of CSF related abnormalities may be encountered during CI surgery in IEMs:

1. **CSF Pressure Pulsation:** This is mostly encountered in IP-II. EVA is a component of this malformation and provides a route for CSF pressure transfer into the inner ear. Before making an incision through the round window membrane, we may observe a pulsation in the membrane. However, upon opening the round window, there is a pulsation in the fluid in the scala tympani (Video 15.1). If the fluid is removed by suction, there is no CSF leakage; neither oozing nor gusher. If we place some fluid such as corticosteroids in the cochleostomy, pulsation is observed. The correct terminology should be "CSF pressure pulsation." In these cases, there is no direct connection of the scala tympani and subarachnoid space.
2. **Oozing:** This is the mild intermittent outflow of clear fluid during cochleostomy [15]. Oozing is the result of a small defect between the malformed inner ear and the IAC. In our practice oozing is intermittent flow of CSF in small quantities which usually stops after a few minutes (Video 15.2). The defect between the IAC and the malformed ear is small and the CSF outflow is easily controlled with soft tissue packing around the electrode. This type of CSF flow is more common in IP-II and EVA. This is compatible with a small direct communication between IAC and cochlea, of the kind found in Mondini deformity. As can be seen in Table 15.1 very rarely children with IP-I malformation had oozing. None of the IP-III cases had oozing.
3. **Gusher:** This is the profuse CSF outflow during cochleostomy. In case of gusher, there is a larger anatomic defect providing a wider communication between the subarachnoid space and the inner ear. In these patients there is profuse CSF outflow upon making the cochleostomy (Video 15.3). It usually lasts between 10 and

20 min. This is the most serious type of CSF leakage with more chance to cause postoperative meningitis. IP-III is the least frequent form of incomplete partition anomalies and cochleostomy produced CSF gusher in all IP-III cases [13, 16–18]. Gusher is observed in a lesser frequency in patients with IP-I and some patients with IP-II. Incesulu et al. [18] also reported CSF gusher in four patients with IP-III. In CC we have observed a CSF leakage very rarely in 2 out of 17 cases (12%) in the form of gusher. In addition, cochlear hypoplasia type II cases also had gusher during cochleostomy.

Between November 2007 and September 2018, 2646 patients underwent CI and ABI operations in the Department of Otolaryngology at Hacettepe University. Two hundred seventy-nine of CI cases had IEMs. One hundred eight (38.7%) had CSF leakage during cochleostomy (Table 15.1). Fifty-four (19%) were in the form of gusher, while 54 (19%) patients had oozing.

As can be seen the greatest frequency of gusher is in IP-III, where there is a 100% incidence of severe gusher. Gusher occurrence is 48% in IP-I and 6.5% in IP-II. Almost all cases of IP-II and EVA have pulsation during cochleostomy.

Not all cases with IEMs have gusher during the surgery. Sometimes in spite of a wide defect at the end of the IAC on imaging (HRCT and MRI), no gusher occurs upon entering the inner ear. It is interesting that none of the 15 patients with common cavity and none of the 24 patients with IP-I that were operated with a defect at the lateral part of IAC had CSF gusher. There are similar reports in the literature as well [19–22]. Most probably there is a fibrotic or thin bony separation between IAC and malformed cochlea not visible with present imaging modalities.

Graham and Ashcroft [23] measured the pressure and flow of a CSF gusher at cochleostomy, in a 4-year-old girl with bilateral Mondini deformity undergoing cochlear implantation. They used a size 23 FG intravenous cannula, which was inserted into the cochlea and connected to a pediatric drip set to form an improvised manometer. If a high pressure is measured with this method, the head of the table can be raised to decrease the pressure of the CSF. Another advantage was the indirect estimation of the size of the lamina cribrosa defect by estimating the duration of the CSF leakage until the meniscus in the manometer stabilizes. Therefore, larger lamina cribrosa defects are a greater risk for CSF leaks and hence meningitis. They concluded that this technique may allow better assessment of the risk of postoperative CSF leakage and meningitis.

Table 15.1 Frequency of gusher and oozing amongst the inner ear malformations

	Gusher	Oozing	No CSF
Common cavity	2	–	15
IP-I	24	2	24
IP-II	6	42	45
IP-III	13	–	–
EVA	–	8	22
Hypoplasia (type I)	1		2
Hypoplasia (type II)	5	–	12
Hypoplasia (type III)	1		24
Cochlear base defect	1	1	

15.4 Radiology

Types of IEMs that have a possibility of CSF leakage: IP-I (Fig. 15.1), IP-II (Fig. 15.2), IP-III (Fig. 15.3), CH-II (Fig. 15.4), CC (Fig. 15.5), and cochlear base defect (Fig. 15.6). Please note the defect between cochlea and IAC.

IP-I and CH-II may have an opacity on the oval window. Usually this may indicate a defective footplate with cyst which has a risk of gusher (Fig. 15.7). Fluid filling middle ear and mastoid in IP-I and CH-II may be CSF (Fig. 15.8).

Radiology demonstrates the defect at the lateral end of the IAC. High resolution computerized tomography is the best method to demonstrate the defect at the lateral end of the IAC. As reported by a number of authors [19–22, 24], not all patients with a defect shown on CT and MRI had CSF leakage during the CI surgery. They reported that no CSF gusher was found during cochlear implantation in common cavity patients in their series. We also have similar view that CSF leakage is very rare in CC. In spite of a wide defect between IAC and the inner ear, none of the 24 patients with IP-I and none of the 15

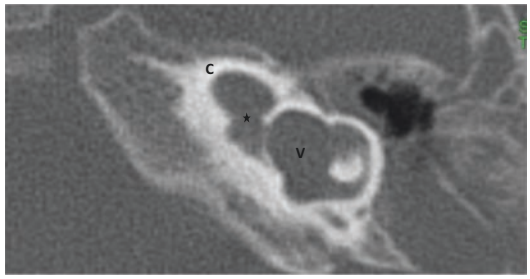


Fig. 15.1 Incomplete partition type I. Note the defect (black star) between cochlea (C) and internal auditory canal (*V* vestibule)

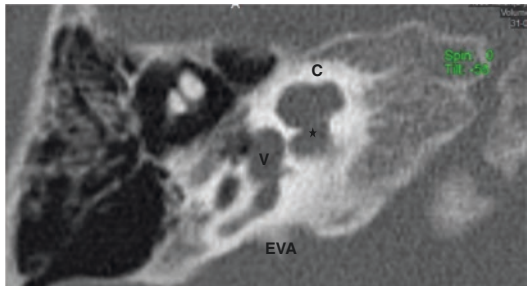


Fig. 15.2 Incomplete partition type II. Note the defect (black star) between cochlea (C) and internal auditory canal (V vestibule, *EVA* enlarged vestibular aqueduct)

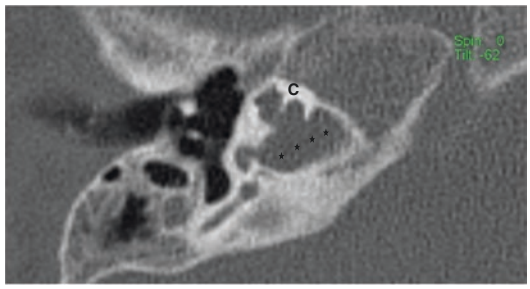


Fig. 15.3 Incomplete partition type III. Cochlea (C) has a defective base (black stars)

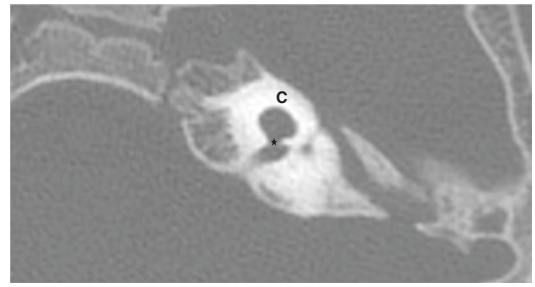


Fig. 15.4 Cochlear hypoplasia type II, with a defect (black star) between cochlea (C) and internal auditory canal

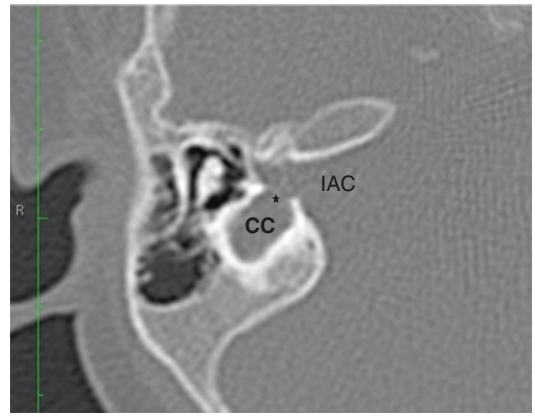


Fig. 15.5 Common cavity (CC) with a defect (black star) between CC and internal auditory canal (IAC)

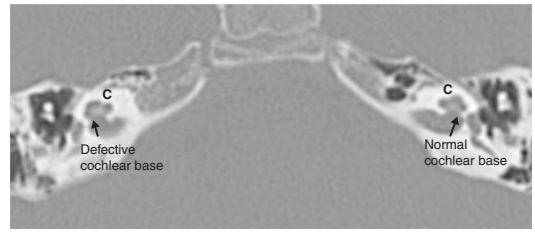


Fig. 15.6 Cochlear base defect on the right. Please note normal cochlear base on the left side

patients with common cavity had gusher or oozing during surgery.

Sometimes the pathology between IAC and the inner ear is subtle but the patient still has CSF gusher or oozing. As clearly demonstrated by Lammerling et al. [25], the CSF leakage in these patients is due to mild modiolar defects and during the operation oozing is observed. Only rarely these patients may have gusher as shown in six of our cases with IP-II. Recently Sennaroglu [26] classified modiolar defects according to histopathological findings. This will be more important in future when the precision of MRI and HRCT will be more detailed in demonstrating modiolar defects.

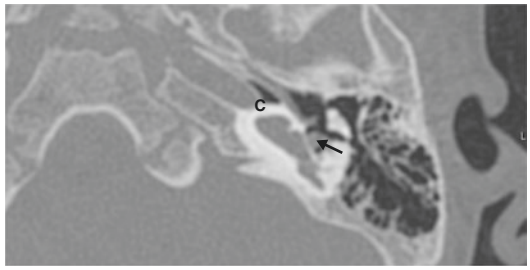


Fig. 15.7 Incomplete partition type I (C) with a soft tissue mass (black arrow) indicating a footplate cyst

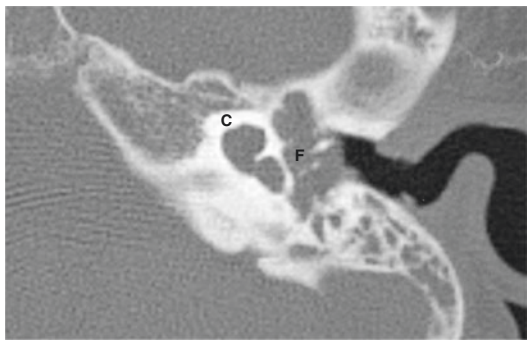


Fig. 15.8 Incomplete partition type I (C) with fluid (F) (cerebrospinal fluid) filling middle ear and mastoid

Fluid in the middle ear and mastoid is not rare in patients with SNHL. There may be two reasons for cases where HRCT or MRI demonstrates fluid in the middle ear and mastoid; otitis media with effusion or spontaneous CSF fistula. In the latter case tympanic membrane is not retracted and looks near normal. In case of IP-I or CH-II, possibility of footplate fistula should be kept in mind in a case of fluid filled middle ear and mastoid. If the patient already had an implant, CT is the only radiological modality to diagnose CSF fistula. If there is a strong possibility of CSF fistula CT-cisternography, which is an invasive method, is the only way to demonstrate the leakage.

In rare cases without the presence of any inner ear anomaly, cochlear base defect may cause gusher. It is always important to inspect the cochlear base for potential defects (Fig. 15.6) [27].

15.5 Treatment

Our management philosophy of CSF gusher during CI surgery changed considerably in the last two decades. Initial cases were managed with a small cochleostomy. We then started to perform slightly larger cochleostomy with occasional CLD. For the present time in case of severe gusher, we perform large cochleostomy, use FORM electrodes with silicon stopper (which is passed through a piece of fascia), always in combination with continuous lumbar drainage (CLD).

Surgeon should not leave operating theater without fully controlling the CSF leakage around the electrode. It is not a good surgical management strategy to rely on subtotal petrosectomy while the leakage continues around the electrode.

Most difficult patients are CSF leakage through the footplate and cochleostomy at the same time. Both windows have to be repaired before terminating the surgery**.**

It is very important to firmly pack a piece of fascia or muscle around the electrode lead at the level of the cochleostomy to prevent CSF fistula in the postoperative period. If there is no watertight seal, there is a risk for permanent CSF leakage with the potential risk of meningitis.

15.5.1 Size of the Cochleostomy

It is advisable to make the facial recess as large as possible. This is very important because severe gusher necessitates prolonged surgical manipulation at the cochleostomy. If the recess is not large, instruments like forceps or claws are difficult to introduce and perform various maneuvers around the electrode. Therefore, first important step is to make a large facial recess.

When we encounter a serious CSF leakage during cochleostomy, it is advisable to stop at this moment and not to use the drill until the CSF outflow decreases considerably. Initially, there is a severe CSF leakage and the surgeon cannot see

the cochleostomy area properly through the fluid and therefore cannot control the tip of the bur. After 10 min when CSF decreases, there is much better visibility of the round window area. We can enlarge the round window to desired size more efficiently.

We do not perform a separate cochleostomy. The reason is that the surgeon may open scala tympani or vestibuli. Our preferred method is to remove the posterior and superior part of the round window niche and enlarge the round window anteroinferiorly. This will remove the hook region and will allow the electrode to be inserted, without doubt, into the scala tympani. As pointed out, slightly larger opening allows the electrode and the fascia around it to be inserted together and more efficiently into the opening.

In 1998 Weber et al. [28] reported their experience in patients with IEMs and recommended a small cochleostomy, allowing the electrode cable to partially block the flow of CSF, reinforced with connective tissue, muscle, and fibrin glue. This was the method we had used initially in our department. Over time we have observed that it was very difficult to insert small pieces of fascia around the electrode inside the cochleostomy. Some of our patients developed rhinorrhea, which necessitated a continuous lumbar drainage (CLD) for 4–5 days. Graham et al. [14] suggested a large cochleostomy for the control of CSF leakage. This has been the method used in our department because it allowed easier insertion of the electrode and more effective insertion of fascia around the electrode. Proper application of muscle tissue around the cochleostomy represents the first line of barrier against CSF leakage and the flow of CSF stops quickly when a proper seal has been established. It is advisable to use fibrin glue at this stage. Papsin [12] also reported the importance of a slightly larger cochleostomy to allow extra mobility with the instruments to pack the cochleostomy. When a CSF leak is encountered, they pack the cochleostomy more tightly than usual and also use fibrin glue to strengthen the seal.

Size of the cochleostomy: This should be tailored according to the type of the electrode used. Active intracochlear part of the FORM electrode is 0.8 mm. End of the stopper is 1.9 mm. Therefore, using a 1.2 mm diamond drill round window is enlarged anteroinferiorly to a size about 1.3 mm. This will allow the passage of the active electrode but not the silicon "cork" stopper. The stopper will squeeze the fascia into the opening.

15.5.2 FORM Electrode with "Cork" Stopper

This is now routinely used in patients with CSF leakage, oozing or gusher. The electrode has a "cork" feature which is a progressive conical stopper at the level of the silicon ring which marks the end of insertion which will effectively block the cochleostomy preventing CSF leakage [29]. The length of the electrode has to be determined according to the type of IEM. Ideally it has to make one full turn around the cochlea.

There are two types [29]:

1. FORM 24 Standard electrode with "cork" stopper: The length of the standard electrode is 25 mm. The contact spacing between active electrodes is 1.7 mm. This is preferred in cochleae such as IP-I, IP-II, and IP-III and in large vestibular aqueduct patients where the outer dimensions of the cochlea are similar to normal. It is also preferred in patients with a large common cavity.
2. FORM 19 Short electrode with "cork" stopper: The length of the shorter electrode is 20 mm. The contact spacing is 1.3 mm. This is preferred in hypoplastic cochlea and in patients with a small common cavity.

In cases where the modiolus is missing (IP-I, IP-III, and common cavity), the exact location of the neural tissue is unknown. Modiolar hugging electrodes may have a disadvantage in this respect. FORM electrode has contact surface on both sides of the electrode array to stimulate the neural tissue present in the anomalous cochlea more effectively. An electrode with full band contacts may also be used efficiently to stimulate the neural tissue.

As already indicated, the diameter of the active part of the electrode is 0.8 mm. The diameter of the cochleostomy must be 1.3 mm in order to allow smooth passage of the active electrode but not the cork stopper. At this stage, if the electrode is passed through a tiny piece of fascia, which covers it circumferentially and positioned at the cochleostomy, it is possible to have an even better seal (Fig. 15.9). It is important to use the tissue glue after each layer of soft tissue is added. Similar to the "first line of defense," the first layer of the soft tissue, which is prepared by perforating a tiny piece of muscle with a sharp pick and passing the electrode through it, is the most important. After the cork stopper is firmly placed into the cochleostomy, a small amount of tissue glue is added to fix this part. Then a thin layer of fascia and a tiny amount of glue is placed to obtain a second layer of barrier. This is continued until the opening around the electrode is properly closed and no CSF leakage is witnessed.

If there is no gusher an electrode with contacts on both sides (Med El) or with full rings (Nucleus 24 k CI24R(ST), Oticon Digisonic EVO, or Classic) can also be used.

Between 2000 and 2008, before using FORM electrodes, severe gusher occurred in 12 patients and 6 of these had postoperative rhinorrhea (50%). Between 2008 and 2012, 16 patients had severe gusher. We have started to use FORM electrodes in this period and only one case had postoperative rhinorrhea. Therefore, there is a dramatic decrease in the postoperative rhinorrhea after using FORM electrodes.

The length of the electrode is particularly important in cochlear hypoplasia [30]. In cochlear hypoplasia thin and short electrode should be preferred. The sizes of the cochlea and scala are smaller than normal cases. If a standard long electrode is used it may not be possible to insert the electrode until the stopper.

Modiolar hugging electrodes should not be used in IP-III and IP-I where there is a defect in modiolus. There is a risk of migration into IAC. The risk of electrode migration into IAC is higher in IP-III. First author has seen many cases where modiolar hugging electrode has migrated into IAC in IP-III. Only one such case was encountered in IP-I. In such a case, electrode removal carries the risk of damage to cochlear and facial nerve.

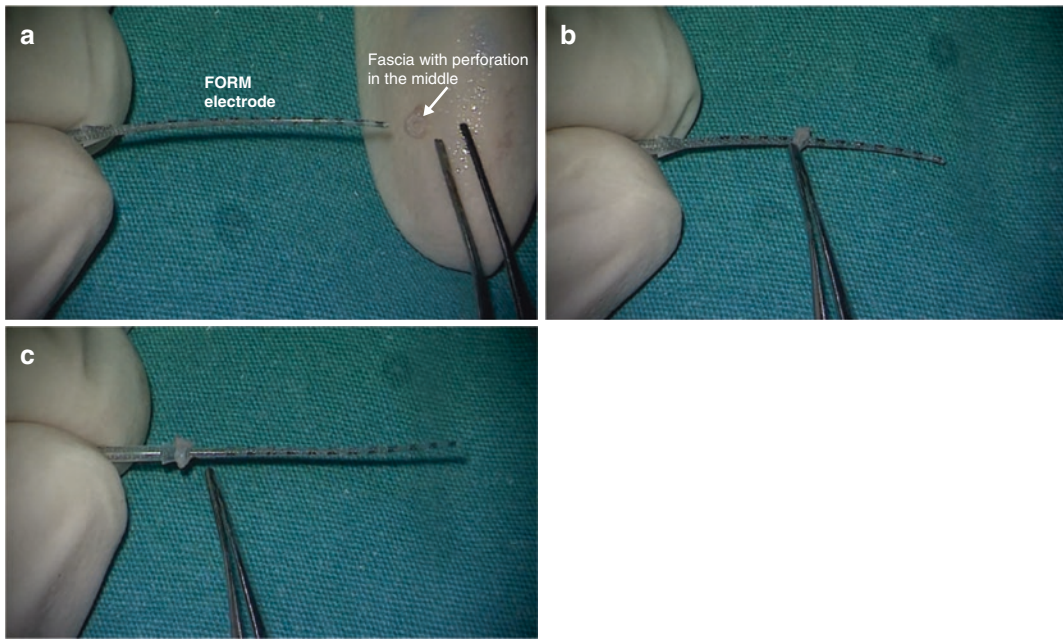


Fig. 15.9 (**a**–**c**) Electrode is passed through a tiny fascia, which is advanced until the stopper of the FORM electrode

15.5.3 Timing of Electrode Insertion

Graham et al. [14] indicated that a CSF gusher increases the technical difficulty of the operation, since refraction through the clear fluid makes it less easy to visualize the electrode as it passes through the cochleostomy. At the same time the surgeon may aim the electrode towards a bony structure and the tip of the electrode may be damaged [31]. Elevating the head of the operating table reduces the flow from the gusher. Because of this it is our practice to wait until the gusher slows down considerably before attempting the electrode insertion. Usually this takes about 10–15 min and at the end, CSF starts to pulsate. This is the appropriate time to insert the electrode. Wootten et al. [13] suggest to place the patient in reverse Trendelenburg position to slow the flow of CSF. Waiting for adequate drainage will aid in visualization and help to ensure successful packing.

Hoffman et al. [5] stated that initial management of CSF gusher is to allow the CSF to flow until it almost finishes and then firmly pack the cochleostomy with soft tissue. This was successful in 16 of 21 patients. In four patients with uncontrollable or recurrent CSF leakage continuous lumbar drainage was necessary.

15.5.4 Use of Fascia Around the Electrode

Application of fascia is important in cases of gusher. Fascia or muscle can be applied as small pieces separately around the electrode. Another way is to harvest a small round piece of fascia, about 2 mm in diameter. The fascia is perforated by a sharp needle and then the electrode is passed through a hole made in the center of the fascia block (Fig. 15.9). The electrode is then inserted with the fascia around it until the silicon stopper reaches the level of cochleostomy. The silicon stopper then squeezes the fascia into the opening and fills the gap of irregularities around the electrode. This will ensure a complete layer of soft tissue around the electrode at the level of cochleostomy.

Firm packing around the electrode is important in patients with gusher. CSF leak may recur after months and may cause partial electrode extrusion. One of our IP-II cases had slow extrusion of the electrode out of the cochlea. There was no CSF leakage but a very strong pulsation. Most probably CSF pulsation was the cause of electrode migration. A case reported by Hoffman et al. [5] had partially extruded electrode with gusher 5 months after the initial surgery. Luntz et al. [8] also described a patient who had a slowly progressive extrusion of all but three electrodes and required reinsertion. They suggested the use of the split bridge technique to prevent extrusion as proposed by Balkany and Telischi [32]. These cases show us that fixation of the electrode array is especially important in cases where there is a risk of profuse CSF leakage, since extrusion of the electrode from the cochlea could pull the seal out of the cochleostomy.

15.5.5 Eustachian Tube Obliteration

After sealing the cochleostomy, Eustachian tube may be temporarily blocked to prevent escape of the CSF into nasopharynx. Oxidized cellulose is the material usually preferred for this purpose. Luntz et al. [8] reported five cases of gusher out of ten various IEMs. Four of them could be controlled easily. One patient required packing of the cochleostomy, vestibule, and mesotympanum with a composite muscle-fascia plug and temporary obstruction of the Eustachian tube with oxidized cellulose for control of the gusher.

15.5.6 Subtotal Petrosectomy

After fully controlling the gusher subtotal petrosectomy can be done in addition to obtain an additional barrier. This should not be seen as a procedure to be done in cases of persistent leak. It is the duty of the surgeon to stop the leakage completely before terminating the surgery. Subtotal petrosectomy can be done as an additional safety procedure. In Hacettepe University this was done in a case of repetitive CSF fistula

through the stapes footplate. In addition, the primary author has seen patients who had CSF coming through the wound or experiencing recurrent meningitis after subtotal petrosectomy. Therefore, the most important factor is to close the cochleostomy effectively.

Subtotal petrosectomy has been used in cases of intractable CSF leakage for more secure sealing of the leak [9, 14, 20, 33]. Bendet et al. [33] have found that subtotal petrosectomy with isolation and obliteration of the tympanomastoid cleft can be used for safe cochlear implantation in patients with an exposed subarachnoid space. They reported a case of Mondini deformity with bilateral CSF fistula at both oval windows. Conservative measures at closing the leak failed in their case and they used bilateral subtotal petrosectomy to close the CSF leak and isolate the inner ear from potential external infections without the need to plug the vestibule with a muscle graft, therefore preserving remaining sensorineural elements.

Weber et al. [9] used the same technique in two implanted children with cochlear dysplasia who had had CSF fistula and meningitis before their cochlear implant surgery. The benefit of subtotal petrosectomy in cochlear implantation is to seal oval window as the site of spontaneous CSF leak besides the cochleostomy site.

Hoffman [5] reported that four patients with uncontrollable or recurrent CSF leakage continuous lumbar drainage became necessary. The authors mentioned that as the continuous lumbar drainage may be a difficult procedure in a child, subtotal petrosectomy should be considered as an alternative procedure.

Saeed et al. [34] reported one case in a two-case series where they planned staged procedures, with obliteration of the middle ear cleft and external ear canal (EAC) at the time of implantation.

This operation has important steps:

1. Lateral part of the skin of the external meatus is everted and closed as a blind sac,
2. Medial part of the skin of the ear canal and the tympanic membrane is removed completely,
3. Canal wall is taken down, and the facial canal, oval, and round windows are exposed,
4. Eustachian tube is blocked, and.
5. Mastoid cavity is obliterated with abdominal fat.

15.5.7 Continuous Lumbar Drainage (CLD)

In the past it CLD was performed if the gusher cannot be controlled effectively during surgery or the child develops rhinorrhea after the surgery.

Over the years there has been a change in our practice and we started to use CLD routinely in all cases of severe gusher. The purpose of the CLD is to divert CSF to a different location and lower its pressure around the cochleostomy so that area around the electrode can start to heal effectively. We should not forget that high CSF has the potential to dislodge the fascia pieces that were placed around the electrode. With CLD, CSF is removed three times a day resulting in less pressure. As the CSF pressure decreases because of the CLD, CSF will exert much less pressure on the cochleostomy area. The function of CLD is to divert CSF to a different area and allow some time for the organization of the soft tissues around the electrode. Typically, it is kept in place for 4–5 days. CLD is definitely not necessary in cases of oozing or pulsation.

It is advisable to inform the family before operation about this procedure. This has not resulted in a meningitis or any other complications in our patients.

In their literature review, Hoffman et al. [5] found eight cases where CSF gusher occurred. The leak was controlled by packing the cochleostomy site in six, but two patients required postoperative CLD to stop CSF leak.

Eisenman et al. [10] found that outflow of CSF from the cochleostomy site was encountered intraoperatively in 7 of 17 children (41%). This method failed in only one patient who had had a profuse leak during the operation and the delayed CSF otorrhea was successfully managed with 48 h of bed rest, a lumbar drain, and oral acetazolamide.

The use of intraoperative CLD was also reported. Tucci et al. [6] experienced three cases of gusher where they placed a spinal drain intraoperatively to effectively control the gusher. The drain was left in place for 4 days postoperatively. Similarly, Syal et al. [35] reported that they used intraoperatively continuous lumbar drainage which continued postoperatively for 7 days in four cases.

15.5.8 Additional Measures

Head elevation and 48 hours of bed rest decrease the CSF leakage. In addition, acetazolamide can be used to decrease CSF production.

Kim et al. [36] reported that in a child with profuse CSF gusher they could not proceed with the surgery. They elevated the head of the child and infused mannitol, to lower the intracranial pressure. When the gusher decreased, they could then insert the electrodes and seal the cochleostomy completely.

Wootten et al. [13] also suggested that the patients should not strain or lift greater than 20 pounds for 2–3 weeks after surgery and to keep their head-of-bed elevated 30° for the same interval. Compression-style head dressings were used for the first 24–72 h. Meticulous three-layer closure has to be performed in these cases with attention to a watertight closure.

15.5.9 Intraoperative Radiology

Electrode position should be checked in every case after CI surgery. This can be done in the afternoon or the evening of the surgery. There are certain situations where the electrode position should be checked intraoperatively. Sennaroglu [1] indicated that a gusher during the surgery of IEMs indicates a wide connection between the subarachnoid space and the inner ear. In these cases, electrode migration into IAC is possible and conventional radiology should be done **intraoperatively** to check the position of the electrode array. Normally the electrode takes the shape of the cochlea or the common cavity and we see the round shape of the electrode according to the dimensions of the cochlea. If the electrode takes a horizontal shape it shows us that the electrode may be in the IAC. In this case the electrode should be repositioned. If left in the IAC, electrode stimulation may cause side effects such as facial and vestibular stimulation. In addition, these electrodes will have to be closed and the benefit from CI will decrease. When Table 15.1 is examined, out of the 108 cases with CSF leakage only 1 case had electrode migration into IAC. This was a case of IP-III. As we have used a straight electrode it was easily repositioned intraoperatively (see Case 25.2 in Chap. 25 **Incomplete Partition Type III).** Another case with IP-III was referred with migration of the electrode into IAC. As modiolar hugging electrode had been used in previous surgery that particular electrode was not removed (Case 25.1 Chap. 25 **Incomplete Partition Type III**). Electrode migration has been observed only once in an IP-I operated in another center (again with modiolar hugging electrode). This never happened in IP-II cases most probably due to the fact that the defect between the cochlea and IAC is smaller. All other cases with CSF gusher or oozing had intracochlear position.

Intraoperative X-ray provides immediate diagnosis of the situation where repositioning of the electrode can be done immediately. If this is not done intraoperatively or in the immediate postoperative period, a revision surgery carries the risk of electrode damage if fibrosis develops around the electrode.

Electrode position should be checked in other situations as well. Graham et al. [14] indicated that in common cavity and cochlear hypoplasia where the correct placement of the electrode cannot be predicted intraoperatively, X-ray is essential. In common cavity in case of CSF gusher there is a risk of the electrode entry into IAC. Even if there is no gusher electrode position should be checked in common cavity. Electrode position is usually unpredictable and intraoperative X-ray is indicated. Cochlear hypoplasia cases may have electrode positioning problems.

Tucci et al. [6] reported two cases of cochlear hypoplasia, where the electrode entered the IAC. Therefore, the same protocol should be done for hypoplasia cases as well. In both of their patients with cochlear hypoplasia, the electrode array was demonstrated with imaging to course through the cochlea into the IAC. They thought that the size of the hypoplastic cochlea did not allow the electrode to curl within the cavity.

Wootten et al. [13] reported a patient with IP-III cochlea where there was a gusher during surgery and the electrode inserted went into the IAC up to the cerebellopontine angle. He was returned to the operating room on postoperative day 2 for electrode repositioning. The prior implant site was sealed without CSF leakage. A complete drill out of the hypoplastic cochlea allowed for specific electrode placement. The external ear canal was left intact. The Eustachian tube was packed with fascia and muscle.

Eisenman et al. [10] however suggested to leave the electrode in place and carefully modify the stimulus levels and the number of active electrodes in an effort to avoid overstimulation of cochlear elements or undesired facial nerve stimulation if the electrode is noted to be in the internal auditory canal on postoperative radiographs.

On the contrary, Copeland et al. [37] found that in severely malformed inner ears, the utility of the plain radiograph is questionable. Of the five malformed inner ears implanted during their study two required CT despite an adequate intraoperative plain radiograph. On the basis of their findings, they made an argument that the CT scan is the study of choice for evaluating cochlear implants in the malformed inner ear when questions arise as to electrode placement because the plain radiograph, again, did not alter management decisions. As can be understood their plain radiographs in those two patients appeared to be sufficient. They concluded that plain radiographs obtained intraoperatively should be reserved for unusual or rare circumstances.

It should be kept in mind that, if the intraoperative radiology is done very rarely, the staff in the operation theater will not be experienced in positioning the patient and other details, so that in difficult cases satisfactory images cannot be obtained. Therefore, it may be a better option to include intraoperative plain radiograph in the surgical theater as a routine part of the surgery in IEMs. Timing is also very important. While plain radiograph is available within minutes after the end of the operation, postoperative CT is usually obtained late (usually the day after) to have an idea about the electrode placement. Therefore, in case of X-ray in the operation theater, a decision for revision will be made immediately after the surgery. Therefore, intraoperative plain radiograph appears to have certain advantages over postoperative CT besides radiation dose.

Fishman et al. [24] suggested the use of fluoroscopic assistance when the intracochlear behavior of the electrode array cannot be predicted. In the case of severely malformed inner ears, there is an increased chance of complications, such as extracochlear array placement, intrameatal array insertion, or kinking or bending of the electrodes. Although we do not use this method, it is advisable to be used in IP-III, IP-I, CC, and CH-II cases. Insertional trauma to the delicate structures can be minimized by avoiding the application of pressure to the electrode after significant resistance to advancement occurs. Insertion end point determination can be precisely defined using fluoroscopy, thereby avoiding both electrode and structural damage. They argued that pushing more electrodes into a common cavity is not necessarily better if excessive damage will occur to the outer wall of the cavity. They preferred a straight array with concentric bands so that outer wall electrode contact may be achieved in these small spherical and ovoid cavities.

15.6 Postoperative Radiology

In patients with IP-II, EVA, CH-III, and CH-IV, the likelihood of the electrode going into the IAC is very low. Therefore, X-ray image can be obtained after the surgery.

15.7 Conclusion

CSF gusher is a challenge to the surgeon. The leak has to be stopped completely before terminating the procedure. A large cochleostomy with FORM electrode passed through a piece of fascia appears to be a satisfactory option to control gusher. In case of severe gusher, a CLD at the end of the surgery decreases CSF pressure and allows the soft tissue packing to stay in place. If there is a gusher, intraoperative X-ray is indicated. Subtotal petrosectomy can be done as an additional precaution only after full control of the leak by the cochleostomy sight in cases of repeated CSF leaks.

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Janssens S, et al. The LAURA multichannel cochlear implant in a true Mondini dysplasia. Eur Arch Otorhinolaryngol. 1996;253(4–5):301–4.
3. Miyamoto RT, et al. Cochlear implantation in the Mondini inner ear malformation. Am J Otol. 1986;7(4):258–61.
4. Silverstein H, Smouha E, Morgan N. Multichannel cochlear implantation in a patient with bilateral Mondini deformities. Am J Otol. 1988;9(6):451–5.
5. Hoffman RA, et al. Cochlear implantation in children with cochlear malformations. Am J Otol. 1997;18(2):184–7.
6. Tucci DL, et al. Cochlear implantation in patients with cochlear malformations. Arch Otolaryngol Head Neck Surg. 1995;121(8):833–8.
7. Slattery WH 3rd, Luxford WM. Cochlear implantation in the congenital malformed cochlea. Laryngoscope. 1995;105(11):1184–7.
8. Luntz M, et al. Cochlear implants in children with congenital inner ear malformations. Arch Otolaryngol Head Neck Surg. 1997;123(9):974–7.
9. Weber BP, et al. Pediatric cochlear implantation in cochlear malformations. Am J Otol. 1998;19(6):747–53.
10. Eisenman DJ, et al. Implantation of the malformed cochlea. Otol Neurotol. 2001;22(6):834–41.
11. Loundon N, et al. Cochlear implantation in children with internal ear malformations. Otol Neurotol. 2005;26(4):668–73.
12. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(1 Pt 2 Suppl 106):1–26.
13. Wootten CT, Backous DD, Haynes DS. Management of cerebrospinal fluid leakage from cochleostomy during cochlear implant surgery. Laryngoscope. 2006;116(11):2055–9.
14. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. J Laryngol Otol Suppl. 2000;25:1–14.
15. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
16. Sennaroglu L, Bajin MD. Incomplete partition type III: a rare and difficult cochlear implant surgical indication. Auris Nasus Larynx. 2018;45(1):26–32.
17. Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27(5):615–23.
18. Incesulu A, Adapinar B, Kecik C. Cochlear implantation in cases with incomplete partition type III (X-linked anomaly). Eur Arch Otorhinolaryngol. 2008;265(11):1425–30.
19. McElveen JT Jr, et al. Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. Laryngoscope. 1997;107(8):1032–6.
20. Mylanus EA, Rotteveel LJ, Leeuw RL. Congenital malformation of the inner ear and pediatric cochlear implantation. Otol Neurotol. 2004;25(3):308–17.
21. Manolidis S, Tonini R, Spitzer J. Endoscopically guided placement of prefabricated cochlear implant electrodes in a common cavity malformation. Int J Pediatr Otorhinolaryngol. 2006;70(4):591–6.
22. Beltrame MA, et al. Double posterior labyrinthotomy technique: results in three Med-El patients with common cavity. Otol Neurotol. 2005;26(2):177–82.
23. Graham JM, Ashcroft P. Direct measurement of cerebrospinal fluid pressure through the cochlea in a congenitally deaf child with Mondini dysplasia undergoing cochlear implantation. Am J Otol. 1999;20(2):205–8.
24. Fishman AJ, et al. Fluoroscopically assisted cochlear implantation. Otol Neurotol. 2003;24(6):882–6.
25. Lemmerling MM, et al. Normal modiolus: CT appearance in patients with a large vestibular aqueduct. Radiology. 1997;204(1):213–9.
26. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
27. Cabbarzade C, Sennaroglu L, Suslu N. CSF gusher in cochlear implantation: the risk of missing CT evidence of a cochlear base defect in the presence of otherwise normal cochlear anatomy. Cochlear Implants Int. 2015;16(4):233–6.
28. Weber BP, et al. Malformations in cochlear implant patients. Am J Otol. 1997;18(6 Suppl):S64–5.
29. Sennaroglu L, Atay G, Bajin MD. A new cochlear implant electrode with a “cork”-type stopper for inner ear malformations. Auris Nasus Larynx. 2014;41(4):331–6.

30. Sennaroglu L, Bajin MD. Classification and current management of ınner ear malformations. Balkan Med J. 2017;34(5):397–411.
31. Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. Otol Neurotol. 2002;23(1):39–42; discussion 42–3.
32. Balkany T, Telischi FF. Fixation of the electrode cable during cochlear implantation: the split bridge technique. Laryngoscope. 1995;105(2):217–8.
33. Bendet E, et al. Cochlear implantation after subtotal petrosectomies. Eur Arch Otorhinolaryngol. 1998;255(4):169–74.
34. Saeed H, Powell HR, Saeed SR. Cochlear implantation in X-linked deafness—how to manage the surgical challenges. Cochlear Implants Int. 2016;17(4):178–83.
35. Syal R, Tyagi I, Goyal A. Cerebrospinal fluid otorhinorrhea due to cochlear dysplasias. Int J Pediatr Otorhinolaryngol. 2005;69(7):983–8.
36. Kim LS, et al. Cochlear implantation in children with inner ear malformations. Ann Otol Rhinol Laryngol. 2006;115(3):205–14.
37. Copeland BJ, Pillsbury HC, Buchman CA. Prospective evaluation of intraoperative cochlear implant radiographs. Otol Neurotol. 2004;25(3):295–7.

16

First Consensus Meeting on Auditory Brainstem Implantation in Children and Non-neurofibromatosis Type 2 Patients

Manuel Manrique, Shakeel R. Saeed, and Levent Sennaroglu

On 18th of September 2009, a group of health care professionals and scientists with extensive experience of auditory implantation attended a meeting convened by the Hacettepe Cochlear Implant Group. This group was comprised of the following professionals representing their cochlear implant groups: Levent Sennaroglu (Hacettepe University, Ankara, Turkey), Vittorio Colletti (Verona University, Italy), Manuel Manrique (University of Navarra, Pamplona, Spain), Roland Laszig (Freiburg University, Germany), Erwin Offeciers (St. Augustinus Hospital, University of Antwerp, Belgium), Shakeel Saeed (Royal National Throat, Nose & Ear Hospital, London, U.K.), Richard Ramsden (Central Manchester University Hospitals, U.K.), Sarp Sarac (Hacettepe University, Ankara, Turkey), Simon Freeman (Central Manchester University Hospitals, U.K.), Helge Rask Andersen (Uppsala University Hospital), Andrzej Zarowski (St. Augustinus Hospital, University of Antwerp, Belgium), Ibrahim Ziyal (Hacettepe University, Ankara, Turkey), Wolf-Peter Sollmann (Neurosurgery Department of Staedtische Klinikum Braunschweig, Braunschweig, Germany), Jan Kaminsky (Freiburg University, Germany), Bartolome Bejarano (University of Navarra, Pamplona, Spain), Ahmet Atas (Hacettepe University, Ankara, Turkey), Gonca Sennaroglu (Hacettepe University, Ankara, Turkey), Esra Yücel (Hacettepe University, Ankara, Turkey), Sebnem Sevinc (Hacettepe University, Ankara, Turkey), Lilli Colletti (Verona University, Italy), Alicia Huarte (University of Navarra, Pamplona, Spain), Lise Henderson (Central Manchester University Hospitals, U.K.), Thomas Wesarg (Freiburg University, Germany), and Konrad Konradsson (Uppsala University Hospital).

The aim of the meeting, based on collective experience was to have a detailed discussion on the pressing and pertinent issues around auditory brainstem implantation (ABI) in children and in non-neurofibromatosis type 2 (NF2) cases and to reach a consensus based on these discussions. The meeting consisted of presentations in four over-arching areas: surgery, experience of individual ABI centers, intraoperative issues, and

M. Manrique
University of Navarra, Pamplona, Spain
e-mail: mmanrique@unav.es

S. R. Saeed (✉)
University College London Ear Institute, Royal National Throat, Nose & Ear Hospital and National Hospital for Neurology and Neurosurgery, London, UK
e-mail: shakeel.saeed@ucl.ac.uk

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_16

rehabilitation with the final session devoted to discussion of the issues raised. The centers presented their experience of ABI in children who had hearing impairment due to congenital abnormalities or acquired pathology where cochlear implantation (CI) was either contraindicated or the possibility of successful placement of the electrode was unlikely. Altogether, 61 children with various types of inner ear malformations, cochleovestibular nerve (CVN) anomalies, cochlear ossification, and bilateral cochlear fractures with cochlear nerve avulsion were presented by different groups. The following topics were discussed, and a consensus was obtained at the end of the meeting in the following domains: Indications, contraindications, qualification of the ABI center, surgical procedure, ABI activation, and rehabilitation.

The agreement achieved by this group underpinned a consensus publication in 2011 [1]. This consensus has stood the test of time as 8 years later, its contents remain valid and the paper serves as a reference document to be used for information pertaining to the main aspects related to the whole ABI process. During this period of time, the indications for ABI have been further consolidated in non-tumor patients, substantiating the reported results in the medical literature with the initial experiences expressed in this publication.

The main contents of the publication are reproduced on the following pages.

16.1 In Which Children and Non-NF2 Patients Is the ABI a Viable Intervention?

Two patient categories were identified:

1. **Prelingual patients** with inner ear malformations and cochlear nerve hypoplasia/aplasia. ABI provides auditory perception in most patients. The potential for speech and language acquisition in the longer term will depend on the age of implantation, the presence or absence of additional disabilities, and the other established factors seen in CI. It also was concluded that open set speech discrimination is possible in selected cases. In addition, prelingually deafened children due to meningitis with total ossification of both cochleae also should be included in this group.
2. **Individuals deafened postlingually** due to meningitis, temporal bone fractures with cochlear nerve avulsion, otosclerosis with gross cochlear destruction, or unmanageable facial nerve stimulation with CI.

16.2 Which Health Care Team Is Best Positioned to Undertake This Intervention?

ABI in children and non-NF2 recipients should only be undertaken by a team experienced in CI, vestibular schwannoma surgery, and adult brainstem implant surgery. The team should comprise the following members: otologist or neurotologist, pediatric neurosurgeon, implantation-experienced audiologist, electrophysiologist, speech and language habilitation/rehabilitation specialist, experienced neuroradiologist, and experienced pediatric anesthesiologist with intensive care unit facilities for children.

16.3 Is It an Appropriate Procedure?

As with any intracranial procedure, ABI has an inherent potential risk of complications that may be serious or life-threatening. For this reason, it is mandatory that the surgical team undertaking such cases has the requisite experience previously described. The current experience of ABI in children and non-NF2 cases has shown the procedure to be safe in appropriate hands, thereby minimizing the risk of compromising the clinical condition or safety of patients. Thus far, none of the centers has reported any permanent serious complications.

16.4 What Are the Radiologic Indications?

16.4.1 Group 1: Well-Defined Congenital Indications

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

16.4.2 Group 2: Possible Congenital Indications

1. Hypoplastic cochleas with cochlear aperture hypoplasia.
2. Common cavity and incomplete partition type I cases if the cochlear nerve is not present.
3. Common cavity and incomplete partition type 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 CVN is a challenge in these cases. 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 the patients with an insufficient response.
5. The hypoplastic cochlear nerve 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. The radiology in these patients should be carefully reviewed with an experienced neuroradiologist. If a sufficient amount of neural tissue cannot be followed into the cochlear space, an ABI may be indicated.

16.4.3 Group 3: Acquired Indications

1. In postlingually deafened children due to meningitis with severe ossification of the cochlea (white cochleas on computed tomography with absolute no cochlear duct signal on magnetic resonance imaging [MRI]), the chance of successful CI placement in the correct location inside the cochlea is very low, and an ABI may be a preferred option in this situation. However, if the cochlea seems to be patent on T2 MRI, CI should be the first option.
2. Bilateral temporal bone transverse fractures with cochlear nerve avulsion.
3. Cochlear otosclerosis with gross destruction of the cochlea which is readily diagnosed on computed tomography and MRI. In addition, some otosclerosis patients have abnormal facial nerve stimulation, which may limit CI use even after appropriate channel programming. These cases also may be an indication for ABI.

Side Selection for ABI: The side with the better developed lateral recess should be preferred:

1. Side where the entrance of the lateral recess is more favorable.
2. Accessibility of the lateral recess, where cerebellar retraction will be less.
3. 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).

16.4.4 ABI Revision

After a certain period, there will be vascularization and scarring around the implanted electrode array. In revision cases due to device failure, it is difficult to explant the electrode array without causing bleeding. Every effort should be made to

avoid revision in such children. Revisions should be performed in an experienced center.

16.5 What Are the Contraindications?

For the present time, no technique demonstrates the presence of the cochlear nuclei efficiently. However, electrical auditory brainstem response (EABR) conducted during ABI surgery demonstrated the presence of all or some of the waves III, IV, and/or V in most cases. This tends to imply that a majority of children with severe malformations have cochlear nuclei. Positive round window EABR, although not 100% sensitive and specific, may predict a good result with CI surgery. Auditory neuropathy seems to be a contraindication to ABI surgery for the present time. Therefore, in these situations, ABI is not indicated initially.

Central pathologies, such as kernicterus, hemosiderosis, corpus callosum agenesis, and severe cerebral palsy, and comorbidities, such as cardiorespiratory disease, must be evaluated with extreme care. In addition, pervasive developmental delays, attention deficit/hyperactivity disorder, significant cognitive impairment, and lack of family support and commitment make it more difficult to obtain a satisfactory outcome from ABI. In general, these result in learning difficulties, which may slow down the progress of ABI patients considerably. It is very important to have a multidisciplinary approach in these cases, and each of them has to be determined according to the severity of the case and on their own merits.

16.6 What Is the Age Limit for ABI in Children?

The newborn infant has a tremendous developmental capacity up to the age of 1 year. Between 1 and 2 years of age, this development is slower. However, children younger than 1 year old have less relative blood volume and cerebrospinal fluid in the posterior fossa. In addition, the entrance of the lateral recess is smaller. Earlier implantation therefore carries the risk of hypovolemic shock due to cerebrospinal fluid and blood loss. In addition, it may be difficult to insert the ABI electrode into the lateral recess. There also is a higher risk of brain swelling below 1 year of age. The optimum age for an elective intracranial surgery in children is considered to be between 18 and 24 months. However, depending on the experience of the team, the minimum age for ABI in children may be as early as 1 year. On the basis that earlier intervention captures the critical period of plastic development of the brain, and in common with the same rationale applied to CI surgery, ABI surgery preferentially should not be done later than 3 years of age. This always needs to be balanced with any other factors that might complicate the adequacy of behavioral feedback which is important for ABI optimization in young children. Perilingual and post-lingual patients have no upper age limit.

16.7 Surgical Procedure, Electrophysiologic Evaluation, and Rehabilitation

16.7.1 Surgical Approach

The retrosigmoid surgical approach is the preferred route to place an ABI in children and non-NF2 cases.

16.7.2 Importance of Electrophysiologic Tests

Older children implanted with an ABI may be able to provide adequate behavioral feedback to enable an effective auditory program to be established similarly to an adult ABI recipient; however, on the premise that younger children will not be able to give such unequivocal feedback, it is necessary to gain help in programming from whichever source is possible. To this end, electrophysiology is deemed a suitable guide if it is possible to record EABRs from the electrode array in situ because this gives objective evidence that the

electrical stimulation is activating the normal auditory pathway. Every effort needs to be taken to obtain meaningful electrophysiologic (EP) data that can be used for this purpose. EP can provide two levels of guidance: First, it may be used to aid optimal electrode positioning during ABI surgery; second, it may be used to decide which electrodes deliver substantially auditory sensations and possibly even provide an estimation of that auditory level.

16.7.3 Electrophysiology at the Time of ABI Surgery

Recording electrodes should be placed on the subject to permit accurate recording of the EABR. Based on the recording electrode montage used for adults and existing experience with children, the following dual channel recordings are advised: Where only a single channel is available, either montage may be selected based on familiarity. Stimulating electrode combinations on the ABI once positioned should be chosen to optimize location. These combinations should allow both the depth and rotation of the array within the lateral recess to be determined and adjusted perioperatively and before the wound is closed.

16.7.4 Electrophysiology Before ABI Activation

Activation is planned after complete wound healing (usually 4–6 weeks after the ABI surgery). Immediately before activation, the child should be anesthetized to perform an accurate EABR mapping of the array. A sufficiently comprehensive series should be performed across the whole array using precisely the electrode combinations and parameters as would be used during the activation, with the exclusion of rate that needs to be appropriate for EABR (e.g., 35 Hz). For each tested electrode combination, the current levels should be increased to obtain the threshold of a recognizable EABR (typically, a 2–3 peak waveform with peaks falling within a 5-ms window after the stimulus). Once a recognizable EABR is established, current levels should be increased to at least double the current to establish if any other non-EABR-like potentials arise. Non-EABR potentials (e.g., peaks beyond 5 ms) should be noted together with their threshold level, and these electrodes excluded from initial activation if they occur below a doubling of current from the EABR threshold level. Based on the above investigation, electrodes should be identified which give the clearest EABRs and these electrodes thus targeted during initial activation.

16.7.5 Activation

Owing both to the possibility of unsuccessful EABR and to the imprecise nature of EABRs recorded from the CNC, activation of the ABI should be based primarily on behavioral feedback but guided by any results from intraoperative monitoring. This must be coupled with experience of typical levels based on an adult ABI population together with general programming principles for a young pediatric CI population.

In preparation for the activation, monitoring leads should be attached to the patient to allow observation of the heart rhythm as the vagus nerve is in close proximity to the intended location of the ABI array. A member of medical staff qualified in resuscitation must be present with any other facilities considered appropriate, which also may dictate the location that the activation takes place (e.g., ward or intensive therapy unit).

Because dizziness is a common side effect in adults, if possible, the child should stand at a table (supported by a parent or carer), with toys appropriate for play audiometry available. A trained therapist familiar with pediatric CI activation is essential.

The current level on each selected electrode with a good EABR should be increased while simultaneously paying careful attention to any behavioral response of the child. This may include generally good signs, such as stilling, distraction, eye widening, blinking, looking up, puzzlement, pointing to the ear, or even smiling.

Alternatively, adverse reactions, such as scratching a part of the body other than the ear, strong blinking, frowning, facial twitching, nystagmus, swaying, or crying. An experienced therapist and audiologist must decide the basis on which to accept or reject reactions to stimulation as evidence of auditory or non-auditory sensations because these must be the primary basis for electing to select an electrode to include in the final map.

After investigation of the array and selection of a group of acceptable electrodes for activation, these electrodes should be ordered in the anticipated biologically appropriate tonotopic order (i.e., proximal electrodes = low frequency, distal electrodes = high frequency). When testing, the mean arterial pressure levels may need to be reduced and increased in live mode. If the program is not tolerated at, a low level, selective deactivation of electrodes may be necessary to achieve a tolerable program.

For the first few days from activation, the aim should be to achieve a program that shows some evidence of distraction when a noise is made but also something which is well tolerated.

16.7.6 Rehabilitation

From the results of different centers, it can be understood that it is possible to restore hearing perception in children with prelingual deafness with severe inner ear malformations and cochlear nerve anomalies. In some selected cases, it also was possible to develop open set speech understanding. However, the family should be warned of different outcomes from this intervention so that their expectations should not be high. When compared with CI surgery, programming and rehabilitation of prelingual children with ABI are much more labor intensive, and the results are not as good as CI. On this basis, the candidacy assessment is much more detailed than in CI patients and requires more experienced staff.

Auditory verbal therapy in these children, where only auditory stimulation is conveyed, may not be as efficient as in CI children. Total communication and speech reading also should be encouraged to convey more linguistic and language information to these children. In this method, speech reading assumes considerable importance as a source of information, whereas tactile and motor kinesthetic stimulation provides supportive avenues for spoken language acquisition. In addition, the involvement in speech reading training programs has a positive effect on postoperative perceptive and expressive linguistic skills. The context and phases of auditory training of these children do not differ from the programs for CI users. However, it may be more appropriate to undertake more intensive training on specific tasks, such as discrimination and identification of suprasegmental features of speech sounds.

The recipients with pervasive developmental delays figure among the worst in terms of subjective auditory performances. Despite their lack of open set discrimination scores, their parents report that the child feels much more confident in their educational settings and in their family. These cases show very clearly that the results of hearing (re)habilitation cannot be defined only in terms of open set speech discrimination, although this should be our ultimate goal.

16.8 Do We Need Any Modification for ABI Electrode in Children?

On balance, the size and shape of the current ABI electrode is considered satisfactory. In the very young child, sometimes the ABI electrode array may be slightly large for the lateral recess, but producing a smaller electrode has the possibility of the plate being too small when these children become adults.

After a certain period, there will be scarring and vascularization around the electrode array. In revision cases due to device failure, it may be extremely difficult to explant the electrode array without causing bleeding.

16.9 How Many Centers in a Country?

This is a challenging intervention and difficult surgical procedure that draws heavily on the experience of auditory implant and lateral Skullbase teams. To gain enough surgical experience, monitoring, programming, and rehabilitation of children with ABI, this intervention should be limited to those centers which meet the criteria previously described. The actual number of centers required in each country will depend on factors, such as population, incidence of congenital deafness, access to health care, resource provision, and expertise developed. As such, individual countries should make their own policy, taking into account these considerations.

16.10 Policy for Foreign Demands for ABI?

When undertaking ABI surgery in another country, the team requirements also apply to that country. If the local team is not very experienced and a serious complication occurs, there is a risk of bringing this intervention into disrepute. It also is very important that there is a full understanding of the audiologic and rehabilitation results of that surgery according to the protocol mentioned here. For some countries, it may be a better option to refer the patient to one of the experienced centers.

16.11 Conclusion

The patient experience and data presented in this meeting demonstrated that it is possible to use the ABI to restore auditory perception in the majority of the patients with severe inner ear malformations, such as labyrinthine and cochlear aplasia, and cochlear nerve agenesis. Some patients may also develop open set discrimination scores. However, the presence of additional disabilities greatly diminishes the auditory outcomes of this procedure.

Reference

1. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.

17 Cochlear Implantation Versus Auditory Brainstem Implantation in the Management of Complex Inner Ear Malformations

Nedim Durakovic, Daniel J. Lee, and Craig A. Buchman

Inner ear malformations account for up to 40% of congenital sensorineural hearing loss cases depending on the diagnostic modality used to evaluate this prevalence [1]. Although cochlear implants (CIs) are the most effective implantable auditory prosthetic device in patients with severe to profound sensorineural hearing loss, inner ear malformations provide a unique challenge in this patient population. Identification and characterization of inner ear malformations is critical in the preoperative period, aiding preparation for potential surgical challenges and postoperative performance expectations. The severity of inner ear malformations can indicate lack of availability or organization of neural elements required for successful cochlear implantation. For a subset of children with profound hearing loss associated with severely anomalous anatomy, placement of an auditory brainstem implant (ABI) is an option. This chapter outlines considerations of CI for inner malformations including potential surgical complications and variability in performance outcomes, as well as consideration for ABI placement.

17.1 Consideration for Different Malformation Types

A variety of inner ear malformations have been described with varying degrees of severity from complete labyrinthine aplasia to near normal cochlear anatomy [2, 3]. Each of these malformations can create unique challenges for successful CI placement. In this chapter, we will focus on three malformations that have unique considerations for cochlear implantation including common cavity malformation, cochlear hypoplasia, and incomplete partition type I. In each of these cases, preoperative imaging with high resolution CT and direct parasagittal T2-weighted MRI images is critical in establishing cochlear nerve integrity as cochlear nerve deficiency (CND) can be associated with these malformations (Fig. 17.1) [4].

In the most severe cases, cochlear nerve aplasia and labyrinthine or cochlear aplasia, there is lack of a neural substrate or even a rudimentary cochlea for placement of a CI. In contrast, common cavity malformations have a cochleovestibular chamber that communicates with the internal auditory canal making implantation feasible. The course of the facial nerve in these cases can be aberrant in the mastoid due to an underdeveloped horizontal semicircular canal. Thus, cochlear

N. Durakovic (✉) · C. A. Buchman
Department of Otolaryngology-Head & Neck Surgery, Washington University in St. Louis, St. Louis, MO, USA
e-mail: durakovicn@wustl.edu; craig_buchman@med.unc.edu

D. J. Lee
Department of Otolaryngology, Harvard Medical School, Boston, MA, USA
e-mail: Daniel_Lee@meei.harvard.edu

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_17

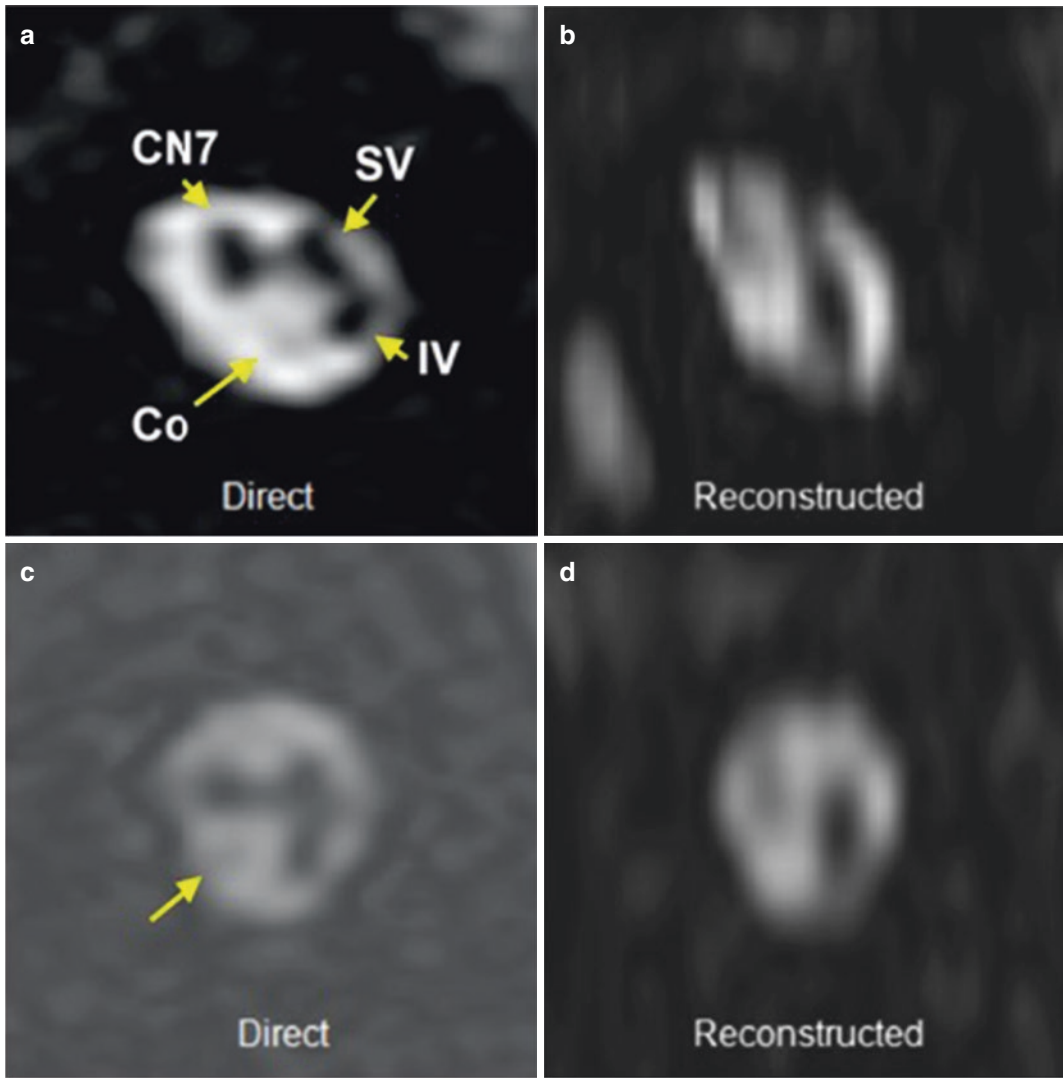


Fig. 17.1 Direct and reconstructed parasagittal T2-weighted MRI sequences illustrate internal auditory canal (IAC) morphology in two pediatric patients with congenital profound hearing loss. Reformatted images perpendicular to the IAC (image **b**) suggest cochlear nerve aplasia but provide poor resolution, whereas *direct* sequences (image **a**) confirm the presence of a small cochlear nerve (Co). Note the normal appearance of the facial nerve (CN7), superior vestibular (SV) nerve, and inferior vestibular (IV) nerve. As with the other example, reconstructed images of the IAC (Image **d**) provide limited visual information and suggest cochlear nerve aplasia, compared to the direct sequence (image **c**) that confirms the presence of cochlear nerve hypoplasia. Accurate imaging data is crucial for the counseling of patients and discussion of management options in this unique patient cohort. (Modified from Noij et al. 2015 [4])

implantation can be challenging through a conventional posterior tympanotomy, requiring either a labyrinthine cochleostomy or a canal down mastoid approach [5].

In contrast to common cavity malformations, cases of cochlear hypoplasia include a spectrum of malformations characterized by a distinct, hypoplastic cochlea with varying degrees of cochlear development. In the most severe form, type I cochlear hypoplasia, a distinct, bud-like cochlea arises anterior to the internal auditory canal. Type II cochlear hypoplasia is characterized

by a normal external cochlear architecture with absent modiolus and interscalar septum. Characteristically these cases have a widened cochlear nerve canal and enlarged vestibular aqueduct with an associated risk of CSF gusher or CI insertion into the internal auditory canal. Type III cochlear hypoplasia is characterized by a hypoplastic modiolus with fewer than two cochlear turns. Finally, in type IV cochlear hypoplasia, the basilar turn has normal development but the middle and apical cochlear turns are hypoplastic [6].

In contrast to cases of cochlear hypoplasia, incomplete partition cases include a spectrum characterized by normal external cochlear dimensions with malformed internal architecture. In type I, the vestibule is dilated and the cochlea appears cystic without a modiolus or interscalar septa. Similar to cochlear hypoplasia type II, the undeveloped modiolus is associated with an enlarged cochlear nerve canal and a risk for CSF gusher or CI insertion into the internal auditory canal. Incomplete partition type II is characterized by a triad of enlarged vestibular aqueduct, minimally dilated vestibule, and aberrant modiolar apex. Finally, type III incomplete partition, or X-linked deafness, is characterized by normal cochlear dimensions, an absent modiolus, but intact interscalar septa, brisk CSF gusher on cochleostomy, and risk of IAC placement and postoperative meningitis [7].

17.2 Potential Surgical Complications

Anticipation of complications with cochlear implantation in inner ear malformations is important in both preoperative planning and family counseling. Cochlear implantation in cases of inner ear malformations poses greater risks even in the most experienced hands. Anticipated complications include facial nerve injury secondary to anomalous facial nerve anatomy, CSF gusher, and electrode misplacement.

Cochlear implantation in non-malformed ears relies on consistent identification of the vertical segment of the facial nerve followed by a posterior tympanotomy with identification of the round window membrane. The short process of the incus aids in identification of the vertical segment with the second genu of the nerve positioned just inferior and slightly medial to the horizontal semicircular canal. A posterior tympanotomy approach allows for CI insertion anterior to the facial nerve.

In malformed inner ears, especially those cases with dysplastic horizontal canals, the facial nerve course can be anomalous relative to normal anatomic expectations. Common variations of facial nerve anatomy include anterior displacement of the nerve, position overlying the promontory (Fig. 17.2) and even overlying the round window membrane, thus placing the facial nerve at higher risk for injury with conventional CI insertion techniques [8]. Preoperative imaging can aid in the diagnosis of aberrant facial nerve anatomy, while intraoperative electromyography (EMG) and early identification of the nerve are critical to safely navigating these cases. Certain cases may even require temporary translocation of the posterior bony ear canal, a combined transcanal and transmastoid approach, or even blind-sac closure of the ear canal in order to safely identify the facial nerve and ensure appropriate CI placement.

While a malformed horizontal semicircular canal may indicate anomalous facial nerve

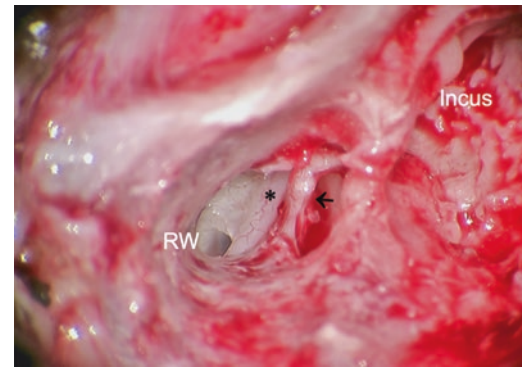


Fig. 17.2 Intraoperative transmastoid view of the left ear through a posterior tympanotomy demonstrates an aberrant facial nerve in cochlear hypoplasia type I (CH-I) in CHARGE association. Absent horizontal semicircular canal with an aberrant facial nerve (*) is identified inferior to the location of the stapes (arrow)

anatomy, a widened cochlear nerve canal or absent modiolus suggests increased risk for CSF gusher as in the case of incomplete partition type I and type III [8]. CSF gushers can be characterized as low flow or high flow leaks and can occur both through the cochleostomy or through a fistula at the stapes footplate [9]. Furthermore, these patients are at an increased risk of meningitis both prior to and following surgery from both the implanted and non-implanted ear.

Identification of a CSF gusher during surgery with appropriate repair rarely results in a postoperative CSF leak [10]. Management of CSF gushers includes a wide cochleostomy with packing of tissue around the CI array as well as use of CI arrays designed with silicone stoppers [11]. Additional steps can be used to seal a CSF leak including obliteration of the Eustachian tube while cerebrospinal fluid diversion is not routinely used in the largest case series of cochlear implantation in malformed inner ears [10].

An underdeveloped modiolus or cochlear nerve canal with associated CSF communication can also predispose to misplacement of the CI into the internal auditory canal or even the vestibule. Malformations at increased risk of these complications include common cavity malformations and incomplete partition type I and III (Fig. 17.3). In contrast, hypoplastic cochleae may cause incomplete CI insertion due to smaller dimensions. In general, if resistance is met, the array should be redirected or insertion stopped. Intraoperative transorbital or reverse Stenvers plain-film X-ray can be used to aid in confirmation of proper electrode placement within the cochlea.

In cases of common cavity malformation or an underdeveloped modiolus such as incomplete partition type I or III, consideration should be made for using a lateral wall array. Use of perimodiolar electrodes can be difficult to deploy effectively in cases of underdeveloped modiolar anatomy [11]. A fully banded electrode design can be used to ensure stimulation in cases where the position of neural elements within the cochlea is inconsistent or occurs along the lateral wall [12]. Finally, use of shorter and thinner electrodes should be considered in cases of hypoplastic cochleae.

Similar to the risk for surgical complications that inner ear malformations pose intraoperatively, they can also pose a challenge in postoperative programming. Although rare, cases of anomalous facial nerve anatomy can result in facial nerve stimulation while common cavity cases can be associated with postoperative vestibular stimulation [13]. In both of these conditions, individual electrode contacts need to be turned off to reduce off-target effects.

17.3 Performance Outcomes

Although several studies have demonstrated adaptation of surgical techniques to allow for safe cochlear implantation in inner ear malformation, very few studies have evaluated CI performance in these patient cohorts. This is primarily due to the scarcity of different types of malformations as well as lack of long-term follow-up data with open-set speech testing [10]. The few studies available with performance data demonstrate variability in CI outcomes depending on the severity of the inner ear malformations. Variability in performance is an important consideration in family counseling and setting appropriate rehabilitation goals. Finally, it is important to interpret outcomes after CI in malformed inner ears in the context of other patient comorbidities and other factors that impact performance beyond device positioning within the cochlea [14, 15].

In general, more severe malformations such as common cavity or those associated with CND have a poorer prognosis with difficulty attaining open-set speech understanding. In the two largest studies evaluating speech outcomes, incomplete partition cases had comparable outcomes to non-malformed ears, while cases of hypoplastic cochleae or CND had worse outcomes [10, 16]. Variability in performance within the incomplete partition spectrum is likely with several case series suggesting satisfactory but worse outcomes for incomplete partition type I compared to type II [17, 18]. Incomplete partition type III is the rarest of the incomplete partition spectrum anomalies with most case studies focused on management of the increased CSF leak rates and

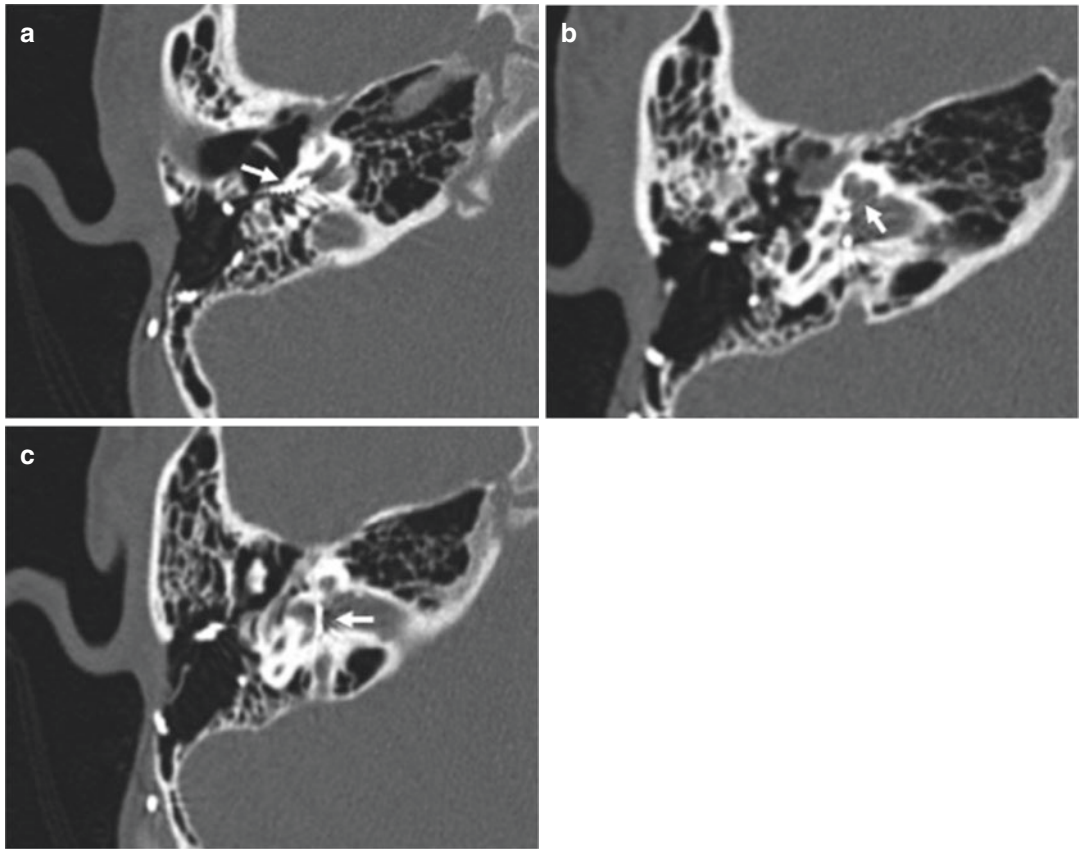


Fig. 17.3 Non-contrast axial CT scan images demonstrate CI placement into the IAC in a case of type III incomplete partitioning. Although the CI is placed through a round window approach (Image **a**), an underdeveloped modiolus and widened cochlear nerve canal (Image **b**) result in displacement of the electrode into the IAC (Image **c**). Use of a lateral wall array is favored in these cases in addition to extending the round window approach in order to achieve placement of the array along the lateral wall of the cochlea. Placement of the array along the lateral wall avoids displacement of the array into the IAC and potentially shearing neural elements in the underdeveloped modiolus

potential for electrode misplacement into the IAC (Fig. 17.3) [11]. Buchman et al. reported that as a group 100% of incomplete partition cases achieved open-set speech perception compared to 50% of hyoplastic cochleae and only 19% of CND cases [10]. Furthermore, visual supplementation was required in 69% of hypoplastic cochleae and 95% of CND cases [10].

17.4 ABI Considerations

Variability in performance of CI in certain inner ear malformations including CND led to the expansion of criteria for ABI to include nontumor cases. The ABI was initially developed for patients with neurofibromatosis type II (NF2) characterized by bilateral vestibular Schwannomas. These patients develop bilateral profound sensorineural hearing loss due to progressive tumor growth or secondary to treatment of their tumors. Traditionally these patients were not CI candidates due to disruption of the cochlear nerve. The ABI was thus designed to be placed at the time of vestibular schwannoma tumor removal with the first device placed in 1979 by Drs. William Hitselberger and William House [19, 20]. While some patients have been able to receive open-set speech perception, the majority of patients gain sound awareness and enhanced

lip-reading. More recently, several groups have demonstrated the benefit of ABI in the non-NF2 patient population, including patients with post-meningitis cochlear obliteration, far advanced otosclerosis, posttraumatic avulsion of both cochlear nerves, and severe inner ear malformations [21].

17.5 ABI Surgical Technique and Potential Complications

Although the receiver-stimulator for an ABI is very similar to a CI, the surgery for device placement is fundamentally different with greater potential risks for the patient than CI surgery. Either a retrosigmoid craniotomy or translabyrinthine approach can be used for ABI placement. In the case of pediatric ABI surgery and especially in the case of very young patients with underdeveloped mastoids, the retrosigmoid approach offers the advantage of a wider view of the posterior fossa in addition to avoiding loss of any residual vestibular function and contamination of the intracranial space with mastoid contents. With either technique, the surgical setup requires cranial nerve monitoring beyond the facial nerve and includes monitoring of CN IX (glossopharyngeal), CN X (vagus), and CN XI (spinal accessory). In addition, setup may require placement of the patient in cranial fixation.

Similar to CI surgery, the receiver-stimulator for the ABI is placed in a subperiosteal pocket. With use of a retrosigmoid incision, the receiver-stimulator may have a more posterior position along the skull compared to traditional CI placement (Fig. 17.4). Furthermore, if a tight subperiosteal pocket cannot be created, suture fixation or placement of an intraosseous seat can be used to ensure the receiver-stimulator remains fixed in position.

The ABI electrode array is a paddle that is designed for placement over the cochlear nucleus. Identification of the cochlear nucleus involves tracing CN IX to its root entry zone. At the root entry zone, the cerebellar flocculus is identified along with the choroid plexus exiting the fourth ventricle. The ABI paddle is placed inside the lateral recess of the fourth ventricle in contact with the ventral surface of the cochlear nucleus, anterior to the choroid plexus (Fig. 17.5). Intraoperative electrically evoked auditory brainstem responses (eABR) can be used to optimize electrode placement over the cochlear nucleus with confirmation of auditory stimulation. In addition, intraoperative eABR testing allows for monitoring for nonauditory stimulation of CN VII, IX, X, and XI by EMG and vital sign monitoring.

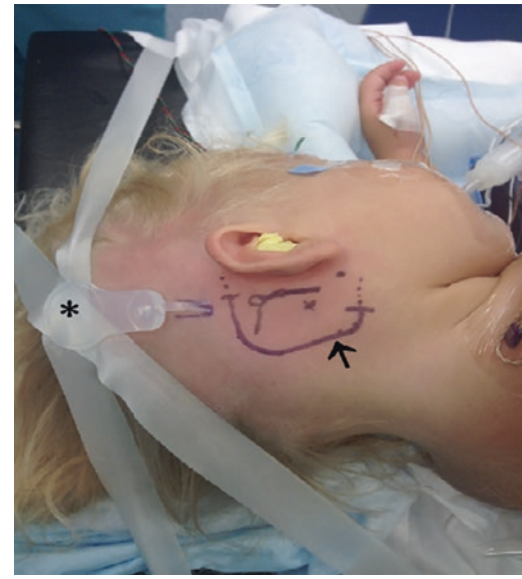


Fig. 17.4 Surgical site incision planning for a right ear retrosigmoid craniotomy with placement of the ABI receiver-stimulator (*) relative to a retrosigmoid craniotomy incision (arrow)

Although several case series have demonstrated the safety and efficacy of ABI placement in patients with inner ear malformations, surgery for ABI placement entails greater risk than CI surgery [22, 23]. Noij et al. conducted a systematic review that identified a major complication rate as high as 21% with the most common complications caused by CSF leak or cerebellar edema [24]. The potential surgical risks of intracranial surgery can be life threatening and include CSF leak, hydrocephalus, nonauditory cranial nerve stimulation, meningitis, and stroke. Other delayed complications include the potential for electrode migration out of the fourth ventricle requiring revision surgery.

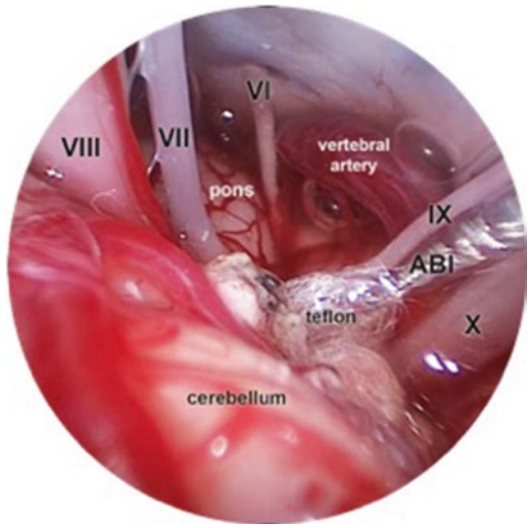


Fig. 17.5 Intraoperative 30° endoscopic view of the right cerebellopontine angle following retrosigmoid craniotomy for a 2-year-old male with congenital deafness and cochlear aplasia illustrates placement of the auditory brainstem implant (ABI) array. The Teflon felt is used to secure the array in the lateral recess of the IVth ventricle. The vestibulocochlear nerve (VIII), facial nerve (VII), glossopharyngeal nerve (IX), and vagal nerve (X) provide indirect landmarks during ABI surgery. Accurate placement is confirmed by electrophysiological measures (eABR). This child has sound detection and is in a total communication learning environment that includes sign language

17.6 ABI Performance Outcomes

Audiometric outcomes of NF2 patients undergoing ABI have demonstrated improved sound awareness, lip-reading, and for some patients even open-set speech perception [25]. In 2001, Colletti et al. reported on two cases with cochlear nerve aplasia implanted with ABIs who were able to obtain speech detection [26]. Subsequent studies at multiple centers revealed that the majority of pediatric ABI users with inner ear malformations are able to attain sound detection and benefit from sound awareness with up to one in two patients developing closed-set speech discrimination and relying on visual communication [22, 27, 28].

Similar to the challenge with interpreting speech outcomes in cases of inner ear malformations after CI, there are no multicenter trials for ABI outcomes in children and so the number of subjects reported for individual studies are limited. In addition, long-term follow-up is limited, and audiometric testing protocols vary across centers in the USA and abroad. In addition, associated nonauditory disabilities are common but underreported and are clearly associated with worse outcomes [24].

While cases of labyrinthine or cochlear aplasia are rare, the lack of a rudimentary cochlea is a contraindication for CI placement and ABI may be a reasonable option. Children with common cavity, cochlear hypoplasia, and incomplete partition deformities may be candidates for a CI or ABI based on the severity of the condition. In a review of 60 pediatric ABI patients, Sennaroglu et al. identified common cavity malformations and other cases with a present cochleovestibular nerve were associated with better outcomes [22]. The presence of a cochlear nerve likely indicated a more well-developed cochlear nucleus, while patients with cochlear hypoplasia or CND undergoing ABI placement had worse performance [22].

Since cases of CND perform poorly with either CI or ABI placement compared to other inner ear malformations, there has been some debate in the literature about the optimal treatment algorithm to pursue [10, 22]. In two separate studies, Colletti et al. retrospectively evaluated auditory perceptual abilities assessed using the Categories of Auditory Performance (CAP) scale in children implanted with CI followed by reimplantation with ABI and age-matched primary CI and ABI patients with CND [29, 30]. In both studies the highest CAP score achieved with CI was three corresponding to an ability to identify environmental sounds while ABI patients were able to achieve up to a CAP score of 7 or an ability to use a telephone with a known speaker. Other studies of CND and outcomes after CI have demonstrated that a limited number of patients as high as 19% can achieve open-set speech perception with a CI alone [10]. With these findings in mind, most authors favor initial CI in the least malformed ear first followed by consideration for ABI placement in the contralateral ear if adequate performance is not achieved [10, 31].

Complex inner ear malformations offer a unique set of challenges for CI centers. A multidisciplinary approach is required with input from families, surgeons, audiologists, neuroradiologists, and speech-language pathologists. In general, cochlear implantation is safe in this patient population but requires careful preoperative planning to avoid complications and guide performance expectations. For select cases with limited benefit after CI placement or severe inner ear malformation, ABI may be a viable alternative approach.

Bibliography

1. McClay JE, Booth TN, Parry DA, Johnson R, Roland P. Evaluation of pediatric sensorineural hearing loss with magnetic resonance imaging. Arch Otolaryngol Head Neck Surg. 2008;134(9):945–52.
2. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
3. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41.
4. Noij KS, Remenschneider AK, Kozin ED, et al. Direct parasagittal magnetic resonance imaging of the internal auditory canal to determine cochlear or auditory brainstem implant candidacy in children. Laryngoscope. 2015;125(10):2382–5.
5. McElveen JT Jr, Carrasco VN, Miyamoto RT, Linthicum FH Jr. Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. Laryngoscope. 1997;107(8):1032–6.
6. Cinar BC, Batuk MO, Tahir E, Sennaroglu G, Sennaroglu L. Audiologic and radiologic findings in cochlear hypoplasia. Auris Nasus Larynx. 2017;44(6):655–63.
7. Ozbal Batuk M, Cinar BC, Ozgen B, Sennaroglu G, Sennaroglu L. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233–8.
8. Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27(5):615–23.
9. Mylanus EA, Rotteveel LJ, Leeuw RL. Congenital malformation of the inner ear and pediatric cochlear implantation. Otol Neurotol. 2004;25(3):308–17.
10. Buchman CA, Teagle HF, Roush PA, et al. Cochlear implantation in children with labyrinthine anomalies and cochlear nerve deficiency: implications for auditory brainstem implantation. Laryngoscope. 2011;121(9):1979–88.
11. Sennaroglu L, Bajin MD. Incomplete partition type III: a rare and difficult cochlear implant surgical indication. Auris Nasus Larynx. 2018;45(1):26–32.
12. Graham JM, Phelps PD, Michaels L. Congenital malformations of the ear and cochlear implantation in children: review and temporal bone report of common cavity. J Laryngol Otol Suppl. 2000;25:1–14.
13. Sennaroglu L, Gursel B, Sennaroglu G, Yucel E, Saatci I. Vestibular stimulation after cochlear implantation in common cavity deformity. Otolaryngol Head Neck Surg. 2001;125(4):408–10.
14. Birman CS, Elliott EJ, Gibson WP. Pediatric cochlear implants: additional disabilities prevalence, risk factors, and effect on language outcomes. Otol Neurotol. 2012;33(8):1347–52.
15. Black J, Hickson L, Black B, Perry C. Prognostic indicators in paediatric cochlear implant surgery: a systematic literature review. Cochlear Implants Int. 2011;12(2):67–93.
16. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(1 Pt 2 Suppl 106):1–26.
17. Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N. Cochlear implant in incomplete partition type I. Acta Otorhinolaryngol Ital. 2013;33(1):56–62.
18. Kontorinis G, Goetz F, Giourgas A, Lenarz T, Lanfermann H, Giesemann AM. Radiological diagnosis of incomplete partition type I versus type II: significance for cochlear implantation. Eur Radiol. 2012;22(3):525–32.
19. Hitselberger WE, House WF, Edgerton BJ, Whitaker S. Cochlear nucleus implants. Otolaryngol Head Neck Surg. 1984;92(1):52–4.
20. Edgerton BJ, House WF, Hitselberger W. Hearing by cochlear nucleus stimulation in humans. Ann Otol Rhinol Laryngol Suppl. 1982;91(2 Pt 3):117–24.
21. Colletti V, Shannon R, Carner M, Veronese S, Colletti L. Outcomes in nontumor adults fitted with the auditory brainstem implant: 10 years' experience. Otol Neurotol. 2009;30(5):614–8.
22. Sennaroglu L, Sennaroglu G, Yucel E, et al. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.
23. Colletti V, Shannon RV, Carner M, Veronese S, Colletti L. Complications in auditory brainstem implant surgery in adults and children. Otol Neurotol. 2010;31(4):558–64.
24. Noij KS, Kozin ED, Sethi R, et al. Systematic review of nontumor pediatric auditory brainstem implant outcomes. Otolaryngol Head Neck Surg. 2015;153(5):739–50.
25. Otto SR, Brackmann DE, Hitselberger WE, Shannon RV, Kuchta J. Multichannel auditory brainstem implant: update on performance in 61 patients. J Neurosurg. 2002;96(6):1063–71.
26. Colletti V, Fiorino F, Sacchetto L, Miorelli V, Carner M. Hearing habilitation with auditory brainstem implantation in two children with cochlear nerve aplasia. Int J Pediatr Otorhinolaryngol. 2001;60(2):99–111.
27. Teagle HFB, Henderson L, He S, Ewend MG, Buchman CA. Pediatric auditory brainstem implantation: surgical, electrophysiologic, and behavioral outcomes. Ear Hear. 2018;39(2):326–36.

28. Puram SV, Barber SR, Kozin ED, et al. Outcomes following pediatric auditory brainstem implant surgery: early experiences in a north American center. Otolaryngol Head Neck Surg. 2016;155(1):133–8.
29. Colletti L, Wilkinson EP, Colletti V. Auditory brainstem implantation after unsuccessful cochlear implantation of children with clinical diagnosis of cochlear nerve deficiency. Ann Otol Rhinol Laryngol. 2013;122(10):605–12.
30. Colletti L, Colletti G, Mandala M, Colletti V. The therapeutic dilemma of cochlear nerve deficiency: cochlear or brainstem implantation? Otolaryngol Head Neck Surg. 2014;151(2):308–14.
31. Sennaroglu L, Colletti V, Lenarz T, et al. 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.

18 Auditory Brainstem Implantation in Children with Inner Ear Malformations

Burcak Bilginer and Levent Sennaroglu

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

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_18].

B. Bilginer (✉)
Department of Neurosurgery, Hacettepe University, Medical Faculty, Ankara, Turkey
e-mail: burcak@tr.net

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_18

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-

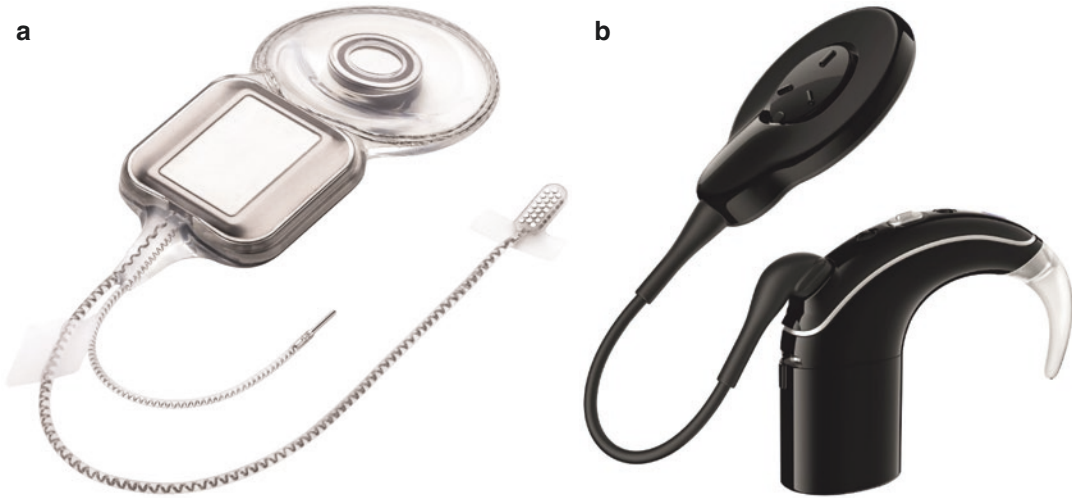


Fig. 18.1 (**a**) Auditory brainstem implant Nucleus ABI 541 (Used with permission of the Cochlear Company), has an array with 21 electrodes that are embedded in a silicone carrier and connected to an implantable internal receiver/stimulator (**b**) Processor CP1000 working with the ABI541

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.

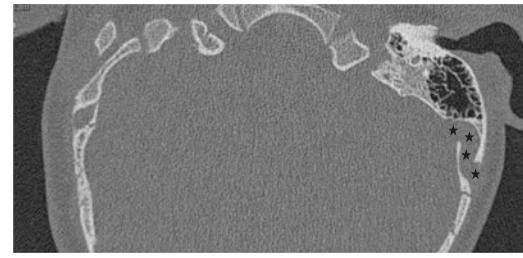


Fig. 18.2 Temporal CT showing a huge emissary vein (black stars) at the craniotomy area

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

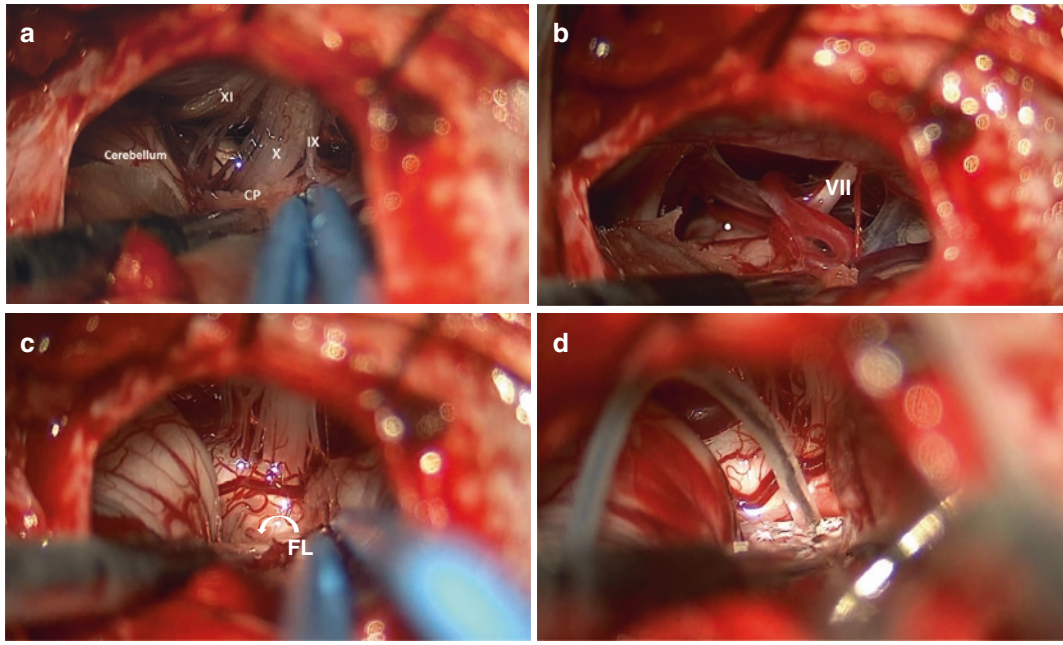


Fig. 18.3 (**a**) Retrosigmoid approach, showing lower cranial nerves (IX = glossopharyngeal nerve, X = Vagus, XI = nervous accessories, CP = choroid plexus), (**b**) facial nerve (VII), (**c**) foramen of Luschka (FL), (**d**) electrode in position

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

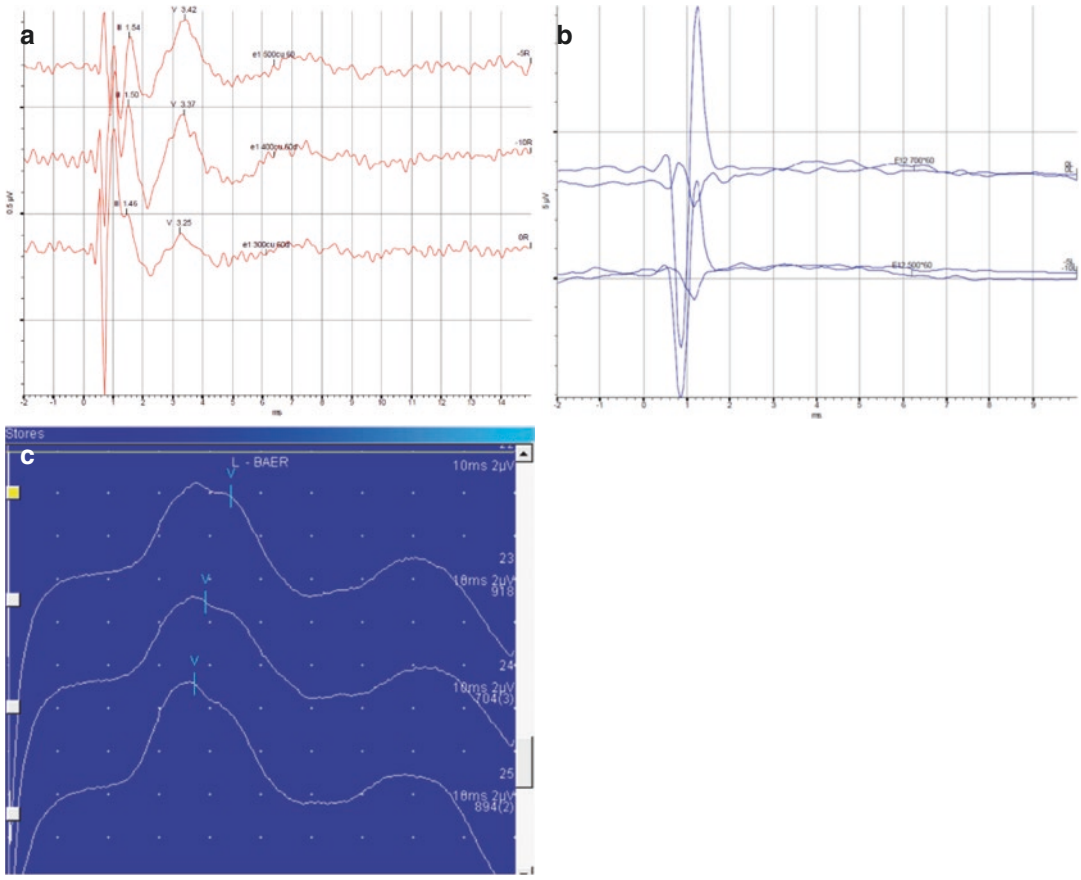


Fig. 18.4 Intraoperative electric auditory brainstem response (eABR): (**a**) eABR recording with waves eIII and eV, (**b**) eABR recording without response, (**c**) eABR recording with both auditory and nonauditory stimulation

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.

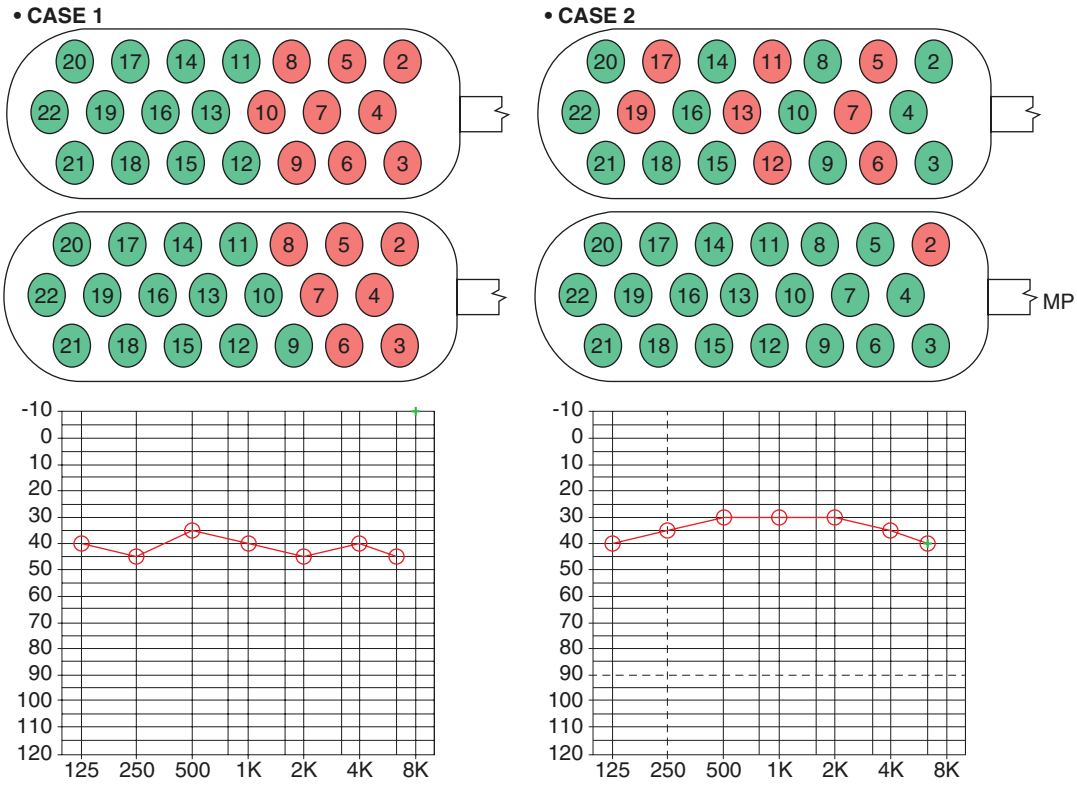


Fig. 18.5 ABI mapping showing side effects. Case 1: mapping showing very little change in the number of active channels causing side effects over time (green = active electrodes without side effects, red = electrodes with side effects) Case 2: Follow-up of a patient which showed decrease in the number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

References

1. Toh EH, Luxford WM. Cochlear and brainstem implantation. 2002. Neurosurg Clin N Am. 2008;19(2):317–29. vii
2. Fayad JN, Otto SR, Brackmann DE. Auditory brainstem implants: surgical aspects. Adv Otorhinolaryngol. 2006;64:144–53.
3. Otto SR, et al. Multichannel auditory brainstem implant: update on performance in 61 patients. J Neurosurg. 2002;96(6):1063–71.
4. Nevison B, et al. Results from a European clinical investigation of the nucleus multichannel auditory brainstem implant. Ear Hear. 2002;23(3):170–83.
5. Colletti V, et al. Hearing habilitation with auditory brainstem implantation in two children with cochlear nerve aplasia. Int J Pediatr Otorhinolaryngol. 2001;60(2):99–111.
6. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
7. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
8. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
9. Casselman JW, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. Radiology. 1997;202(3):773–81.
10. Sennaroglu L, et al. 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.
11. Sennaroglu L, Ziyal I. Auditory brainstem implantation. Auris Nasus Larynx. 2012;39(5):439–50.
12. Colletti V, et al. Hearing restoration with auditory brainstem implant in three children with cochlear nerve aplasia. Otol Neurotol. 2002;23(5):682–93.
13. Colletti V, et al. Auditory brainstem implant (ABI): new frontiers in adults and children. Otolaryngol Head Neck Surg. 2005;133(1):126–38.
14. Klose AK, Sollmann WP. Anatomical variations of landmarks for implantation at the cochlear nucleus. J Laryngol Otol Suppl. 2000;27:8–10.
15. Abe H, Rhoton AL Jr. Microsurgical anatomy of the cochlear nuclei. Neurosurgery. 2006;58(4):728–39; discussion 728–39
16. Friedland DR, Wackym PA. Evaluation of surgical approaches to endoscopic auditory brainstem implantation. Laryngoscope. 1999;109(2 Pt 1):175–80.
17. Brackmann DE, et al. Auditory brainstem implant: I. Issues in surgical implantation. Otolaryngol Head Neck Surg. 1993;108(6):624–33.
18. Laszig R, et al. Central electrical stimulation of the auditory pathway in neurofibromatosis type 2. Ear Nose Throat J. 1999;78(2):110–1, 115–7.
19. Terr LI, et al. Cochlear nucleus anatomy related to central electroauditory prosthesis implantation. Otolaryngol Head Neck Surg. 1990;102(6):717–21.
20. Kuchta J. Twenty-five years of auditory brainstem implants: perspectives. Acta Neurochir Suppl. 2007;97(Pt 2):443–9.
21. Bento RF, et al. Retrolabyrinthine approach for surgical placement of auditory brainstem implants in children. Acta Otolaryngol. 2012;132(5):462–6.
22. Behr R, et al. The high rate CIS auditory brainstem implant for restoration of hearing in NF-2 patients. Skull Base. 2007;17(2):91–107.
23. Lenarz T, et al. Auditory brainstem implant: part I. Auditory performance and its evolution over time. Otol Neurotol. 2001;22(6):823–33.
24. Cervera-Paz FJ, Manrique MJ. Auditory brainstem implants: past, present and future prospects. Acta Neurochir Suppl. 2007;97(Pt 2):437–42.
25. Sollmann WP, Laszig R, Marangos N. Surgical experiences in 58 cases using the nucleus 22 multichannel auditory brainstem implant. J Laryngol Otol Suppl. 2000;27:23–6.
26. Grayeli AB, et al. Auditory brainstem implant in neurofibromatosis type 2 and non-neurofibromatosis type 2 patients. Otol Neurotol. 2008;29(8):1140–6.
27. Sennaroglu L, et al. Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. Otol Neurotol. 2009;30(6):708–15.
28. Colletti V, et al. Complications in auditory brainstem implant surgery in adults and children. Otol Neurotol. 2010;31(4):558–64.
29. Bayazit YA, et al. Complications of pediatric auditory brain stem implantation via retrosigmoid approach. ORL J Otorhinolaryngol Relat Spec. 2011;73(2):72–5.

Complete Labyrinthine Aplasia (Michel Deformity)

19

Levent Sennaroglu and Mehmet Yaralı

Special Features

1. Definite indication for ABI.
2. Three subtypes present radiologically.
3. Facial nerve can be identified in cases with otic capsule.
4. Needs to be differentiated from complete labyrinthine ossification usually seen after meningitis.
5. Patients demonstrate response with ABI indicating normal cochlear nuclei development.

Definition: Complete labyrinthine aplasia (CLA) is the absence of the cochlea, vestibule, semicircular canals, vestibular and cochlear aqueducts [1, 2]. This is known as the Michel deformity. The petrous bone is reported to be hypoplastic, whereas the otic capsule may be partially developed or completely absent [3]. In the majority of patients, the internal auditory canal (IAC) does not develop normally; it may consist only of facial canal, or there may be no IAC at all. In the former situation the labyrinthine, tympanic, and mastoid segments of the facial nerve can be recognized in the temporal bone. However, in some patients, it may not be possible to identify and trace the facial canal in the temporal bone even though there is no facial paralysis. Malleus and incus are usually present and normally developed. Stapes may show certain abnormalities.

19.1 Histopathology and Pathophysiology

No postmortem specimen with CLA has been examined or reported. However, based on radiology, three different types of CLA are present. Some subtypes have otic capsule formation. It is not possible however to comment on the layers of otic capsule without histopathological examination.

This is the developmental arrest before the formation of the otocyst and the membranous labyrinth. In CLA there are subtypes where otic capsule is normally developed or is absent. This suggests that development of the membranous labyrinth and the otic capsule may be independent from each other. Even if the membranous labyrinth is completely absent, it is possible to have the otic capsule present or absent. Therefore, there must be separate mechanisms for the development of membranous labyrinth and otic capsule. This is most probably due to the fact that the outer two layers of the otic capsule have their vascular supply from the middle ear, while the membranous labyrinth receives its vascular supply from the IAC [4]. According to Donaldson [4], the inner endosteum receives its vascular

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

M. Yaralı (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_19

supply from the IAC, and the enchondral and outer periosteal layers get their vascular supply from the middle ear mucosa; if both vascular supply pathways are damaged, it may result in CLA without otic capsule formation; if only the vascular supply from the IAC is damaged, it may result in CLA with otic capsule formation (see Chap. 3 on Pathophysiology of IEM's).

(a) **CLA with aplastic/hypoplastic petrous bone:** In this group, CLA is accompanied by aplasia/hypoplasia of the petrous bone (Fig. 19.1a, b).
(b) **CLA without otic capsule:** In this subtype, formation of the petrous bone is normal, but the otic capsule is completely absent. Thus in this subgroup of CLA, the vascular supply from the IAC and middle ear appears to be severely damaged, resulting in the absence of all three layers of the otic capsule (Fig. 19.1c).
(c) **CLA with otic capsule:** These are cases of CLA where the formation of the petrous bone and the otic capsule is normal; there is only a canal for the facial nerve (Fig. 19.1d). Only in this group of CLA with otic capsule development, the facial canal occupies its normal location. This may show us that otic capsule formation is essential for the facial canal to obtain its normal position.

19.2 Literature Review

CLA may be present with some other skull base or middle ear anomalies. Ozgen et al. [3] reported that among 14 ears with CLA, most of the ears had middle ear and mastoid volume decrements. Also stapes was aplastic in one ear and dysplasia in five ears. Internal acoustic canal also showed anomalies such as aplasia in four and stenosis in ten ears. Similar to internal acoustic canal anomalies, facial nerve anomalies and deviated course were observed in 11 ears. Jugular bulb bony covering was defective in nine ears, tegmen tympani was defective in three ears.

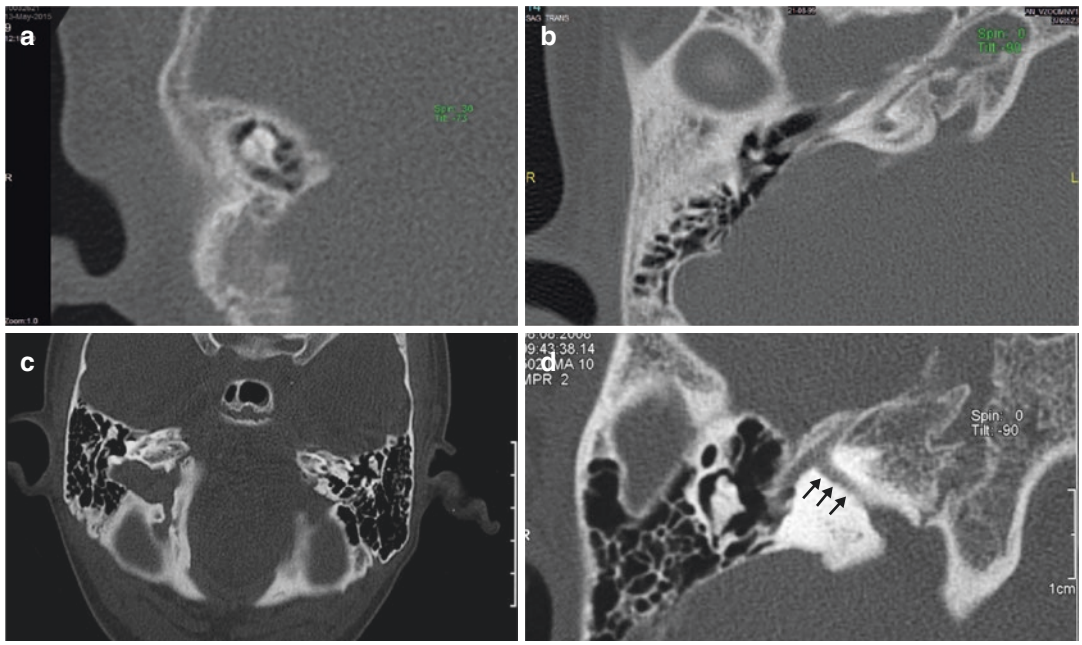


Fig. 19.1 Complete labyrinthine aplasia (CLA). Cochlea, vestibule, semicircular canals, vestibular, and cochlear aqueducts are absent. CLA can be seen with aplasia (**a**) and hypoplasia (**b**) of the petrous bone. (**c**) CLA without otic capsule. (**d**) CLA with the otic capsule. Labyrinthine (black arrows) and tympanic segments can be seen and followed in the temporal bone

As the crucial inner ear structures are not developed in patients with CLA, the observed hearing status is profound SNHL. No detectable hearing thresholds were obtained by objective (otoacoustic emissions, acoustic reflex testing, and auditory brainstem response testing) or subjective (pure tone audiometry) audiometry for either ear among five bilateral CLA patients.[3] Moreover, among four unilateral CLA patients, hearing thresholds were severely decreased in the contralateral ear. This is due to the fact that they had contralateral ear dysplasias.

19.3 Clinical Findings

In addition to profound SNHL, CLA has been associated with some other anomalies and syndromes. Most frequently observed anomalies are skull base [3, 5] and posterior fossa anomalies [5]. Some of these observed anomalies are petrous bone hypoplasia, otic capsule aplasia, decreased middle ear and mastoid volume, aplasia and dysplasia of stapes, internal acoustic canal aplasia, anomalies of facial nerve canal, and abnormal facial nerve course [3]. In terms of syndromes with CLA, Tekin et al. [6] defined a syndrome in 2008 and named it as LAMM (labyrinthine aplasia, microtia, microdontia) where CLA is observed. In this syndrome delay in motor skills during infancy, type-I microtia, and microdontia along with mild micrognathia in some cases has been observed. In addition, a 2-year-old case with CLA, aplasia of sixth, seventh, and eighth cranial nerves, internal carotid artery aplasia, and horizontal gaze anomalies together with developmental delays have been reported by Highley et al. in 2011 [7]. The authors gave conjectural diagnosis of Athabascan Brainstem Dysgenesis Syndrome (ABDS) for this case. One example of microtia in a patient with CLA is given in Fig. 19.2. This patient received an ABI.

Thalidomide induced external ear deformation, facial palsy, and atrial-septal defect together with CLA have been also reported [8]. Moreover, CLA may be present within families, Daneshi et al. [9] have reported CLA in two brothers and their cousin. Our series has a similar finding, two of our CLA cases are cousins (Cases 19.2 and 19.3 below).

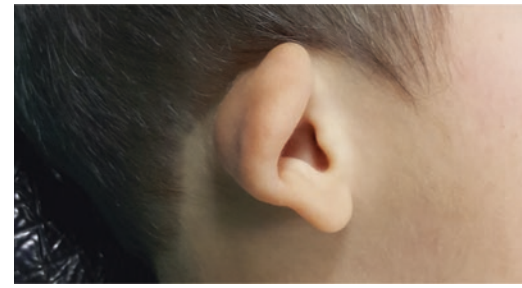


Fig. 19.2 Microtia seen in CLA

19.4 Radiological Findings

In CLA cochlea, vestibule, semicircular canals vestibular, and cochlear aqueducts are absent. Until September 2018, 776 patients with various IEMs were evaluated in the Department of Otolaryngology at Hacettepe University. Out of 1552 ears there are 64 CLA (3.9%) on our database (34 on the right side and 30 on the left). Twenty-three cases are bilateral. Nonsymmetric cases have rudimentary otocyst, IP-I, cochlear hypoplasia, and common cavity on the contralateral side. Most common of these are IP-I and rudimentary otocyst.

According to radiological findings, Sennaroglu et al. [10] reported three subgroups of CLA:

(a) **CLA with aplastic/hypoplastic petrous bone:** No inner ear development is present. Petrous bone may be aplastic (Fig. 19.1a) or hypoplastic (Fig. 19.1b). Due to severe hypoplasia/absence of petrous bone dura may be adjacent to the middle ear in some locations. Middle ear ossicles are always present even though there is most severe inner ear malformation.

(b) **CLA without otic capsule:** As can be seen petrous bone development is normal, but the otic capsule is completely absent (Fig. 19.1c). There is no IAC development. Facial nerve canal cannot be identified. Middle ear ossicles are present. In this subgroup of CLA, the

vascular supply from both IAC and middle ear appears to be severely damaged, resulting in the absence of all three layers of the otic capsule.

(c) **CLA with otic capsule:** This group appears to be the most developed of CLA subtypes. Facial nerve canal (with meatal, labyrinthine, tympanic, and mastoid segments) can be seen on HRCT of the temporal bone (Fig. 19.1d). Middle ear ossicles are present. It may be due to complete absence of vascular supply only from the IAC.

19.5 Differential Diagnosis

It is possible to misdiagnose CLA as complete ossification of cochlea and vestibular system after meningitis. Although cochlea vestibular labyrinth may be completely ossified, bulging of the promontory, and air in the round window niche can be noticed immediately in total ossification after meningitis (Fig. 19.3a, b). This is not found in CLA.

19.6 Audiological Findings

Audiological examination of patients with CLA reveals either no response at all or profound sensorineural hearing loss (SNHL) at 125, 250, and 500 Hz at the upper limits of the audiometer. As there is no inner ear development, this low frequency response demonstrates that this cannot be true hearing, but rather it has to be accepted as a vibrotactile sensation. This can also be seen in patients with a rudimentary otocyst and cochlear aplasia. In fact, reliable results from audiometric test may not always be derived from very young children as this test may be difficult to perform in young children. Audiometric results from a case are given in Fig. 19.4, showing bilateral profound hearing loss.

19.7 Management

As there is no inner ear development, it is not possible to perform cochlear implant (CI) surgery in these children. According to First Consensus Meeting [11], ABI indications are divided into two groups. CLA constitutes a definite ABI indication. Auditory brainstem implantation (ABI), in which the electrode is placed into the lateral recess to directly stimulate the cochlear nuclei, is

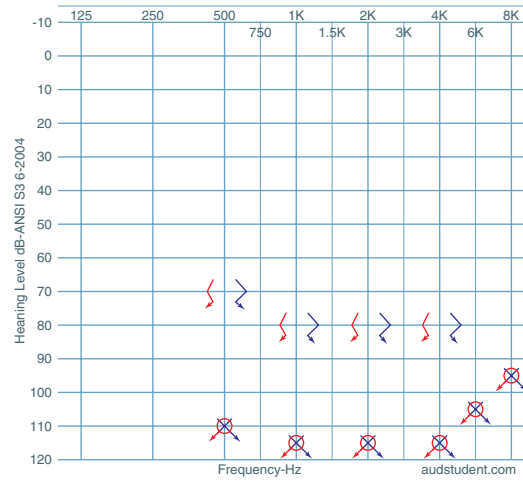


Fig. 19.4 Profound hearing loss in CLA

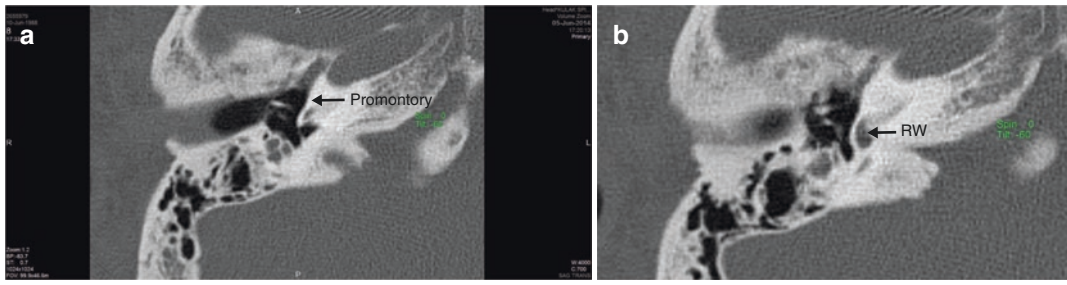


Fig. 19.3 Labyrinthine ossification after meningitis (**a**, **b**). Please note the round window (RW) niche which is absent in CLA. Promontory is produced by the basal turn which is absent in CLA

the only surgical option for hearing habilitation. Although translabyrinthine, retrosigmoid, and retrolabyrinthine approaches can be used for ABI surgery, the retrosigmoid approach is preferred in children [12]. Temporal bone is much smaller in children of 2–3 years of age when compared to that of an adult. As a result, the translabyrinthine approach provides a much more limited surgical exposure than the retrosigmoid approach. In addition, drilling the temporal bone with a translabyrinthine approach to expose the brainstem requires longer surgical times compared to retrosigmoid craniotomy. Therefore, we favor the retrosigmoid approach for ABI surgery in children.

Based on the findings in 124 children who have undergone ABI surgery in Hacettepe University, there is no real correlation between the type of inner ear malformation and the development of the cochlear nuclei. Even in the absence of an inner ear, the cochlear nuclei is developed and it is possible to obtain electrically evoked far-field responses of the upstream auditory pathways (EABR) during ABI surgery. An example of intraoperative eABR recording is given in Fig. 19.5. As can be seen even though whole inner ear is absent, it is possible to obtain Vth wave with intraoperative e-ABR.

Unfortunately, there is no way of preoperatively determining whether the cochlear nuclei are present and functional in children with CLA based on current magnetic resonance imaging (MRI) techniques and evoked response testing methods. Indeed, children with labyrinthine aplasia have demonstrated hearing benefits that mimic those of children with other inner ear pathologies who underwent ABI surgery.

Until September 2018, among 124 children with IEM who had an ABI in Hacettepe University

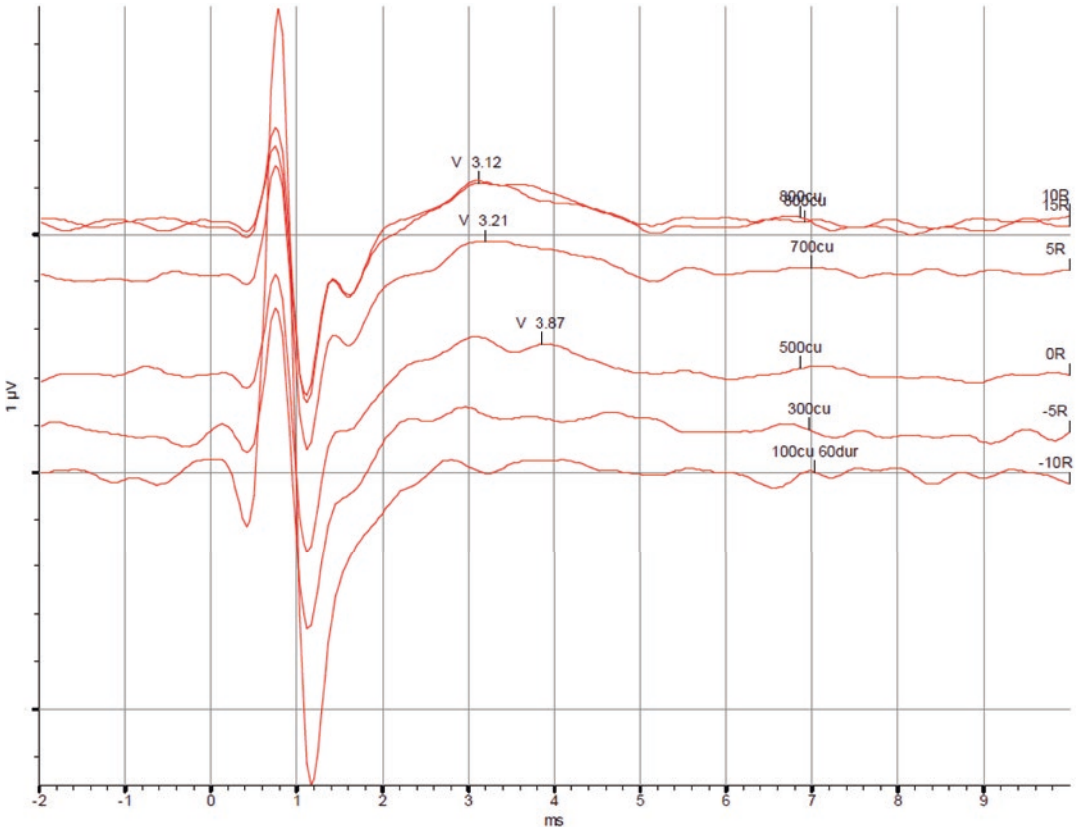


Fig. 19.5 Intraoperative e-ABR recording. Even though whole inner ear is absent, it is possible to obtain Vth wave with intraoperative e-ABR. Recordings are obtained through stimulation of the sixth electrode during the operation of 1-year old male patient

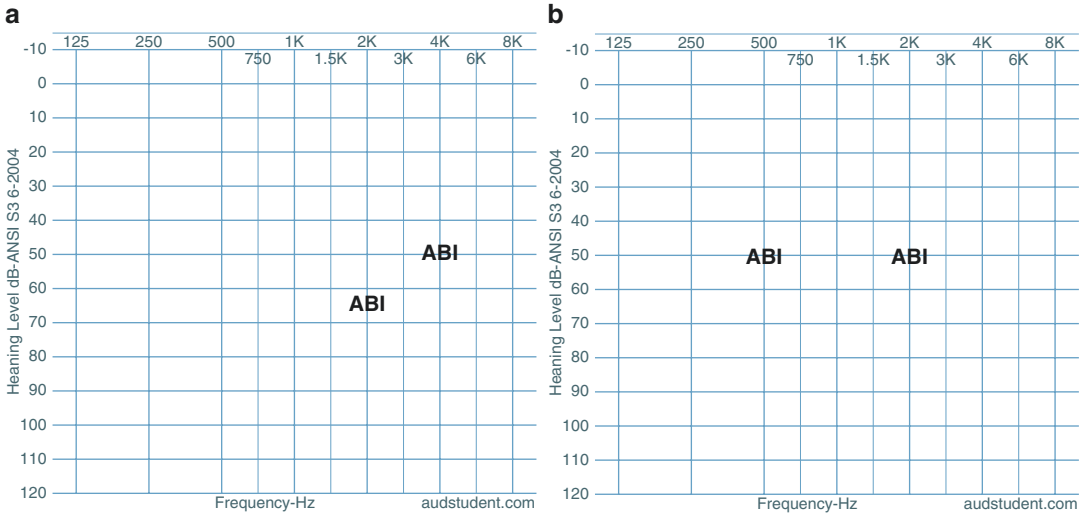


Fig. 19.6 Thresholds 6 months (**a**) and 1 year (**b**) after initial programming. Stability of thresholds is most probably due to lack of compliance of the family with the rehabilitation program

there are 16 CLA. They all had a response with their ABI. This shows that none of these patients with CLA had cochlear nucleus aplasia.

19.8 Outcome with ABI

In terms of free field aided thresholds ABI users with CLA may show various outcomes. Usually thresholds reach better levels with implant use over time. Some cases are given below.

Case 19.1: BEO 1 Year Old Female Patient Operated on 15.11.2016

She had an uneventful ABI surgery in 2016. Six months after the first fitting her thresholds were between 50 and 65 dB (Fig. 19.6). After the first device activation user has very rarely attended our program. Aided thresholds 1 year after the first fitting demonstrated very little improvement. There was a long time gap between the fitting sessions, which is not compatible with our follow-up. Normally in our department fitting sessions are done every 2–3 months within the first year. Stability of thresholds most probably was due to lack of rehabilitation sessions. Ideally, better thresholds are expected over time.

This case shows the importance of compliance with the rehabilitation program comprised of device fitting sessions and auditory training. Not attending to regular programming and therapy sessions may result in sub-optimal aided thresholds with the device.

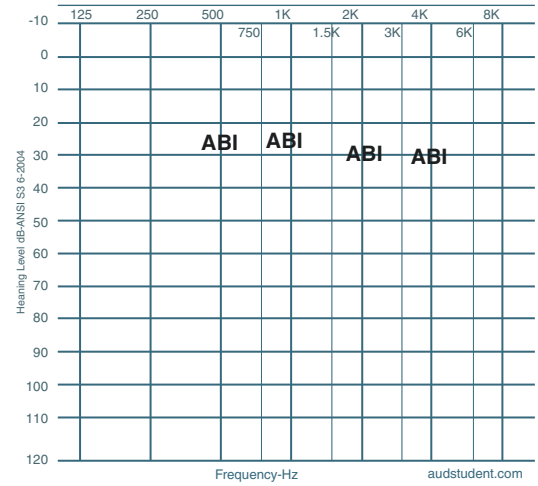


Fig. 19.7 Aided thresholds around 25 to 30 dB after 5 years of ABI use

Case 19.2: SO, 2 Year Old Male Patient, Operated on 24.1.2013

His audiogram after using ABI for more than 5 years demonstrates aided thresholds of 25–30 dB for both speech sounds and pure tones of 500–1000–2000–4000 Hz (Fig. 19.7).

Case 19.3: FRO, 1-Year-Old Female Patient Operated on 18.5.2016

She was implanted in 2016. After successive fitting sessions, her aided thresholds got better both due to increment in device charge levels and the user is getting used to sounds. Test performed at 2 years after programming demonstrated better thresholds when compared to the fitting session done around 6 months after the first fitting (Fig. 19.8). Compared to Case 19.2, who has been using his ABI for more than 5 years, it can be noticed that latter has worse thresholds.

Case 19.4: SO 2 Year Old Male Patient Operated on 24.1.2013

It is common to observe side effects during individual electrode stimulation during testing. This is due to proximity of the implant electrode to the cranial nerves. An interesting observation for this case was disappearance of side effects in time, so the deactivated channels were re-activated (Fig. 19.9). The MAP on left belongs to a fitting session on 2014, Electrodes 2, 3, 12, 13, 14, 15, 16 were deactivated due to vestibular side effects upon individual electrode stimulation (shown in box as AE-active electrode). The MAP on right

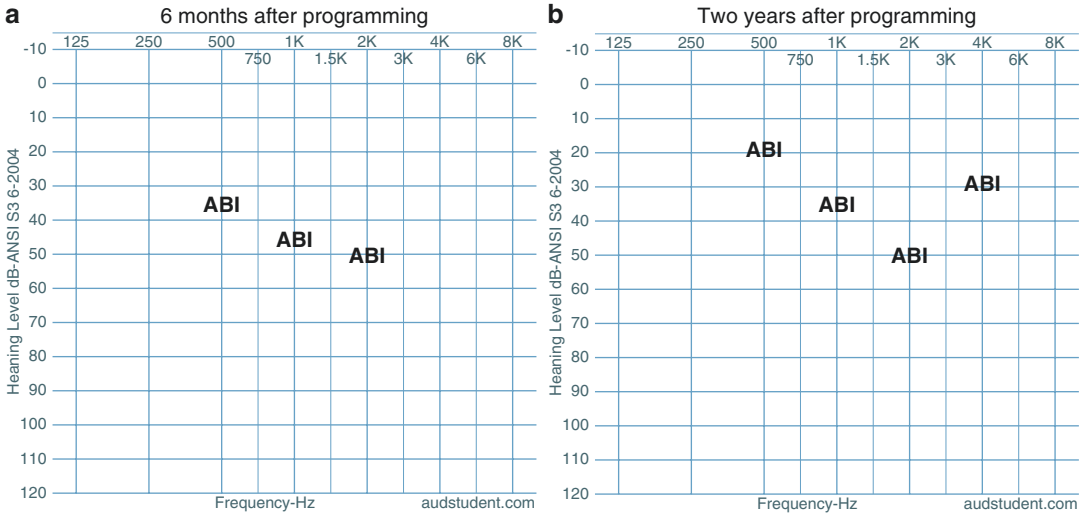


Fig. 19.8 Thresholds 6 months (**a**) and 2 year (**b**) after initial implant programming. She attended fitting sessions and auditory training regularly and showed improvement in thresholds over time

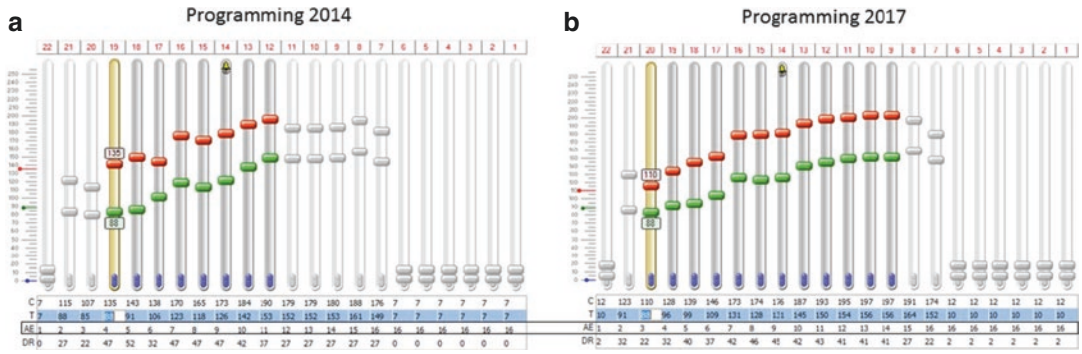


Fig. 19.9 (**a**) The MAP from a fitting session in 2014. (**b**) The MAP from a fitting session in 2017. Electrodes initially causing vestibular side effects were re-stimulated individually in 2017, and no side effect was observed. Upon observing clear responses to stimulation from these electrodes without any side effect, they were re-activated

belongs to a fitting session on 2017. When these electrodes were re-stimulated individually, no side effects were observed. Upon observing clear responses to stimulation from these electrodes, they were re-activated.

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
3. Ozgen B, et al. Complete labyrinthine aplasia: clinical and radiologic findings with review of the literature. AJNR Am J Neuroradiol. 2009;30(4):774–80.
4. Donaldson JA, Duckert LG, EW LPMR, editors. Surgical anatomy of the temporal bone. 4th ed. New York: Raven Press; 1992.
5. Marsot-Dupuch K, et al. CT and MR findings of Michel anomaly: inner ear aplasia. AJNR Am J Neuroradiol. 1999;20(2):281–4.
6. Tekin M, et al. Homozygous FGF3 mutations result in congenital deafness with inner ear agenesis, microtia, and microdontia. Clin Genet. 2008;73(6):554–65.
7. Higley MJ, et al. Bilateral complete labyrinthine aplasia with bilateral internal carotid artery aplasia, developmental delay, and gaze abnormalities: a presumptive case of a rare HOXA1 mutation syndrome. AJNR Am J Neuroradiol. 2011;32(2):E23–5.
8. Jorgensen MB, Kristensen HK, Buch NH. Thaladomide-induced aplasia of the inner ear. J Laryngol Otol. 1964;78:12.
9. Daneshi A, Fahradi M, Asghari A, Emamjomeh H, Abbasalipour P, Hasanzadeh S. Three familial cases of Michel's aplasia. Otol Neurotol. 2002;23:346–8.
10. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
11. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
12. Sennaroglu L, Sennaroğlu G, Atay G. Auditory brainstem implantation in children. Curr Otorhinolaryngol Rep. 2013;1:80–91.

Rudimentary Otocyst

20

Levent Sennaroglu and Mehmet Yaralı

Special Features

1. Pathology between a complete labyrinthine aplasia and common cavity.
2. Definite indication for ABI.

20.1 Definition

A rudimentary otocyst consists of incomplete millimeter-sized representations of the otic capsule (round or ovoid in shape) without an IAC (Fig. 20.1a, b) [1]. Sometimes parts of the semicircular canals may be present. Within the spectrum of inner ear anomalies, this pathology is between a complete labyrinthine aplasia (CLA) and common cavity (CC): In CLA, there is no inner ear development, while in CC there is an ovoid or round cystic space instead of a separate cochlea and vestibule. The CC communicates with the cerebellopontine cistern via the IAC. The rudimentary otocyst is a few millimeters in size and does not communicate with the subarachnoid space via an IAC.

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

M. Yaralı (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

20.2 Histopathology and Pathophysiology

There was no specimen of rudimentary otocyst in the Massachusetts Eye and Ear Infirmary and University of Minnesota temporal bone collections. In addition, no report has been published in the literature. Between the third and fourth week, the inner ear is in the form of an otocyst (otic vesicle). The insult probably occurs at the beginning of the formation of the otocyst and results in rudimentary otocyst deformity (see Chap. 3 Pathophysiology of IEM for more details) [2].

20.3 Clinical Findings

They present with profound sensorineural hearing loss (SNHL).

20.4 Radiology

Until September 2018, 776 patients with various IEMs were evaluated in the Department of Otolaryngology at Hacettepe University. Out of 1552 ears 13 had rudimentary otocyst deformity (0.8%). Only one patient had bilateral rudimentary otocyst. Remaining 11 cases had unilateral RO.

On HRCT and MRI, there is a millimetric inner ear structure with an oval or round shape (Fig. 20.1a, b), and IAC is absent. It is surrounded by otic capsule. The otocyst deformity may be

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_20

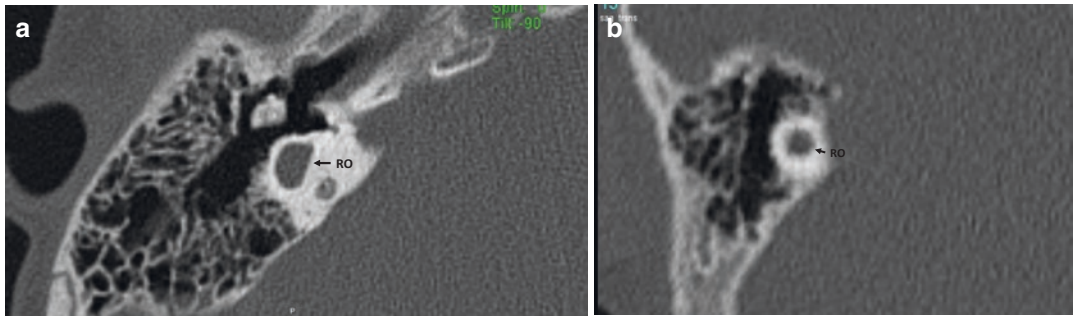


Fig. 20.1 (**a**, **b**) Rudimentary otocyst consisting of millimetric otic capsule remnant surrounded by otic capsule but without an internal **auditory** canal

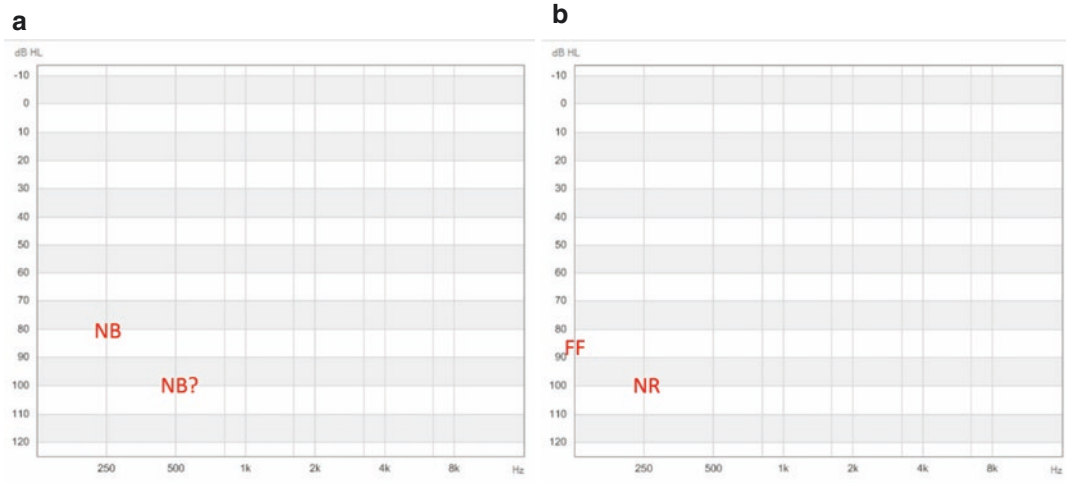


Fig. 20.2 Audiological configuration in rudimentary otocyst. Response in both cases should be accepted as vibrotactile stimulation. (**a**) Pre-op free field audiometric results obtained via narrow band (NB) stimuli. (**b**) Pre-op free field (FF) audiometric results. *NR* no response

accompanied by rudimentary SCC formations. This represents a pathology between CLA and common cavity.

20.5 Audiological Findings

Audiological findings are similar to those found in complete labyrinthine aplasia. There is non-progressive profound SNHL (Fig. 20.2a, b). As discussed before, it is possible to obtain thresholds in low frequencies possibly indicating vibrotactile response. The response should not be regarded as hearing.

20.6 Management

The fact that there is no connection between the otocyst and the brainstem is a contraindication to CI surgery. As a result, auditory brainstem implantation (ABI) is the only option for hearing restoration in these patients. In the first Consensus Meeting there were two categories of ABI indications. Rudimentary otocyst had not yet been defined at that time, but it is now accepted as a definite indication for an ABI [3, 4].

Two patients with rudimentary otocyst had ABI surgery so far in Hacettepe University. Responses to environmental and audiological test

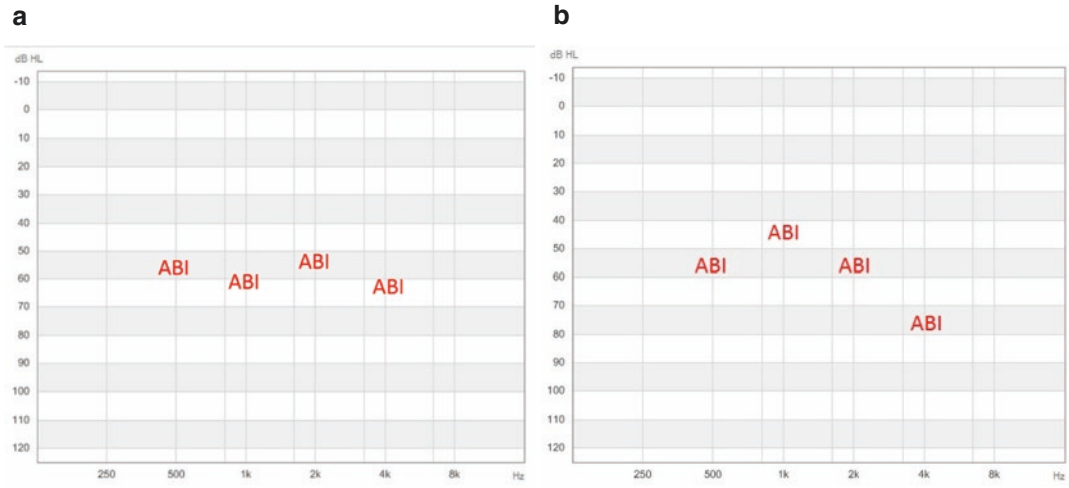


Fig. 20.3 Aided thresholds with auditory brainstem implantation: (**a**) Aided thresholds after 3 years of ABI use, (**b**) Aided thresholds after 2 years of ABI use

stimuli developed in time. Aided thresholds of these ABI users are given in Fig. 20.3a, b.

20.7 Cases

Case 20.1: ZK 1.5 Year Old Female Patient, Operated on 16.12.2014

Audiological testing upon consultation to our department confirmed severe to profound hearing loss for this patient (audiometric results were given in Fig. 20.2a). Based on radiological and audiological findings ABI decision was given. Surgery was done when she was 1.5 years old. After consecutive fittings for 3 year aided thresholds are given in Fig. 20.3a.

Case 20.2: BD 3.5 Year Old Female Patient, Operated on 19.11.2014

Patient presented with severe to profound hearing loss (Fig. 20.2b). Based on audiological and radiological findings ABI decision was given. Surgery was done when she was 3.5 years old. Aided thresholds after 2 years of ABI use are given in Fig. 20.3b.

References

1. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
2. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
3. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
4. Sennaroglu L, et al. 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.

Common Cavity

21

Mehmet Yaralı and Levent Sennaroglu

Special Features

1. A single chamber representing cochlea and vestibule.
2. Two options for hearing rehabilitation: CI or ABI.
3. Meningitis possible but rarer than IP-I.
4. Can be misdiagnosed as cochlear aplasia with vestibular dilatation which is a contraindication to CI surgery.
5. Transmastoid labyrinthotomy or double labyrinthotomy are the methods for cochlear implantation. No need to open facial recess.
6. Electrode position must be checked with transorbital view after all cases. If there is gusher X-ray must be done intraoperatively. If a complication is suspected, a CT is strongly advisable.

21.1 Definition

A common cavity (CC) is defined as a single chamber (ovoid or round in shape) representing the cochlea and vestibule [1] (Fig. 21.1a, b). Theoretically, this structure has cochlear and vestibular neural structures within this single chamber. CC is the last anomaly before a clear separation between cochlea and vestibular system is evident. This is the reason why the term "common cavity" is used to define this abnormality. IAC usually enters the cavity at its center. Semicircular canals (SCC) or their rudimentary parts may accompany CC. Cases with vestibular dilatation are occasionally termed as "vestibular common cavity," which is, however, not a correct term.

CC is believed to contain cochlear and vestibular neural elements, and according to the radiological findings, they have a round or ovoid structure, with an IAC opening into the center. This represents development arrest before there is a clear differentiation into cochlea and vestibule: it is in between rudimentary otocyst and cochlear aplasia [2]. The timing of the insult must be around fourth–fifth week. It must be differentiated from cases of rudimentary otocyst and cochlear aplasia with a dilated vestibule. **Rudimentary otocyst** is an earlier anomaly without an IAC, where performing an ABI surgery is the only way to restore hearing [3]. In cases of CC, if there is a well-developed cochleovestibular nerve (CVN), the patient may benefit from CI; otherwise, ABI is the only treatment option for hearing restoration. **Cochlear aplasia with a dilated vestibule** (CADV) is a more developed anomaly, where the vestibule forms as a completely separate structure, located in its normal position, which is the posterolateral part

M. Yaralı (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_21

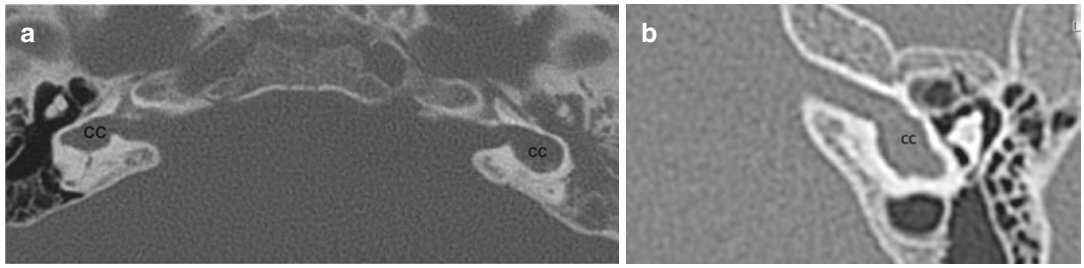


Fig. 21.1 (**a**, **b**) Examples of large common cavity (CC) seen as ovoid structure

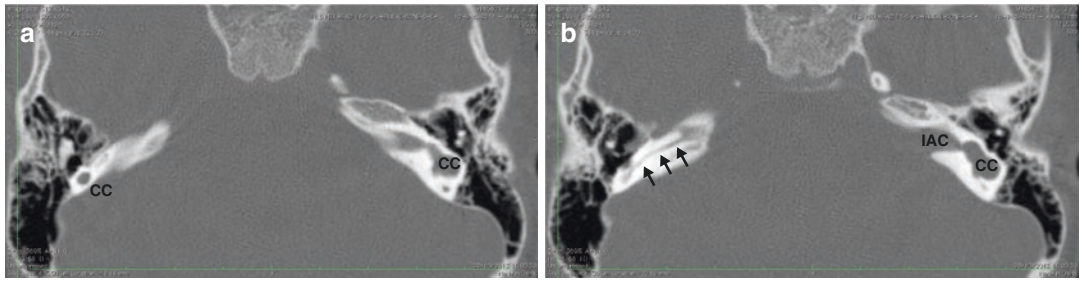


Fig. 21.2 (**a**, **b**) A small CC on the right side with long and narrow IAC. On the left side a normal sized CC and IAC

of the IAC [1]. As the cochlea is absent, an enlarged vestibule can be misdiagnosed as CC.

Clinical Significance of Proper Classification: It is very important to differentiate between rudimentary otocyst and CC for choosing the appropriate treatment modality. While an ABI is the only treatment option for rudimentary otocyst, patients with CC may benefit from CI if there is a well-developed CVN. Similarly, CADV is a definite contraindication for CI, leaving ABI as the only treatment modality. CI surgery in a patient with CADV may be catastrophic as there will be no hearing.

21.2 Radiology

According to the IEMs database of the Hacettepe ENT Department, common cavity represents 6,7 % of the 776 patients with 1552 ears that were evaluated until September 2018.

A **common cavity (CC)** (Fig. 21.1a, b) is an ovoid or round structure. Both images represent large CC. It can also be a small cavity (Fig. 21.2a). The location of a CC may be anterior or posterior to the normal location of the labyrinth, but usually it is located posteriorly. SCCs or their rudimentary parts may accompany a CC. IAC is usually oriented backwards towards the CC and usually enters the cavity at its center. IAC may be normal (Fig. 21.1a, b) or narrow (Figs. 21.2b and 21.3). It is highly possible to have CVN hypoplasia in cases of long and narrow IAC.

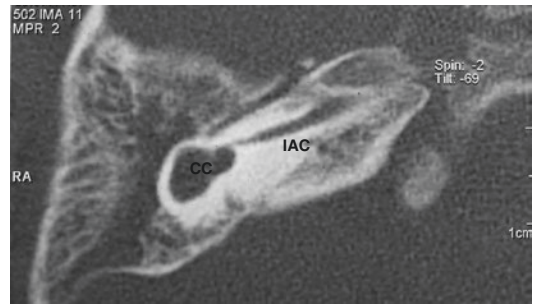


Fig. 21.3 A normal sized CC with a long and narrow IAC

As noted above, it is very important to differentiate between **cochlear aplasia with vestibular dilatation** (CADV) and common cavity. IAC in **CADV** is normal in size, location, and angulation (Fig. 21.4a, b). The vestibule and semicircular canals are located in their normal position which is at the posterolateral part of IAC. The accompanying SCCs may be enlarged or normal,

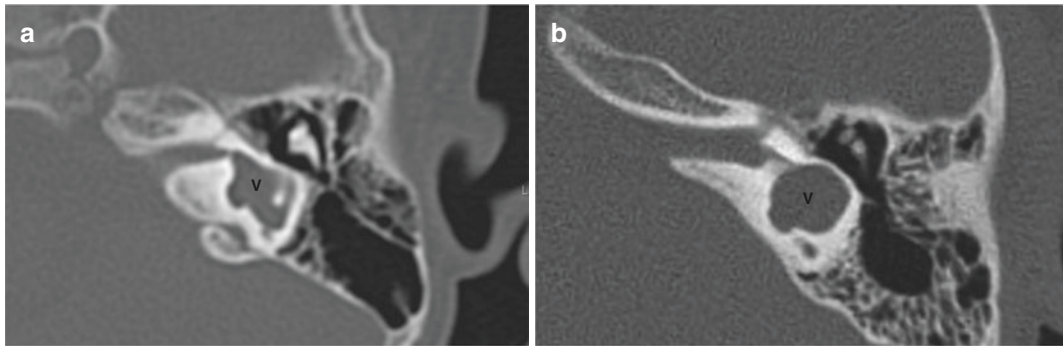


Fig. 21.4 (**a**, **b**) Cochlear aplasia with vestibular dilatation (CADV) where IAC is normal in size, location, and angulation. The dilated vestibule (V) and semicircular canals are located in their normal position which is at the posterolateral part of IAC

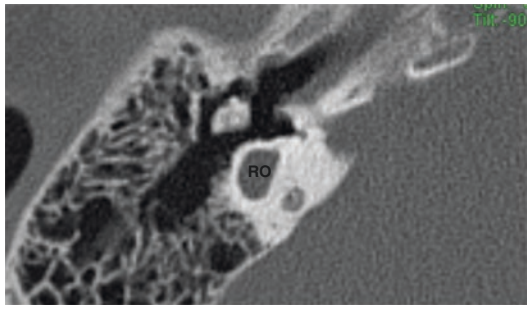


Fig. 21.5 A rudimentary otocyst deformity: rudimentary otocyst (RO) is ovoid or round structure which is a few millimeters in dimensions without any internal auditory canal development

but external outline of vestibule and semicircular canals is similar to normal. The only difference is the dilated vestibule in addition to cochlear aplasia. It is very important to differentiate these malformations from each other, because cochlear implantation in a CC may result in acoustic stimulation, but in CADV, CI will fail with no functional stimulation. In spite of these factors, it may sometimes be difficult to differentiate between CC and CADV.

A **rudimentary otocyst deformity** (Fig. 21.5) is a more primitive malformation than the common cavity deformity. There is a rudimentary, cystic ovoid, or round structure which is a few millimeters in dimensions without any internal auditory canal development.

As CC represents cochlea and vestibule, separate CN and VN are not to be expected. As there is no separate cochlea and vestibule, the single nerve, entering the cavity is most appropriately termed as common cochleovestibular nerve (CVN) (Fig. 21.6a). On sagittal oblique sections typically only two nerves are present in IAC; the nerve anteriorly located is FN, and posterior one is CVN (Fig. 21.6c). If CVN is 1.5 times larger in diameter than FN it can be accepted as normal. Sometimes CVN is hypoplastic if it is thinner in diameter to FN (Fig. 21.6b). In such cases it is difficult to expect a good outcome with CI. With today's technology, it is not possible to determine the amount of cochlear fibers within CVN.

21.3 Clinical Findings

These patients have non-progressive, stable profound SNHL. Recurrent meningitis is possible in this group. Cerebrospinal fluid (CSF) gusher occurred only in two cases out of 19 cases. CSF gusher is much rarer when compared to IP-I and IP-III cases.

Audiological findings: Depending on the cochlear fiber components in CVN, these patients may have detectable hearing thresholds only at low frequencies and at the upper limits of the audiometer. In fact, presence of detectable thresholds during audiometric testing usually signals the presence of cochlear neural tissue, making the patient a candidate for CI. However, no detectable thresholds in audiometric testing, combined with radiologic imaging may make the patient a candidate for ABI, which will be dis-

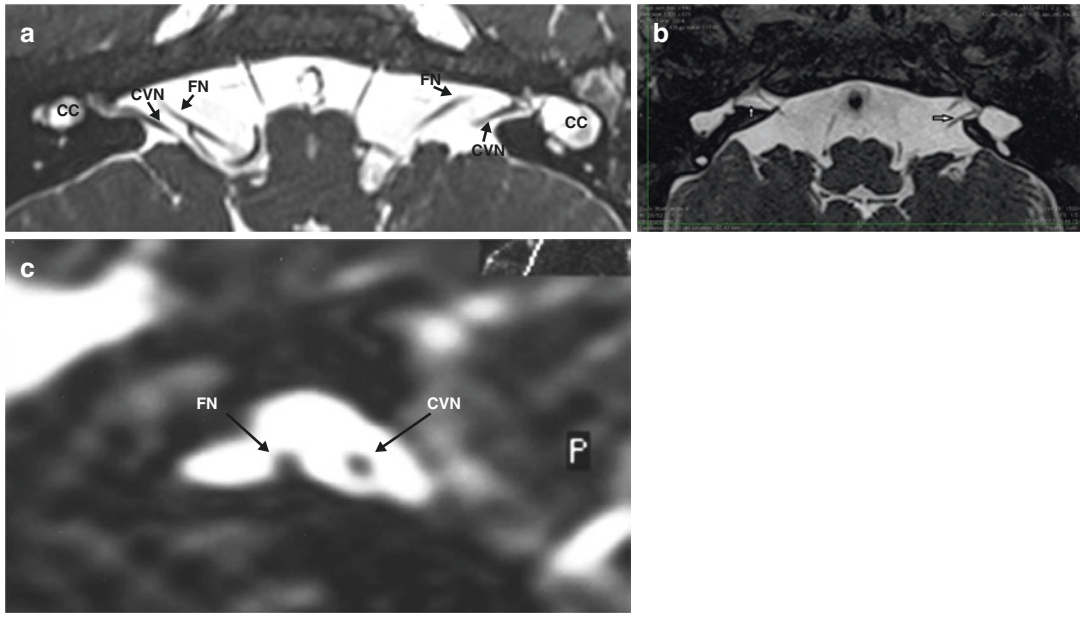


Fig. 21.6 (**a**) Bilateral common cavity (CC) deformity. CVN is slightly larger than facial nerve (FN) and can be accepted as normal on both sides. (**b**) Bilateral CC with normal CVN on the left and hypoplastic CVN on the right. Please note that CVN on the right is much thinner in diameter than ipsilateral FN. (**c**) Sagittal oblique section showing CVN (posterior) and FN (anterior) in the IAC

cussed more in detail in the following section. Figure 21.7a shows the audiometric thresholds of a patient with CC, who has thresholds on the right side and subsequently implanted with a CI. Conversely, a patient with no detectable threshold on audiological evaluation underwent ABI surgery (Fig. 21.7b). Recently it has become our policy to perform CI on the side with thresholds and CVN on MRI, and ABI on the side without auditory response and hypoplastic/aplastic CVN on MRI (Fig. 21.7c).

21.4 Management

The presence of a cochleovestibular nerve (CVN) is important when discussing management options with the family. High resolution 3 Tesla MR imaging with direct sagittal oblique views (rather than reformats) of the internal auditory canal should demonstrate the presence of a CVN before the decision of cochlear implantation. As there is no separate cochlea and vestibule, the nerve entering the CC cannot be named as CN, and it should be termed as CVN. At the present time, there is no method to determine the amount of cochlear fibers in CVN [4]. If the size of CVN is 1.5 times larger than ipsilateral FN it can be accepted as normal. The exact location of the neural distribution is not precisely known in CC. Therefore, audiological outcomes cannot be predicted precisely before CI surgery. Initially the family should be informed that if there is very limited progress with CI, ABI should be done on the contralateral side to provide the best possible bilateral hearing stimulation to the child. This decision should be done as early as possible. CI is always done to the side with better auditory response and better CVN on MRI. CC is among the possible indications of ABI because of this reason. If the child is evaluated between 2 and 3 years of age, which is slightly late for language development, CI and ABI can be considered simultaneously to avoid delay in language development. Simultaneous CI and ABI were done in two cases of CC.

If a behavioral audiometric response or language development is present with hearing aid

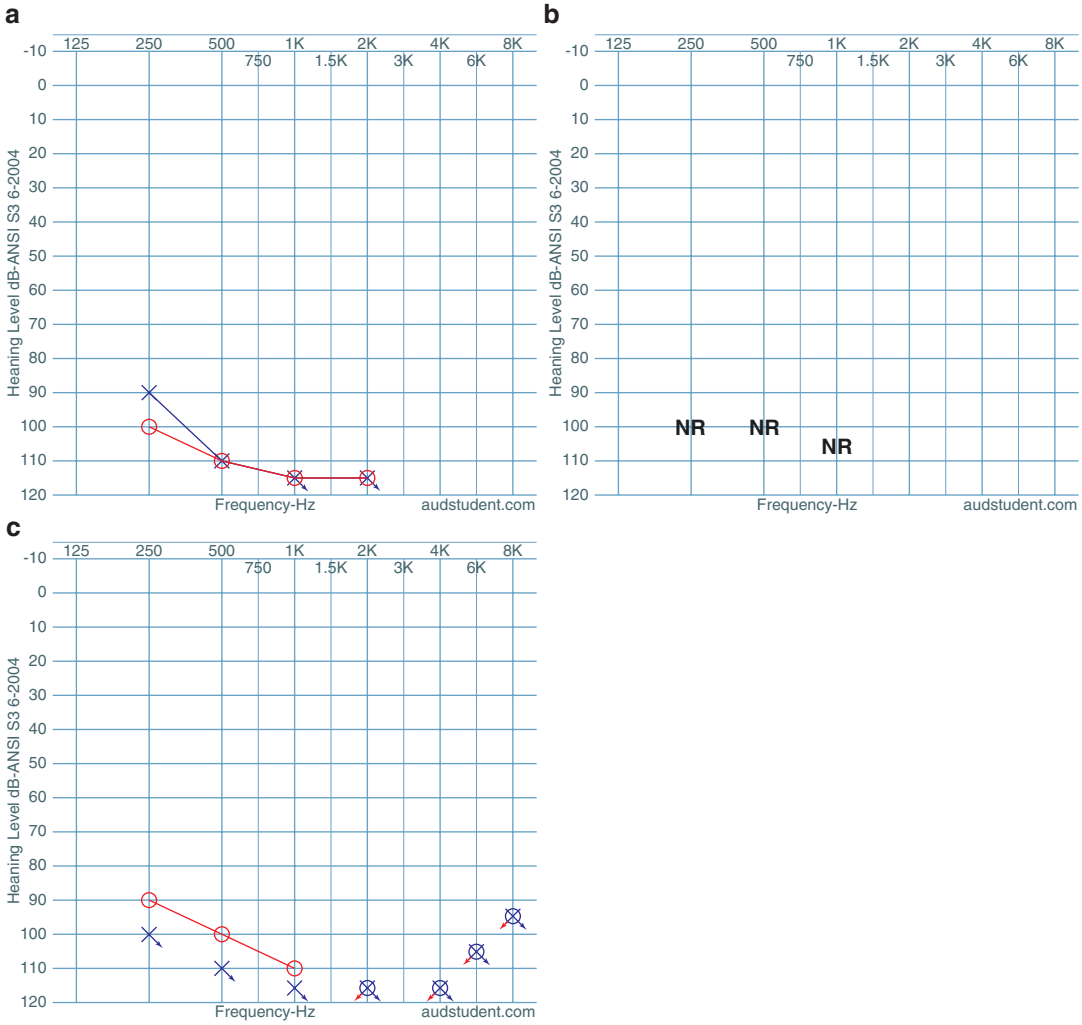


Fig. 21.7 Audiometric configurations in CC. (**a**) Detectable audiometric thresholds on the right side. He received a CI on the right side, (**b**) no detectable threshold on audiological evaluation (*NR* no response). She received an ABI on the right side. (**c**) Thresholds on the right side only. This patient underwent CI on the right and ABI on the left side

use, a meaningful population of cochlear fibers is assumed to be existent and the patient is a candidate for CI. Among audiological test, insert ear phone testing brings valuable information. Insert earphone test results of a patient are given in Fig. 21.7c. Due to clear responses with insert earphone testing a CI was planned on the right side. On the left side there was no audiological response with extremely hypoplastic CVN on MRI. In 2016 she received a CI on right, ABI on left simultaneously.

21.5 Surgery

There are different surgical approaches used for CI surgery in CC. Most popular approach is via a **transmastoid labyrinthotomy** as described by McElveen [5]. After a simple mastoidectomy, air cells are removed to identify the common cavity. Originally the position of the labyrinthotomy was described to be located in the lateral SCC; however, experience in our department has shown that any position along the cavity allows for safe entry

into the CC. Therefore, a labyrinthotomy location at the periphery of the CC, away from the facial nerve, is preferred. A straight (non-modiolar hugging) electrode is preferred, resulting in a lateral position of the electrode. A pre-curved electrode has contacts located medially and will be curved at the center. As a result, it may not stimulate the periphery of the CC efficiently.

It is not necessary to open the facial recess. Usually it will not add any benefit to surgical exposure. As indicated before, CC is usually located posterior to ear canal, and mastoidectomy is sufficient to identify the CC and perform electrode insertion via labyrinthotomy. During the first transmastoid labyrinthotomy operation in Hacettepe University (1999), facial recess was opened but no promontory could be identified. This is because there is no separate cochlear formation anteriorly which will produce the bulging of promontory.

Postoperative transorbital X-ray is absolutely necessary after CI surgery and it should demonstrate circular shaped electrode placement that follows the internal curvature of the common cavity. In a non-complicated surgery, a CT is not necessary after CI surgery. If there is CSF gusher, transorbital X-ray should be taken intraoperatively. The presence of a CSF gusher confirms a connection between CC and IAC, which carries the possibility of electrode migration into IAC. If the electrode is in the IAC, it should be repositioned intraoperatively using a double labyrinthotomy approach. Recently we operated a CC where the electrode was misplaced into IAC even though there was no gusher (see below).

In patients with a very narrow or long IAC where the presence of cochlear fibers is questionable (Figures right side on 21.2) and 21.3), an ABI may be a more appropriate option from the outset. Additionally, if there is no CVN demonstrable by MRI, ABI is definitely indicated.

Double labyrinthotomy technique: This technique was described by Beltrame et al. [6] to avoid the tip of the electrode entering the IAC. Two close labyrinthotomy holes 1–2 mm apart are created. After insertion the tip is directed outside with a tiny 90° hook. This maneuver is done gently to avoid damage to the tip. The electrode is then inserted into the CC while the tip is stabilized at the second hole. This will prevent inadvertent entry of the electrode into the IAC. The surgeon must handle the electrode delicately to avoid damaging the tip.

Later Beltrame et al. [7] developed a special electrode for cochlear implant surgery in for CC. This electrode has a non-stimulating tip which is inserted into one labyrinthotomy hole and then delivered through a second labyrinthotomy with a microhook. This is done to avoid damage to the active part of the electrode during insertion via double labyrinthotomy technique. In this technique two labyrinthotomy holes are produced. They are separated by a few millimeters. Electrode is inserted from the posteroinferior one and inactive tip is grasped and taken out via the superior hole. Then the electrode is pushed backwards into the cavity. This is to avoid entry into IAC and provide stimulation around the periphery of the cavity where the neural elements are thought to be located.

Electrode choice: Because exact location of neural tissue in CC is unpredictable, electrodes with contacts on both sides or full ring electrodes should be used. In case of CSF gusher, FORM electrode with a cork type silicon stopper is advantageous in controlling the CSF leakage. The length of the electrode can be calculated by measuring the diameter of the CC and using the formula $2\pi r$ to estimate the perimeter [1]. This roughly determines the length of the electrode that will make one full turn inside the CC. There are two types of FORM electrodes: FORM 19 (19 mm in length) is ideal for small CC, while FORM 24 (active length 24 mm) can be used in large CCs. Pre-curved electrodes with contacts facing the modiolar side will coil within the open lumen of the CC and not likely stimulate the neural elements found along the lateral wall. Therefore, it is not advisable to use pre-curved electrodes in CC.

21.6 Cases

Between November 1997 and September 2018, 2639 CI and ABI have been performed by Hacettepe Implant team. Four hundred and two of these had IEM. Forty had CC.

CI Cases:19 patients with CC have been implanted with CI so far in our series (2 of which had revisions). Progress in terms of aided thresholds with CI vary from case to case. They usually reach to aided hearing levels of daily speech sounds, but further progress in terms of auditory skills may be limited in some cases (will be discussed in Chap. 30 Audiological outcome with CI). One such patient is presented in Fig. 21.8a. She has been using a CI for 2 years, and her aided thresholds are within daily conversation limits but in terms of auditory skills her progress is suboptimal. The best decision would have been an ABI for the contralateral ear, as no CVN is detectable on the other ear. This case shows the importance of monitoring the auditory and speech skills by a specialist, so that appropriate measures can be taken when required.

Most update treatment option is to use CI on the better ear and ABI on the worse ear (where there is no audiological response and aplastic/hypoplastic CVN) applied in the same surgical session. One case is presented below. She had a CI and ABI in the same surgical session. Nine months after the surgery aided thresholds with CI and ABI are given in Fig. 21.8b. With both devices she responds to speech sounds at 35–40 dB HL. Around 2 months after the initial fitting, her Meaningful Auditory Integration Scale score was 25 over 40, showing that she had lowest scores. Nevertheless, she was observed to be developing her communication skills in auditory-verbal situations. Further information about rehabilitative outcomes of this case will be discussed in Chap. 32.

21.6.1 CI Cases

Case 21.1: MU, 5-Year Old Male Patient, Operated on November 1999

He had three episodes of meningitis prior to his application to our department. His HRCT and MRI revealed bilateral CC without any fluid in the middle ear and mastoid. His preoperative CT showed bilateral CC (Fig. 21.9a). He had a CI surgery via transmastoid labyrinthotomy approach in November 1999. He experienced severe nystagmus during initial programming of CI 1 month after surgery. His postoperative CT showed the electrode placement inside CC (Fig. 21.9b). His current levels were lowered until there was hearing with no nystagmus. He was followed weekly and every week it was pos-

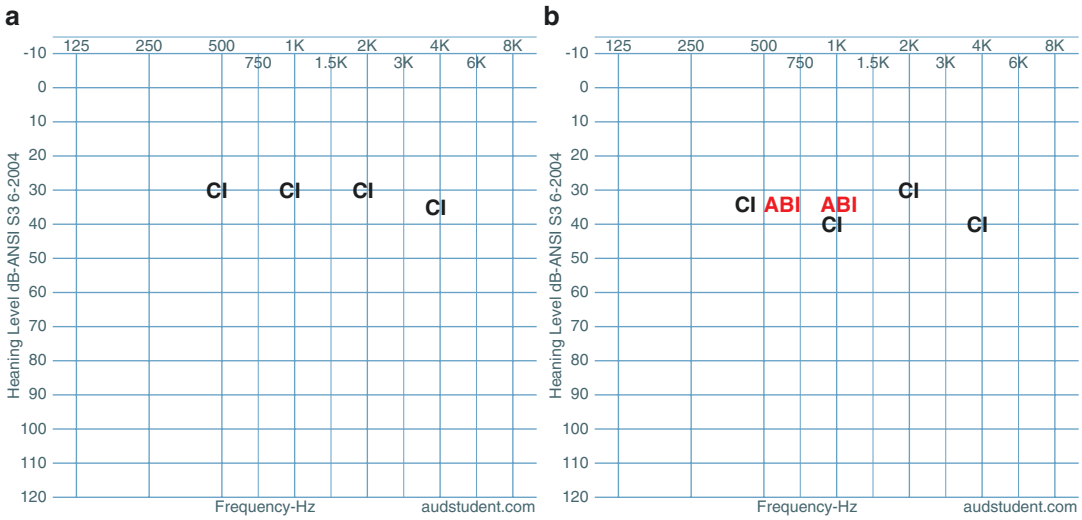


Fig. 21.8 (**a**) After using CI for 2 years aided thresholds reached around 30–35 dB within daily conversation limits, but language skills are suboptimal, making her a candidate for ABI on the contralateral ear. (**b**) Aaided thresholds of a patient with simultaneous CI and ABI 9 months after surgery; with both devices she responds to speech sounds at 35–40 dB HL

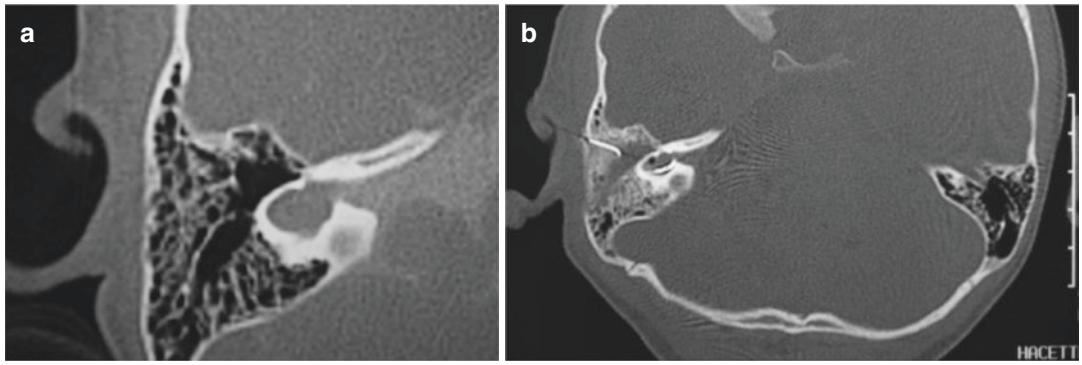


Fig. 21.9 (**a**) Preoperative CT showing CC on the right side, (**b**) HRCT showing electrode placement in CC

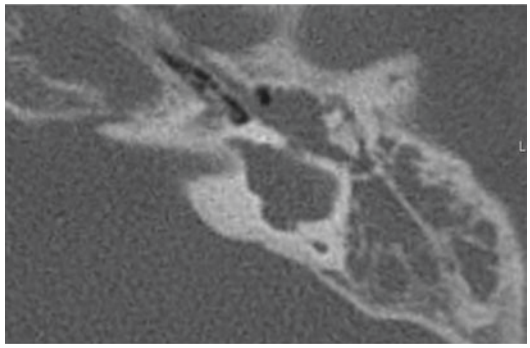


Fig. 21.10 HRCT showing CC with a soft tissue mass in the middle ear on the left side

sible to raise the current levels slightly without causing nystagmus. After 3 months there was no nystagmus even with high current levels. This patient showed that it is possible to stimulate vestibular components of the CVN with CI. Fortunately, this showed progressive adaptation over time and nystagmus completely disappeared after 3 months. He is one of the best performers in the whole CI group in terms of language development.

Case 21.2: FEÇ 4-Year-Old Male, Operated on 31 July 2015

He had recurrent meningitis before applying to our department. His HRCT and MRI showed a soft tissue mass in the middle ear on the left side, in addition to bilateral CC (Fig. 21.10). He had a CI on July 2015 on the right side via transmastoid labyrinthotomy. In September 2015 his left ear was explored via endaurally and a congenital cholesteatoma was discovered. It was completely removed. Most probably it is a coincidental finding to have a congenital cholesteatoma together with CC. CI surgery was done initially because of his late application not to delay language development more. The side with cholesteatoma was not preferred for CI surgery because of the risk for recurrence.

Case 21.3: NSC 2-Year-Old Female, Operated on December 2015

She had a CI surgery via transmastoid labyrinthotomy. There was no CSF gusher intraoperatively. Postoperative X-ray and then HRCT demonstrated electrode migration into IAC (Fig. 21.11a, b). This is a very interesting case as it is the first case where electrode migrated into IAC without gusher. Electrode migration can be expected in case of gusher. Next day electrode was repositioned via double labyrinthotomy (Fig. 21.11c).

Case 21.4: DED 2-Year Old Female, 23 June 2016

She had bilateral CC with a good sized CVN on the right side (Fig. 21.12a) and a hypoplastic CVN on the left side (Fig. 21.12b). Her audiological examination with insert ear phones revealed clear response on the right side but no response on the left side (Fig. 21.7c). She underwent simultaneous CI and ABI surgery. During transmastoid labyrinthotomy there was a CSF gusher on the right side. Nucleus 24 K straight electrode was placed into the cavity. Intraoperative

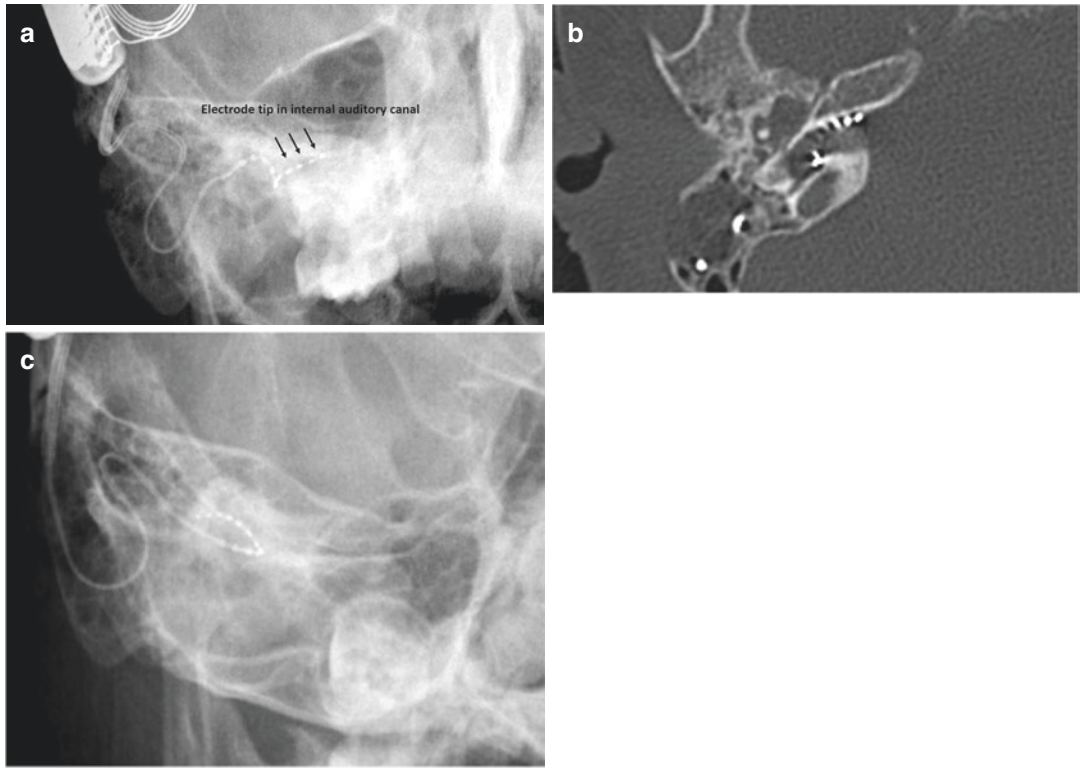


Fig. 21.11 Postoperative imaging demonstrating electrode migration into IAC: (**a**) transorbital X-ray, (**b**) CT findings, (**c**) repositioned electrode after double labyrinthotomy

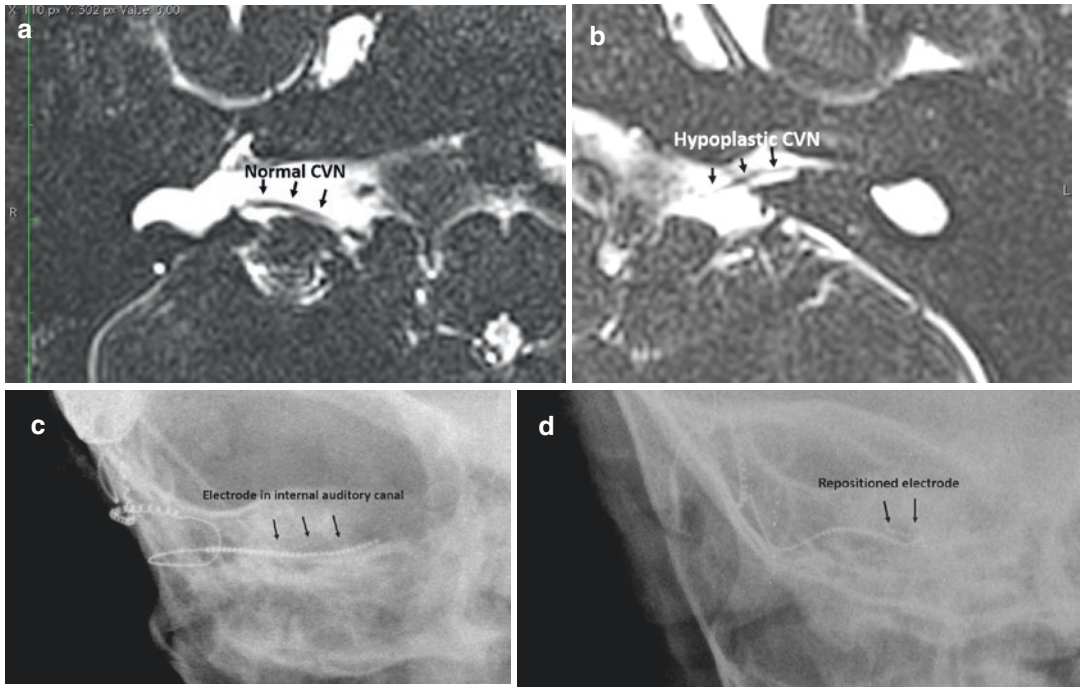


Fig. 21.12 Patients with bilateral CC who underwent simultaneous CI and ABI; (**a**) a good sized CVN on the right side, (**b**) hypoplastic CVN on the left side, (**c**) Intraoperative transorbital X-ray demonstrating a straight electrode indicating electrode migration into IAC, (**d**) view after repositioning using double labyrinthotomy approach

transorbital X-ray demonstrated electrode migration into IAC (Fig. 21.12c). Electrode was immediately repositioned using double labyrinthotomy approach (Fig. 21.12d).

Case 21.5: MB 2.5-Year-Old Male. Operated on March 2011

He had CI surgery on the right side via transmastoid labyrinthotomy approach on March 2011 (Fig. 21.13a). Electrode placement was close to labyrinthine segment on the FN. He demonstrated good audiological development at the beginning with his CI. After 10 months, he started to have FN stimulation. At the beginning it was on few electrodes but soon it was present on almost all electrodes even with very low current levels so that he was unable to use the CI. HRCT demonstrated the tip of the electrode to be in the vicinity of the labyrinthine segment (Fig. 21.13b). On May 2012 he underwent a revision surgery where electrode was repositioned via double labyrinthotomy (Fig. 21.13c). Although electrode was placed away from labyrinthine segment, FN stimulation occurred once again. In January 2013 he underwent a contralateral ABI (Fig. 21.13d).

21.6.2 ABI Cases

Tweny-one cases with CC have been implanted with ABI so far in our series. Despite the fact that individual variations exist, similar to other users of ABI with different inner ear malformations, ABI users with CC usually start to respond to environmental sounds within first 6 months to 1 year. This time interval may shorten based on individual factors and family adherence to rehabilitation program or lengthen due to the presence of comorbid problems such as autism spectrum disorder, mental retardation, and possible learning disabilities along with low compliance with rehabilitation. The aided thresholds improve to a better level in time, reaching to daily conversation limits. In a previous study of our group, among the pediatric ABI users with IEMs the best language outcomes and aided thresholds were found to belong users with CC [8]. This is

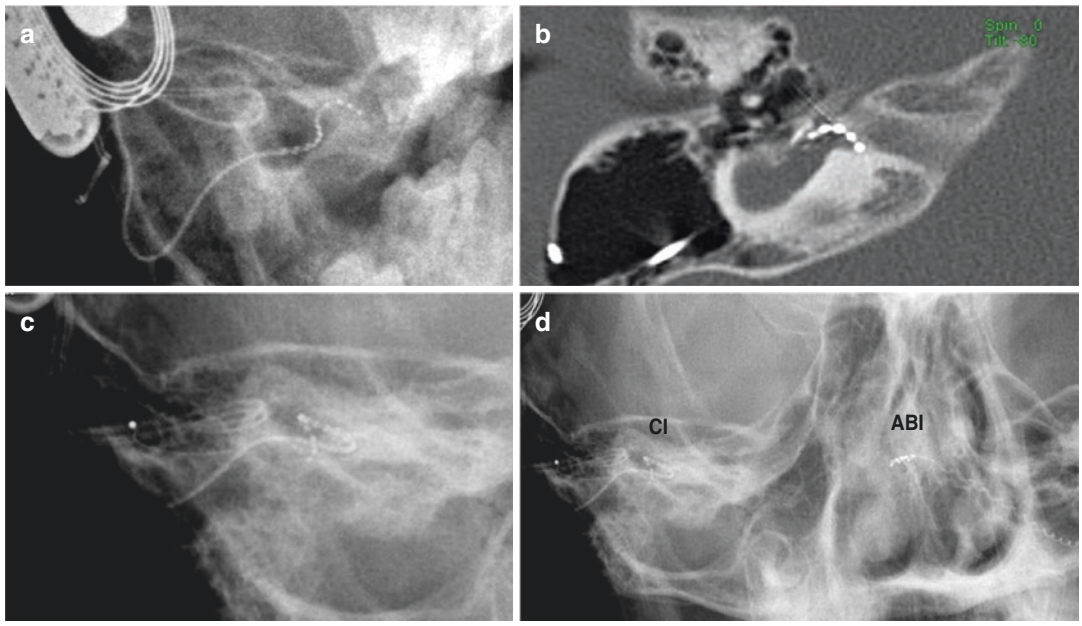


Fig. 21.13 (**a**) Transorbital X-ray demonstrating electrode placement towards IAC, (**b**) HRCT showing the tip of the electrode to be in the vicinity of the labyrinthine segment, (**c**) transorbital view after repositioning via double labyrinthotomy approach, (**d**) after contralateral ABI

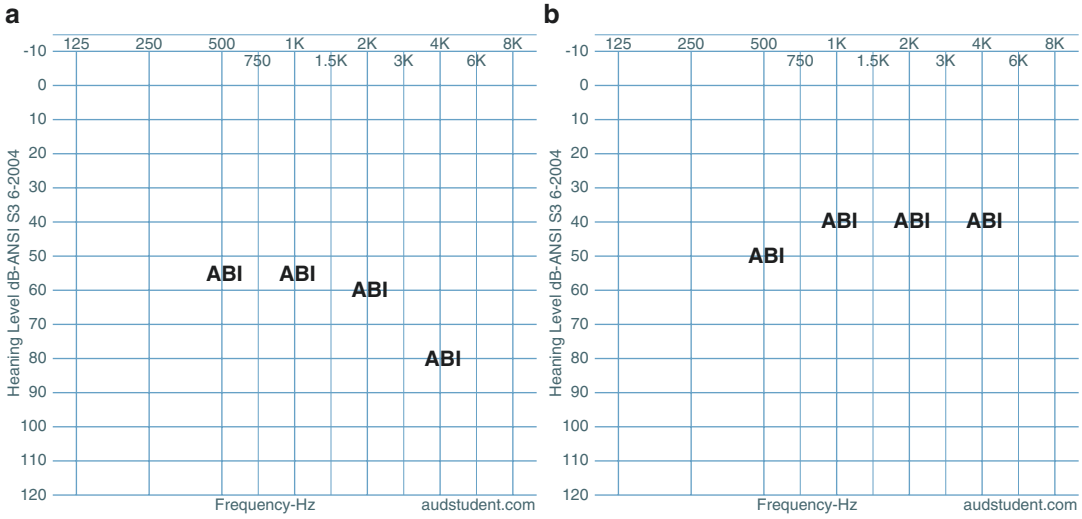


Fig. 21.14 An ABI user with improvement in aided thresholds in time: Aided thresholds after 1 year (**a**), and 3.5 years (**b**) of implant use

most probably due to the fact that the nerve supplying CC is CVN and contains some cochlear fibers. It is possible that they may provide some stimulation to the brainstem even before implantation. Some case examples who use ABI are given below. Please note the improvement in aided thresholds with longer implant use.

Case 21.6: ZDE 4-Year-Old Female Patient Operated on 8.5.2015

After her ABI surgery and first fitting she complied very well with the training sessions and rehabilitative program. This resulted in fast progress in terms of responding to environmental sounds and auditory development. Her aided thresholds with ABI in two different fitting sessions are given in Fig. 21.14. Audiogram on the left shows aided thresholds after 1 year of implant use, and audiogram on right shows aided thresholds after three and a half year of implant use. Please note improvement in aided thresholds in time.

Case 21.7: ST 13-Year-Old Female Patient Operated on 6.2.2008

She has been using ABI for 10 years. Her aided thresholds showed improvement over time (Fig. 21.15). Similar to Case 21.1, she attended fitting sessions and rehabilitative program regularly.

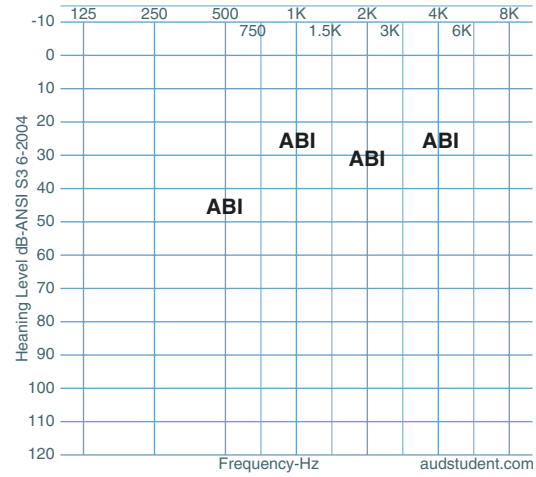


Fig. 21.15 ABI user for 10 years. It is possible to observe thresholds around 25–30 dB in cases who have been using their implants for more than 5 years

Case 21.8: AG 6-Year-Old Female Patient Operated on 19.2.2014

This is another case who attends regularly to our program. In Fig. 21.16, aided thresholds at two different fitting sessions are given. Audiogram on left shows aided thresholds 6 months after the surgery, and audiogram on right shows aided thresholds 1 year after the surgery. The reader may notice improvement in aided thresholds in time with regular device use.

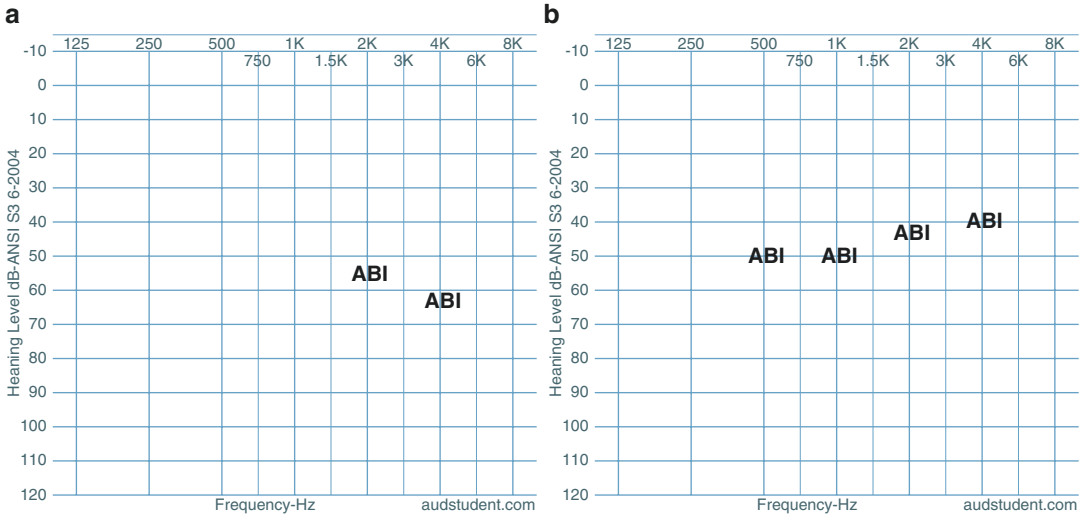


Fig. 21.16 Aided thresholds with ABI 6 months (**a**) and 1 year (**b**) after the surgery

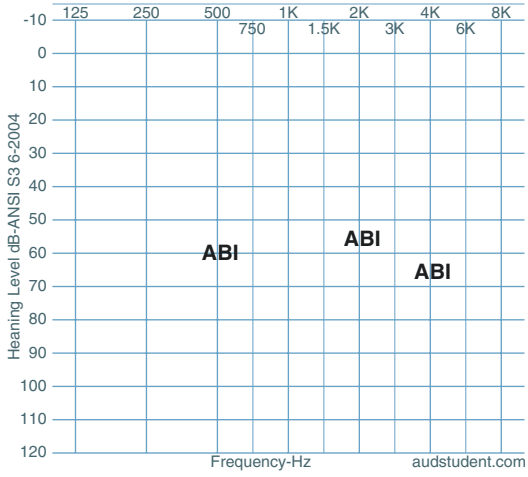


Fig. 21.17 Case 21.9. He has been using ABI for 1 year. He was recently diagnosed as autism spectrum disorder. Although he is not compliant with tests and fitting sessions due to inattention, he responds to test stimuli and environmental sounds

Case 21.9: D.D. 6-Year-Old Male Patient Operated on 20.04.2016

This case has been diagnosed with autism spectrum disorder recently in 2018. Although he is not well compliant with fitting sessions and aided gain tests due to his comorbid problem, he responds to test stimuli and environmental sounds. The aided thresholds may be suboptimal, but keeping in mind his comorbid problem these thresholds may in fact be supra-thresholds. Despite his comorbid problem he derives benefit from the implant as he uses his device all day and responses to environmental sounds and test stimuli are improving (Fig. 21.17).

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
3. Sennaroglu L, Bajin MD. Classification and current management of Inner ear malformations. Balkan Med J. 2017;34(5):397–411.
4. Sennaroglu L, Sennaroglu G, Atay G. Auditory brainstem implantation in children. Curr Otorhinolaryngol Rep. 2013;1:80–91.
5. McElveen JT Jr, et al. Cochlear implantation in common cavity malformations using a transmastoid labyrinthotomy approach. Laryngoscope. 1997;107(8):1032–6.
6. Beltrame MA, Bonfioli F, Frau GN. Cochlear implant in inner ear malformation: double posterior labyrinthotomy approach to common cavity. Adv Otorhinolaryngol. 2000;57:113–9.
7. Beltrame MA, et al. Double posterior labyrinthotomy technique: results in three Med-El patients with common cavity. Otol Neurotol. 2005;26(2):177–82.
8. Sennaroğlu L, Sennaroğlu G, Yücel E, Bilginer B, Atay G, Bajin MD, Mocan BÖ, Yaralı M, Aslan F, Çınar BÇ, Özkan B, Batuk MÖ, Kirazlı ÇE, Karakaya J, Ataş A, Saraç S, Ziyal İ. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.

Cochlear Aplasia

22

Betul Cicek Cinar, Mehmet Yaralı, and Levent Sennaroglu

Special Features

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

22.1 Definition

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

22.2 Histopathology and Pathophysiology

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

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

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

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

B. C. Cinar (✉) · M. Yaralı
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_22

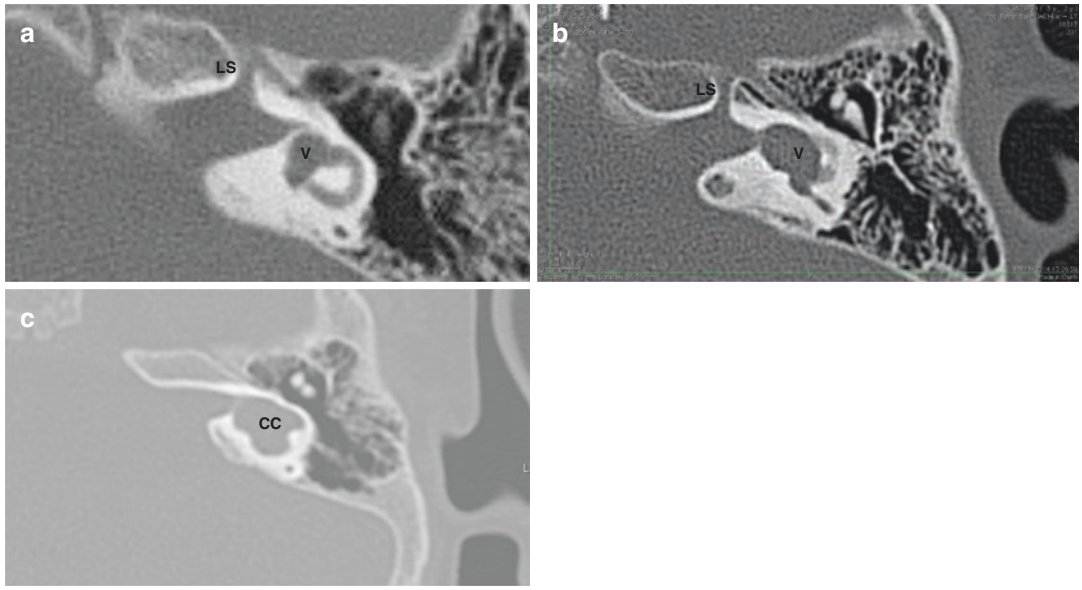


Fig. 22.1 (**a**) Cochlear aplasia with a normal vestibule (V), (**b**) cochlear aplasia with a dilated vestibule, (**c**) common cavity (CC). Please note that labyrinthine segment of the facial nerve (LS) is anteriorly dislocated to the usual location of cochlea

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

22.3 Literature Review

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

22.4 Clinical Findings

They present with nonprogressive profound SNHL.

22.4.1 Radiology

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

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

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

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

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

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

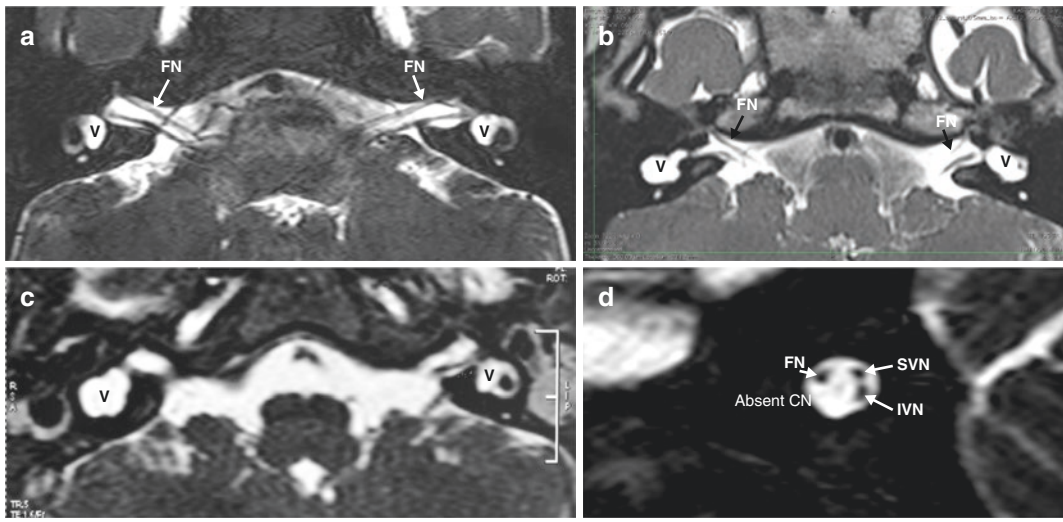


Fig. 22.2 Axial MRI demonstrating bilateral symmetric cochlear aplasia with normal vestibule (**a**) and dilated vestibule (**b**). It is also possible to encounter cochlear aplasia with normal vestibule (left) and dilated vestibule (right) in the same patient (*V* vestibule, *FN* facial nerve) (**c**).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

22.4.2 Audiological Findings

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

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

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

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

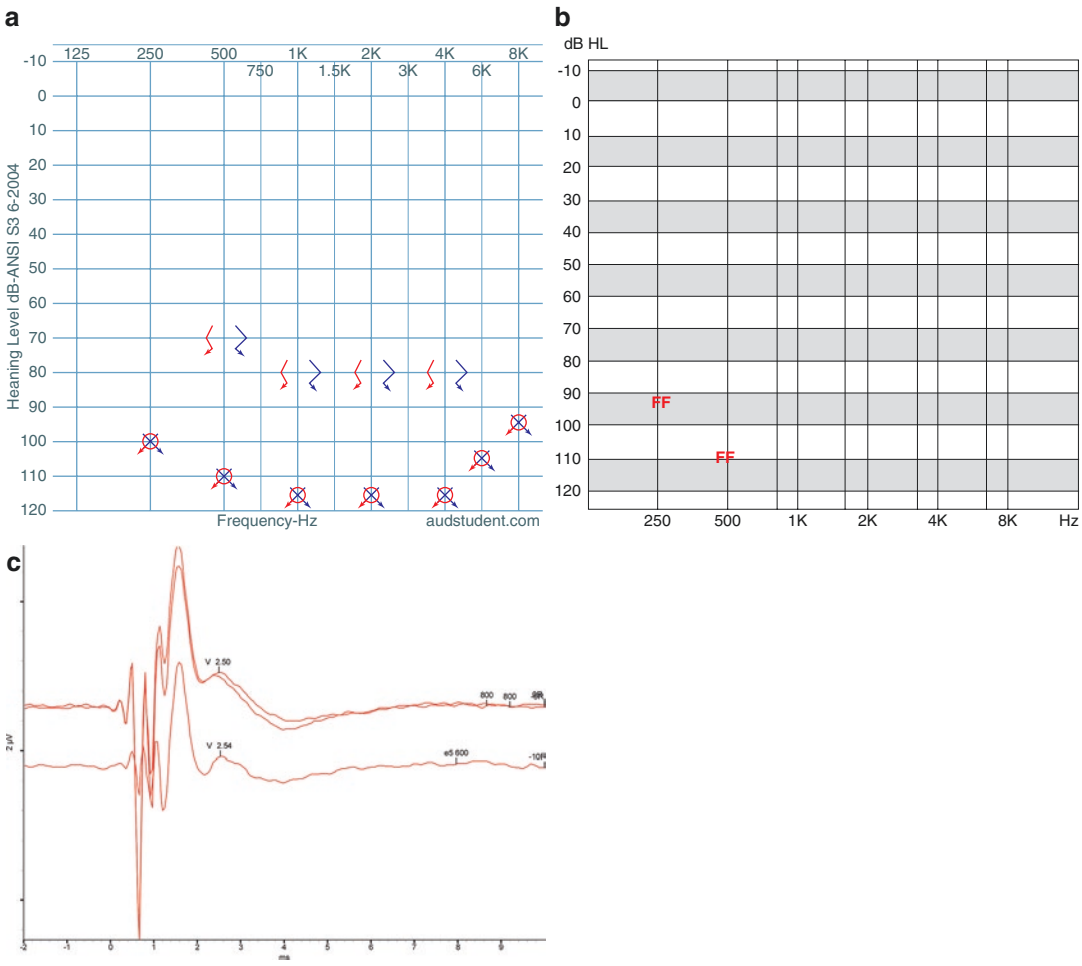


Fig. 22.3 Audiological findings in cochlear aplasia; No response (**a**), profound hearing loss at low frequencies (**b**). An example of intraoperative eABR wave from ABI (**c**)

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

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

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

22.5 Cases

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

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

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

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

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

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

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

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

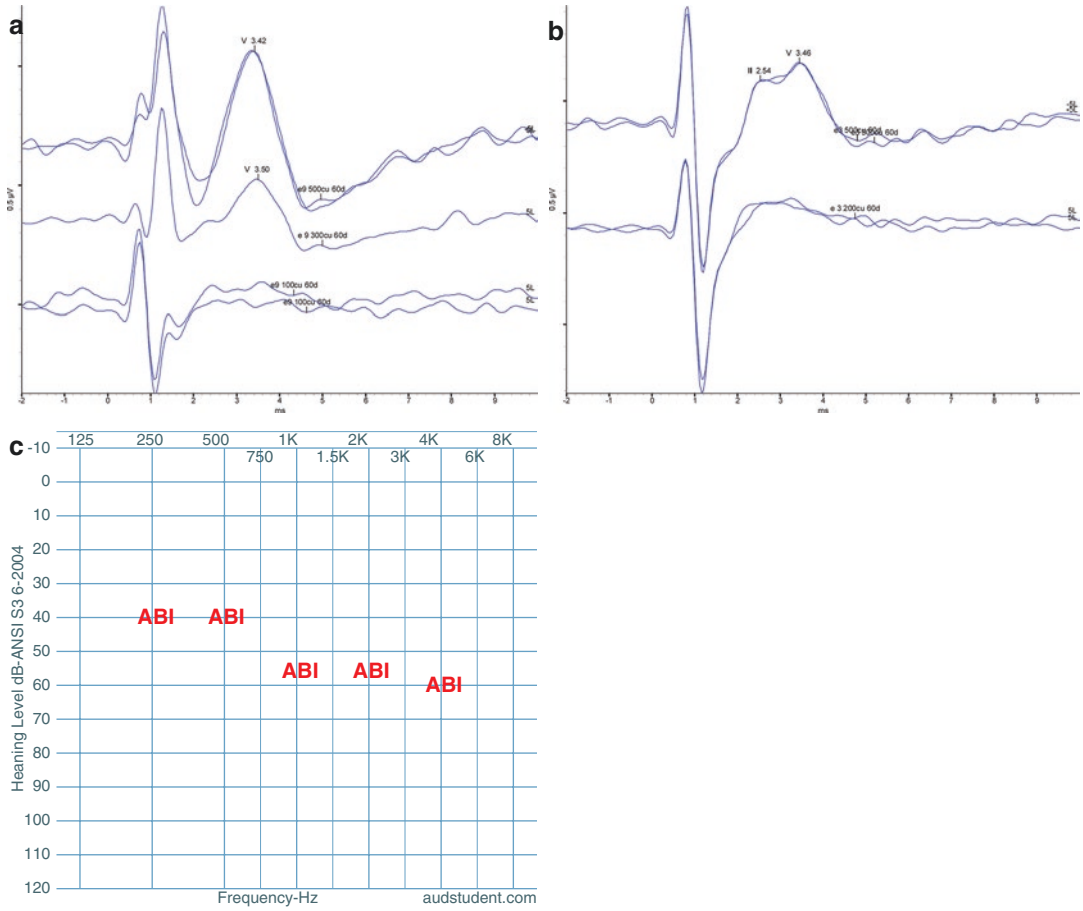


Fig. 22.4 (**a**) and (**b**) Case 22.1. Intraoperative eABR recordings from ninth (*A*) and third (*B*) electrodes. (**c**) Free field thresholds with ABI

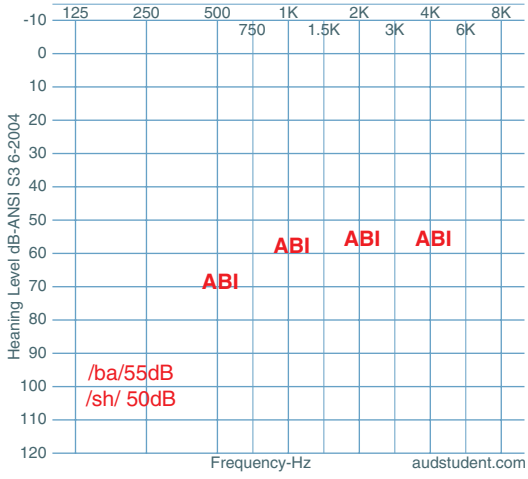


Fig. 22.5 Case 22.2. Free field thresholds with ABI

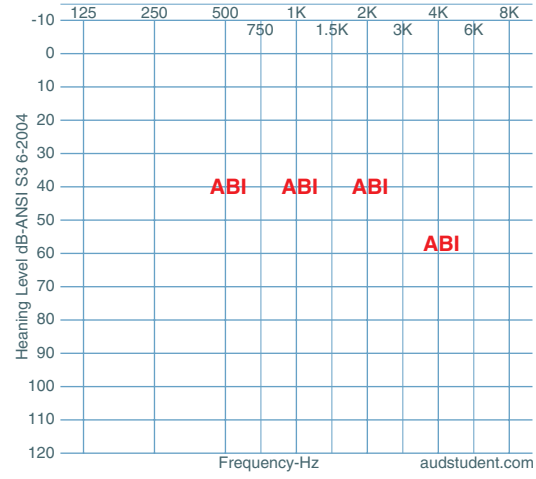


Fig. 22.6 Case 22.3. Free field thresholds with ABI

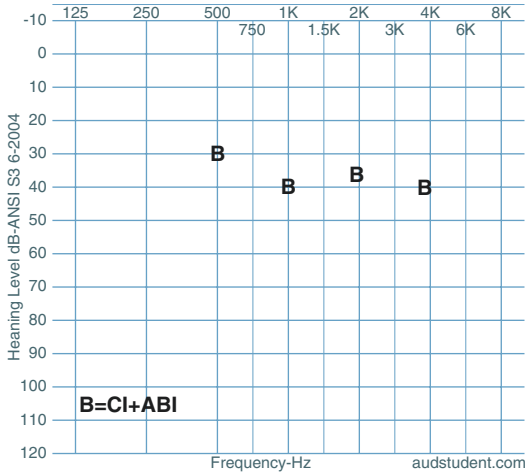


Fig. 22.7 Case 22.4. Free field thresholds with CI and ABI

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

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

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

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Sennaroglu L. Cochlear implantation in inner ear malformations - a review article. Cochlear Implants Int. 2009;11:4.
3. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
4. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
5. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
6. Jeong SW, Kim LS. Cochlear implantation in children with cochlear aplasia. Acta Otolaryngol. 2012;132(9):910–5.
7. Kontorinis G, et al. Aplasia of the cochlea: radiologic assessment and options for hearing rehabilitation. Otol Neurotol. 2013;34(7):1253–60.
8. Sennaroglu L, et al. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.

Incomplete Partition Type I

23

Levent Sennaroglu and Merve Ozbal Batuk

Special Features

1. One of the most important causes for recurrent meningitis in children.
2. Spontaneous fistula at stapes footplate.
3. Nonprogressive, profound SNHL requiring CI.
4. Facial nerve anomaly.
5. Observed deformities probably result from defective vascular supply from IAC.
6. It is possible to have CN deficiency, necessitating ABI.

23.1 Definition

Characteristics of incomplete partition type I (IP-I) were described in 2002 by Sennaroglu and Saatci [1] and named as "**cystic cochleovestibular malformation.**" They represent approximately 11.5% of inner ear malformations (IEM). The cochlea can be clearly differentiated from vestibular system and is located in its usual location in the anterolateral part of the fundus of the internal auditory canal (IAC). In IP-I, cochlea lacks the entire modiolus and interscalar septa (Fig. 23.1a), giving the appearance of an empty cystic structure. IP-I cochlea has been reported to have external dimensions (height and length) similar to normal cochlea [2]. It is accompanied by an enlarged, dilated vestibule (Fig. 23.1b). Vestibular aqueduct enlargement is very rare. Due to developmental abnormality of the cochlear aperture and absence of the modiolus, there is a defect between the IAC and the cochlea (Fig. 23.1c), and CSF may completely fill the cochlea. Consequence of this is a CSF gusher during cochleostomy or a recurrent meningitis through spontaneous oval window fistula. IP-I constitutes the most important etiology for recurrent meningitis in children.

23.2 Histopathology and Pathophysiology

An IP-I cochlea has normal external dimensions with defective modiolus and interscalar septa (ISS) [3]. It is accompanied by a grossly dilated vestibule. Histopathological studies showed a wide connection between the cochlea and the vestibule [4]. Although the otic capsule around the IP-I cochlea consisted of three layers, the endosteum (inner periosteal lining) is much thinner than

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_23].

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

M. Ozbal Batuk (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_23

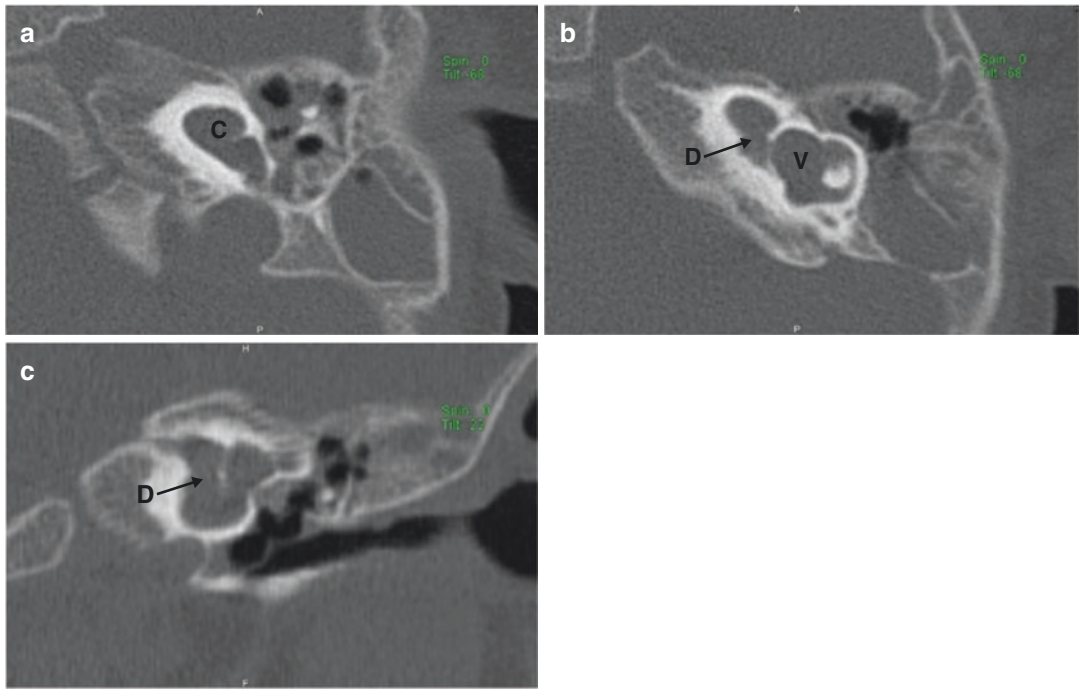


Fig. 23.1 Incomplete partition type I (IP-I) Cochlea (C) which lacks the entire modiolus and interscalar septa, with the appearance of an empty cystic structure (**a**), accompanied by an enlarged, dilated vestibule (v) (**b**). Please note the defect (D) between IP-I cochlea and internal auditory canal on axial (**b**) and coronal sections (**c**)

a normal cochlea, and at some locations it was completely absent [3]. However, the middle enchondral and outer periosteal layers appeared to have developed normally. This is valid for vestibule and semicircular canals as well.

The stapes footplate is frequently defective. The bony footplate was partially replaced either by a fibrotic tissue or a thin membrane. In Massachusetts Eye and Ear Infirmary specimens there was no connection of the cochleovestibular space with CSF-containing IAC, because the basal part of the modiolus was intact (Fig. 23.2).

All of the defective areas have their vascular supply from IAC. Therefore, it is possible to explain all of the observed abnormalities seen in the defective endosteum in IP-I as a result of defective vascular supply coming from the IAC (see Chap. 3). When compared to normal subjects, endosteum in IP-I is thinner and defective. All five patients with IP-I pathology had a very thin and defective endosteum (innermost layer of the otic capsule) all around the cochleovestibular space, while the middle enchondral and outer periosteal layers were normal.

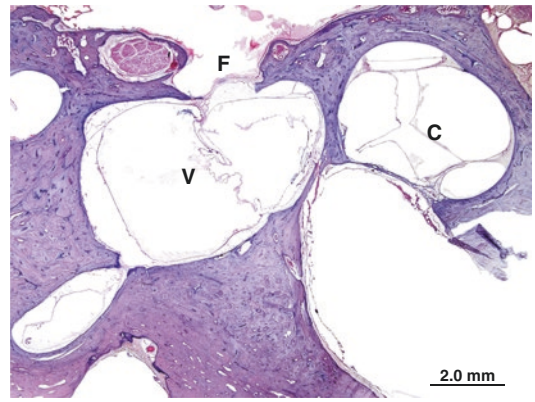


Fig. 23.2 IP-I Cochlea (C) and dilated vestibule (V). The bony footplate (F) was partially replaced either by a fibrotic tissue or a thin membrane. Note that there was no connection of the cochleovestibular space with CSF-containing IAC, because the basal part of the modiolus was intact. This makes CSF pressure not responsible for stapes footplate defect. (With permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary)

Three of the specimens had a thin, but intact modiolar base, and a stapes footplate defect at the same time. High CSF pressure cannot be

held responsible for observed pathologies because it is possible to have footplate defect even in the presence of intact partition between IAC and cochlea. Embryologically, the vestibular part of the stapes footplate is derived from the endosteum, and a defective endosteum may be responsible for this defect. In addition, the fundus defect in IP-III was larger and present in all cases, but no fistula was observed in the oval window. As a result, these rare patients demonstrate that high CSF pressure cannot be held responsible for the defective development at the stapes footplate.

As the external dimensions of the cochlea are similar to those of a normal cochlea, it can be assumed that the vascular supply from the middle ear is normal, and hence the outer two layers of the otic capsule develop normally. This is most probably the factor preventing abnormal dilatation of the cochlea. The pathology is at the fundus, modiolus and interscalar septa. The vascular supply of the modiolus and endosteum comes from the arteries of the IAC. Therefore, in IP-I the etiology appears to be the defective vascular supply coming from the IAC causing endosteal deformities.

23.3 Literature Review

IP-I malformation was first defined as "cystic cochleovestibular malformation" in 2002. During the 16 years, many studies were published about the clinical experience, radiological findings, and audiological outcomes of patients with IP-I malformation.

Berrettini et al. [5] emphasized the surgical difficulties, such as high risk of CSF gusher, otogenic recurrent meningitis, modification in surgical approach, and audiological outcomes in four IP-I cases. They specified the negative factors affecting the audiological outcomes in patients with IP-I such as late diagnosis, late amplification, low sociocultural level, and bilingualism.

An et al. [6] published their cochlear implant experience in 23 children with IP-I malformations. They reported a high rate of cochlear nerve aplasia (63%) in their IP-I series. They also reported that after 2 years CI use, auditory performance and benefit of the CI in children with IP-I are similar to the children with normal cochlea.

Kontorinis et al. [7] investigated the features of the IP malformations. They reported that IP-I malformation is a more severe malformation and auditory performance is worse than IP-II malformation. They recommended cochlear implantation as the best hearing rehabilitation option in patients with IP-I. They also emphasized the heterogeneous auditory performance after CI in IP-I.

Studies have shown that the audiological performance of patients with IP-I malformations is worse than the other IP malformations [5, 7–10]. Due to the developmental abnormality of the cochlea and inadequate residual neural activity they perform worse than the other IP malformations. Different than other IP malformations, it is possible to encounter cochlear nerve hypoplasia in patients with IP-I. This is an important factor affecting the auditory performance.

IP-I malformation can be seen bilaterally or unilaterally. In unilateral IP-I cases, it is possible to have other types of IEMs and in some cases a normal cochlea on the contralateral ear. In the study of Batuk et al. [10] the most common IEM on the contralateral ear was cochlear hypoplasia type II (34%) in cases with unilateral IP-I and in one case normal cochlea was reported.

23.4 Clinical Findings

They have profound SNHL. Hearing loss is stable, without showing progression. Hearing aids are in general not sufficient. They need a CI or an ABI for hearing habilitation.

It is very common for IP-I cases to have recurrent meningitis. More commonly they may be consulted for recurrent meningitis by department of pediatrics. As explained in the pathophysiology section, this is due to stapes footplate fistula.

First author operated 15 patients with stapes footplate fistula (one revision). Nine of these cases had IP-I anomaly. Two additional cases with IP-I anomaly had had a prior CI surgery and the leakage was around the electrode (See Case 23.2 below). Two patients were particularly interesting. One patient had recurrent meningitis as a result of contralateral oval window fistula 3 years after CI surgery, having no relationship to CI surgery (see Case 23.4 below). Second patient developed meningitis 5 months after surgery as a result of CSF leakage, on the same side with CI surgery, but there was no CSF leakage around the electrode. There was a separate defect in the stapes footplate and CSF leakage came from the defective footplate (See Case 23.3 below).

If a patient is consulted for recurrent meningitis and hearing loss (unilateral or bilateral), IP-I anomaly should be suspected. To a lesser extent cochlear hypoplasia type II (CH-II) may be responsible for recurrent meningitis. It is mandatory to explore the middle ear and repair the possible leaking stapes fistula to avoid further attacks of meningitis.

23.5 Radiology

According to the IEMs database of the Hacettepe ENT Department, IP-I represents 11.5% of the 776 patients with 1552 ears that were evaluated until September 2018 and 29% of incomplete partitions.

Cochlea looks like an empty bony cyst (Fig. 23.1a). External size of IP-I cochlea is similar to normal cochlea [2]. Vestibule is grossly dilated sometimes involving the lateral SCC on axial sections (Fig. 23.1b). Cochlear aperture (bony cochlear nerve canal) may appear as defective (Fig. 23.1b). It is very important to see the cochlear aperture on coronal sections as well (Fig. 23.1c). In the latter case it appears defective indicating a strong possibility of intraoperative CSF gusher.

IP-I anomaly looks very similar to cochlear hypoplasia type II (CH-II). The only difference between them is the smaller dimensions of CH-II. It is very important to compare the two sides on a symmetric axial CT to understand the difference. Figure 23.3a–f demonstrates such a case and postoperative X-ray image of this case can be seen in Fig. 23.3g. On the right side there is an IP-I cochlea, which is larger in size and appears earlier on higher axial sections before the CH-II on the left side (see Case 23.7 below for more details). It is very important to notice this difference and choose an appropriate length of an electrode. Recent unpublished study by Pamuk [11] uses certain measurements to differentiate between IP-I and CH-II (see Chap. 26).

In IP-I, cochlear nerve can be normal, hypoplastic, or aplastic (Fig. 23.4a–c). It is also very important to see the cochlear nerve on direct sagittal oblique sections. Figure 23.4d, e shows normal and aplastic CN, respectively.

It is also possible to observe the cyst at the oval window. Normally oval window niche is aerated without any soft tissue. If there is a fistula with a cyst, a soft tissue mass can be seen at the oval window area (Fig. 23.5a). Sometimes there is a fluid filling middle ear and mastoid (Fig. 23.5b). **If there is a history of a recurrent meningitis in a case with IP-I, these findings are definite indications for middle ear exploration via endaural approach**.

23.6 Audiological Findings

Audiological findings showed poorer auditory responses in IP-I patients compared to other IP malformation types. Majority of IP-I patients have severe to profound SNHL (Fig. 23.6a). Hearing aids are rarely sufficient to support oral language development, and virtually all patients with IP-I need CI or ABI surgery for hearing habilitation. At Hacettepe University, 50 patients with IP-I have undergone CI surgery. In our patient population only three patients could use hearing aids unilaterally. Specifically, these children used the hearing aid on the side with severe hearing loss and had CI on the contralateral side with profound hearing loss (Fig. 23.6b). In the study of Batuk et al. [10], all ears with IP-I malformations

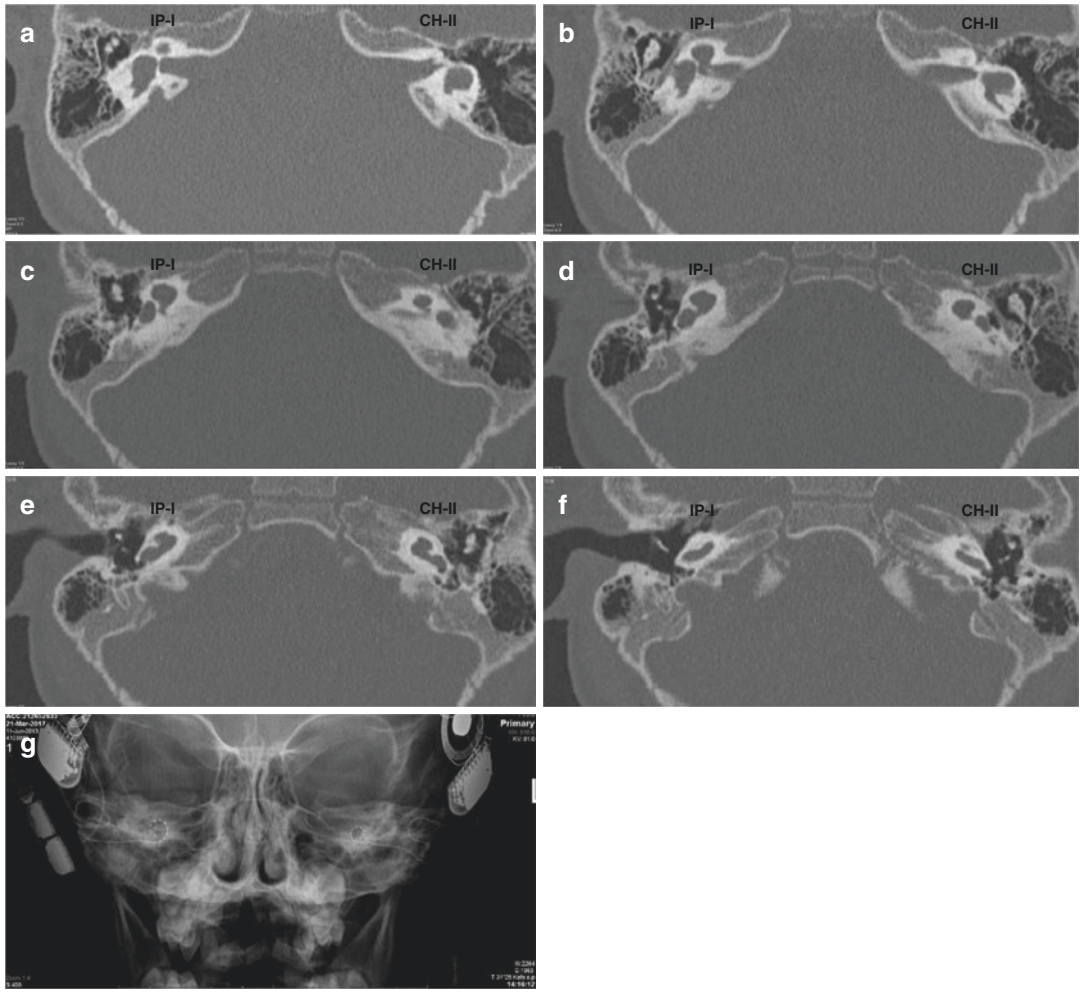


Fig. 23.3 (**a–f**) Comparison of IP-I and CH-II. On the right side there is an IP-I cochlea, which is larger in size and appears earlier on higher axial sections before the CH-II on the left side. It is very important to notice this difference and choose an appropriate length of an electrode. On the right side 24 mm length FORM 24 electrode was used. On the left shorter electrode (FORM 19) was used. On the postoperative X-ray image both electrodes make one turn around the base of the cochlea (**g**)

were diagnosed with severe to profound SNHL. According to our experience main treatment option for IP-I malformations is cochlear implantation if CN is present. In cases of IP-I patients with an aplastic CN, ABI should be recommended. Depending on the status of the CN, audiological outcome with CI can change. In cases with normal CN, speech and language development will be better than the cases with hypoplastic CN. In cases with aplastic CN on one side and a hypoplastic CN on the contralateral side, testing with insert earphones may demonstrate thresholds on the side with hypoplastic CN (Fig. 23.6c). This is a very important finding to choose the better side for CI surgery and nonresponsive side for ABI. It is also possible to have unilateral IP-I anomaly with completely normal anatomy and hearing on the contralateral side (Fig. 23.6d).

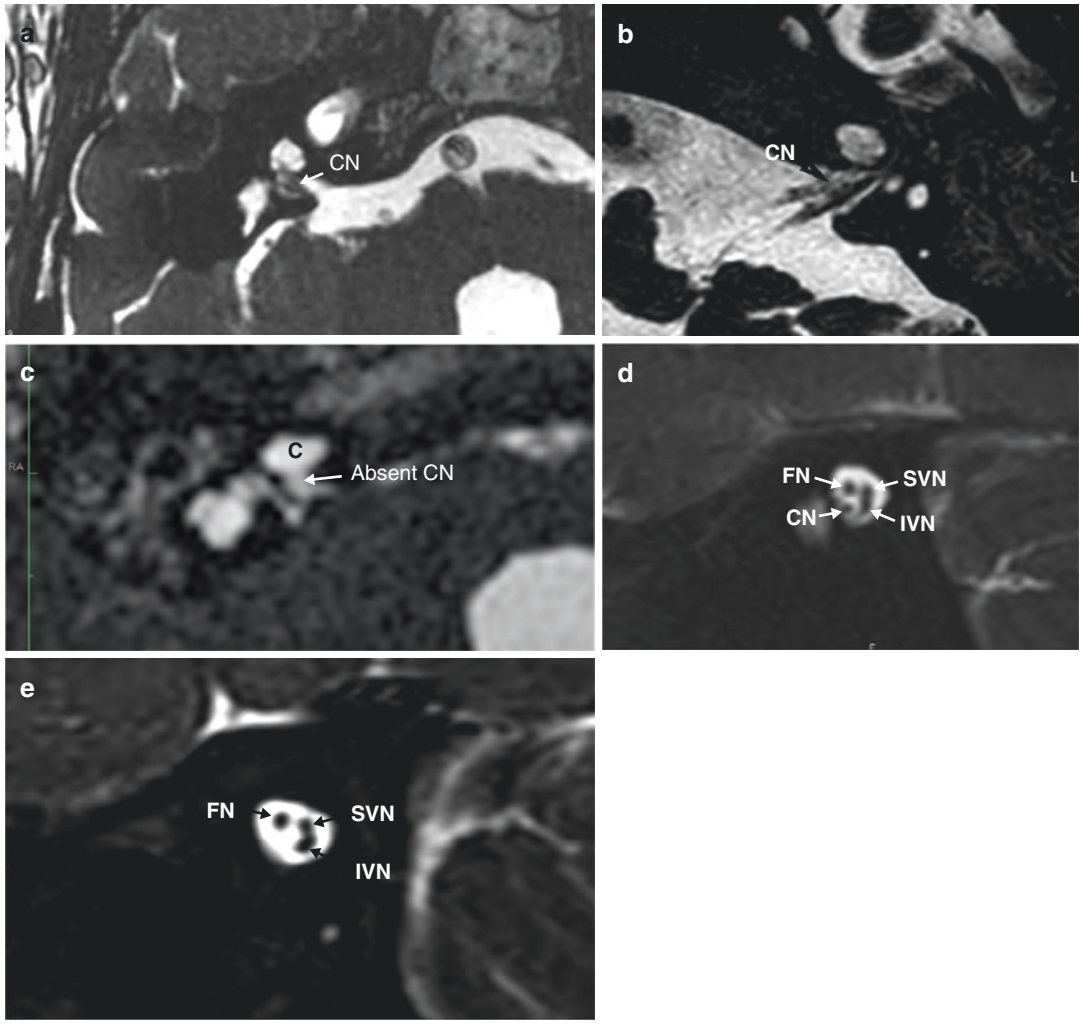


Fig. 23.4 Magnetic resonance imaging of cochlear nerve (CN) in IP-I. CN can be normal (**a**), hypoplastic (**b**), or aplastic (**c**). Parasagittal section perpendicular to the IAC showing normal (**d**) and aplastic CN (**e**) (*CN* cochlear nerve, *FN* facial nerve, *SVN* superior vestibular nerve, *IVN* inferior vestibular nerve)

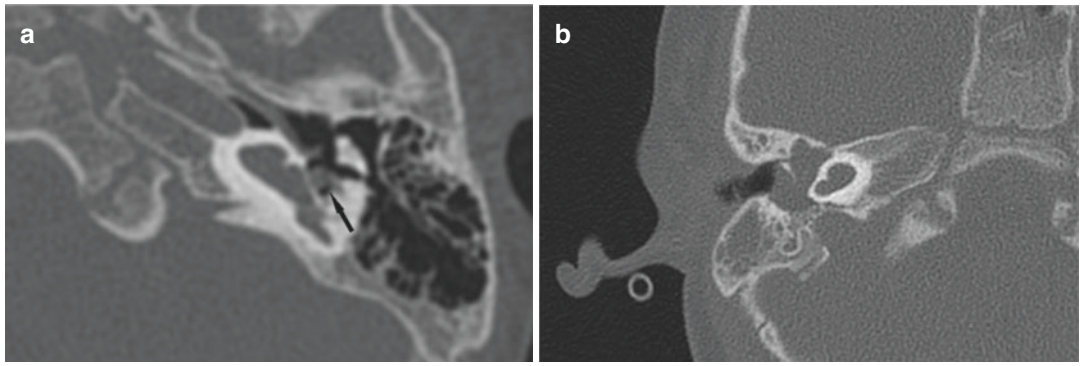


Fig. 23.5 Imaging findings indicating a CSF fistula in IP-I: (**a**) a soft tissue mass at the oval window area (arrow), (**b**) fluid filling middle ear and mastoid

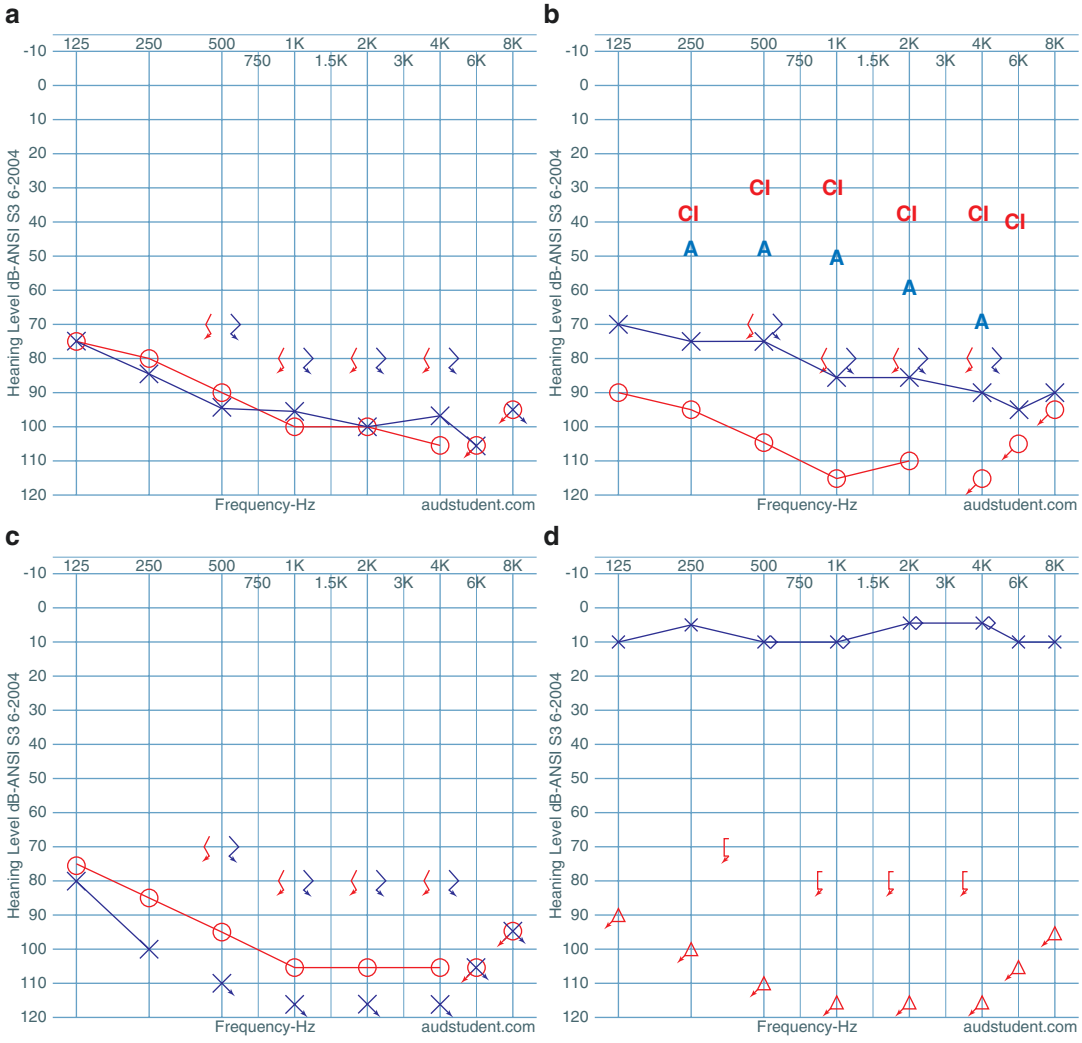


Fig. 23.6 Hearing configuration in IP-I: (**a**) Bilateral profound SNHL, (**b**) asymmetric hearing loss: severe hearing loss on the left and profound hearing loss on the right. Patient is using hearing aid on the left and cochlear implant on the right side (A and CI aided thresholds with hearing aid and CI, respectively). (**c**) Thresholds obtained during testing with insert earphones in a patient with hypoplastic CN on the right side. Left side aplastic CN. (**d**) Single sided deafness in unilateral IP-I with completely normal anatomy and hearing on the contralateral side

23.7 Management

1. If a patient with IP-I is diagnosed with bilateral moderate to severe hearing loss, hearing aids can be used for rehabilitation. However, we have not had experience with such a case, as none of the patients had any residual hearing bilaterally. For patients with asymmetrical hearing loss (i.e. one side with moderate to severe hearing loss and the other side with profound hearing loss), the best option is to use bimodal stimulation, with a hearing aid on the side with moderate hearing loss and a CI on the contralateral side with profound hearing loss (Fig. 23.6b).
2. If there is bilateral severe to profound SNHL in a patient with IP-I (Fig. 23.6a), CI is indicated. It is very important to demonstrate the cochlear nerve with MRI in order for a patient to be a candidate for CI. If CN is present, then

a CI should be done; if CN is absent, ABI surgery is the only surgical option. In children with IP-I, results of unilateral CI are not as good as CI in children with normal anatomy. Therefore, for better hearing and language outcome, bilateral CI should be offered to these patients. Because of the risk of CSF gusher, bilateral CI surgery should not be done simultaneously.

3. ABI is indicated in IP-I patients with aplastic CN. Eight patients with IP-I and aplastic CN have received ABI in our department.
4. In cases of bilateral IP-I where CN is present unilaterally, CI is done on the side where CN is present and ABI on the side with CN aplasia. It is possible to perform this surgery simultaneously in experienced centers.

23.8 Surgery

Two major complications are possible to occur during CI surgery in patients with IP-I.

23.8.1 Gusher

Out of 50 cases with IP-I anomaly, 24 patients had CSF gusher. Two patients had oozing. Therefore, CSF gusher is expected roughly in 50% of cases. Although the partition between IAC and cochlea looks defective on HRCT and MRI, there may be a separation between the cochlea and IAC, and CSF may not be communicating with the cochlea in all cases. Therefore, gusher does not occur in every IP-I patient even though the fundus appears to be defective. This is because radiological modalities are not as precise as histopathologic sections for the present time. If the partition between cochlea and IAC is defective (Fig. 23.1b, c), cochlea is usually filled with CSF and there will be an egress of CSF as soon as the cochleostomy is made. Therefore, cases of gusher occur in cases which have a true histological defect between IAC and cochlea.

Histopathological support for the latter hypothesis was present in the specimens, because four of them had a thin, bony modiolar base separating cochlea from the IAC (Fig. 23.2); at today's level of radiological precision it may not be possible to determine this on HRCT or MRI. Therefore, the cochlea is not always filled with pulsating CSF and CSF gusher is not seen in roughly 50% of the cases.

There are different stages of developmental pathology in the modiolus and cribriform area. It is possible to observe a very thin layer of bone between the IAC and IP-I cochlea (a subtotal modiolar defect). This is the reason why there is no oozing and no gusher in patients where the base of the cochlea appears to be defective on imaging—¼ lower part of the modiolus may be present. The reason for not having any CSF leakage must be the thin layer of basal modiolus, which cannot be demonstrated with HRCT and MRI. It is possible to explain this finding through developmental embryology. According to Gulya [12], ¾ superior part of the modiolus receives its vascular supply from the internal radiating arteriole of the main cochlear artery. The ¼ basal part of the modiolus receives the vascular supply from the cochlear ramus of the vestibulocochlear artery. In cases with a thin layer of bone at the fundus, the former artery must be damaged while the latter is intact. In cases where the modiolus is completely absent, with a wide connection between the cochlea and the IAC, both arteries are affected.

All these features are valid for CH-II as well. The only difference is the size of the cochlea, which is less in CH-II when compared to IP-I (Fig. 23.3a–f). The occurrence of a gusher, a spontaneous CSF fistula at the footplate, and all other clinical findings can be observed here as well. The author has seen a spontaneous CSF fistula in two, and a gusher during CI surgery in four CH-II cases. One important clinical difference is the fact that in CH-II shorter electrodes (around 20 mm in length) should be used, whereas in IP-I, it is preferable to use electrodes with a length of 25 mm (Fig. 23.3g). If longer electrodes are used in cochlear hypoplasia, they will make more turns in the cochlea, and this may cause electrode displacement into the IAC. In addition, there is a possibility of not being able to insert the electrode until cork stopper, therefore,

failing to obtain satisfactory control of CSF leakage. If a short electrode is used in IP-I, it may not provide full turn around basal turn and satisfactory stimulation of the neural tissues.

It is very important to properly seal the leak or the patient will be prone to recurrent meningitis. The surgeon who is operating on patients with an elevated risk of gusher should adopt the principle that **it is essential to fully control the gusher prior to completing the surgery**. FORM electrodes with a cork-type stopper have been developed to achieve this principle. Less severe leakage of CSF, known as oozing, can be usually managed by simply packing the cochleostomy with soft tissue. If there is a defect at the fundus of the IAC between cochlea and IAC on both sides, bilateral CI should not be done simultaneously. There is a risk for CSF leakage and in case there is a postoperative rhinorrhea, it would be impossible to know which side is causing the leakage.

There are several options for managing a CSF gusher to avoid a postoperative leak:

(a) Small cochleostomy: The size of the cochleostomy can be made small enough such that the electrode fits tightly and there is minimal space to place soft tissue around the cochleostomy. Based on our experience, this technique, however, is usually not effective in controlling the CSF leakage around the electrode as there is no sufficient space around the electrode to insert fascia (Video 23.1).
(b) Large cochleostomy: The size of the cochleostomy is larger (approximately twice the size of the electrode). Although it seems paradoxical, this is more effective in controlling the CSF leakage than a small cochleostomy because pieces of soft tissue can be placed inside the cochleostomy around the electrode.
(c) Electrode with a cork-type stopper (FORM electrodes): Sennaroglu L developed this electrode to have better control of CSF leakage at the cochleostomy. It has a progressive conical shape and is much more effective at controlling CSF leak than the standard silicon ring at the end of the electrode [13]. To make it more effective, it is passed through a piece of soft tissue prior to insertion into the cochleostomy. Ideally the cochleostomy should be circular but in reality there are irregularities around it. The soft tissue therefore serves the purpose of filling these irregularities around the cochleostomy and the stopper stabilizes the system at the cochleostomy. As explained in Chap. 15 use of FORM electrodes with cork stopper dramatically decreased postoperative rhinorrhea. It is strongly advisable to use 1.2 mm diamond bur to enlarge round window niche and apply electrode with a tiny piece of fascia into the cochleostomy during gusher. If the electrode is applied at a point with no gusher the surgeon may not be confident if the system is functioning properly (Video 23.2).

For IP-I cases it is advisable to use FORM 24 which will make one full turn around the cochlea. If FORM 19 is used, it is shorter and it will not make a one full turn in an IP-I cochlea. It will not produce the desired stimulation effect. If FORM 24 is used in CH-II it may not be possible to obtain full insertion because the cochlea is smaller in size. There is a risk that stopper will not be at the cochleostomy if longer version of FORM electrode is used in hypoplastic cochlea. Therefore, preoperative HRCT is very important to choose correct length of electrode in asymmetric pathologies (see Case 23.7).

Even with electrodes having cork-type silicon stopper, surgical manipulation is usually necessary to completely control CSF leakage. If CSF continues to ooze around the electrode, there will be an increased risk of recurrent meningitis. In this situation, slight enlargement of the cochleostomy may allow better placement of soft tissue at the cochleostomy. If full control of the leakage cannot be accomplished, it is not correct to rely on subtotal petrosectomy. The leakage can find points of drainage, as it happened in three cases operated in other centers. Subtotal petrosectomy can be done once the leakage is fully controlled for additional safety. **Therefore, surgeon should not leave the**

operating room without complete control of the leak.

(d) Subtotal petrosectomy: This technique includes complete removal of the skin of the outer ear canal, blind sac closure of the ear canal, obliteration of the cavity with fat, and plugging the Eustachian tube in addition to the procedures mentioned above. By plugging the Eustachian tube the procedure provides additional protection against meningitis. However, it should be emphasized that if the leakage is not fully controlled, complications may still occur. The author was consulted with three patients having complications even after subtotal petrosectomy. Two had recurrent meningitis while the second patient had CSF leakage through the wound.

Once a subtotal petrosectomy is done in a child with a CI, it is very difficult to check the condition of the ear. MR imaging cannot be done because of the CI without magnet removal or in cases where the device is approved for 1.5 T MRI scanning with the magnet in place. High resolution CT cannot differentiate between the fat obliteration and CSF coming from the leakage. If high CSF pressures produces a fistula at the oval window, it may not be possible to detect such a defect. Therefore, we prefer proper control of the leak at the cochleostomy, leaving the ear canal and tympanic membrane intact. During follow-up, any abnormality in the tympanic membrane may alert the surgeon to a CSF leakage. In that situation endaural exploration of the middle ear may be necessary. Endaural exploration allows drilling and enlarging ear canal without damaging the implant electrode. This provides better exposure of the stapes footplate and manipulation when compared to endomeatal approach.

(e) Continuous lumbar drainage (CLD): In patients with severe CSF leakage, 4–5 days of CLD decrease CSF pressure and allow for better healing of the cochleostomy site to promote healing. It is our practice to perform this in every patient with severe CSF leakage. One case with severe leakage who did not have CLD had recurrence of rhinorrhea 2 days after surgery.

Electrode choice: Exact location of the neural tissue in IP-I cochlea is not known precisely. Therefore, electrodes with contacts only on the modiolar side may not produce the desired effect. Electrodes with contacts on both sides or full rings may provide better stimulation. In addition, an electrode with cork-type silicon stopper (FORM) stops CSF leakage more efficiently.

Postoperatively, these patients should have an X-ray to demonstrate the position of the electrode. If there is severe gusher, transorbital X-ray should be taken in the operating theater. If the X-ray demonstrates that the electrode is in the IAC, the electrode must be repositioned. Electrodes that are modiolar hugging may have a higher likelihood of facial or cochlear nerve damage when they are being removed from IAC for repositioning. None of the IP-I patients operated in Hacettepe University had electrode migration into IAC. The author has seen one case of IP-I operated in another center with a modiolar hugging electrode where electrode migrated into IAC. As the stylet had been removed during initial operation, no attempt was made to reposition the electrode because of the risks to nerves in the IAC. The possibility of electrode misplacement into IAC in IP-I is much less than IP-III. When the histopathological findings are taken into account IP-I has smaller defect at the cochlear base when compared to IP-III. This most probably prevents electrode misplacement into IAC in IP-I. Larger defect in cochlear base in IP-III makes it more probable for electrode misplacement into IAC.

23.8.2 Facial Nerve Anomaly

As there is abnormal development of the labyrinth, the facial nerve may have an abnormal course. As such, the facial recess approach may not provide sufficient exposure for round window

insertion. In this situation, a transcanal approach in addition to postauricular approach may be necessary. This has been done in 2 patients (out of 50) who were operated in our department [14]. In cases where this is not possible, subtotal petrosectomy (where the skin is totally removed and the bony external auditory canal is taken down) may provide a better view of the landmarks.

Split ear canal technique [14]: This approach can be used in cases of facial nerve abnormality. The procedure involves transmastoid facial recess approach combined with transcanal exploration. A slit is produced in the ear canal; this connects mastoid and ear canal. Cochleostomy is done via transcanal approach: cochlea is in direct view. After managing CSF leakage around the cochleostomy, electrode lead is transferred to mastoid via the split ear canal (Video 23.3). This method has been named as **split ear canal technique**. A thin cartilage is used to cover the defect in the ear canal (see Case 23.6).

23.9 Meningitis in IP-I

Meningitis can occur in IP-I patients even prior to their CI surgery or originating in their nonoperated ear. High CSF pressure filling the cochlea disrupts the already deficient thin membranous stapes footplate, leading to a CSF fistula at the oval window and meningitis. Several cases of this have been reported in the literature [9, 15–17].

Usually, these children have recurrent meningitis and HRCT reveals a small opacity in the oval window area (Fig. 23.5a). Imaging may also reveal fluid filling middle ear and mastoid (presumably CSF) (Fig. 23.5b). **All patients with IP-I and recurrent meningitis who have**

1. an opacity at the oval window,
2. normal tympanic membrane with HRCT showing fluid filling the middle ear and mastoid, should have an exploration of the middle ear with special attention to the stapes footplate. Best route is via endaural approach.

It is interesting to note that meningitis is often reported in IP-I cases. One Hundred percent of IP-III cases have CSF gusher during CI surgery or stapedotomy but meningitis is very rarely reported in these patients [9, 15]. This is most likely due to the fact that the stapes footplate is thicker in these patients due to possible thickened endosteum and thus it is much less likely to develop a footplate defect and CSF fistula. This is another factor showing different mechanisms in IP-I and IP-III.

The author has operated 15 cases of recurrent meningitis with IEMs. Twelve had IP-I anomaly. Two cases were operated in other centers and had CSF leakage around electrode. Nine patients had oval window fistula (one of the patient was revised because of recurrent rhinorrhea). All cases had a spontaneous CSF fistula with a cystic structure present at the stapes footplate. Once it was punctured, egress of CSF came from this defect. This was repaired by inserting a piece of fascia through the defect into the vestibule, in a dumbbell shape (see Chap. 12). It is possible that in IP-I cochlea, where the modiolus is completely absent, high CSF pulsations acting on the thin membrane at the footplate may easily produce an oval window fistula. If there is a bony separation between the IAC and the cochlea, the footplate defect may not be noticed at all during the patient's lifetime. One of these cases had immediate recurrence of rhinorrhea after surgery as postoperative CLD was not performed. After his revision surgery with CLD he never had any leakage or meningitis. One case had recurrence of meningitis and she was operated with subtotal petrosectomy (See Case 23.4).

None of the 13 spontaneous CSF fistulas due to IEM operated on by the author had a fistula at the round window. All of the reported spontaneous CSF fistula cases in IP-I in the literature are located at the oval window [15–18]. This also shows that this is observed in cases with a defective footplate. In IP-III there is a larger defect in the fundus, a high pulsating CSF pressure in the cochlea, and always a severe gusher upon cochleostomy; but a spontaneous CSF fistula has never been encountered. The reason may be that the endosteum, which is deficient in IP-I (probably due to defective vascular supply coming from the IAC), is properly formed in IP-III. In IP-III, it looks possible that the endosteum is well developed (and may even be thicker than normal),

causing no fistula at the stapes footplate and preventing spontaneous CSF fistula formation.

Stapes footplate defect is most probably present at birth. High CSF pressure can cause a fistula through the defective footplate, or otitis media during childhood may result in recurrent meningitis. All cases that have been operated on by the author were children. No adult patient has been operated on so far: it appears that it is not a progressive disease, and that it must be present during childhood. If it had been the result of high pressure only, it would have been possible to see this clinical entity at all ages. This shows us that high pressure is not necessary all the time for the development of the oval window fistula. The defective development is most probably a result of a deficient periosteum present at birth, but high CSF pressure may produce a fistula in this already defective area.

Management: Meningitis in IP-I necessitates an endaural exploration of the middle ear. Usually there is a cystic structure of variable size originating from the stapes footplate. The cyst is carefully removed. **It is strongly advisable to keep the stapes in place and insert a piece of fascia or muscle tissue through the footplate defect into the vestibule, keeping the stapes in place to hold the soft tissue in position.** Tissue glue is then used after soft tissue placement to further anchor the tissue. In some cases, it may be necessary to remove the stapes and obliterate the defect with more pieces of fascia and bony tissue. The incus can be inserted into the oval window and may provide a tight seal if all else fails. In situations with severe CSF leakage, postoperative continuous lumbar drainage is the routine procedure to allow safe healing of the footplate or cochleostomy. Subtotal petrosectomy can also be done as an additional procedure once CSF leakage has been fully controlled.

Ten cases had fistula at the stapes footplate. Eight cases developed fistula at the footplate spontaneously. They were not operated before. Two cases had CI operation before and developed fistula ipsilaterally; there was no leakage at the cochleostomy but a fistula was present at the footplate. Two cases were operated for CI but they developed a fistula on the contralateral nonoperated ear.

23.10 Clinical Experience

Between November 1997 and September 2018, 2639 patients underwent CI and ABI. Four hundred and two had IEMs. Fifty-eight had IP-I type IEM. Fifty had CI (5 were revision) and 8 had an ABI.

23.10.1 Revisions CI Surgery in IP-I

(a) Two patients had device failure. Both CIs were replaced without any difficulty.
(b) Two had CSF leakage around electrode. Both had their initial CI operations in other centers. They were explored and fascia was applied around the electrode into the cochleostomy. In one case this was successful. Remaining case necessitated an electrode change from Cochlear Nucleus CI24RE with Contour Advance to Med El FORM electrode. There has been no leakage or meningitis afterwards (Case 23.2).
(c) Cochlea could not be found during the initial operation. It was successfully located during revision surgery with full insertion of CI.

Eight cases had CN aplasia and underwent an ABI surgery.

23.11 Cases

Case 23.1: PK 6-Year, Female. Operated in Aug 2006

She had rudimentary otocyst on the right side and IP-I on the left side (Fig. 23.7). Left side was

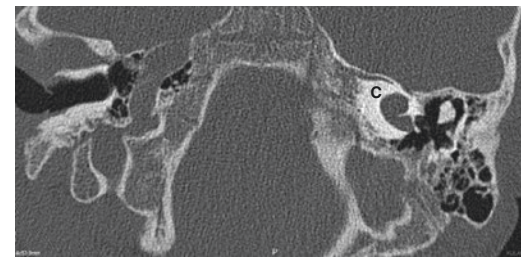


Fig. 23.7 Case 23.1. IP-I on the left side with rudimentary otocyst on the right side

operated in August 2006. There was a severe CSF gusher. Cochlear Nucleus CI24RE with Full-Band Straight Electrode was passed through a tiny piece of fascia and electrode was inserted into cochleostomy. Cochleostomy was closed completely so that there was no CSF leakage. Three months after surgery she was admitted to our hospital in a comatose situation and diagnosed with meningitis. She was hospitalized in intensive care unit, her situation deteriorated and she died after 1 month.

This patient is the main reason for the development of an electrode with "cork" type stopper (later called FORM electrodes) to close cochleostomy more efficiently [13] by the first author. It was not possible to learn whether she had a stapes footplate fistula causing the CSF leakage and meningitis. The reason for including her in this book is to remind that management of IEMs, particularly IP-I, carries the risk of meningitis which may be fatal.

Case 23.2: YY 5-Year-Old Female Patient, Operated in January 2011

She had bilateral IP-I anomaly. Left ear was operated in another center with CI. Operation note mentioned severe gusher during surgery. She had recurrent meningitis before coming to our department. On her initial examination, her left tympanic membrane was dull and vascularized. HRCT revealed fluid filling the middle ear on the side with CI (Fig. 23.8a). On January 2011 her left ear was explored via endomeatal approach. There was a CSF leakage around Cochlear Nucleus CI24RE with Contour Advance electrode. Electrode was kept in place and fascia was applied around the electrode and inside the cochleostomy. She had one more attack of meningitis. Her left tympanic membrane was dull and vascularized. In April 2012 she had a revision surgery via original postauricular approach. It was observed that leakage around the Contour advance electrode persisted. Implant was removed totally. Med El FORM 24 electrode was passed through a tiny piece of fascia and fully inserted into cochlea. Postoperative X-ray showed the electrode inside IP-I cochlea (Fig. 23.8b). She had no more meningitis after this surgery. She demonstrated limited language development because of hypoplastic cochlear nerve on that side. The cause of meningitis in this patient was leakage at the cochleostomy site.

Case 23.3: MBP 1-Year-Old Female Patient, Operated in June 2010

She had bilateral IP-I. On her initial HRCT there was no fluid in the middle ear and mastoid (Fig. 23.9a). Right side was operated in June 2010. There was severe gusher once cochleostomy was made. Cochleostomy was closed effectively with Med-El FORM electrode with cork stopper and fascia. Five months later she had recurrent meningitis. HRCT demonstrated fluid filling middle ear and mastoid (Fig. 23.9b). Endomeatal exploration was done in Jan 2011. Middle ear exploration revealed continuous CSF leakage coming from a fistula at the stapes footplate. There was no leakage around electrode.

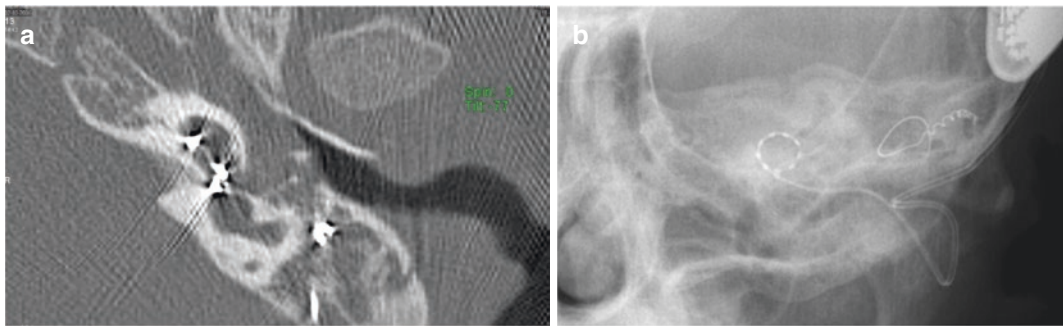


Fig. 23.8 Case 23.2. (**a**) Tomography showing fluid filling the middle ear on the side with CI. (**b**) Postoperative X-ray showed FORM 24 electrode inside IP-I cochlea

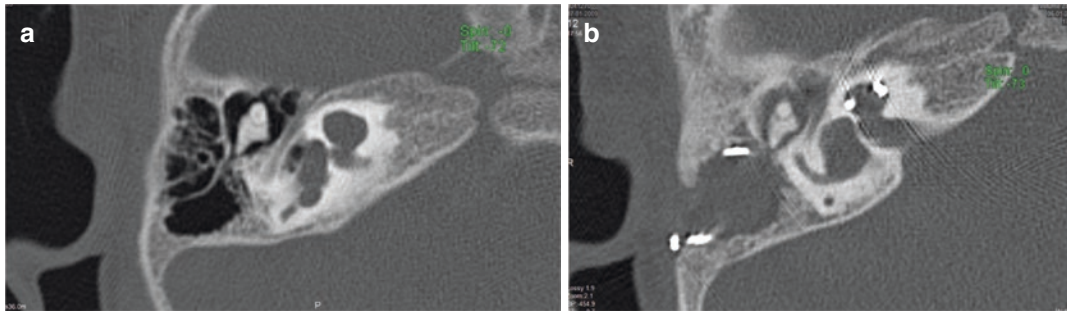


Fig. 23.9 Case 23.3. (**a**) Preoperative tomography showing IP-I anomaly without any fluid in middle ear and mastoid. (**b**) Postoperative tomography 6 months after surgery with fluid in the middle ear and mastoid

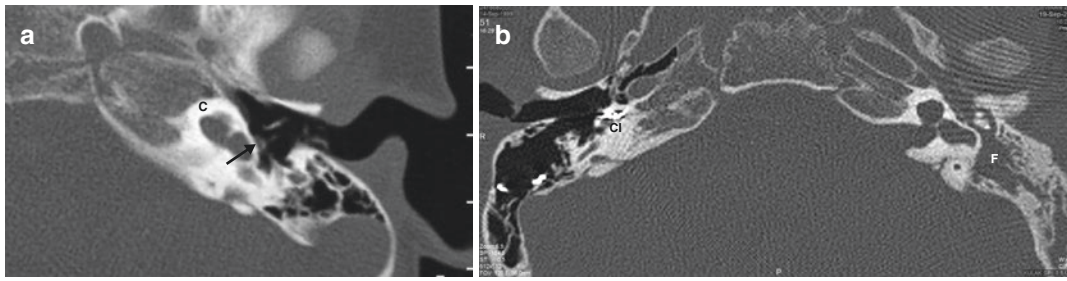


Fig. 23.10 Case 23.4. (**a**) Bilateral IP-I deformity without any fluid in the middle ear, but with a soft tissue on the left ear in the oval-round window area (black arrow). (**b**) Postoperative CT showing CI on the right side with fluid (F) in the middle ear and mastoid on the contralateral left side

Footplate defect was obliterated with fascia keeping stapes intact (Video 12.3).

She had meningitis via stapes footplate fistula and it was not related to CI surgery. There are two cases like this who developed meningitis at the same side with CI but via stapes footplate. It is now our practice to examine footplate area at the time of CI surgery in IP-I cases. If a fistula is identified it has to be repaired during CI surgery.

Case 23.4: CK 6-Year-Old Female Patient, Operated in September 2005

She had bilateral IP-I deformity. Her initial HRCT showed bilateral IP-I deformity without any fluid in the middle ear (Fig. 23.10a). More careful **retrospective** examination demonstrated a soft tissue which was present at that time on the left ear in the oval window area. She received a CI for right side in September 2005. She developed two attacks of meningitis. HRCT revealed fluid in the middle ear and mastoid on the contralateral left side (Fig. 23.10b). Treatment options were discussed and her left side was explored for a footplate fistula. There was a cystic structure coming from the stapes footplate. Once it was punctured there was a severe CSF leakage. Stapes was kept in place and footplate defect was obliterated with fascia. She had recurrence of meningitis and later subtotal petrosectomy was performed. She had no more attacks of meningitis.

Case 23.5: EP 2-Year-Old Male Patient, Operated in July 2013

His sister was operated for ABI because of severe IEM. Her mother said she had a brother who had two attacks of meningitis. I asked her about his hearing and she said he was deaf on one side only. I told her that I want to examine him immediately. Next day he was brought from Istanbul to Ankara. His HRCT revealed an IP-I with fluid filling middle ear and mastoid on the right side. Left side had normal cochlea and well aerated middle ear and mastoid (Fig. 23.5b). Next day he was operated endaurally and the stapes footplate

cyst was removed and defect was sealed with fascia. He had no further attack of rhinorrhea or meningitis (Video 12.2).

Case 23.6: DY 2-Year-Old Female Patient, Operated in June 2008

She had CLA on the right side and IP-I on the left side. Left side was operated with a CI in June 2008. During facial recess development it was discovered that facial nerve was located laterally preventing entry into the middle ear via facial recess. It was decided for a combined approach and tympanomeatal flap was elevated and middle ear was entered through the canal (See Fig 11.2). Ear canal was split with a tiny diamond burr. During cochleostomy there was a severe CSF gusher. Electrode with cork stopper was inserted into cochleostomy through the ear canal. Electrode was then transferred into mastoid via the split ear canal (Video 23.3). A thin cartilage was used to cover the defect in the ear canal.

This is the second case where a split ear canal technique was employed. First case was reported previously in 2001 [14]. Facial recess approach could not be used because of facial nerve anomaly. The reason for the split ear canal is not to leave the electrode in the ear canal. This carries the risk of infection which may eventually lead to removal of the implant. Therefore, by transferring the electrode from ear canal into mastoid, this potential complication is avoided.

Case 23.7: BAK, 2-Year-Old Male Patient, Operated in July 2015

He had IP-I on the right side and CH-II on the left side (Fig. 23.3a–f). Right side IP-I was operated in July 2015 with FORM 24. There was severe gusher which was controlled effectively with cork-type stopper. Left side CH-II was operated in March 2017 with FORM 19. Postoperative transorbital X-ray showed the two sides with at least one turn of the electrode (Fig. 23.3g).

The two anomalies are coincidental often. It is very important to choose the correct length of electrode with stopper. For IP-I FORM 24, for CH-II FORM 19. If the FORM 19 is chosen for IP-I, it will not make one full turn around the cochlea, resulting in inefficient stimulation. If we choose FORM 24 for CH-II we may not be able to insert the electrode until stopper therefore, it will not be effective in controlling CSF gusher.

23.12 Outcomes with CI and ABI

Auditory performance of the patients with IP-I showed variance between individuals. After CI and ABI, outcomes were variable depending on the presence of any additional handicaps, age at implantation, duration of the implant use, placement of the internal electrode, condition of the cochlear nerve, and the other factors.

Especially in patients using CI, the status of the cochlear nerve plays an important role during follow-up. During the CI mapping sessions in IP-I patients audiologists are faced with some difficulties such as inadequate stimulation or facial nerve stimulation. The reason for inadequate stimulation may be associated with the residual nerve fibers. To overcome the mapping difficulties, it is important to observe the child carefully during the stimulation and make necessary changes in their maps. Programming modifications should contain wider amplitude, higher pulse width, higher stimulation levels, and deactivation of the electrodes with facial nerve stimulation.

Although auditory perception abilities of the patients with IP-I were better than the common cavity and cochlear hypoplasia malformations, they show worse performance when compared to other incomplete malformations. In case of a cochlear nerve hypoplasia, auditory performance was affected negatively and the results were poorer in the children with normal cochlear nerve.

In generally, patients with IP-I showed good performance after ABI if they had early diagnosis, early implantation, good family support, and no additional handicap. Although language development of children with ABI was slower than the children with cochlear implant, they can show improvement with the time. For better outcome, more frequent follow-up is advised for the patients with IP-I after CI and ABI (for further details see Chaps. 30–32).

References

1. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41.
2. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25(4):520–9.
3. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
4. Khan AM, Levine SR, Nadol JB Jr. The widely patent cochleovestibular communication of Edward Cock is a distinct inner ear malformation: implications for cochlear implantation. Ann Otol Rhinol Laryngol. 2006;115(8):595–606.
5. Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N. Cochlear implant in incomplete partition type I. Acta Otorhinolaryngol Ital. 2013;33(1):56.
6. An YS, Lee JH, Lee K-S. Surgical outcomes after cochlear implantation in children with incomplete partition type I: comparison with deaf children with a normal inner ear structure. Otol Neurotol. 2015;36(1):e11–7.
7. Kontorinis G, Goetz F, Giourgas A, Lenarz T, Lanfermann H, Giesemann AM. Radiological diagnosis of incomplete partition type I versus type II: significance for cochlear implantation. Eur Radiol. 2012;22(3):525–32.
8. Sennaroglu G, Sennaroglu L. Hearing loss in inner ear malformations. In: Encyclopedia of otolaryngology, head and neck surgery. Berlin: Springer; 2013. p. 1143–50.
9. Sennaroglu L. Cochlear implantation in inner ear malformations–a review article. Cochlear Implants Int. 2010;11:4–41.
10. Batuk MÖ, Çınar BÇ, Özgen B, Sennaroğlu G, Sennaroğlu L. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233.
11. Pamuk G. Measurement of cochlear dimensions in cochlear hypoplasia using temporal bone computerized tomography and magnetic resonance imaging and its effects on cochlear implant selection. Ankara: Hacettepe University Medical Faculty; 2018.
12. Gulya, editor. Gulya and Schuknecht's anatomy of the temporal bone with surgical implications. 3rd ed. New York: Informa Healthcare; 2007.
13. Sennaroğlu L, Atay G, Bajin MD. A new cochlear implant electrode with a "cork"-type stopper for inner ear malformations. Auris Nasus Larynx. 2014;41(4):331–6.
14. Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. Otol Neurotol. 2002;23(1):39–43; discussion 42–3
15. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
16. Shetty PG, Shroff MM, Kirtane MV, Karmarkar SS. Cerebrospinal fluid otorhinorrhea in patients with defects through the lamina cribrosa of the internal auditory canal. Am J Neuroradiol. 1997;18(3):478–81.
17. Syal R, Tyagi I, Goyal A. Cerebrospinal fluid otorhinorrhea due to cochlear dysplasias. Int J Pediatr Otorhinolaryngol. 2005;69(7):983–8.
18. Ahmed S, Moffat D. An alternative method for dealing with cerebrospinal fluid fistulae in inner ear deformities. Am J Otol. 1998;19(3):288–91.

Incomplete Partition Type II

24

Merve Ozbal Batuk and Levent Sennaroglu

Special Features

1. No typical audiological configuration. Hearing loss usually fluctuating and progressive throughout lifetime with occasional sudden SNHL. Air-bone gap is usually present at the low and middle frequencies.
2. Usually they become a candidate for cochlear implantation during follow-up.
3. During cochleostomy, pulsation is common; oozing is seen occasionally; gusher is rare.
4. Facial nerve anomaly or meningitis are very uncommon.
5. Progressive SNHL is probably due to CSF pressure transmission into inner ear via EVA.
6. Different stages of modular defects as a result of CSF pressure.

24.1 Definition

In incomplete partition type II cochlear malformation (IP-II), the apical part of the modiolus is defective but the basal part is anatomically normal (Fig. 24.1a) [1, 2]. This is the type of cochlear anomaly which was originally described by Carlo Mondini and together with a minimally dilated vestibule and an enlarged vestibular aqueduct (EVA) (Fig. 24.1a, b) constitutes the triad of the **Mondini deformity.** Unfortunately, the term "Mondini" has been used inappropriately to describe different inner ear malformations (IEM). This name has to be used only if the above triad of malformations are present [2–4]. The apical part of the modiolus and the corresponding interscalar septa are defective. This gives the apex of the cochlea a cystic appearance due to the confluence of middle and apical turns. The external dimensions of the cochlea (height and diameter) are not different from that seen in normal cases. As this study pointed out, it is not correct to define this anomaly as a cochlea with 1.5 turns [5]. This description should only be used for cochlear hypoplasia.

24.2 Histopathology and Pathophysiology

Sennaroglu [6] recently reported on the histopathological findings in IP-II cases. Four specimens in Massachusetts Eye and Ear Infirmary (MEEI) had a cochlea with an enlarged scala vestibuli (SV). The interscalar septum (ISS) was pushed upwards in all four specimens (See Figure 3.10). The way that the ISS is expanded upwards

M. Ozbal Batuk (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_24

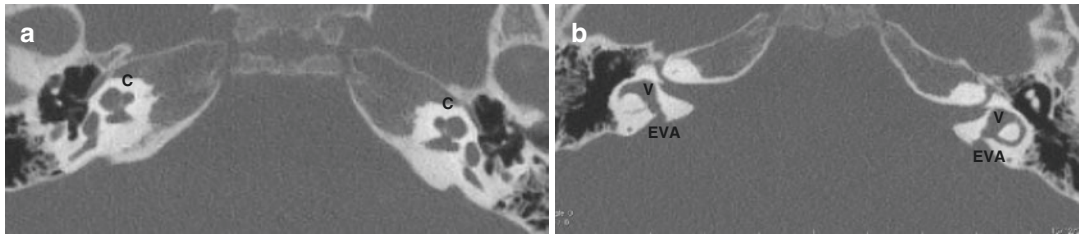


Fig. 24.1 Incomplete partition type II (IP-II). (**a**) Cochlea (C) with defective apical part of the modiolus with normal basal turn. (**b**) Minimally dilated vestibule (V) and an enlarged vestibular aqueduct (EVA)

in all four specimens gives a strong impression that it is the result of high pressure inside the SV which was present in the embryological period. The scala tympani was normal in all four specimens, and all four specimens had a very large endolymphatic sac and EVA. None of the four cases had a defective stapes footplate. All three layers of the otic capsule were completely normal around the cochlea and vestibule. There was no defect in the modiolus, connecting the cerebrospinal fluid (CSF)-containing IAC and the scala vestibuli.

None of the four cases with IP-II had underdevelopment in the internal architecture (modiolus and ISS) compared to other specimens, suggesting vascular insufficiency. In addition, the SCCs were normal in all four specimens.

First author also investigated IEM specimens of University of Minnesota in 2018. There was only one patient with bilateral IP-II with symmetric findings. Both specimens had EVA and type V: partial modiolar defect.

Pathophysiology can be explained in terms of pressure transfer into inner ear via EVA [6]. Cochlea, with its excellent bony otic capsule, is well protected from the high pulsating pressure of the CSF. Any bony opening in the otic capsule or fundus may allow the transmission of this pressure into the cochlea. This abnormally high and pulsating pressure has been observed almost on every occasion by surgeons when they perform a cochleostomy in cases of EVA. Even though there is no CSF leakage, pulsation is frequently observed (Video 15.1). This may be the reason for the continuous pressure transmission into the inner ear, which results in vestibular dilatation and SV dilatation, modiolar defects, and progressive SNHL. The high pulsating CSF pressure may also be responsible for further enlargement of EVA. A large sac with more endolymph production and/or high pressure may cause EVA. Recently radiological evidence was also provided for this pressure transfer hypothesis in a case of unilateral IP-II [7].

24.3 Literature Findings

IP-II constitutes the most common form of the incomplete partition malformations. In IP-II, the internal organization of the cochlea is more developed than in IP-I malformations. While the modiolus appears to be defective, particularly in the apical parts, the basal part is present [8].

Reinshagen et al. [9] analyzed retrospectively the MRI images from their 10 years' database showing the features of the IP-II malformation. At the initial diagnosis 27 patients were diagnosed with EVA (10 of them only EVA, 17 of them IP-II and EVA together). Although in 17 of 27 patients IP-II and EVA were seen together, after the retrospective analysis 4 out of the remaining 10 patients were also diagnosed as IP-II. They emphasized the importance of the detailed analysis of the internal architecture of the cochlea in order not to miss the exact diagnosis of the IEMs. They stressed the importance of the radiological evaluation for proper classification.

The degree of the severity of the IEM affects the audiological outcomes. In IP-II the audiological performance was reported to be similar to

cases with normal cochlea [10, 11]. In IP-I malformation the modiolus did not develop and due to the cystic empty cavity the residual neural activity was worse. However, in IP-II malformation, the modiolus could be observed with the basal development of the cochlea and residual hearing is better than IP-I [12].

An examination of the literature revealed that the hearing loss associated with IP-II and EVA malformations is heterogeneous, and the conductive component is not associated with middle ear pathologies [11, 13–23]. It was reported that IP-II could be seen isolated or with EVA and generally seen in both ears bilaterally [22]. In IP-II malformations the degree of the hearing loss varies from moderate to severe/profound hearing loss [8, 22].

Although tympanogram measurement and otoscopic examination were normal, mixed type hearing loss can be diagnosed in cases with IP-II. Boston et al. [14] found that the air-bone gap seen in IP-II may be associated with the structural anomalies of the third window, membranous labyrinth, or bony spiral lamina. They also emphasized that the size of the vestibular aquaduct may affect the amount of the air-bone gap and the air-bone gap was seen especially at the low frequencies. It was thought that it is possible to explain air-bone gap with third window phenomenon as seen in superior semicircular canal dehiscence.

Ahadizadeh et al. [24] reported the audiological outcomes (word recognition scores, speech reception threshold, and pure tone audiometry) in patients with EVA and IP-II in their retrospective longitudinal study. They mentioned that the severity of the hearing loss in patients with EVA was not associated with the IP-II malformation. The main reason for the severity of hearing loss was indicated as the EVA.

Batuk et al. [8] reported that different types and degrees of hearing loss were observed in patients with IP-II malformations. The rate of the severe/profound hearing loss was high (76%) in IP-II patients. This could be the result of the progressive characteristics of IP-II malformation. In light of these findings, patients with IP-II were usually diagnosed with mixed type hearing loss and, due to the progressive hearing loss during follow-up care, cochlear implantation could be recommended.

Cinar et al. [25] studied the objective test methods used in cochlear implant recipients with inner ear malformations including IP-II. They reported that electrically compound action potentials (ECAP) and electrically stapedial reflexes (ESRT) showed difference between IP-I and IP-II malformations. In IP-II malformation, the rate of observing ECAP was higher than IP-I due to the presence of the functional neural structures in IP-II. Despite the possibility of observation of the ECAP and ESRT for patients with IP-II, they recommended the use of electrically auditory brainstem response (E-ABR) rather than ECAP and ESRT during the programming. They showed the increased threshold levels and prolonged latencies of the wave V in E-ABR measurements of cochlear implant users with IEM when compared with the normal cochlear anatomy.

24.4 Clinical Findings

Patients with IP-II usually apply with hearing loss. In IP-II there is no characteristic type of hearing loss. Hearing may be normal at birth; in such a case, hearing screening may not detect this anomaly. They usually show progressive SNHL usually becoming candidates for CI sometimes in their life. They may have sudden SNHL attacks as well. Patients with progressive HL usually have very good language development. Head trauma has been linked with progression of HL and these children are advised to wear helmet during sports to avoid head trauma. The latter may cause increase in CSF pressure which may be transmitted into inner ear via EVA. Recurrent meningitis is very uncommon in IP-II because footplate defect has not been reported. In spite of air-bone gap (ABG), it is not advisable to explore these patients to perform stapedotomy. If HRCT shows IP-II anomaly, stapedotomy should be avoided as ABG may be due to the third window phenomenon.

24.5 Radiology

According to the IEM database of Hacettepe University Department of Otolaryngology out of 776 patients with various IEMs, 372 of 1652 ears had IP-II (22.5%). 98% of these were bilateral, and only 2% had a different pathology on the contralateral ear.

Sennaroglu and Saatci [5] reported the external size of the cochlea in incomplete partitions to be similar to normal cochlea. Therefore, external size of the labyrinth is normal. As a result, it is not common to encounter facial nerve abnormalities in IP-II cases.

According to defects of the modiolus a classification of modiolar defects has been proposed [6] (Fig. 24.2). Enlarged scala vestibuli, superior, partial, subtotal modiolar defects, and complete absence of modiolus can be seen in IP-II depending on the effect of CSF pressure on the developing modiolus. For the present time, this is more valid for histopathological specimens as HRCT and MRI are not precise enough to detect these changes. In future when radiological evaluation methods will have more detailed resolution, it will be applicable for radiology as well.

Earliest deformity that can be observed is scala vestibuli dilatation (Fig. 24.3a). Cystic apex is the most frequent anomaly detected by radiology (Fig. 24.3b). Gusher is likely to occur in cases of complete modiolar defects (Fig. 24.3c). It is very difficult to notice other defects in between with the precision of imaging for the present time.

Size of the enlarged vestibular aqueduct changes: Vestibular aqueduct is accepted to be normal when its width is less than 1.5 mm on midpoint between opening into vestibule and posterior fossa on axial sections. If it is larger than 1.5 mm it is accepted as enlarged. Vestibular aqueduct enlargement can be mild (1.5–3 mm), moderate (3–5 mm), or massive (>5 mm) (Fig. 24.4a–c). Recently, Sennaroglu L and Bajin D added vertical dimensions as well for the diagnosis (Fig. 24.4d, e). MRI is important in demonstrating the endolymphatic sac (Fig. 24.4f).

It is usually symmetric (Fig. 24.5a). Rarely unilateral or asymmetric EVA can be seen (Fig. 24.5b).

Cochlear nerve is always present on axial and sagittal oblique sections (Fig. 24.6a, b). Therefore, in IP-II there is no indication for an ABI.

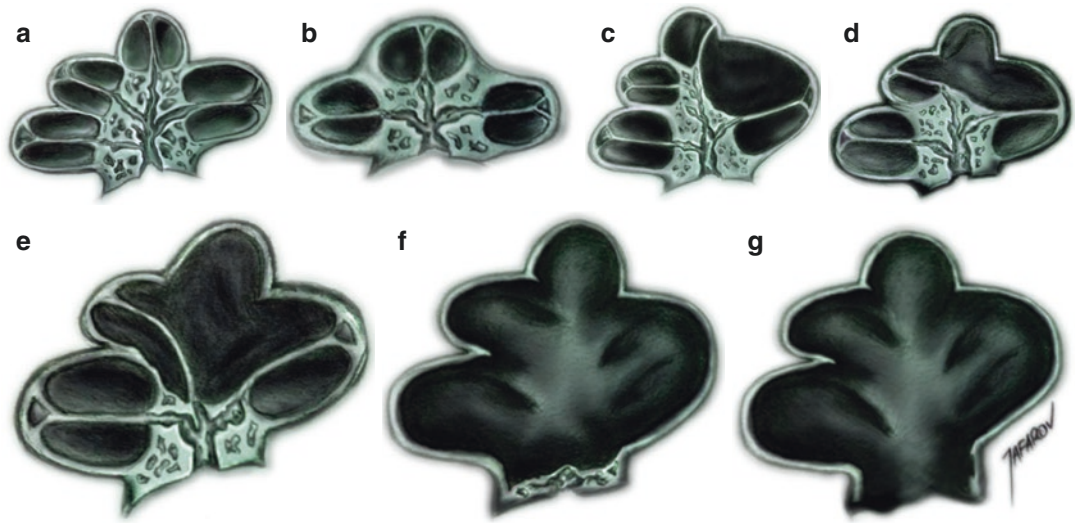


Fig. 24.2 Classification of modiolar defects: (**a**) Normal modiolus, (**b**) Shortened modiolus, (**c**) Enlarged scala vestibuli, (**d**) Superior modiolar defect, (**e**) partial modiolar defect, (**f**) Subtotal modiolar defects, (**g**) Complete absence of modiolus. In IP-II any of the defects between C-G can be seen. (With permission of CI International)

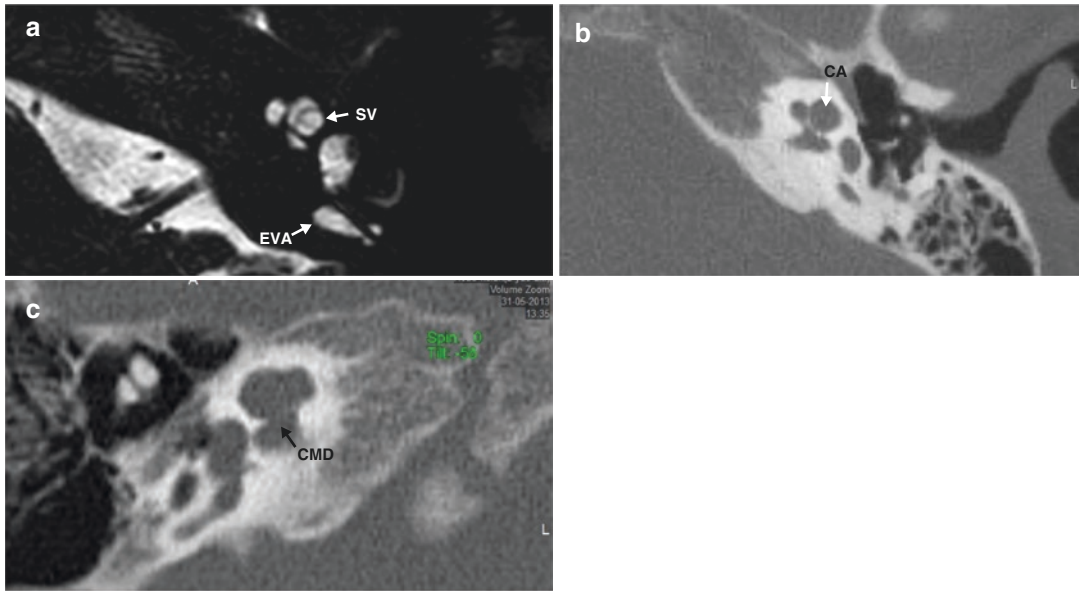


Fig. 24.3 (**a**) Scala vestibuli (SV) dilatation (*EVA* enlarged vestibular aqueduct), (**b**) Cochlea with cystic apex (CA), (**c**) complete modiolar defect (CMD)

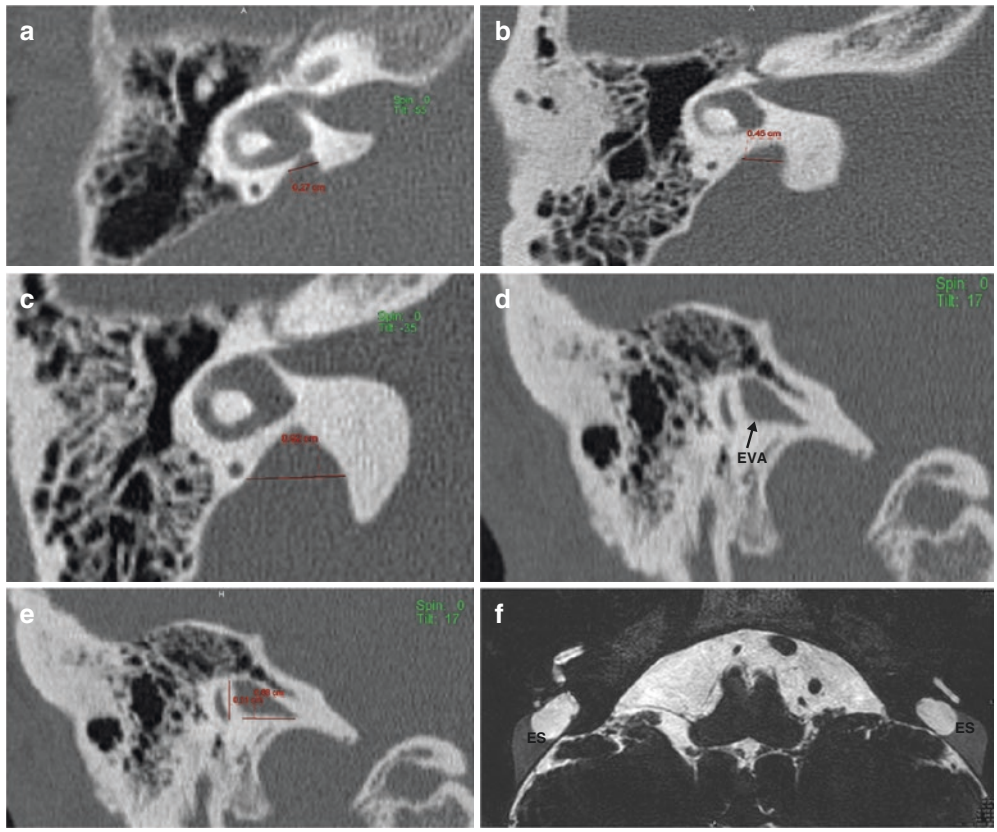


Fig. 24.4 Vestibular aqueduct enlargement: (**a**) Mild (1.5–3 mm), (**b**) Moderate (3–5 mm), (**c**) Massive (>5 mm), (**d**, **e**) measurement of vertical and horizontal dimensions of vestibular aqueduct on coronal sections, (**f**) Bilateral massive enlargement of endolymphatic sacs (ES)

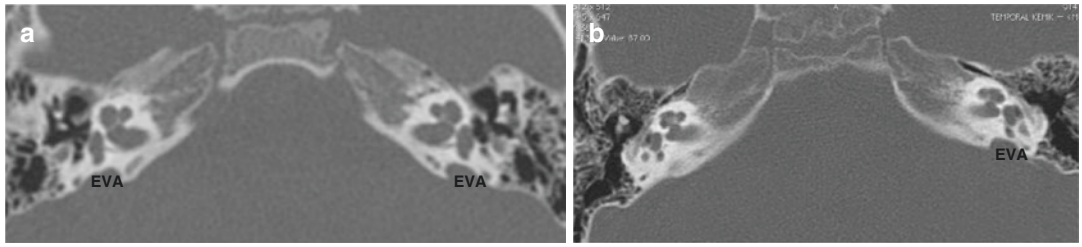


Fig. 24.5 Symmetric (**a**) and asymmetric (**b**) enlarged vestibular aqueduct (EVA)

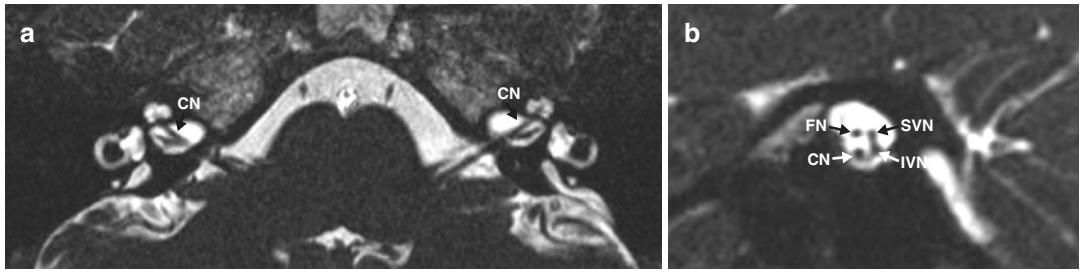


Fig. 24.6 Magnetic resonance imaging of cochlear nerve in IP-II: (**a**) axial section (*CN* cochlear nerve, *white star* modiolus), (**b**) parasagittal section perpendicular to the IAC (*CN* cochlear nerve, *FN* facial nerve, *SVN* superior vestibular nerve, *IVN* inferior vestibular nerve)

24.6 Audiology

These patients do not have a characteristic hearing level, as their audiometric threshold testing varies from normal to profound. The hearing loss can be symmetric or asymmetric (Fig. 24.7a, b). The most characteristic audiological finding is air-bone gap (ABG) particularly present at low frequencies (Fig. 24.7c), but sometimes ABG can be seen in all frequencies (Fig. 24.7d). The degree of the hearing loss can vary from normal hearing (Fig. 24.7e) to profound hearing loss (Fig. 24.7f). The configuration of the hearing loss may be increasing towards high frequencies (Fig. 24.7g) or be flat (Fig. 24.7h). Govaerts et al. [26] indicated that the conductive component could not be explained by middle ear impedance problems, such as effusion. Tympanometry is normal in the absence of otitis media and acoustic reflexes are generally present. The cause of the ABG in these children is likely to be due to a "third window" effect from the enlarged vestibular aqueduct and can resemble the audiometric findings in superior canal dehiscence syndrome.

These patients can experience progressive hearing loss throughout their lifetime. At birth, they may have normal hearing but usually there is progressive hearing loss over time. The EVA transmits the high CSF pressure to the inner ear and may cause progressive hair cell damage. Pulsation seen during the surgery when the cochleostomy is created demonstrates the high intracochlear pressure transmitted by EVA. Head trauma may exacerbate this hearing loss and these children are advised to avoid head trauma as much as possible. Sudden hearing loss can also be seen. Because of the progressive nature of the hearing loss, these patients usually have good language development. Papsin [27] also reported that children with incomplete partition are typically implanted older than other cases for the aforementioned reason.

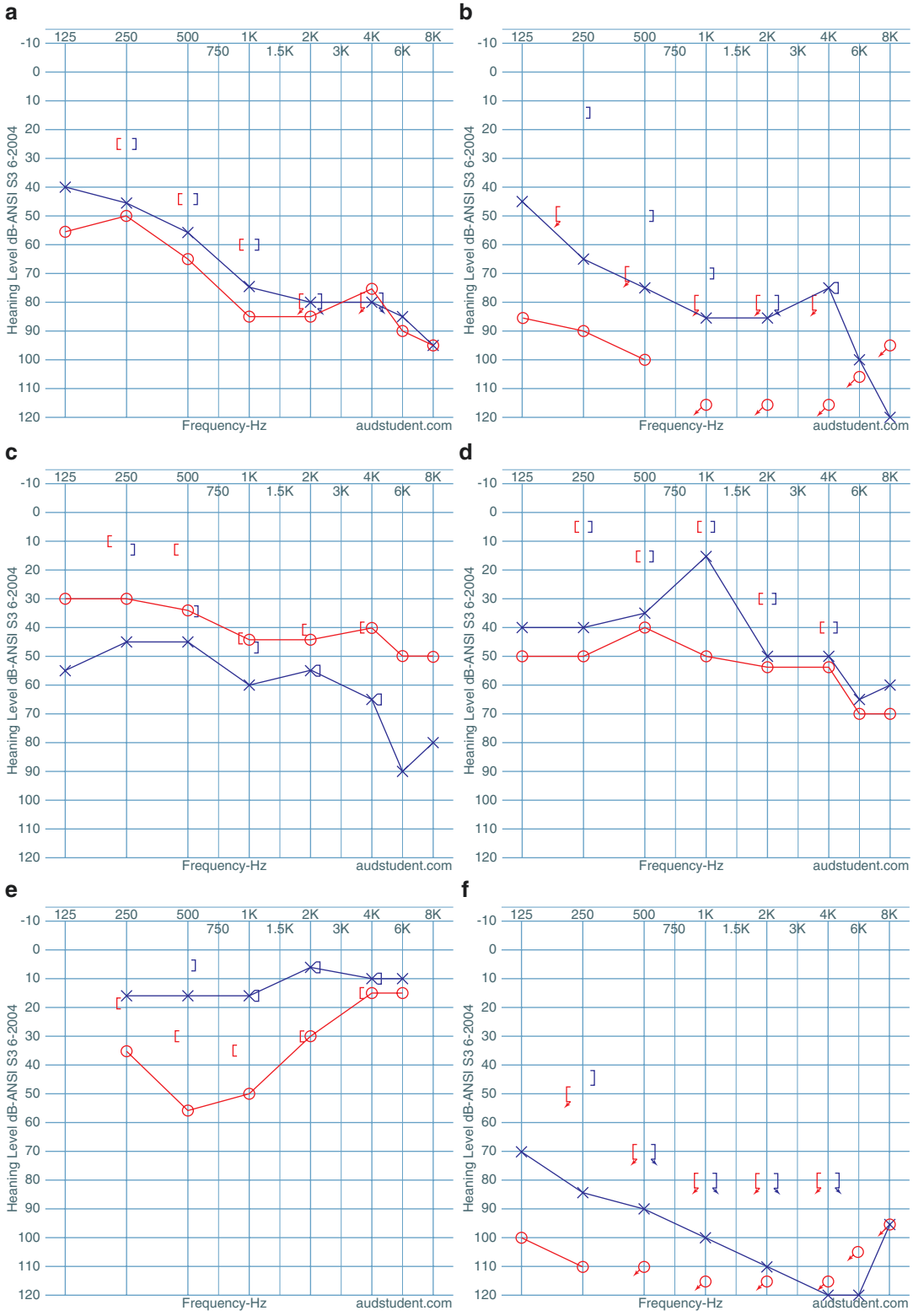


Fig. 24.7 Types of hearing loss (HL) in IP-II: (**a**) Symmetric mixed HL, (**b**) asymmetric mixed (HL), (**c**) air-bone gap at low frequencies, (**d**) air-bone gap in all frequencies, (**e**) normal hearing on the left side, (**f**) bilateral asymmetric profound hearing loss, (**g**) hearing loss increasing towards high frequencies, (**h**) Flat type HL

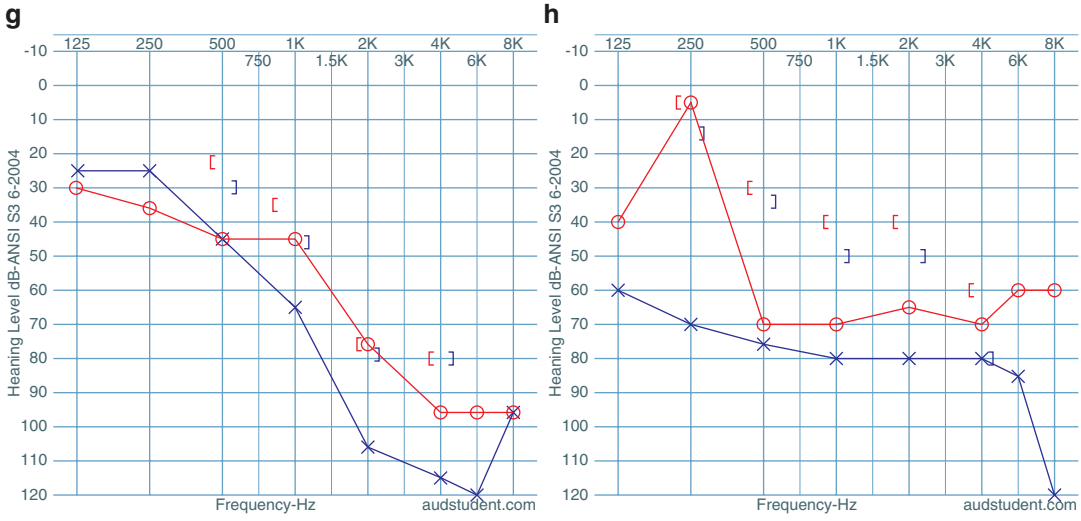


Fig. 24.7 (continued)

24.7 Management

If profound hearing loss is present at birth, child may fail in hearing screening and hearing aids may be indicated within a few months. More typically, these patients may have near normal hearing at birth and usually do not require amplification initially. In this situation, the hearing screening will fail to detect hearing loss in IP-II deformity. With progressive hearing loss, they become candidates for hearing aid.

In case of sudden deafness, they are advised to go to hospital immediately for steroid treatment and intravenous dextran infusion (see Case 24.1 below). If started early it is possible to have recovery to the previous thresholds. In case of late treatment hearing loss may be permanent. **All IP-II patients should be warned about the possibility of sudden hearing loss and immediate treatment to avoid progression of hearing loss as much as possible**. Every effort should be shown to preserve the natural hearing.

Usually the progression in hearing loss continues, ultimately creating a need for CI at some point in their life. They usually demonstrate very good language development. What causes this progression remains unknown. A role for head trauma has been suggested, and these patients are advised to avoid trauma by wearing helmets during sports and avoiding contact sports completely. It looks possible that transmission of CSF pressure via EVA plays a role in progressive hearing loss.

They have excellent language development. As the progression occurs usually during adolescence, they are usually reluctant to accept CI surgery during adolescence because they have developed excellent language until that time.

24.8 Surgery

In our department between 1997 and September 2018, 2646 patients underwent CI and ABI operations. 279 of these had IEMs. Ninety-three had IP-II deformity. During surgery, a facial recess approach was successfully used in all 93 patients who underwent CI surgery at Hacettepe University. This standard approach could be used in all patients because the cochlea and labyrinth had normal external dimensions. As a result, the facial nerve does not typically have an abnormal course that would prevent using the facial recess approach.

Four of these operations are revision surgery. One patient had two revisions because of progressive extrusion of CI electrode (see Case 24.6 below) and device failure. Other causes of revision were device failure and wound dehiscence.

Out of 93 patients operated on in our department, 45 patients had no CSF leakage, 42 patients had CSF oozing, and 6 patients had gusher (see Case 24.5 below). Almost all patients demonstrated pulsation at the time of cochleostomy (see Case 24.4 below). Pulsation without CSF leakage is used to explain pathophysiology and progressive nature of the disease. Head trauma may cause increase in CSF pressure resulting in progressive or sudden HL.

Gusher is more common in IP-I and IP-III, but it may occur in IP-II as well. This observation demonstrates that there may be a type VII modiolar defect (complete absence of the modiolus) (see Case 24.5). An EVA cannot be responsible for CSF leakage during cochleostomy. As we know endolymphatic sac is part of endolymphatic system but when we open round window we expose scala tympani which is part of the perilymphatic system. The most possible explanation for the gusher is the modiolar defect which is caused by high pulsating CSF pressure transmitted into cochlea via EVA during embryological development. This defect may allow CSF leakage at the time of round window opening. The cochleostomy should be closed completely because there is a risk of recurrent meningitis if CSF leakage persists around the electrode in the cochleostomy.

Electrode choice is important in IP-II. The basal part of the modiolus is normal. All kinds of electrodes (modiolar hugging, straight) can be used. Because of the risk of CSF leakage and occasional severe gusher, FORM electrodes with cork type stopper may be advantageous. As the external dimensions of the cochlea is similar to normal cases FORM 24 is advisable. The surgeon must be prepared to use the measures described in Chap. 15 to manage the CSF gusher.

As there is a low risk for the electrode to enter the IAC, X-ray can be taken after surgery. Six patients had severe gusher during surgery. In case of severe gusher there is wider connection between cochlea and IAC. In such cases it is advisable to obtain intraoperative transorbital X-ray to check the position of the electrode. If there is device malposition, it has to be revised during the surgery. No such complication occurred in 93 IP-II patients undergoing CI surgery.

None of the IP-II cases had any facial nerve abnormality during surgery. In addition no case of meningitis was observed in any of the IP-II patients.

24.9 Experience with CI

As they all have cochlear nerve, IP-II cases do not need an ABI.

Among the inner ear malformation subgroups, cases with IP-II malformations are the best performers with CI. They can improve their speech perception and auditory skills after CI. Their performances with CI are nearly same as the CI users with normal cochlear anatomy. In some cases, there can be side effects such as facial nerve stimulation or no auditory perception in some electrodes. For these side effects we recommend to use increased duration or pulse width and to deactivate affected electrodes. Due to the progressive hearing loss characteristics of IP-II, they have been followed with hearing aids before CI surgery. As a result they have auditory experience before CI surgery and this is the reason why they can achieve better auditory perception scores in early period after CI (see Chap. 30 for more details).

24.10 Cases

Case 24.1: 5-Year Old Male Patient, Attacks of Sudden Hearing Loss

He was diagnosed with bilateral severe hearing loss (Fig. 24.8a) and HRCT demonstrated bilateral IP-II deformity. Despite the fact that his family did not want CI, fluctuation in his hearing was determined during audiological follow-up. He had experienced three sudden hearing loss attacks on the right ear. After the sudden hearing loss (Fig. 24.8b), he was hospitalized for medical treatment with steroids and dextran for 10 days. His hearing improved after hospitalization (Fig. 24.8c). Two weeks later family applied with sudden hearing loss (Fig. 24.8d) on both sides. In spite of early admission and similar medical treatment, hear-

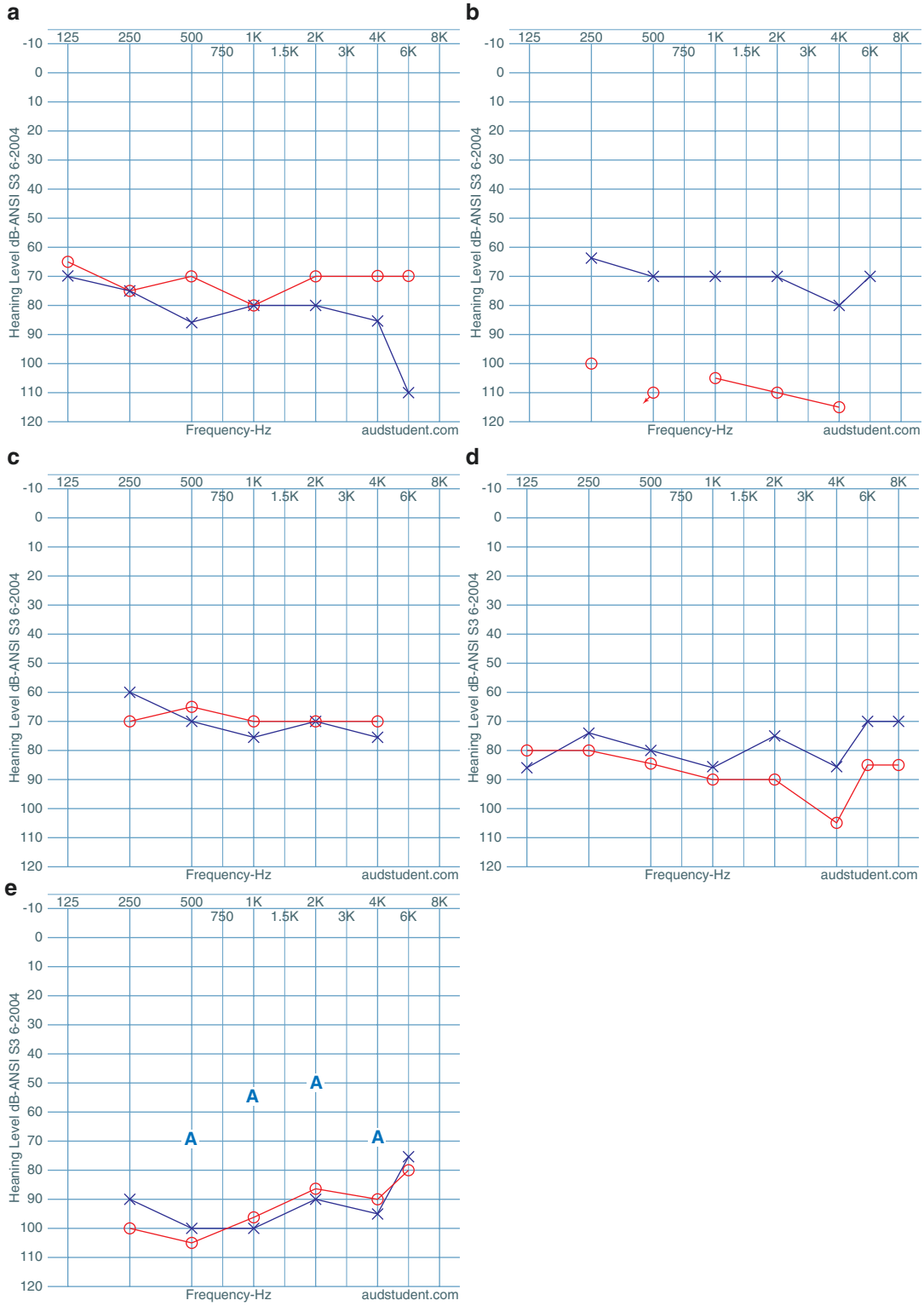


Fig. 24.8 Case 24.1: (**a**) Bilateral severe hearing loss (HL), (**b**) right side profound HL after sudden HL, (**c**) hearing recovery on the right side after medical treatment, (**d**) sudden SNHL on both sides 2 weeks after recovery, (**e**) No recovery after last attack. Profound HL requiring cochlear implantation

ing recovery was not as good as before. As a result hearing deteriorated and CI was suggested (Fig. 24.8e).

This is a typical presentation with attacks of sudden hearing loss and progressive hearing loss. It is probably due to transmission of CSF pressure into inner ear via EVA.

Case 24.2: 33-Year-Old Female Patient, Unilateral Hearing Loss and Tinnitus

She applied with the complaint of hearing loss and tinnitus on the right side (Fig. 24.9a). HRCT revealed right side EVA (Fig. 24.9b). Vestibular aqueduct and cochlea was normal on the left side. Her MRI was very interesting revealing enlarged scala vestibuli on the side with EVA (Fig. 24.9c). She was recently published as a radiological evidence for pressure transfer via EVA [7]. Side with normal vestibular aqueduct did not have any abnormality in the cochlea.

This is the lightest form of deformity that can be seen in IP-II. This patient shows radiologically that EVA transmits pressure into inner ear causing scala vestibuli dilatation during fetal development.

Case 24.3: 8-Year-Old Male Patient, Progresive Hearing Loss

He applied to our clinic at the age of 2 years old when he was using bilateral hearing aid. His first audiogram showed profound SNHL on the right and severe SNHL on the left (Fig. 24.10a). Cochlear implantation was suggested for the right ear. Family did not want CI due to his good performance with the hearing aids inspite of the detailed explanation of the possible progression of the hearing loss. During follow-up his hearing showed progression on the left side (Fig. 24.10b) and he did not respond to sounds as good as he did before at home. After this progressive hearing

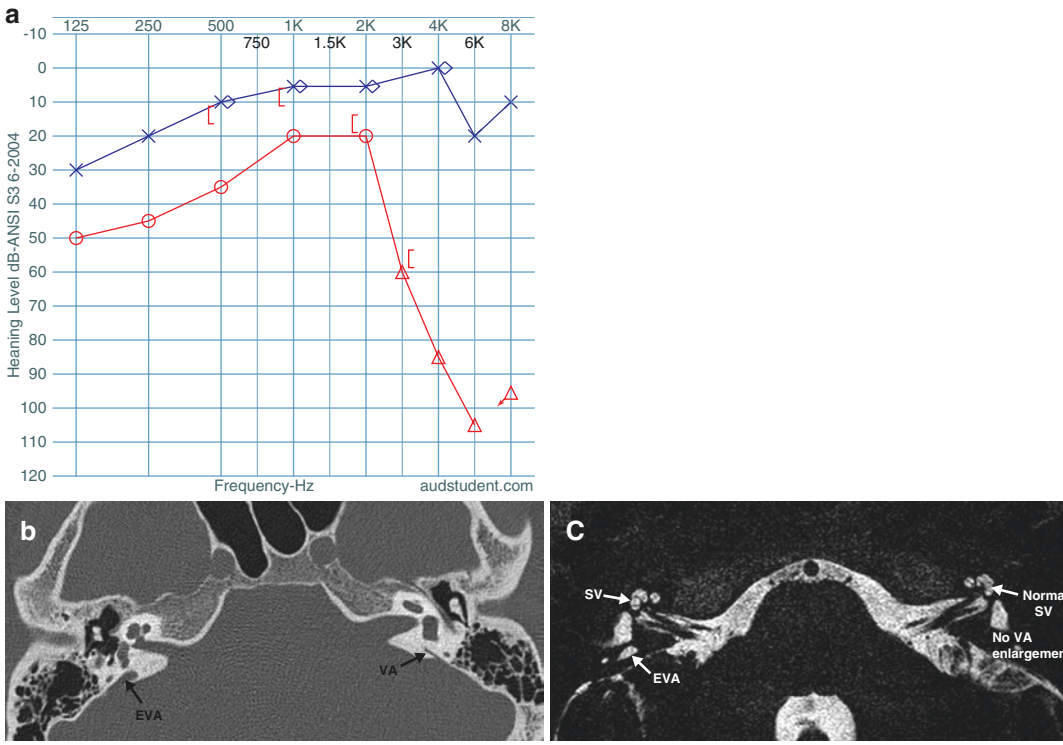


Fig. 24.9 Case 24.2: (**a**) Unilateral mild SNHL, (**b**) enlarged vestibular aqueduct (EVA) on the right, normal vestibular aqueduct on the left side (VA), (**c**) enlarged scala vestibuli (SV) on the side with enlarged vestibular aqueduct while left side reveals normal cochlear findings where vestibular aqueduct was normal. (With permission of CI International)

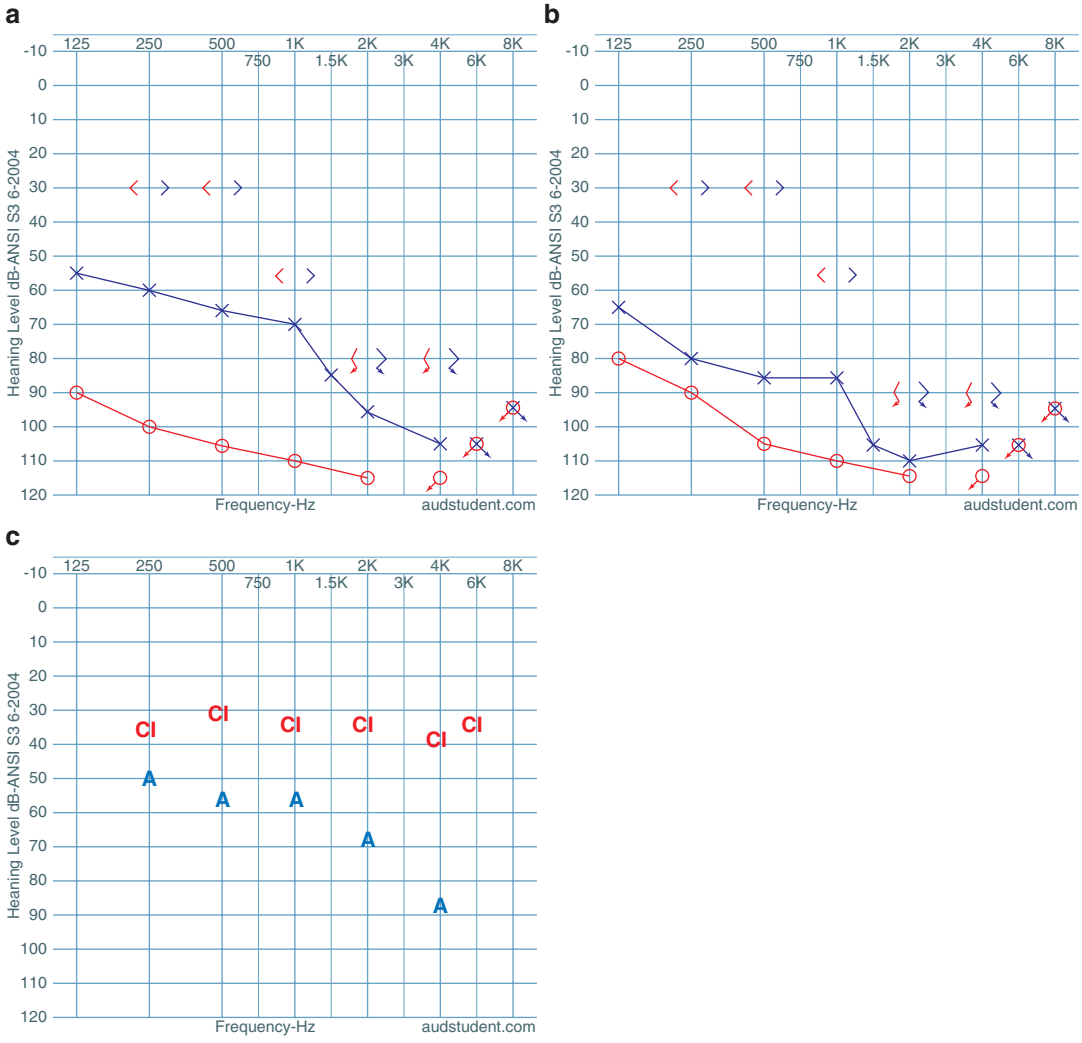


Fig. 24.10 Case 24.3: (**a**) profound SNHL on the right and severe SNHL on the left, (**b**) progressive SNHL resulting in bilateral profound SNHL, (**c**) Aided thresholds with right CI

loss on the left ear, he was implanted on the right ear and left ear was followed with the hearing aids. Postoperative hearing thresholds with CI are given in Fig. 24.10c.

Case 24.4: 14-Year-Old Female Patient, Operated on January 2014

She had progressive SNHL necessitating a CI. HRCT of the patient revealed bilateral IP-II deformity with type V: partial modiolar defect (Fig. 24.11). During cochleostomy there was no CSF leakage (gusher or oozing) but pulsation was present (Video 15.1). When the fluid accumulating at the cochleostomy was removed cochleostomy site appeared to be normal. When dexamethasone was introduced strong pulsation was observed.

This is the most common observation in IP-II patients. We think this strong pulsating CSF is the reason for modiolar destruction during embryological development also causing progressive hearing loss later on.

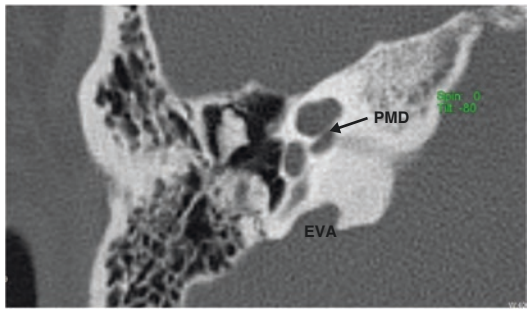


Fig. 24.11 Case 24.4: Partial modiolar defect (PMD) where there is a thin partition between cochlea and internal auditory canal (type V modiolar defect)

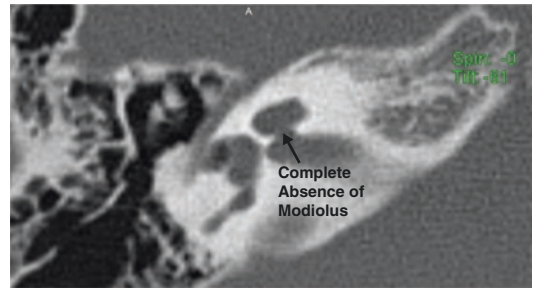


Fig. 24.12 Case 24.5: Complete absence of the modiolus (type VII modiolar defect) no partition between cochlea and internal auditory canal

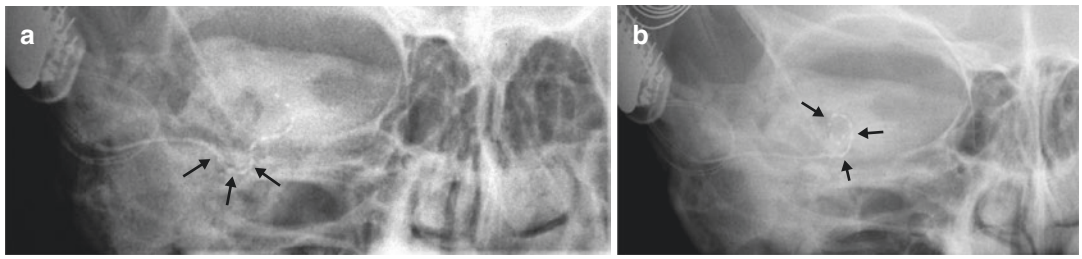


Fig. 24.13 Case 24.6: (**a**) Electrode extrusion out of the cochlea, (**b**) full insertion after revision

Case 24.5: 7-Year-Old Female Patient, Operated on March 2010

She had bilateral profound SNHL and right side CI was planned. Her HRCT demonstrated bilateral IP-II deformity with type VII: complete absence of the modiolus (Fig. 24.12). During surgery there was a profuse CSF gusher. We used a FORM 24 electrode passed through muscle tissue to stop the leakage.

Gusher is very rare in IP-II but still can be experienced. In IP-II HRCT should be examined from this perspective to see the possibility of CSF gusher. If there is type VII modiolar destruction gusher is possible. Surgeon then should be ready for gusher. Appropriate electrode should be prepared and the family should be informed about the possibility of CSF leakage and risk of meningitis.

Case 24.6: 27-Year Old Female Patient, Operated on 9 Sep 2008

She had bilateral IP-II with progressive SNHL. She was operated in 2008. There was pulsation during surgery and FORM 24 was inserted fully into cochlea. Postoperative transorbital X-ray showed good placement of the device into the cochlea. She made good benefit from CI. Then her benefit started to decrease and she started to have pain during CI use. Her transorbital film was repeated. It was surprising to see the extrusion of the electrode out of the cochlea (Fig. 24.13a). She was revised in November 2009. Again there was pulsation during cochleostomy. Full insertion was obtained (Fig. 24.13b).

This is rare but it may be due to CSF pulsating pressure pushing the electrode out of the cochlea. It is advisable to pass the electrode through a piece of fascia and have a firm application at the cochleostomy.

References

1. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope. 2002;112(12):2230–41. https://doi.org/10.1097/00005537-200212000-00019.
2. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41. https://doi.org/10.1002/cii.416.

3. Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. Am J Otol. 1994;15(4):551–7.
4. Lo WW. What is a 'Mondini' and what difference does a name make? AJNR Am J Neuroradiol. 1999;20(8):1442–4.
5. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25(4):520–9.
6. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20. https://doi.org/10.1179/1754762815Y.0000000016.
7. Sennaroglu L. Another evidence for pressure transfer mechanism in incomplete partition two anomaly via enlarged vestibular aqueduct. Cochlear Implants Int. 2018;19(6):355–7. https://doi.org/10.1080/14670100.2018.1489938.
8. Batuk MÖ, Çınar BÇ, Özgen B, Sennaroğlu G, Sennaroğlu L. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233.
9. Reinshagen K, Curtin H, Quesnel A, Juliano A. Measurement for detection of incomplete partition type II anomalies on MR imaging. Am J Neuroradiol. 2017;38(10):2003–7.
10. Kamogashira T, Akamatsu Y, Kashio A, Ogata E, Karino S, Kakigi A, Iwasaki S, Yamasoba T. Development of auditory skills after cochlear implantation in children with inner ear malformations. Acta Otolaryngol. 2016;136(1):78–82.
11. Wu CC, Chen YS, Chen PJ, Hsu CJ. Common clinical features of children with enlarged vestibular aqueduct and Mondini dysplasia. Laryngoscope. 2005;115(1):132–7.
12. Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N. Cochlear implant in incomplete partition type I. Acta Otorhinolaryngol Ital. 2013;33(1):56.
13. Attias J, Ulanovski D, Shemesh R, Kornreich L, Nageris B, Preis M, Peled M, Efrati M, Raveh E. Air-bone gap component of inner-ear origin in audiograms of cochlear implant candidates. Otol Neurotol. 2012;33(4):512–7.
14. Boston M, Halsted M, Meinzen-Derr J, Bean J, Vijayasekaran S, Arjmand E, Choo D, Benton C, Greinwald J. The large vestibular aqueduct: a new definition based on audiologic and computed tomography correlation. Otolaryngol Head Neck Surg. 2007;136(6):972–7.
15. Clark JL, Roeser RJ. Large vestibular aqueduct syndrome: a case study. J Am Acad Audiol. 2005;16(10):822–8.
16. Gopen Q, Zhou G, Whittemore K, Kenna M. Enlarged vestibular aqueduct: review of controversial aspects. Laryngoscope. 2011;121(9):1971–8.
17. Ha JF, Wood B, Krishnaswamy J, Rajan GP. Incomplete cochlear partition type II variants as an indicator of congenital partial deafness: a first report. Otol Neurotol. 2012;33(6):957–62.
18. Hirai S, Cureoglu S, Schachern PA, Hayashi H, Paparella MM, Harada T. Large vestibular aqueduct syndrome: a human temporal bone study. Laryngoscope. 2006;116(11):2007–11.
19. Mamıkoğlu B, Bentz B, Wiet RJ. Large vestibular aqueduct syndrome presenting with mixed hearing loss and an intact mobile ossicular chain. Otorhinolaryngol Nova. 2000;10(5):204–6.
20. Nakashima T, Ueda H, Furuhashi A, Sato E, Asahi K, Naganawa S, Beppu R. Air–bone gap and resonant frequency in large vestibular aqueduct syndrome. Otol Neurotol. 2000;21(5):671–4.
21. Pritchett C, Zwolan T, Huq F, Phillips A, Parmar H, Ibrahim M, Thorne M, Telian S. Variations in the cochlear implant experience in children with enlarged vestibular aqueduct. Laryngoscope. 2015;125(9):2169–74.
22. Roesch S, Moser G, Rasp G, Toth M. CT-scans of cochlear implant patients with characteristics of Pendred syndrome. Cell Physiol Biochem. 2013;32(7):166–72.
23. Sennaroglu G, Sennaroglu L. Hearing loss in inner ear malformations. In: Encyclopedia of otolaryngology, head and neck surgery. Berlin: Springer; 2013. p. 1143–50.
24. Ahadizadeh E, Ascha M, Manzoor N, Gupta A, Semaan M, Megerian C, Otteson T. Hearing loss in enlarged vestibular aqueduct and incomplete partition type II. Am J Otolaryngol. 2017;38(6):692–7.
25. Cinar BC, Atas A, Sennaroglu G, Sennaroglu L. Evaluation of objective test techniques in cochlear implant users with inner ear malformations. Otol Neurotol. 2011;32(7):1065–74.
26. Govaerts PJ, Casselman J, Daemers K, De Ceulaer G, Somers T, Offeciers FE. Audiological findings in large vestibular aqueduct syndrome. Int J Pediatr Otorhinolaryngol. 1999;51(3):157–64.
27. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(1 Pt 2 Suppl 106):1–26. https://doi.org/10.1097/00005537-200501001-00001.

Incomplete Partition Type III

25

Levent Sennaroglu, Merve Ozbal Batuk, and Munir Demir Bajin

Specific Features

1. Mixed or sensorineural hearing loss.
2. Labyrinthine segment of the facial nerve above cochlea.
3. Irregular vestibular aqueduct enlargement.
4. Severe gusher.
5. Spontaneous CSF fistula or recurrent meningitis uncommon.
6. Thinner otic capsule due to missing outer two layers; most probably due to defective vascular supply from middle ear.
7. Avoid stapedotomy.

25.1 Definition

The cochlea in incomplete partition type III (IP-III) has an interscalar septa but the modiolus is completely absent (Fig. 25.1) [1, 2]. IP-III cochlear malformation is the type of anomaly present in X-linked deafness, which was described by Nance et al. [3] for the first time in 1971. Phelps et al. [4] initially described the high resolution computerized tomography (HRCT) findings associated with this condition and this characteristic deformity was included under the category of incomplete partition deformities for the first time by Sennaroglu et al. in 2006 [5]. This anomaly is the rarest form of incomplete partition cases. According to the radiological database in Hacettepe University Department of Otolaryngology, IP-III constitutes 2.9% of the inner ear malformation group.

25.2 Histopathology and Pathophysiology

No specimen involving IP-III has been reported in the literature. As we have no histopathological specimen involving IP-III cochlea, information obtained from the radiology of the temporal bone is used to explain pathophysiology.

Radiological evaluation of cases with X-linked deafness revealed a normal sized cochlea with a very specific appearance where interscalar septa are present but modiolus is absent. Otic capsule around the membranous labyrinth is thinner in IP-III when compared to that in a normal cochlea (Fig. 25.1). If a specimen of a normal cochlea is examined under light microscopy, the inner endosteal layer of the otic capsule follows the outline of the membranous labyrinth (Fig. 25.2). The middle

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_25].

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

M. D. Bajin
Department of Otolaryngology, Hacettepe University, Medical Faculty, Ankara, Turkey
e-mail: dbajin@hacettepe.edu.tr

M. Ozbal Batuk (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_25

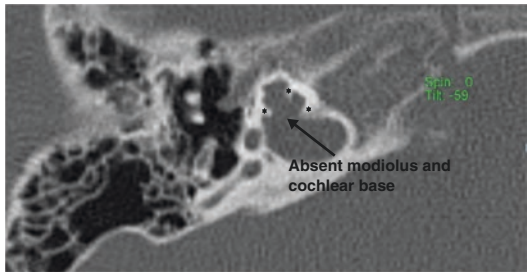


Fig. 25.1 Cochlea in incomplete partition type III (IP-III) with interscalar septa (black stars) but absent modiolus

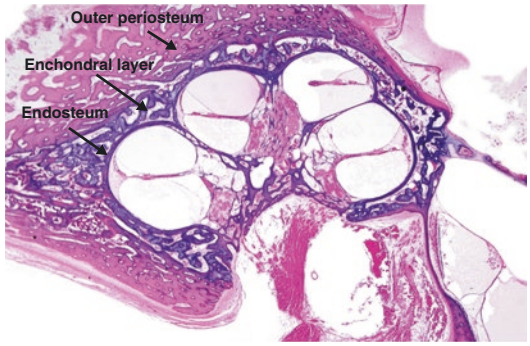


Fig. 25.2 Histopathological specimen showing normal cochlea. Note that endosteum follows the membranous labyrinth, while enchondral and outer periosteal layers increase the thickness of the otic capsule, without following the contour of the membranous labyrinth. (With permission from Massachusetts Eye and Ear Infirmary)

enchondral and outer periosteal layers increase the thickness of the otic capsule, without following the contour of the membranous labyrinth. HRCT demonstrates that in IP-III, the otic capsule around the cochlea is thin and follows the outline of the membranous labyrinth as if it is formed only by a thick endosteal layer. Instead of the usual three layers, probably the second and third layers are either absent or very thin. The otic capsule most probably consists of a thickened inner endosteal layer without enchondral and outer periosteal layers.

Sennaroglu L recently explained pathophysiology of IEMs by embryological development of the otic capsule [6]. Outer two layers of the otic capsule receive their vascular supply from the middle ear. It appears that pathology in X-linked deafness may be due to abnormal vascular supply from the middle ear mucosa as a result of a genetic abnormality resulting in an otic capsule consisting of only thicker endosteal layer.

Absence of outer two layers resulting in a thinner otic capsule has other distinctive features which are specific for IP-III. In normal anatomical conditions the cochlea is located at the anterolateral part of the IAC. Vestibule occupies the posterolateral part of the IAC. In IP-III, it looks as if the cochlea is directly located at the lateral end of the IAC, almost in a straight line. This is a unique finding not present in any of the other anomalies. However, this is a misinterpretation, because of the absence of enchondral and outer periosteal layers at the base of the cochlea. In a normal cochlea, the middle enchondral layer constitutes the major part of the cochlear base, with a contribution from the inner periosteal layer (Fig. 25.2). Therefore, the missing bony layer or layers give a false impression that the cochlea is situated directly lateral to the IAC (Fig. 25.1).

The cochlear base and modiolus are completely absent. The cochlear base consists of two layers: the endosteum and the middle enchondral layer. The middle enchondral layer provides the bulk to the cochlear base. If this layer is absent, the endosteal layer may not be sufficient to provide a thick base where the modiolus will have its support. Therefore, the absence of layers 2 and 3 results in a defective cochlear base and absent modiolus, in spite of normal vascularization from the IAC to the modiolus. It is quite possible that, if the base is defective, the modiolus cannot form the attachment points and develop appropriately.

Spontaneous CSF leakage through the oval window is frequently seen and reported in IP-I, even though both IP-I and IP-III both are associated with high volume CSF leakage on cochleostomy. This is most probably due to stapes footplate defect as a result of endosteal developmental anomaly in IP-I. In IP-III endosteum is normally developed with thinner otic capsule due to defective outer layers. Therefore, spontaneous CSF leakage is very rare in IP-III.

25.3 Literature Findings

Incomplete partition type III malformation (IP-III) is the type of cochlear anomaly seen in X-linked stapes gusher syndrome or X-linked

deafness in the literature. X-linked stapes gusher syndrome is the result of a mutation in the POU3F4 gene of the X chromosome. Due to the X-linked inheritance, IP-III malformation is expected to be seen only in males instead of females. Nevertheless, some studies showed that it may rarely be observed in females [7, 8]. This was explained with the possibility of the autosomal recessive form of the pathology or the non-randomized inactivation of the normal X gene with a mutation of the X gene.

Sennaroglu et al. [9] stated that IP-III is the least common anomaly among incomplete partition anomalies. They suggested that cases with IP-III are usually diagnosed with severe to profound mixed type hearing loss and the most appropriate approach is cochlear implantation.

Sennaroglu et al. [2] observed mixed hearing loss where sensorineural component was dominant in their patients and cochlear implant was applied to all the 7 patients. Air bone gap was not significant and no stapes surgery was done. They stated that if stapes surgery was done, there would be severe gusher, with a very high risk of losing the residual hearing. In such patients, a hearing aid or CI can be recommended according to the level of the hearing loss. However, stapes surgery should be avoided especially in the patients with a SNHL component at an early grade. Otherwise hearing loss will increase or may become profound.

Choi et al. [10] retrospectively analyzed the medical records of five patients with IP-III in order to determine the reason for the air bone gap in IP-III malformations. All five patients were diagnosed with moderate to profound mixed type hearing loss, which was usually progressive. Although stapes fixation was observed in 3 out of 5 patients, stapes was mobile in the remaining two patients. They mentioned that stapes fixation cannot be the reason for the air bone gap in IP-III due to the inconsistency of the findings of these patients during the middle ear surgery. They concluded that the main reason for the air bone gap in IP-III is the pathologic third window phenomenon which is the result of an abnormal connection between cochlea and internal acoustic canal.

Jeong and Kim [11] mentioned that audiologic features and pathogenesis of the IP-III were different than other IEMs. They recommended bone conduction hearing aids as the initial choice when the inner ear function is good and conductive hearing loss is observed. They mentioned that in case of large air bone gap with good bone conduction thresholds, follow-up can be done with bone conduction hearing aids.

In the study by Stankovic et al. [12], audiological outcomes of 4 boys with IP-III were evaluated with speech recognition tests and behavioral thresholds with CI. Although one of these developed closed set speech perception, remaining three only developed speech detection and were using primarily sign language after cochlear implantation. They also found out that it was not possible to observe electrical compound action potentials in the basal part of the cochlea in this group despite the functioning internal device. They thought that cognitive and behavioral problems could also affect the speech development of these four cases but they suggested hearing aids rather than cochlear implantation in cases of IP-III for better sound perception. As a result, they advised preoperative gene mutation analysis and long term follow-up in patients with IP-III.

Choi et al. [13] analyzed the database of the 1200 CI users retrospectively and reported the audiological, radiological, and surgical results of 11 patients with IP-III. Eight of these 11 patients underwent CI. They evaluated the auditory perception abilities of all patients before and after CI and found out that the auditory performance of the patients with IP-III was similar to patients with normal cochlea after CI at the first 3 months. But between postoperative third months and 24th months, it was found that the auditory progression was slower and their speech perception abilities are worse than children with normal cochlear anatomy.

In the recent study by Kanno et al. [14], IP-III was diagnosed in six out of 1004 patients with a prevalence of 0.6%. Even though enlarged vestibular aqueduct was seen in all patients, none of them had fluctuation in hearing during follow-up. Degree of the hearing loss was reported to be

mild to profound in their series, all of which were mixed type hearing loss.

Kamogashira et al. [15] analyzed the frequency of intraoperative cerebrospinal fluid (CSF) gushers and the frequency of facial nerve stimulation for each type of malformation in their study. They mentioned that the prevalence of CSF gusher was higher for the malformations with poorly shaped modiolus. While CSF gusher was seen in one IP-III case, postoperative facial nerve stimulation was not observed for this case.

Smeds et al. [16] published their results in 15 IP-III patients. Nine of the children had a mutation affecting the gene POU3F4 on Xq21. In three cases the electrode was found to be in the internal auditory canal on intraoperative X-ray and repositioned successfully. One child had a postoperative rhinorrhea, which was confirmed to be cerebrospinal fluid but this resolved with conservative treatment. No severe complications occurred. They concluded that cochlear implantation is a safe and beneficial procedure for this patient group.

25.4 Clinical Findings

The patients present with pure SNHL or more commonly with mixed hearing loss. Hearing loss is not progressive. We have not encountered any IP-III case with pure CHL.

CHL component may be due to a fixed footplate. In our experience, during CI surgery palpation of the stapes usually revealed fixation. In the past, stapedotomy attempts in such cases have resulted in a CSF gusher; **stapedotomy should be avoided in these patients** [17]**.** The vestibular surface of the stapes develops from the endosteum, just like the ISS. Endosteal thickening is most probably present here as well, causing stapes fixation and hence a conductive component on an audiogram.

It is possible to detect bone conduction levels in IP-III, in spite of the absence of the modiolus; the levels found may not be true bone conduction levels. There may be two mechanisms to explain this:

1. **Thickness of the otic capsule:** These cases always have a normal sized cochlear nerve demonstrated on MRI (Fig. 25.8). Absence of the outer two layers may make the cochlea more sensitive to bone conduction.
2. **Third window phenomenon:** The other explanation for bone thresholds is that the defective cochlear base is acting like a third window. A defective cochlear base is present in IP-I as well, resulting in a gusher and an oval window fistula. But the cochlear base defect cannot be the reason for sensitive bone conduction, as none of the IP-I patients has good BC levels. On MRI, all patients with IP-III have a well-developed cochlear nerve. In contrast, in IP-I cases, cochlear nerve deficiency is frequently observed. Therefore, a normal cochlear nerve supply may result in better bone conduction levels with a thinner otic capsule, in spite of the absence of the modiolus. The thin otic capsule most probably makes the cochlea more sensitive to sound.

There has been no record of a spontaneous CSF fistula through the stapes footplate, in spite of the fact that there is always a severe CSF gusher during the cochleostomy. On HRCT, the otic capsule is thin, but no fistula is reported. As a result, no case of recurrent meningitis has been reported in IP-III. It is possible to explain this by embryology. In IP-III, there is a normally developed inner endosteal layer. In contrast to IP-I, this high CSF pressure never causes footplate erosion. In IP-I, enchondral development is deficient, probably due to vascular insufficiency. This results in absent ISS and defective footplate. High CSF pressure then produces a fistula and recurrent meningitis in IP-I. As endosteal development is normal in IP-III no stapes fistula and recurrent meningitis has been reported in spite of CSF filling every IP-III cochlea.

25.5 Radiology

According to the IEM database of Hacettepe University Department of Otolaryngology out of 776 patients with various IEMs, 44 of 1552 ears had IP-III (2.9%). One hundred percent of these

were bilateral and symmetric. It is interesting to note that none of the IP-III had a different pathology on the contralateral ear.

Cochlear deformity in X-linked deafness is always symmetrical and bilateral and very similar in different individuals (Fig. 25.3).

Phelps et al. [4] reported that there is a bulbous IAC, incomplete separation of the coils of the cochlea from the internal auditory canal and widened first and second parts of the intratemporal facial nerve canal with a less acute angle between them. Talbot and Wilson [18] later added that the modiolus is absent and there is a more medial origin of the vestibular aqueduct with varying degrees of dilatation.

Sennaroglu et al. [19] reported that in this deformity the size of the cochlea is similar to normal cases with interscalar septa present but the modiolus is completely absent (Fig. 25.1). From an earlier study, the external dimensions of the cochlea (height and diameter) were found to be similar to the normal cochlea [5]. As the size is normal, it was included in the incomplete partition anomalies of the cochlea [5]. The cochlea is located directly at the lateral end of the internal auditory canal instead of its usual anterolateral position (Figure 25.4a). This gives the cochlea a characteristic appearance. This may be due to absence of cochlear base as mentioned before.

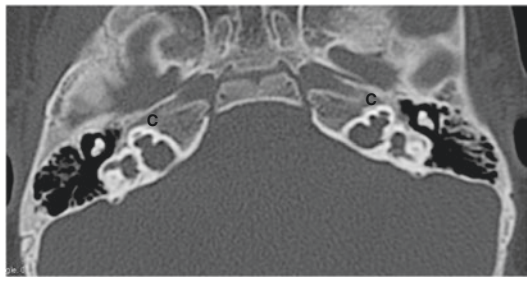


Fig. 25.3 IP-III cochlea deformity which is always symmetrical and bilateral

Cochlear base has a large defect (Figs. 25.1 and 25.4a, b). This defect is larger than type 7 complete absent modiolus. This increases the possibility of electrode misplacement into IAC more than IP-I cases. Therefore, modiolar hugging electrodes should not be used in IP-III (modiolus is absent in IP-III).

Some cases have quite thick interscalar septum. This decreases the volume of intracochlear space (Fig. 25.5). This may result in electrode misplacement into IAC if a long electrode (>25 mm in length) is used. It is advisable to use an electrode with a shorter length (<20 mm) in this situation.

Labyrinthine segment of the facial nerve has a more superior position in relation to cochlea [19]; the labyrinthine segment is located almost above the cochlea (Fig. 25.6). This is very specific for IP-III. If the axial sections are followed from top to bottom, the first structure that is identified, above the cochlea, in IP-III is the labyrinthine segment of the facial nerve. The labyrinthine segment of the facial nerve normally courses around the basal part of the cochlea, but in this anomaly it is always located above the cochlea. Recently Sennaroglu [6] added that there is a much thinner otic capsule around the cochlea and vestibule where interscalar septum appears to be thicker than normal The labyrinthine segment of the

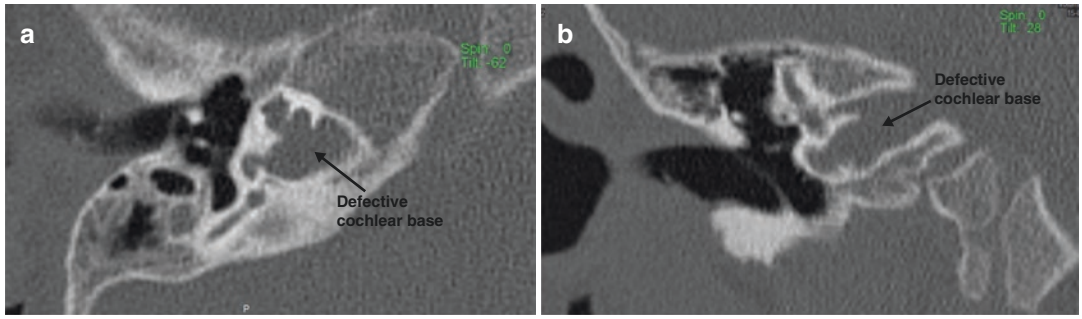


Fig. 25.4 Large defect at the cochlear base: (**a**) axial and (**b**) coronal views. This is not just absence of the modiolus but whole cochlear base is defective which makes it possible for the modiolar hugging electrodes to migrate into IAC

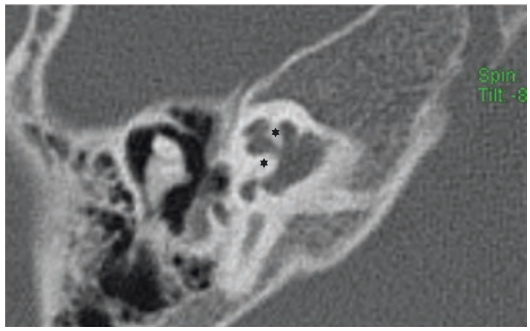


Fig. 25.5 IP-III cochlea with thick interscalar septum (black stars), resulting in a decrease in volume of intracochlear space

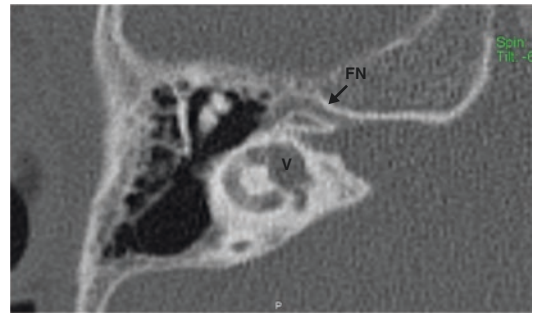


Fig. 25.6 Labyrinthine segment of the facial nerve (FN) located superior in relation to cochlea (*V* vestibule)

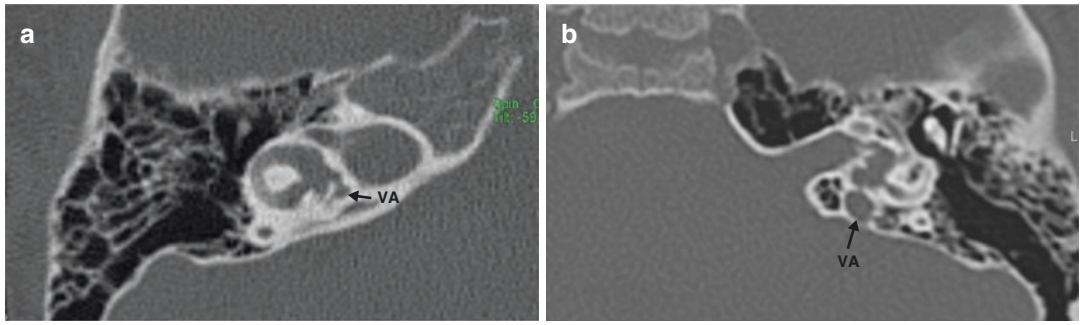


Fig. 25.7 (**a**, **b**) Vestibular aqueduct (VA) dilatation which is irregular and dysplastic, different from characteristic EVA

facial nerve is not in the normal position, because missing layers of the otic capsule prevent the nerve to obtain its normal position. The outer layers of the otic capsule, therefore, play an important role in the position of the labyrinthine segment of the facial nerve.

Vestibular aqueduct shows dilatation (Fig. 25.7a, b). It is irregular and dysplastic; it is not like characteristic, smooth dilatation observed in EVA. This feature may also be due to defective outer two layers of the otic capsule.

MRI demonstrates cochlear nerve in all IP-III cases (Fig. 25.8). Therefore, ABI is not indicated in IP-III.

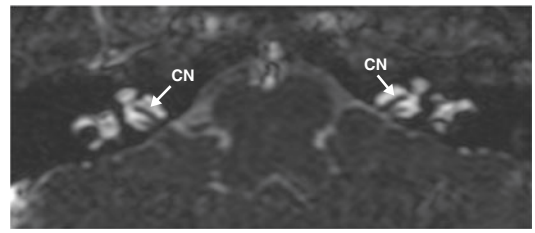


Fig. 25.8 Bilateral normal cochlear nerve

25.6 Audiological Findings

There may be two types of hearing loss associated with this malformation:

1. Mixed type hearing loss (Fig. 25.9a): The SNHL component is most likely due to the modiolar defect, whereas the conductive component may be due to stapedial fixation or third window phenomenon. The air bone gap usually involves high frequencies as well as low frequencies. Snik et al. [20] suggest that the air bone gap is associated with a third window phenomenon. They reported that because of the congenital malformation, the audioves-

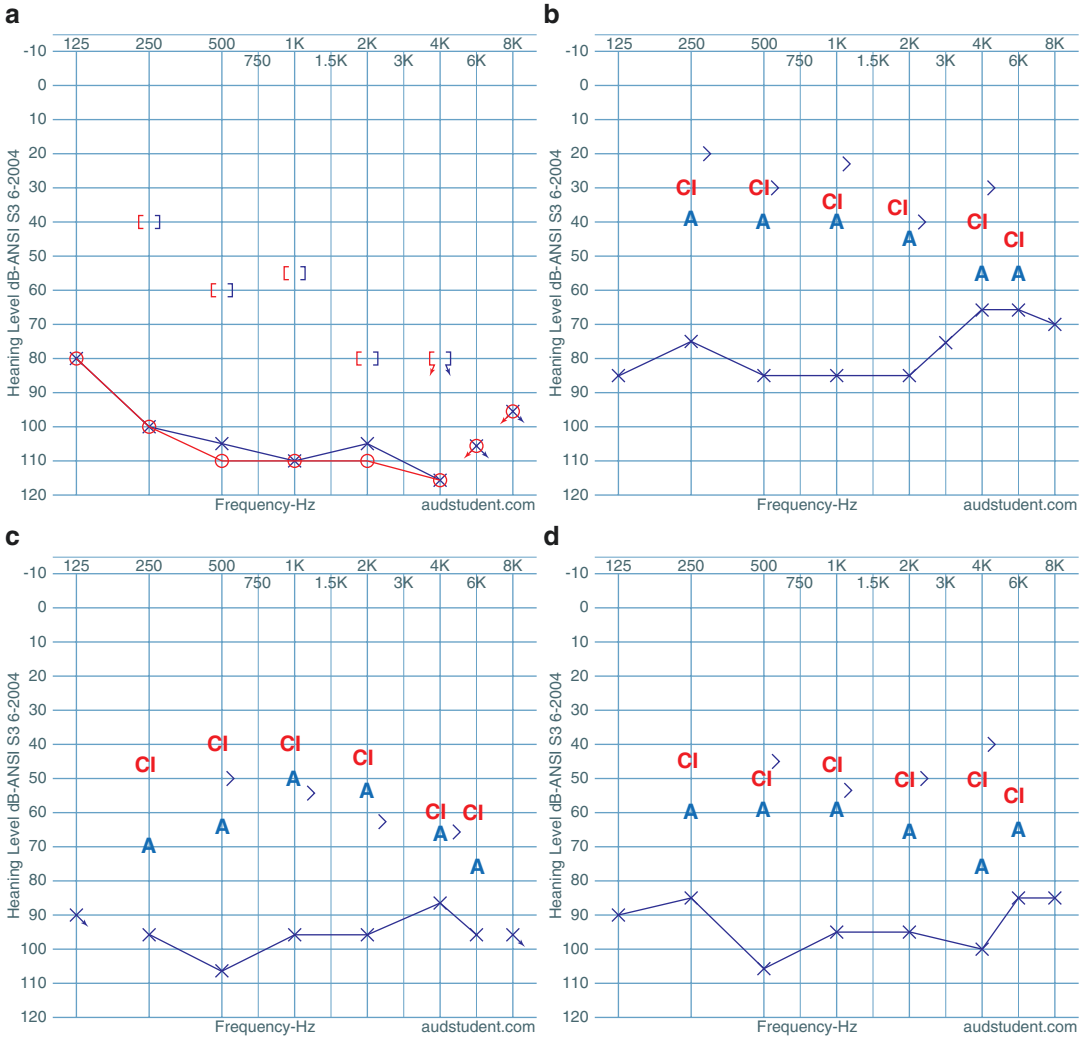


Fig. 25.9 Audiological findings in IP-III: (**a**) Profound mixed type hearing loss, (**b**) Hearing thresholds with right cochlear implant and severe mixed type hearing loss with contralateral hearing aids, (**c**, **d**) Hearing thresholds with cochlear implant on the right side and hearing aid on the left side of twin brothers (*A* thresholds with hearing aids, *CI* thresholds with CI)

tibular system functioned more effectively than it normally would, thus leading to better bone conduction levels. Their study revealed that audiological studies were in accordance with pure sensorineural hearing loss and air bone gap in the audiogram did not signify a conductive hearing loss component.

In the study of one of the authors [21], all patients with IP-III were diagnosed with severe to profound mixed type hearing loss. The air conduction thresholds were at the range of 70–110 dB and better in the low frequencies. Conversely, bone conduction thresholds were at the level of 50–80 dB and in general air bone gap was observed in all frequencies.

2. Profound SNHL: In the study of Snik et al. [20], audiological findings (such as pure tone audiometry, stapedial reflex, and auditory brainstem responses) in patients with X-linked deafness showed similar findings with pure sensorineural hearing loss. In our clinical experience, however, no patient had pure sensorineural hearing loss. All patients with IP-III were diagnosed with mixed type hearing

loss, with various degrees of air bone gap. The severity of this type of hearing loss may vary. It is most likely due to the absence of the modiolus and in this situation, CI surgery is the primary means of restoring hearing.

25.7 Management

Patients with severe mixed type hearing loss can be habilitated with hearing aids until they reach the plateau. It is possible to develop speech and language with hearing aids. Nearly all of our patients are rehabilitated with cochlear implants and most of them prefer to use hearing aids on the contralateral ear. Despite profound hearing loss, patients with IP-III can benefit from hearing aids on the contralateral ear for bimodal stimulation. Figure 25.9b–d shows audiological findings of three brothers with IP-III.

In mixed type hearing loss, bone anchored hearing devices were also shown as an option for the management of the hearing loss in the literature [11]. According to our clinical experience, it is difficult to recommend bone conduction devices to the cases with IP-III. Due to the increased bone conduction thresholds (nearly 50–70 dB), it is difficult to obtain benefit in hearing with bone anchored hearing devices. None of our patients was diagnosed with pure conductive hearing loss. All have mixed type hearing loss with high bone conduction thresholds.

Mixed hearing loss gives the impression of stapedial fixation. Stapedotomy results in severe gusher and further SNHL, and thus, should be avoided. There were many reports in the past where a stapedectomy was performed in IP-III cases resulting in dead ears. Patients with severe-profound HL are candidates for CI.

As CN is always present ABI is never indicated in IP-III.

25.8 Surgery

Because of the absent modiolus and cochlear base, during the surgery of IP-III two serious problems may occur:

1. Gusher: These patients may have severe gusher during surgery, because of the large defect between the cochlea and the IAC. If it is not properly sealed, the postoperative CSF leakage may lead to recurrent meningitis. Ideally, the size of the cochleostomy should be slightly larger than the electrode in order to allow for soft tissue to be placed around the electrode. Passing the electrode through a tiny piece of fascia and inserting this together with the electrode may further improve the seal at the cochleostomy site.

   In case of severe CSF leakage, a continuous lumbar drainage is always performed. This decreases CSF pressure acting on the cochleostomy and allows the soft tissue to stay in place providing safer initial healing of the cochleostomy site.

   An electrode with a “cork” type stopper (FORM 24 which is 24 mm) provides proper sealing of the cochleostomy (to prevent CSF fistula postoperatively) and also makes one full turn around the cochlea. In case of thick ISS with reduced intracochlear space (Fig. 25.5) shorter electrode (such as FORM 19) is preferred.
2. Electrode misplacement into the IAC: Because of the defective modiolus, electrodes with contact surface on both sides or complete rings may provide better stimulation. The probability of the longer electrodes entering the IAC is more than the shorter electrodes. Therefore, an electrode with contact surfaces on both sides or with full rings that will make only one turn around the cochlea appears to be more appropriate.

   The base of the cochlea and modiolus is absent. The IAC is enlarged and opens directly into the cochlea. As noted before, there is no angulation between the cochlea and the IAC, and they are almost in a straight line. This increases the possibility of an electrode misplacement into the IAC. This risk is more if a modiolar hugging electrode is used, because there is no modiolus in these patients [22]. Modiolar hugging electrodes tend to coil towards the center of the cochlea. As this area is defective, they may easily be misplaced into the

IAC, which necessitates removal and repositioning. As it is a curled electrode in the IAC and cerebellopontine angle, during removal there is a high possibility of damaging the cochlear and facial nerves, both of which may have terrible consequences to the patient. A straight electrode has no possibility of creating these complications during removal and should be preferred for surgery in IP-III (see Case 25.2 below).

Electrode choice: Modiolar hugging electrodes have a tendency to go towards the center of the cochlea. As there is no modiolus in IP-III, this may result in misplacement into the IAC. The fact that the cochlea is located directly at the lateral end of the IAC rather than the usual anterolateral position may facilitate displacement of the electrode into the IAC during insertion. Longer electrodes may also be misplaced into the IAC. If a modiolar hugging electrode is used and postoperative X-ray demonstrates that the electrode is inside the IAC, the facial and cochlear nerves may be damaged when the electrode is removed. Thus, straight electrodes that are 25 mm in length and provide one full turn around the cochlea are preferable.

It is interesting that these cases have an interscalar septum. It appears as thicker than normal and can be observed on HRCT. In rare cases, they are so thick that they decrease the intracochlear space. In these cases, electrodes with normal length (>25 mm) can be misplaced into the IAC because of the decreased intracochlear space. Therefore, shorter electrodes, approximately 20 mm in length, should be preferred in these particular cases.

FORM electrodes are ideal for these cases. It is passed through a 1.5 × 1.5 mm fascia. The silicon stopper which tapers smoothly is inserted with fascia and squeezes the fascia in the cochleostomy.

Size of the cochleostomy is very important. Outer diameter of the silicon stopper is 1.9 mm. If the round window is enlarged with 1.2 mm burr to a size of 1.3 or1.4 mm stopper may effectively fix the fascia into the opening without allowing the stopper to pass through. Very small or large cochleostomy may not be effectively closed.

An X-ray should be taken in the surgery to confirm placement of the electrode. If the electrode is discovered to be in the IAC, it appears to be straight and it should be repositioned during surgery. To reposition the electrode, cochleostomy is slightly enlarged anteriorly and the electrode tip is gently bent towards the lateral wall (See Case 25.2 below).

As mentioned before, spontaneous CSF leakage through the oval window is frequently seen and reported in IP-I, even though both IP-I and IP-III are associated with high volume CSF leakage on cochleostomy. This may be secondary to the thicker stapes footplate that is present in IP-III.

As a principle the surgeon should not leave the surgical theater without completely controlling the CSF leakage around the electrode. If there is a severe gusher postoperative continuous lumbar drainage may divert the CSF away from the cochleostomy area allowing the healing of the cochleostomy area. In case of difficult control of CSF gusher, subtotal petrosectomy can be an additional precaution to eliminate the connection between the nasopharynx and the middle ear to prevent meningitis. However, it should never be forgotten that subtotal petrosectomy is an additional measure and it is not the first line of defense in controlling the CSF gusher. Also we would like to emphasize the importance of continuous lumbar drainage. The authors had to revise two patients where they controlled the gusher intraoperatively with FORM electrode, tissue packing, and tissue glue without continuous lumbar drainage. It might be time consuming after a long and problematic surgery but it is crucial for the healing period and highly recommended for every gusher case.

25.9 Audiological Outcomes

Rehabilitation options for patients with IP-III were variable depending on the type and degree of the hearing loss. In a condition of good bone conduction thresholds, bone conduction hearing devices can be selected for the initial follow-up. When the bone conduction thresholds are worse than 40 dB, it is not possible to benefit from bone conduction devices. Hearing aids could be one of the choices for the cases with moderate hearing loss. During follow-up, progressive hearing loss

can be observed. In case of severe to profound hearing loss in IP-III malformation, cochlear implantation should be recommended. Though speech development of the patients with IP-III is improved after cochlear implantation, their speech perception abilities are not as good as the children with normal cochlea. Their speech development abilities are affected by the age at diagnosis, age at implantation, presence of additional handicap, family support, and cognitive abilities like the other hearing impaired children. In our clinical experience with 13 IP-III malformations, we did not observe any side effects such as facial nerve stimulation during follow-up.

In our department between 1997 and September 2018, 2646 patients underwent CI and ABI operations. Two hundred and seventy-nine of CI cases had IEMs. Thirteen had IP-III deformity. During surgery, a facial recess approach was successfully used in all 13 patients. All had severe gusher.

We have performed two revisions: one device failure, one electrode misplacement into IAC. We also noticed one electrode misplacement into IAC on intraoperative X-ray and corrected immediately by extending the cochleostomy anteriorly (Case 25.2 below). All had continuous lumbar drainage in the postoperative period.

None of the patients had rhinorrhea.

Case 25.1: BZ 14-Year-Old Male, Operated May 2013

He had been operated four times before in another center. Initially he had excellent hearing. After final revision his hearing was lost completely and he was referred to our department. His HRCT demonstrated modiolar hugging electrode in the IAC and cerebellopontine angle (Fig. 25.10a–c). Revision was planned. The family was informed that it was planned to cut and leave the active electrode because of risk to facial and cochlear nerves. During revision surgery patient's implant was removed and the electrode was cut at the level of the cochleostomy (Video 25.1). Another cochleostomy was done at the level of the round window. There was a profuse CSF leakage and a new electrode was fully inserted. X-ray demonstrated rotation of the electrode in the cochlea (Fig. 25.10d).

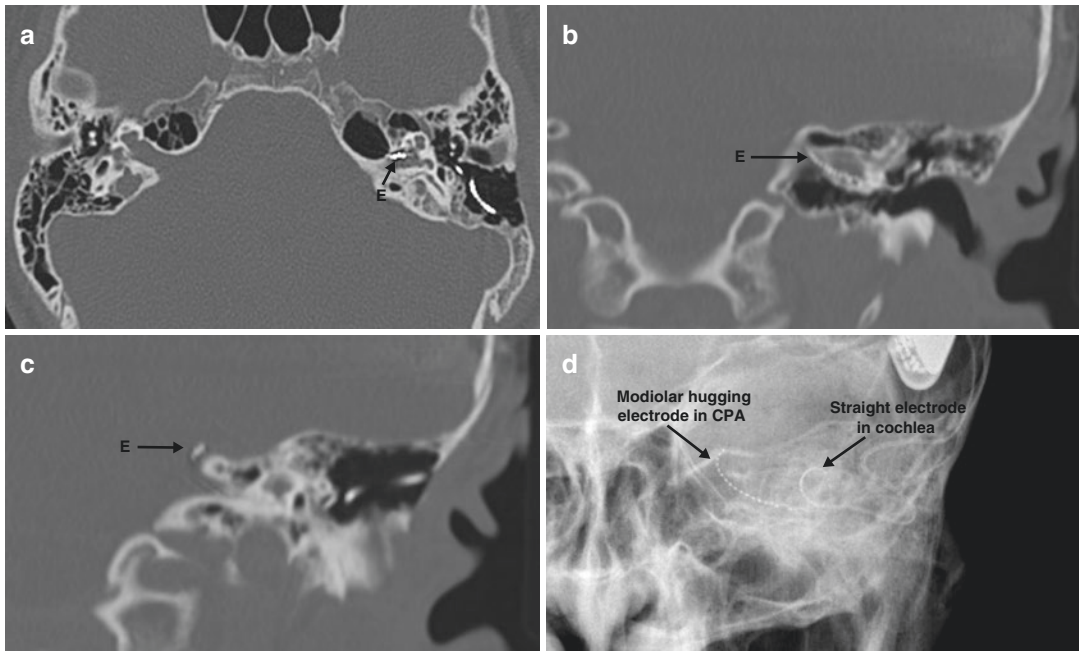


Fig. 25.10 Case 25.1 Preoperative tomography demonstrating modiolar hugging electrode (E) in the IAC and cerebellopontine angle (**a** axial, **b**, **c** coronal views). (**d**) X-ray demonstrating rotation of the new electrode in the cochlea, together with previous modiolar hugging electrode

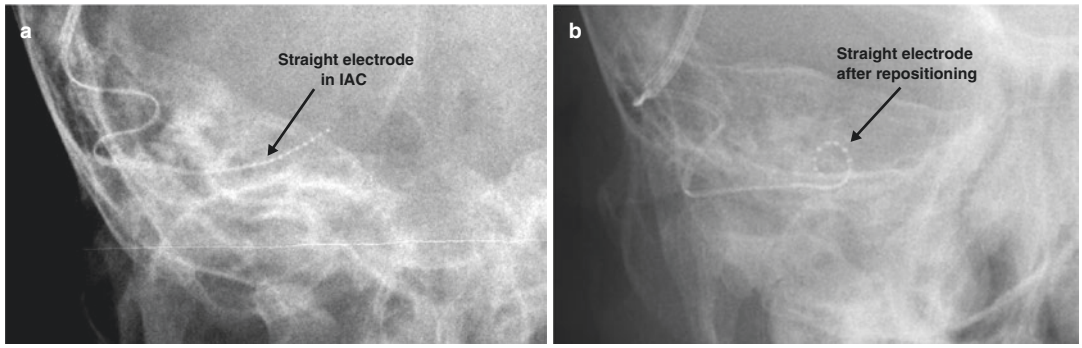


Fig. 25.11 Case 25.2 (**a**) Intraoperative X-ray taken showing a straight electrode: electrode migration into IAC, (**b**) satisfactory electrode placement around cochlea after repositioning

Unfortunately hearing did not recover. Most possible explanation is the cochlear nerve damage during the last revision before coming to our center. Therefore, modiolar hugging electrodes are not advised to be used in IP-III. If used and it is misplaced into IAC, it should not be removed as there is a risk of damage to cochlear and facial nerves.

Case 25.2: MS 2-Year-Old Male Patient Operated March 2015

He had bilateral symmetric IP-III with profound SNHL. His right ear was operated. Facial recess was opened and there was profuse CSF gusher. Form 24 with muscle was inserted into the cochleostomy. Leakage was controlled. Intraoperative X-ray taken at this stage demonstrated a straight electrode: electrode migration into IAC (Fig. 25.11a). Electrode was removed immediately. Cochleostomy was extended slightly anteriorly (Video 25.2). Electrode was bent slightly towards lateral wall (away from the modiolus). Electrode was inserted into cochlea and intraoperative X-ray demonstrated satisfactory electrode placement around cochlea (Fig. 25.11b).

Because of the large defect at the base of the cochlea, electrode migration is possible even with straight electrodes. It is very important to aim the electrode away from the modiolus. Unlike modiolar hugging electrodes, straight electrode can be removed and repositioned without any damage to the nerves and vessels in the IAC. All IP-III patients should have intraoperative X-ray to see the position of the electrode.

References

1. Sennaroglu L. Cochlear implantation in inner ear malformations--a review article. Cochlear Implants Int. 2010;11(1):4–41.
2. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
3. Nance WE, et al. X-linked mixed deafness with congenital fixation of the stapedial footplate and perilymphatic gusher. Birth Defects Orig Artic Ser. 1971;07(4):64–9.
4. Phelps PD, et al. X-linked deafness, stapes gushers and a distinctive defect of the inner ear. Neuroradiology. 1991;33(4):326–30.
5. Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27(5):615–23.
6. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
7. Papadaki E, et al. X-linked deafness with stapes gusher in females. Eur J Radiol. 1998;29(1):71–5.
8. Saylisoy S, et al. Computed tomographic findings of X-linked deafness: a spectrum from child to mother, from young to old, from boy to girl, from mixed to sudden hearing loss. J Comput Assist Tomogr. 2014;38(1):20–4.
9. Sennaroglu G, Sennaroglu L. Hearing loss in inner ear malformations. In: Encyclopedia of otolaryngology, head and neck surgery. Berlin: Springer; 2013. p. 1143–50.
10. Choi BY, et al. Audiological and surgical evidence for the presence of a third window effect for the conductive hearing loss in DFNX2 deafness irrespective

of types of mutations. Eur Arch Otorhinolaryngol. 2013;270(12):3057–62.
11. Jeong S-W, Kim L-S. A new classification of cochleovestibular malformations and implications for predicting speech perception ability after cochlear implantation. Audiol Neurotol. 2015;20(2):90–101.
12. Stankovic KM, et al. Cochlear implantation in children with congenital X-linked deafness due to novel mutations in POU3F4 gene. Ann Otol Rhinol Laryngol. 2010;119(12):815–22.
13. Choi BY, et al. Clinical observations and molecular variables of patients with hearing loss and incomplete partition type III. Laryngoscope. 2016;126(3):E123–8.
14. Kanno A, et al. Frequency and specific characteristics of the incomplete partition type III anomaly in children. Laryngoscope. 2017;127(7):1663–9.
15. Kamogashira T, et al. Prediction of intraoperative CSF Gusher and postoperative facial nerve stimulation in patients with cochleovestibular malformations undergoing cochlear implantation surgery. Otol Neurotol. 2017;38(6):e114–9.
16. Smeds H, et al. X-linked malformation and cochlear implantation. Otol Neurotol. 2017;38(1):38–46.
17. Kumar G, Castillo M, Buchman CA. X-linked stapes gusher: CT findings in one patient. AJNR Am J Neuroradiol. 2003;24(6):1130–2.
18. Talbot JM, Wilson DF. Computed tomographic diagnosis of X-linked congenital mixed deafness, fixation of the stapedial footplate, and perilymphatic gusher. Am J Otol. 1994;15(2):177–82.
19. Sennaroglu L. Special article: incomplete partition type III. In: Naito Y, editor. Pediatric ear diseases diagnostic imaging atlas and case reports. Basel: Karger; 2013. p. 106–8.
20. Snik AF, et al. Air-bone gap in patients with X-linked stapes gusher syndrome. Am J Otol. 1995;16(2):241–6.
21. Batuk MÖ, et al. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233.
22. Sennaroglu L, editor. Special article: incomplete partition type III in pediatric ear diseases diagnostic imaging atlas and case reports. Basel: Karger; 2013. p. 106–8.

Cochlear Hypoplasia

26

Levent Sennaroglu, Betul Cicek Cinar, Burce Ozgen, and Gozde Pamuk

Special Features

1. Four subtypes present.
2. Hearing spectrum ranges from conductive, mixed to sensorineural hearing loss. Most common is profound sensorineural hearing loss.
3. Gusher and meningitis possible in CH-II.
4. Facial nerve abnormality most commonly seen in cochlear hypoplasia. Specific labyrinthine segment abnormality is encountered in CH-IV.
5. Frequent stapes fixation or oval window atresia, making stapedotomy a treatment option.
6. During cochlear implantation thin and short electrode should be used.
7. Auditory brainstem implantation is necessary in cases of cochlear nerve aplasia.

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-83674-0_26].

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

G. Pamuk
Department of Otolaryngology, Hacettepe University, Medical Faculty, Ankara, Turkey

B. C. Cinar (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

B. Ozgen
Department of Radiology, Hacettepe University, Medical Faculty, Ankara, Turkey

26.1 Definition

Cochlear hypoplasia (CH) represents a group of inner ear malformations (IEM) in which dimensions are less than those of a normal cochlea with various internal architecture deformities. In cochlear hypoplasia, there is a clear differentiation between cochlea and vestibule and both of these structures occupy their respective locations relative to the IAC. In this regard, they are later, more developed malformations when compared to rudimentary otocyst and common cavity. In smaller cochlea, it is usually difficult to count the number of turns with computerized tomography (CT) and/or magnetic resonance imaging (MRI). But definition "cochlea with 1.5 turns" should be used for hypoplasia (particularly CH-III), rather than for incomplete partition type II (IP-II) cochlea. According to our own radiological data and literature, four different types of cochlear hypoplasia can be identified [1, 2] (Fig. 26.1):

CH-I: bud type cochlea, round or ovoid in shape, arising from the IAC. Internal architecture is severely deformed; no modiolus or interscalar septa can be identified.

CH-II: cystic cochlea where modiolus demonstrates various levels of defect resulting in cystic cochlea.

CH-III: cochlea with less than two turns but normal internal architecture.

CH-IV: cochlea with normal basal turn, but smaller middle and apical turns.

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_26

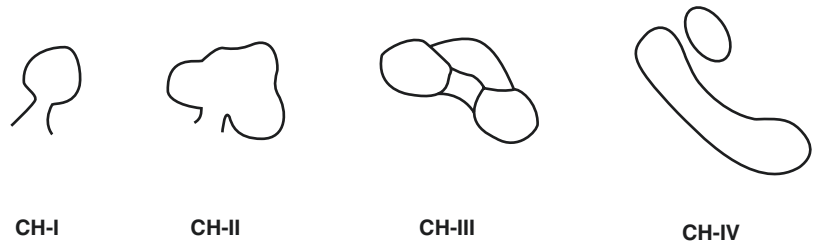


Fig. 26.1 Diagrammatic representation of four types of cochlear hypoplasia (CH). CH-I, CH-II, CH-III are midmodiolar views, while CH-IV is the section passing through the round window in order to give their most characteristic views

26.2 Histopathology and Pathophysiology

When the specimens in Massachusetts Eye and Ear Infirmary and University of Minnesota were examined they showed certain characteristic findings. They were all smaller in size when compared to normal cochlea. CH-I cochlea was round or ovoid in shape with completely absent modiolus or with a thin bony partition (subtotal modiolar defect) between the cochlea and IAC. No neural structure was present in the cochlea. All CH-II cochlea had partial modiolar development. Interscalar septa was absent or hypoplastic, giving the cochlea a shape rounder than normal. The defective area of the modiolus was always in the upper ½ segment. In CH-III modiolus, scala and ISS were intact. The only difference from normal was that the cochlea consisted of approximately one and a half turns. Otic capsule showed normal development with all three layers. Modiolar development was normal, but shorter in height.

It looks possible to explain the pathophysiology with genetics and decreased vascular supply. Genetics determines the length of the cochlear duct. Cochlear duct completes one turn by sixth week and 2.5 turns by eighth week. Further growth is by caliber only where first basal, then middle and apical turns reach adult size by midterm. By 20th week, membranous labyrinth is maximum in size and it is housed within a bony capsule. By 25th week inner ear displays an adult configuration. If development of the cochlear duct stops earlier than eighth week, this results in CH-III. If in addition vascular deficiency from IAC is present it results in CH-I or CH-II. If development stops between 10 and 20 week, resulting deformity is CH-IV (see Chaps. 2 and 3 for more details).

26.3 Literature Review

The studies in literature mostly focus on IEM in general and they did not specify subtypes. Besides, most of the studies focus on CI or ABI outcomes. Both audiological thresholds and CI-ABI outcomes show variability among individuals with different anomalies. In a recent retrospective study by Melo et al. [3], it was reported that out of 329 patients, 26 patients had IEM and in their study CI outcomes of these patients were reported. Preoperatively all their patients had profound SNHL without any improvement with hearing aids before CI. Distribution of IEMs was 5 IP-II, 8 CH-III, 3 EVA, 3 EVA with partial SCC aplasia, 5 partial SSC aplasia, and 1 cochlear nerve hypoplasia. They used CAP and SIR score to measure CI outcomes. Outcomes of IEM group were lower than control group but the difference was not statistically different. They stated that similar results with control groups could be related to absence of severe malformations in the study group. Even though they divided groups according to IEMs, they did not report CI outcomes separately according to IEM groups so that there was no information related to CH groups.

Feng et al. [4] reported one cochlear hypoplasia case with cochlear implant. They described cochlear hypoplasia as "severe" with rudimentary cochlear bud. This child was diagnosed with bilateral profound hearing loss and recommended hearing aids at first. And after 1 year follow-up, cochlear implantation was applied. Two years

after CI, free field thresholds were in between 30 and 55 dB for 125–4000 Hz. CAP and SIR scores were 5. They concluded that cases with severe cochlear hypoplasia could benefit from CI.

Puram et al. [5] reported a case of bilateral CH, 16 months of age who received an ABI, together with his follow-up results with ABI. This child had bilateral CH-II malformation and severely hypoplastic cochlear nerves. Preoperative audiological testing showed that there was no response at the limits of audiometers. They first tried to cochlear implant but CI was aborted due to abnormal inner ear anatomy and then they decided to ABI surgery. After ABI this child showed free field thresholds at 30–50 dB in between 250 and 4000 Hz and also developed some awareness to parental voice and environmental noise.

Unlike previous studies, Cinar et al. [6] reported audiologic and radiologic findings of cochlear hypoplasia. Their results focus on four different types of cochlear hypoplasia and gave comprehensive audiological results according to the subgroups of CH. It was reported that four types of cochlear hypoplasia showed differences in both type and degree of hearing loss. In CH-I, CH-II, and CH-III groups, degree of hearing loss varied from moderate to profound and also some cases had no response to sound on subjective testing and most of them had SNHL. Although SNHL can be seen in CH-IV group as well, some cases presented with mild and pure conductive hearing loss and also all cases in this group had responses to sound at subjective testing. Mixed type hearing loss was more common in CH-III. However, CI or ABI outcomes were not reported in this study.

26.4 Clinical Findings

Hearing loss in CH is usually nonprogressive but there may be progressive HL over time. Some cases of CH-III and CH-IV made benefit from hearing aids initially but later they were implanted with CI. It is possible to have a spectrum of different types and thresholds of hearing loss in CH: mild, moderate to profound conductive, mixed or SNHL. They may rarely have pure CHL. This makes it possible to use variety of methods in the habilitation of hearing loss.

Although much rarer than IP-I, cerebrospinal fluid (CSF) fistula and recurrent meningitis are possible and seen mainly in CH-II.

26.5 Radiology

According to the IEMs database of Department of Otolaryngology at Hacettepe University, 776 patients with 1552 ears were evaluated until September 2018. Three-hundred and ninety of the 1552 ears had CH (25%); 84 had CH-I (22%), 86 had CH-II (22%), 174 had CH-III (45%), and 46 had CH-IV (11%).

(a) **CH-I (Bud-like cochlea)**: The cochlea is like a small bud, round or ovoid in shape, located in the anterolateral part of IAC (Fig. 26.2a). Internal architecture is severely deformed; no modiolus or interscalar septa can be identified.

(b) **CH-II (Cystic hypoplastic cochlea):** The cochlea is smaller in its dimensions with defective modiolus and interscalar septa, but its external architecture is similar to normal cochlea (Fig. 26.2b). Fundus of IAC is usually defective providing a connection between cochlea and IAC. The vestibular aqueduct may be enlarged and the vestibule may be dilated. In this type of hypoplasia, recurrent meningitis, gusher and unintentional entry of the CI electrode into IAC is possible.

(c) **CH-III (Cochlea with less than two turns):** The cochlea has a short modiolus and the overall length of the interscalar septa is reduced, resulting in fewer turns (i.e. less than two turns). The internal (modiolus, interscalar septa) and external architecture are similar to that of a normal cochlea, but the dimensions are smaller and number of turns are fewer (Fig. 26.2c). There is a type of hypoplastic cochlea consisting of basal turn only with normally developed modiolus (Fig. 26.2d). This is also included in CH-III because modiolus and internal architecture of the cochlea are normally developed except the length of the basal turn. Vestibule is usually hypoplastic and semicircular canals may be hypoplastic or aplastic (Fig. 26.2e).

(d) **CH-IV (Cochlea with hypoplastic middle and apical turns)**: The cochlea has a basal turn which is nearly normal in size and appearance; however, the middle and apical turns are severely hypoplastic and located anterior and medially rather than in their normal central position (Fig. 26.2f–i). The labyrinthine segment of the facial nerve is usually

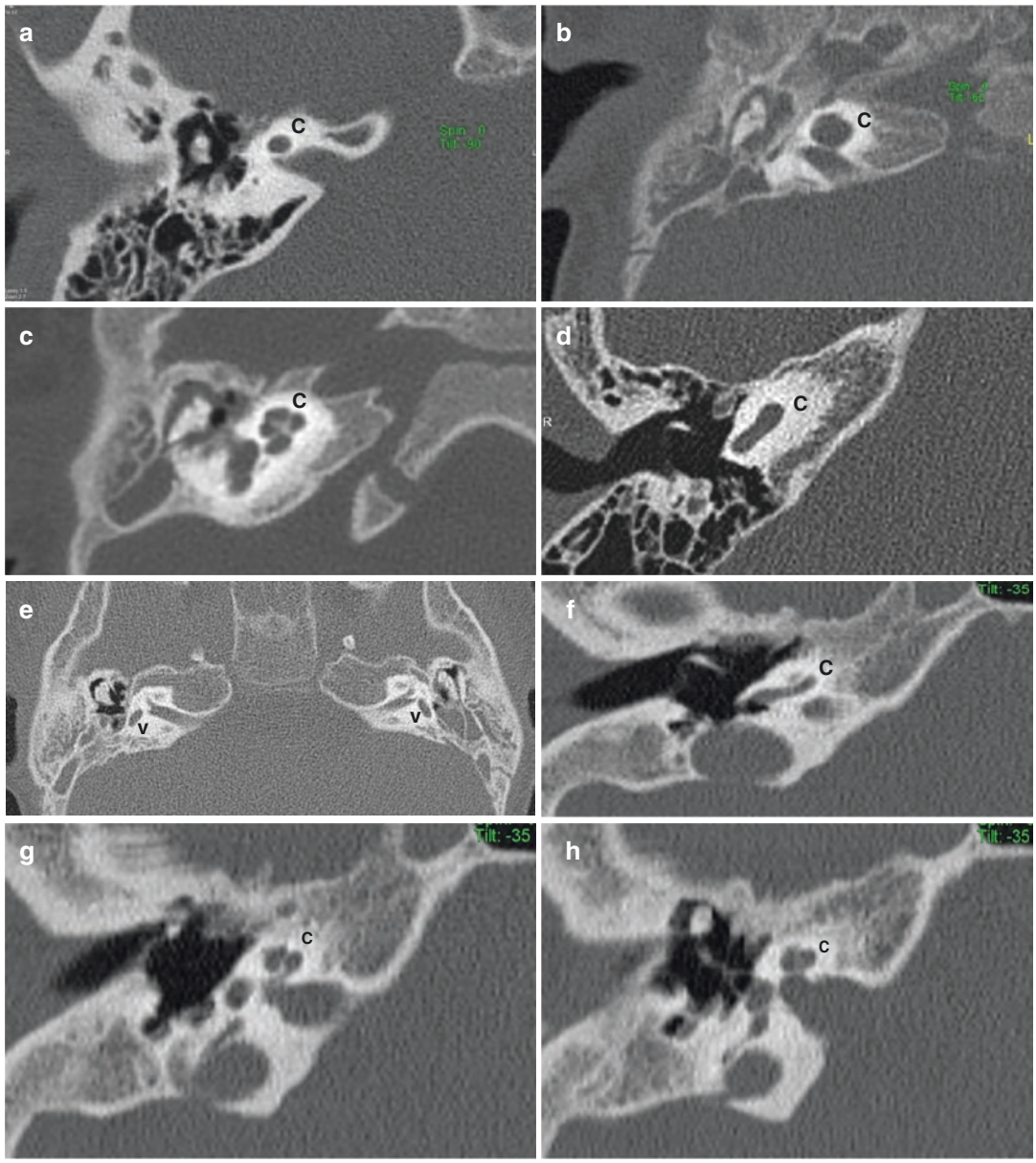


Fig. 26.2 Types of cochlear hypoplasia (CH): (**a**) CH-I: bud type cochlea, round or ovoid in shape, without modiolus and interscalar septa, (**b**) CH-II: cystic cochlea with modiolus deformity resulting in cystic cochlea, (**c**) CH-III: cochlea with less than two turns but normal internal architecture, (**d**) CH-III consisting of basal turn only with normally developed modiolus, (**e**) hypoplastic vestibule and aplastic semicircular canals accompanying a CH-III cochlea. (**f–i**) CH-IV: cochlea with normal basal turn, but smaller middle and apical turns. (**j–l**) Labyrinthine segment of the facial nerve is located anterosuperior to the cochlea rather than in its usual posterolateral location which is pathognomonic sign for CH-IV (arrows pointing labyrinthine and tympanic segments of the facial nerve)

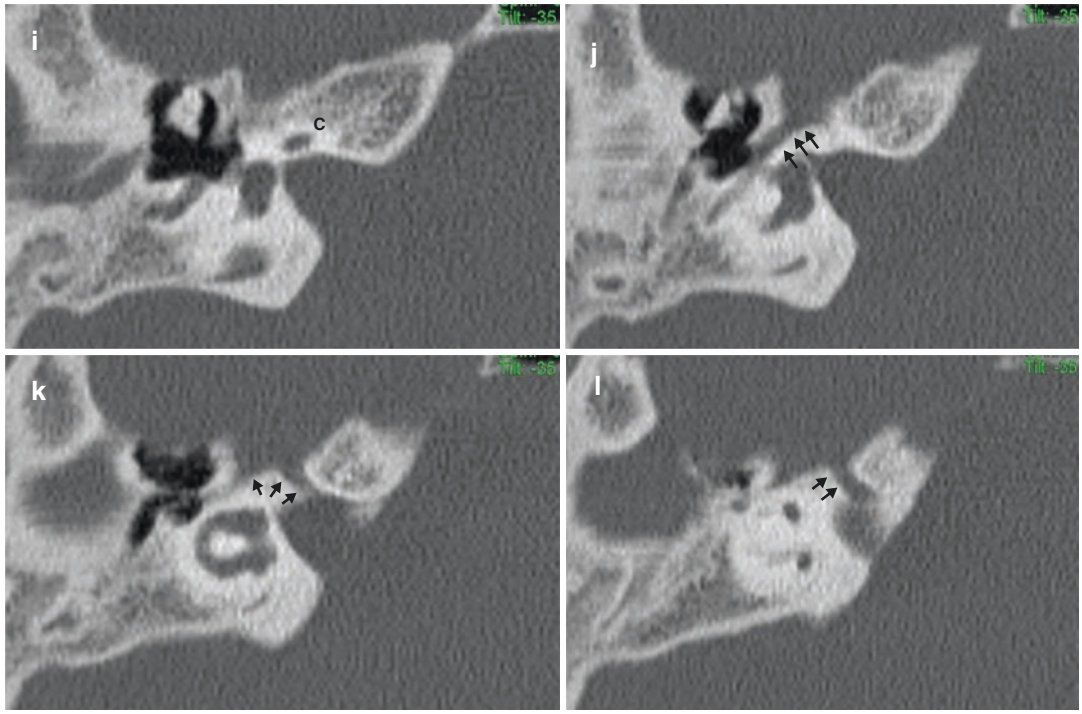


Fig. 26.2 (continued)

located anterosuperior to the cochlea rather than in its usual posterolateral location [7] (Fig. 26.2j–l). This is pathognomonic sign for CH-IV. Vestibule and semicircular canals are usually normal.

The vestibule and semicircular canals demonstrate various abnormalities. Vestibule is frequently hypoplastic, semicircular canals may be hypoplastic or aplastic (Fig. 26.2e).

In our department Pamuk [8] has performed a radiological study about dimensions of CH. In her thesis, it was aimed to obtain objective values for CH by measuring and comparing dimensions of healthy and hypoplastic cochleas. Using temporal CT, basal turn length, basal turn height, and midmodiolar cochlear height were measured axial section through round window using the 3D MPR (multiplanar reconstruction), the cochlear canal mid-scalar and lateral wall length marked from the round window to the helicotrema were measured. There was a significant difference in basal turn length, basal turn height, midmodiolar height, cochlear canal mid-scalar, and lateral wall length between control groups and CH I, II, and III ($p < 0.001$). The basal turn height and the cochlear canal lateral wall length in CH IV were significantly lower when compared with the control group ($p < 0.001$ and $p = 0.002$, respectively). Cochlear hypoplasia should be suspected if length of the basal turn is <7.5 mm in axial section through round window (Fig. 26.3a), and height of cochlea is <3.5 mm on midmodiolar section (Fig. 26.3b). These two measurements can easily be done by clinicians. Once hypoplastic cochlea is diagnosed, sub-typing should be performed, and appropriate length of the cochlear implant electrode should be selected. While there is not enough benefit from selecting a short electrode according to the size of the cochlea, long electrodes may also increase apical damage.

The cochlear aperture may be normal, hypoplastic, or aplastic (Fig. 26.4a, b). Sometimes cochlear aperture is extremely narrow which makes CI surgery a difficult decision (Fig. 26.4c). It is very important to demonstrate CN in case of profound

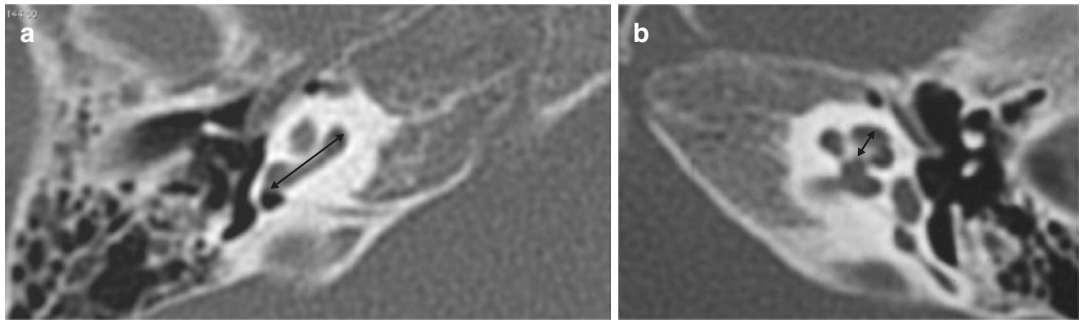


Fig. 26.3 Measurements in order to differentiate cochlear hypoplasia from normal cases and incomplete partition anomalies. (**a**) basal turn length: if length of the basal turn is <7.5 mm in axial section through round window CH is diagnosed, (**b**) height of cochlea: if the height of cochlea is <3.5 mm on midmodiolar section, CH can be diagnosed

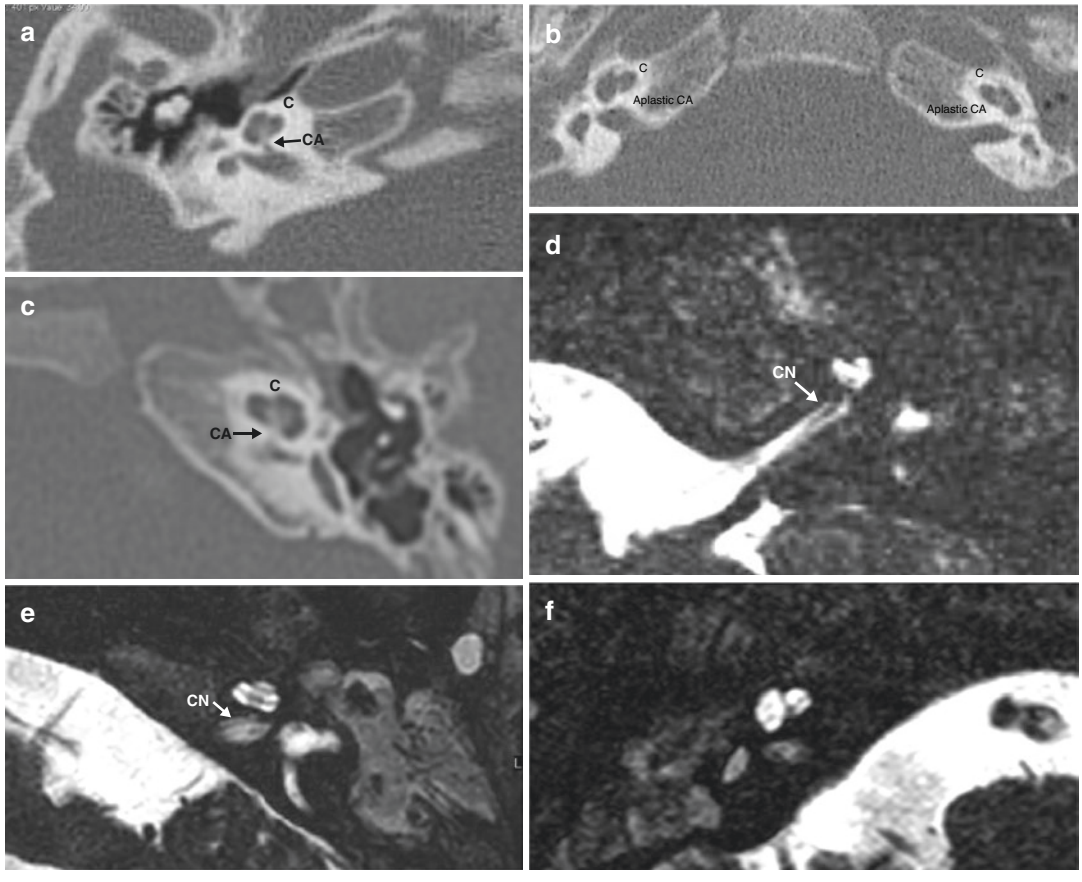


Fig. 26.4 Cochlear aperture (CA) and cochlear nerve (CN) abnormalities in CH: (**a**) Hypoplastic CA, (**b**) Aplastic CA, (**c**) extremely narrow CA, (**d**) Normal CN, (**e**) Hypoplastic CN, (**f**) Aplastic CN (aperture is completely aplastic, no CN is seen in the usual location). Direct parasagittal oblique cuts perpendicular to IAC may demonstrate hypoplastic (**g**) or aplastic (**h**) CN

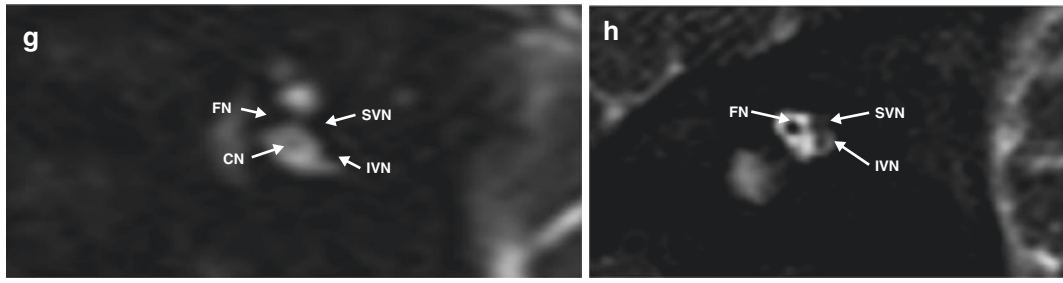


Fig. 26.4 (continued)

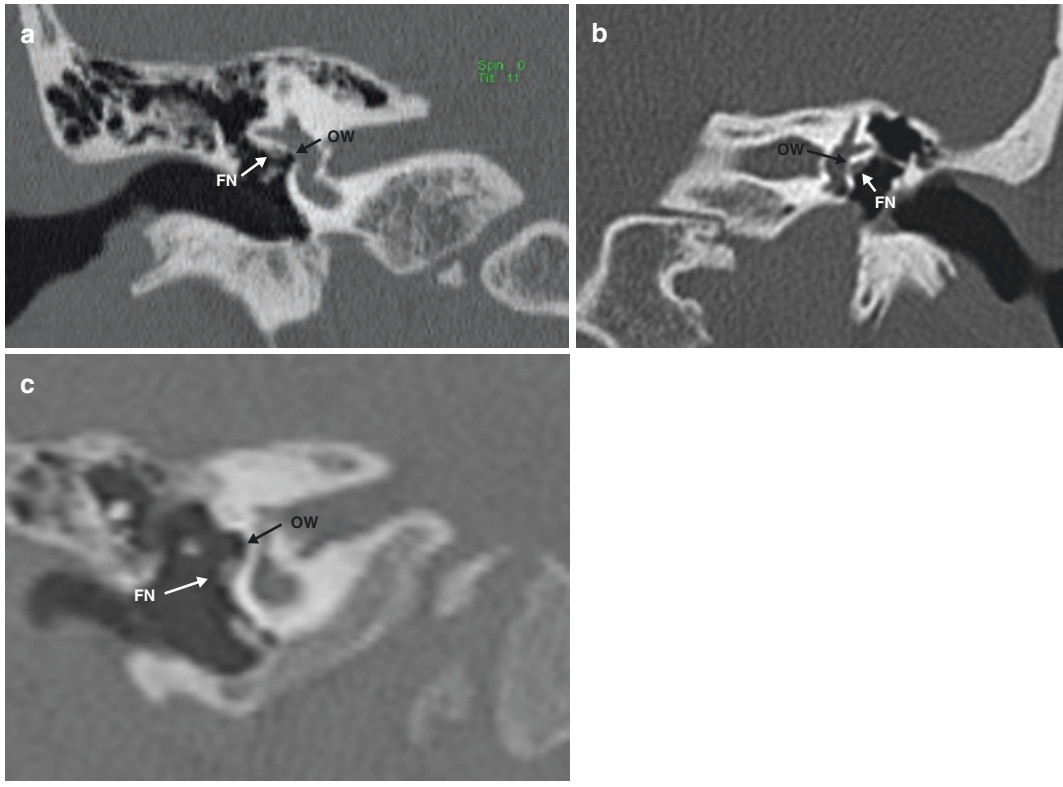


Fig. 26.5 Facial nerve (FN) abnormalities in cochlear hypoplasia on coronal sections in relation to oval window (OW). (**a**) Normal location of FN inferior to lateral semicircular canal and lateral to the OW, (**b**) FN located at the OW, (**c**) FN located inferior to the OW

SNHL. Cochlear nerve (CN) may be normal, hypoplastic, or aplastic (Fig. 26.4d–f). Direct parasagittal oblique cuts perpendicular to IAC may demonstrate hypoplastic or aplastic CN (Fig. 26.4g, h).

Cochlear hypoplasia is the group of IEMs where FN anomaly is most frequently encountered. This is best visualized on the coronal sections of temporal bone HRCT. Normally FN is superior lateral to the oval window (Fig. 26.5a). It is possible to have the FN at the oval window (Fig. 26.5b) or inferior to the oval window (Fig. 26.5c). If the FN is at the oval window, this makes middle ear exploration for congenital conductive hearing loss extremely difficult. If it is

inferiorly located it may lie over the promontory or round window making cochleostomy difficult. This makes the CI surgery extremely challenging (see cases below).

26.6 Audiological Findings

There is no characteristic audiological configuration for CH. These patients may present with different audiological configurations. Decision-making about the amplification options may be difficult, particularly in patients with a hypoplastic cochlear nerve. Patients with mild to moderate hearing loss can be habilitated with hearing aids and usually they have near-normal language development (Fig. 26.6a). Majority of cochlear hypoplasia patients have severe to profound hearing loss where a CI would be a reasonable option if they have a cochlear nerve (Fig. 26.6b). Some cases have cochlear nerve hypoplasia where the size of CN is less than ipsilateral FN. CI is again the best option as long as the hypoplastic nerve can be followed until cochlea.

Some patients have cochlear aperture aplasia with cochlear nerve aplasia and thus, an ABI would be the best hearing habilitative option. Other patients with cochlear hypoplasia have extremely hypoplastic cochlear nerves. CN is very thin and can be traced with difficulty until cochlea. The best option in these cases is to use CI in the better-developed side, better audiological response or the side that is more suitable to CI surgery (Fig. 26.6c). If there is limited hearing and language development, an ABI should be considered for the contralateral side.

Some cases of hypoplasia (CH-III and particularly CH-IV) may have pure conductive or mixed hearing loss in which the conductive component is due to stapedial fixation (Fig. 26.6d). They may benefit considerably from stapedotomy in case of conductive or mixed type hearing loss. A case of severe mixed hearing loss whose HRCT consisted of only basal turn made much better use of hearing aids in the postoperative period and her speech improved considerably after bilateral stapedotomy (Fig. 26.6e).

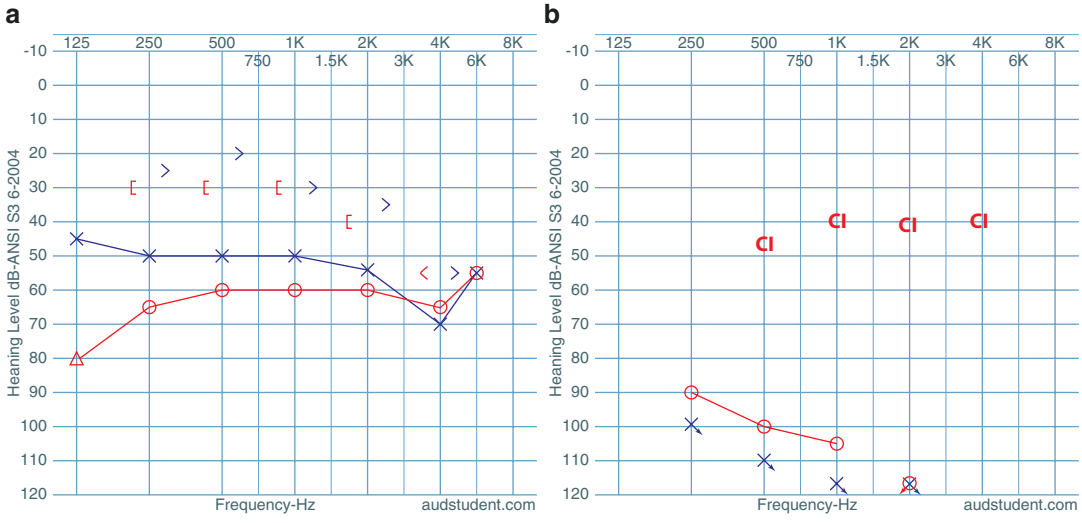


Fig. 26.6 Audiological configurations in cochlear hypoplasia: (**a**) moderate to moderately severe mixed HL, (**b**) profound sensorineural HL, (**c**) bilateral profound HL with response with insert earphones on the left side, (**d**) bilateral mild conductive HL, (**e**) bilateral severe mixed HL, (**f**, **g**) thresholds obtained with ABI in two different patients with CH

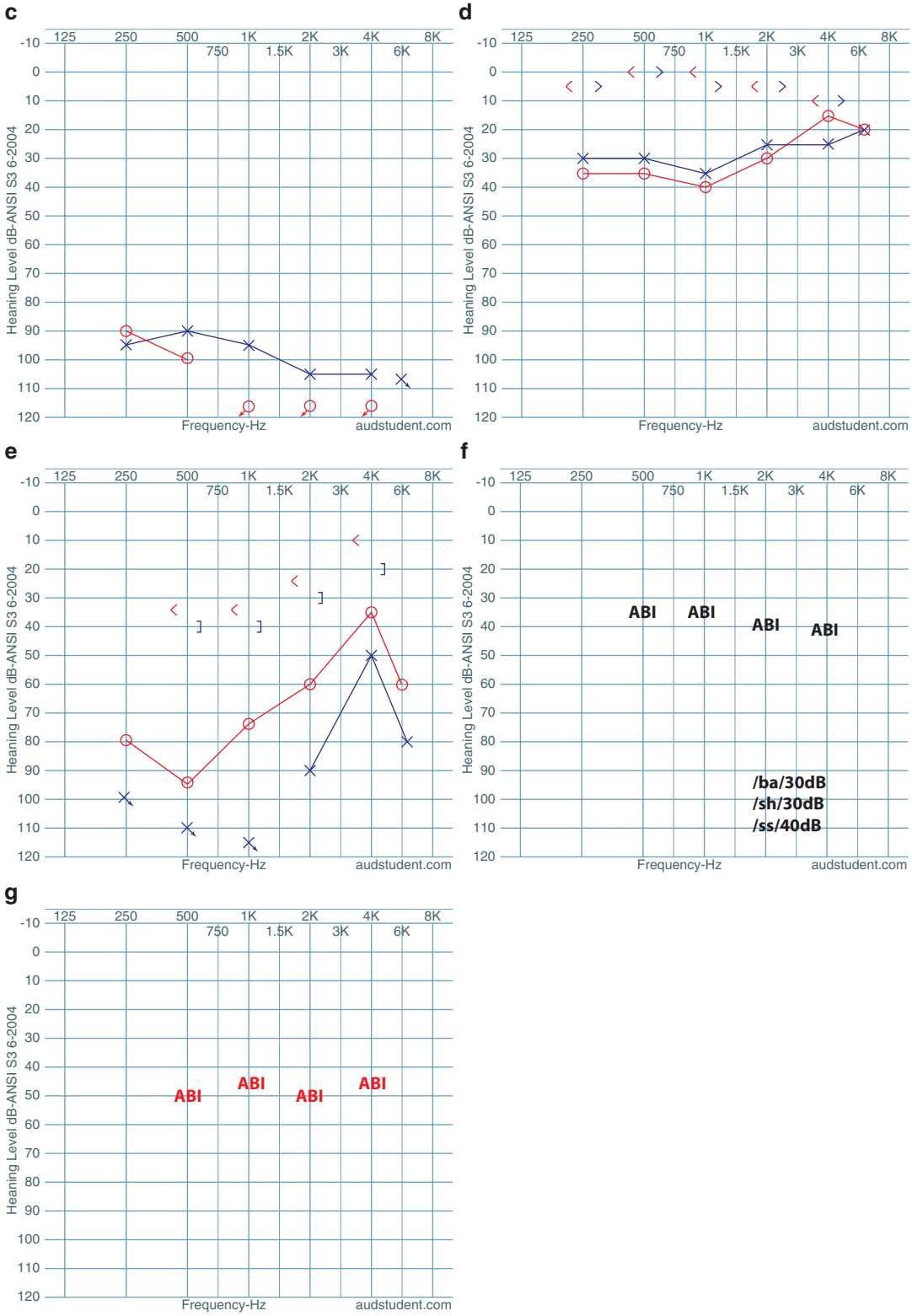


Fig. 26.6 (continued)

26.7 Management Options

1. Mild pure conductive HL: This is usually seen in CH-IV. They may have mild to moderate pure CHL. **Stapedotomy** may result in near-normal hearing.
2. Cases with moderate to severe mixed type of hearing loss: These patients may benefit from **stapedotomy and hearing aid use**. This can be done in childhood if the family is motivated and can result in enhanced oral language development with or without hearing aid use depending on air and bone conduction thresholds after surgery.
3. **Cochlear implantation:** CH cases with profound hearing loss who have a cochlear nerve demonstrated on MRI are candidates for CI. In cases of CN deficiency if there is a threshold with insert earphones, CI is preferred even in cases with extremely hypoplastic nerves. This is the most challenging group of IEMs for CI surgery (see below: Difficulties during CI surgery in CH).
4. **ABI:** ABI is indicated for patients with aplasia of cochlear aperture and CN. As indicated before, if the child does not demonstrate significant improvement with CI, ABI should be considered as soon as possible on the contralateral side. If there is hardly visible CN on MRI with no response with insert earphones during behavioral testing, ABI may be given priority over CI. It is possible to obtain thresholds around 30–40 dB with ABI (Fig. 26.6f, g).
5. **Simultaneous CI and ABI:** If there is CH with CN on one side and a definite indication for ABI on the contralateral side, CI and ABI can be done together.

Between November 2007 and September 2018, 2643 patients had CI and ABI in our department. Four-hundred and six of these cases were with various IEMs. Eighty-eight had cochlear hypoplasia: 52 received CI, while 36 were implanted with an ABI.

Three patients had revision CI surgery for device failure.

26.8 Outcome After Stapedotomy

Cases with CH-IV may have conductive hearing loss due to ossicular chain pathology. In these cases stapedotomy is the first choice for hearing restoration. Conversely, if they refuse operation, hearing aid fitting should be done especially in pediatric cases. After stapedotomy postoperative thresholds in a patient with mild conductive hearing loss may come within normal limits where hearing aids may not be necessary any more (See Chap. 14 for further details).

26.9 Outcome with CIs and ABIs

Patients with CH show variations in auditory performances with CI because of anatomical challenges, additional handicaps, and placement of intracochlear electrode.

CI users with CH have some programming problems due to anatomical difficulties. It is common to have inactive electrodes with CI. Although inactive electrodes can be seen in apex as well, most common location is the basal part of the intracochlear electrode. Facial stimulation, pain in the ipsilateral ear, dizziness due to electrical stimulation, poor sound quality, and narrow dynamic range are most common causes of inactive electrodes. For facial stimulation modification of duration/pulse width may be effective to keep electrodes active. Especially electrodes in basal part of the intracochlear electrodes could have narrow dynamic ranges. These electrodes could be active or inactive depending on free field thresholds at higher frequencies.

Performance with CI mostly depends on status of CN. If CN is normal, outcomes with CI are usually better. It can be inferred that CI outcomes of CH-IV are better than other CH subtypes since they mostly have normal CN. However, if CN is hypoplastic they make limited benefit from CI resulting in very poor language development. Even if their pure tone thresholds are between 25 and 35 dB ranges, their speech understanding and production do not progress further. This is indication for contralateral ABI. CI and ABI users can

differentiate between CI and ABI benefits separately. One specific patient defined her situation with the phrase "*With CI I can hear, but with ABI, I can understand the speech*."

Early diagnosis, early implantation, absence of additional disabilities, and good family support are predictive of better ABI performance. ABI user with CH shows good performance, but their progress is slower when compared to a CI user. With regular follow-up and educational support, they make improvement with time (See Chaps. 30, 31, and 32 for further details).

26.10 Surgical Approach

26.10.1 Difficulties During CI Surgery in Cochlear Hypoplasia

Difficulties can be divided into three groups:

1. **FN anomaly:** A transmastoid facial recess approach can be used in the majority of these patients. During surgery, facial nerve malposition is to be expected because of labyrinthine abnormalities. We have noticed that if the lateral semicircular canal is not developed properly, the facial nerve may be in an abnormal location. In one patient, for example, we found that the facial nerve was already dehiscent and lying in the area of the facial recess.

   FN may be located inferior to the oval window, lying on the promontory. This was present in ten cases. In these cases cochleostomy is usually located behind the FN (Case 26.5). In one case cochleostomy was done anterior to the FN as the promontory was prominent. In two patients hypoplastic vestibule was found and the electrode was inserted into the scala vestibuli through the vestibule (Case 26.8).
2. **Difficulty in reaching the hypoplastic cochlea** If the cochlea is small, the promontory may not have the usual protuberance and it may be difficult to visualize the promontory and round window through the facial recess. In these situations, modifications in the surgical approach may be necessary to provide better access to the hypoplastic cochlea.

   (a) **Facial Recess Approach**: This is the area between FN and chorda tympani and is sufficient in majority of the cases.

   (b) **Combined Approach (Facial Recess and Transcanal)**: This approach adds the advantage of observing the footplate area better than in standard FR approach. In CH-II cases if there is a footplate fistula this can be managed better with this approach during CI surgery. If there is CSF leakage through the stapes footplate during CI surgery, this is the best approach to repair the leakage at the oval window and enable electrode insertion through the round window.

   (c) **Anterior mobilization of ear canal:** This provides slightly more visibility of the medial wall of the middle ear when compared to standard facial recess approach. It can be used in mild CH cases. Two cuts are done in the bony ear canal (roughly at 8 and 12 o'clock positions for the right ear). Once the cuts are complete ear canal is mobilized anteriorly (without detaching the bone from the skin).

   (d) **Temporary removal of ear canal:** Ear canal can be removed keeping the skin intact. In this way it is possible to have better view of the oval and round window area. At the end of the surgery, ear canal can be reconstructed without any difficulty. It is advisable to thin the ear canal with appropriate burrs but make the actual cut with a thin diamond burr (such as 0.8 mm) so that bony part of the ear canal is preserved. If we use a larger burr, the bone removed will be more and bone of the ear canal will be smaller. With larger bone the resulting ear canal reconstruction will be better. This provides the opportunity to inspect the ear in the postoperative period (Case 26.6).

   (e) **Split ear canal** [9]: If the facial nerve prevents the view of the round window area and hence electrode insertion, one alternative technique is to use transcanal-

transmastoid approach. We produce a full length cut in the ear canal. The electrode is inserted into the cochleostomy via transcanal approach and then it is transferred into the mastoid via split ear canal. In this way electrode cable is not in the ear canal.

(f) **Blind sac closure of the ear canal:** If the cochlea cannot be found with above mentioned methods, skin and posterior wall of the ear canal are removed and a direct approach to the medial wall of the middle ear is provided (Case 26.3).

3. **Gusher**
   This is mostly seen in CH-II. Gusher is managed in the usual way mentioned in Chap. 15. Size of the cochlea is much smaller in hypoplasia when compared to incomplete partitions. Therefore, if we use FORM 24 electrode with stopper in case of gusher in CH-II, electrode may not be inserted fully into the cochlea and stopper will not be at the cochleostomy. We developed FORM 19 electrodes with cork type stopper for these cases. In CH-II it will make one full turn around the cochlea and stopper will be at the cochleostomy site.

Electrode choice: As the cochlea is smaller than normal, the length and cross sectional area of the cochlear duct are smaller when compared to that of a normal cochlea. Thinner and shorter electrodes should therefore be used. A standard electrode may be too long for the cochlea and it may not be possible to obtain a full insertion. In this case the stopper will not be at the cochleostomy and leakage will not be controlled efficiently. This happened in one case who also had gusher and the author had difficulty in controlling the gusher as the stopper could not be inserted until the stopper. It is advisable to use electrodes less than 20 mm in length.

CH-II has the possibility of CSF leakage. A short electrode with a stopper type silicon ring may be used along with other measures for managing a CSF gusher. Because of the possibility that a full insertion may not be obtained, we have developed a shorter version of the electrode (19 mm) with a cork type silicon stopper hypoplastic cochlea (particularly type II). FORM 19 is ideal for these patients.

It is very important to notice cochlear hypoplasia preoperatively and choose the appropriate length of electrode. During CI surgery it is important to use thin (0.8 mm) and short electrodes (<20 mm). A larger and longer electrode may not be inserted into a short and narrow scala, particularly in the middle and apical turns. In the case of a gusher, it is the aim of the surgeon to insert the electrode up to the silicon stopper immediately after the active part. If long electrodes are chosen, they may not be inserted fully into the cochlea: this may result in insufficient control of the CSF leakage, because the stopper will not be at the level of the cochleostomy. Excessive manipulation of the electrode may also break the thin layer of bone between the IAC and the hypoplastic cochlea, and the electrode may inadvertently be inserted into the IAC.

It should be noted that the anatomy is particularly distorted in CH-I and CH-II. Even though CI can be successfully placed into a hypoplastic cochlea, limited language development is expected. During the initial consent, the family should be informed that ABI surgery will most likely be required on the contralateral side in the future.

26.11 Cases

Case 26.1: DK 22-Year-Old Female Patient Operated July 2013

She had bilateral hearing loss since birth. Her audiogram revealed severe mixed HL on the right side and moderate conductive HL on the left side (Fig. 26.7a). Her HRCT demonstrated bilateral CH-IV malformation with anterior and superior dislocation of the labyrinthine segment of FN (Fig. 26.2f–l). She underwent right stapedotomy with closure of air-bone gap (Fig. 26.7b).

She is the daughter of Case 26.3. Interesting aspect is that both mother and daughter have exactly the same IEM type CH-IV, but mother had bilateral profound SNHL necessitating a CI surgery, while her daughter had bilateral conductive hearing loss which was managed with stapedotomy.

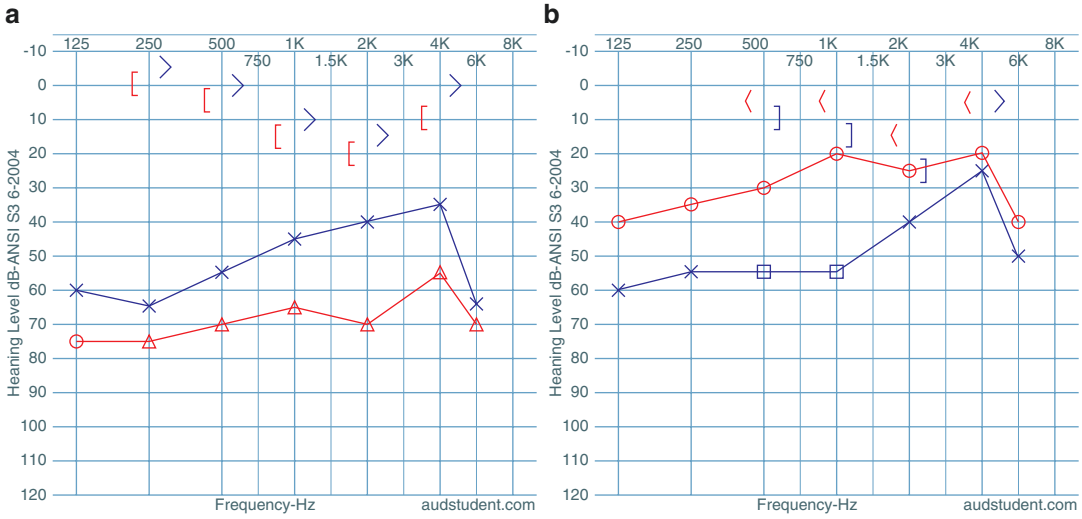


Fig. 26.7 Case 26.1. (**a**) Preoperative severe mixed HL on the right side and moderate conductive HL on the left side, (**b**) postoperative thresholds after right stapedotomy with closure of air-bone gap

Case 26.2: HS 11 Year-Old Female Patient Operated in January 2013

The reader is advised to examine Case 14.2 in Chap. 14. She had bilateral severe mixed type hearing loss (Fig. 26.6e). Her air and bone conduction threshold average were 110 and 41 dB on the left and 76 and 26 dB on the right ear. The family was informed that the operation was planned to make her benefit more from hearing aid with the aim of decreasing the air-bone gap. She had bilateral stapedotomy; left side Jan 2013 and right side on April 2014. On both sides, all of her ossicles were found to be fixed bilaterally. In both ears malleus and incus were mobilized after atticotomy and stapedotomy was performed. Because of the decrease in ABG postoperatively, she made better use of her hearing aids and her speech showed tremendous improvement.

Case 14.1 (İH) in Chap. 14 had bilateral CH-III with mild conductive hearing loss (Fig. 26.6d). He underwent bilateral stapedotomy. FN was located inferior to the oval window which complicated the surgery in addition to CSF gusher. After both surgeries he was able to hear bilaterally at 25–30 dB without hearing aids and family did not want to use hearing aids.

Case 26.3: NK 38-Year-Old Female Patient Operated in July 2010

She had bilateral profound SNHL. HRCT revealed bilateral symmetric CH-IV (Fig. 26.8a). Labyrinthine segment was superiorly dislocated on both sides. During CI surgery it was discovered that the recess was very narrow. FN was identified, posterior wall of the ear canal was thinned considerably but it was not possible to visualize promontory or the round window. After reexamining the HRCT scans at this stage, it was realized that it was almost impossible to expose the round window through facial recess on both sides (Fig. 26.8a). It was decided to perform subtotal petrosectomy to increase surgical exposure of the round window. After subtotal petrosectomy RW could be easily identified and full insertion of CI was possible. Postoperative HRCT demonstrated angle of insertion into cochlea which would be impossible to obtain without subtotal petrosectomy (Fig. 26.8b).

This patient demonstrates that labyrinthine segment anomaly may also affect the position of tympanic and mastoid segments decreasing the visibility of the round window through the facial recess approach. In addition CH-IV anomaly may present with conductive or sensorineural hearing loss.

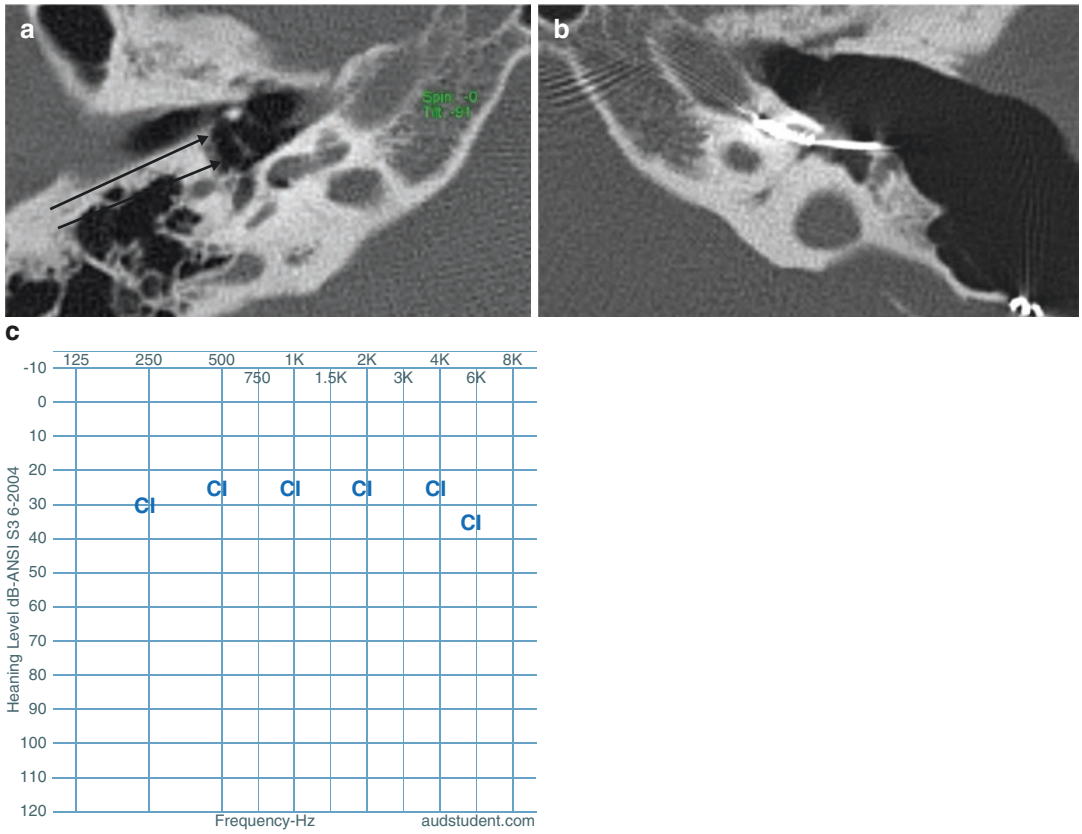


Fig. 26.8 Case 26.3. Facial nerve abnormality preventing the use of facial recess approach: (**a**) Right side axial CT showing that round window cannot be seen through the facial recess. (**b**) Postoperative HRCT showing electrode insertion into the basal turn. Please note the angle of insertion into cochlea which would be impossible to obtain without subtotal petrosectomy. (**c**) Postoperative thresholds with CI

With CI on the left ear, she had good performance (Fig. 26.8c).

Case 26.4: BAK, 2-Year-Old Male Patient, Operated in July 2015

This patient was already reported as Case 23.7 in Chap. 23. He had IP-I on the right side and CH-II on the left side (Fig. 22.2a–f). Right side IP-I was operated in July 2015 with FORM 24. There was severe gusher which was controlled effectively with cork type stopper. Left side CH-II was operated in March 2017 with FORM 19. Postoperative transorbital X-ray showed the two sides with at least one turn of the electrode (Fig. 22.2g).

The two anomalies are coincidental often. It is very important to choose the correct length of electrode with stopper. For IP-I, FORM 24, for CH-II FORM 19 are ideal electrodes. If the FORM 19 is chosen for IP-I, it will not make one full turn around the cochlea, resulting in inefficient stimulation. If we choose FORM 24 for CH-II we may not be able to insert the electrode until stopper; therefore, it will not be effective in controlling CSF gusher.

Case 26.5: MII 2-Year-Old Male Patient, Operated September 2017

He had bilateral CH-II and FN lying on the promontory on both sides (Fig. 26.9a). During CI surgery FN was lying vertically on the promontory. FR was extended posteriorly towards the usual location of FN and visibility is increased. Pyramidal eminence and stapedial tendon were sacrificed to have full vision of the area. Cochleostomy was done immediately posterior to the FN. There was a gusher. FORM

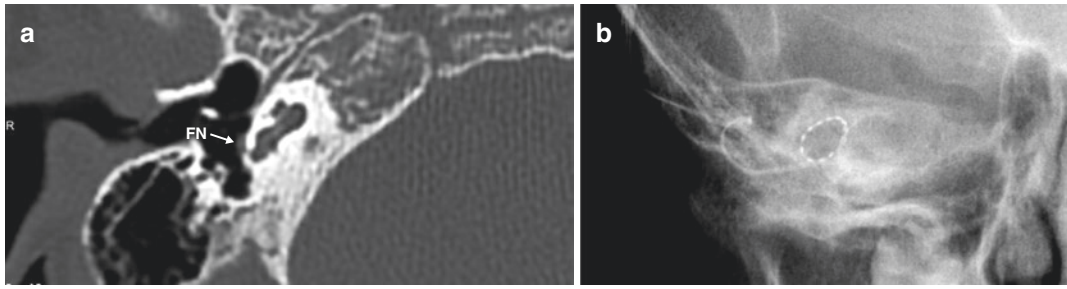


Fig. 26.9 Case 26.5. (**a**) Axial section showing CH-II and facial nerve (FN) lying on the promontory on the right side. (**b**) Postoperative implant position

19 was fully inserted into the cochlea (Fig. 26.9b) (Video 26.1).

Case 26.6: BSA 2-Year-Old Female Operated March 2015

She had bilateral profound SNHL. HRCT demonstrated bilateral CH-II (Fig. 26.2b). It was very difficult to identify cochlea through facial recess. Facial nerve was identified; ear canal was thinned. However, promontory could not be recognized. Two radial cuts were done in the ear canal with a very thin diamond burr and the ear canal was removed. Excellent exposure of the middle ear was obtained. A cochleostomy was performed on the promontory and there was a severe CSF gusher. Only partial insertion of the FORM 24 was obtained. Postop view demonstrates one turn in the cochlea but 1/3 of the electrode was out of the cochlea (Fig. 26.10).

In CH it may be difficult to locate round window through facial recess and anterior mobilization or temporary removal of posterior ear canal may be necessary. In addition, choosing the correct length of the electrode for each IEM is extremely important. It would have been more appropriate to use FORM 19 for this case. It would be more likely to have full insertion with the stopper at the level of cochleostomy if FORM 19 had been used.

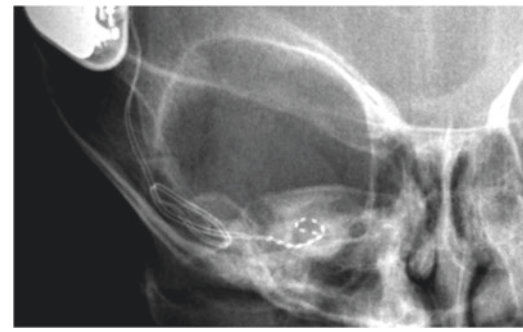


Fig. 26.10 Case 26.6. Postoperative X-ray showing partial insertion of FORM 24 where electrode makes one turn in the cochlea but 1/3 was out of the cochlea

Case 26.7: NO 2-Year-Female Patient Operated May 2011

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. 26.11a). She had CI surgery in May 2011 on the left side. Initially she showed some progress with CI. Her speech and language development then came to a plateau after which point she did not demonstrate a progress. Our team decided to make an ABI on the contralateral side. In May 2013 she underwent right ABI surgery. She showed very good progress after ABI both in CAP scores and SIR (Fig. 26.11b).

She is one of the first patients who demonstrated marked improvement after ABI surgery. After seeing her progress, duration between CI and ABI is decreased. In some cases the two procedures are performed simultaneously.

Case 26.8: ARR, 2-Year-Female Patient, Operated May 2015

She had bilateral profound SNHL. HRCT demonstrated bilateral CH-III, with hypoplastic vestibule and absent semicircular canals (Fig. 26.12a). Facial nerve was located inferior

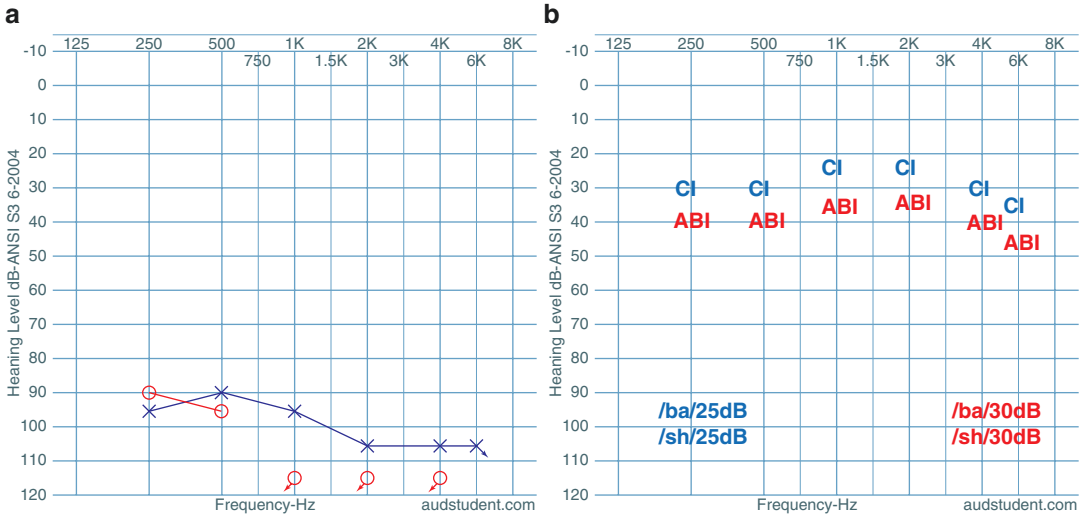


Fig. 26.11 Case 26.7. (**a**) Bilateral profound SNHL with a response on the left side using insert earphones, (**b**) postoperative CI and ABI thresholds

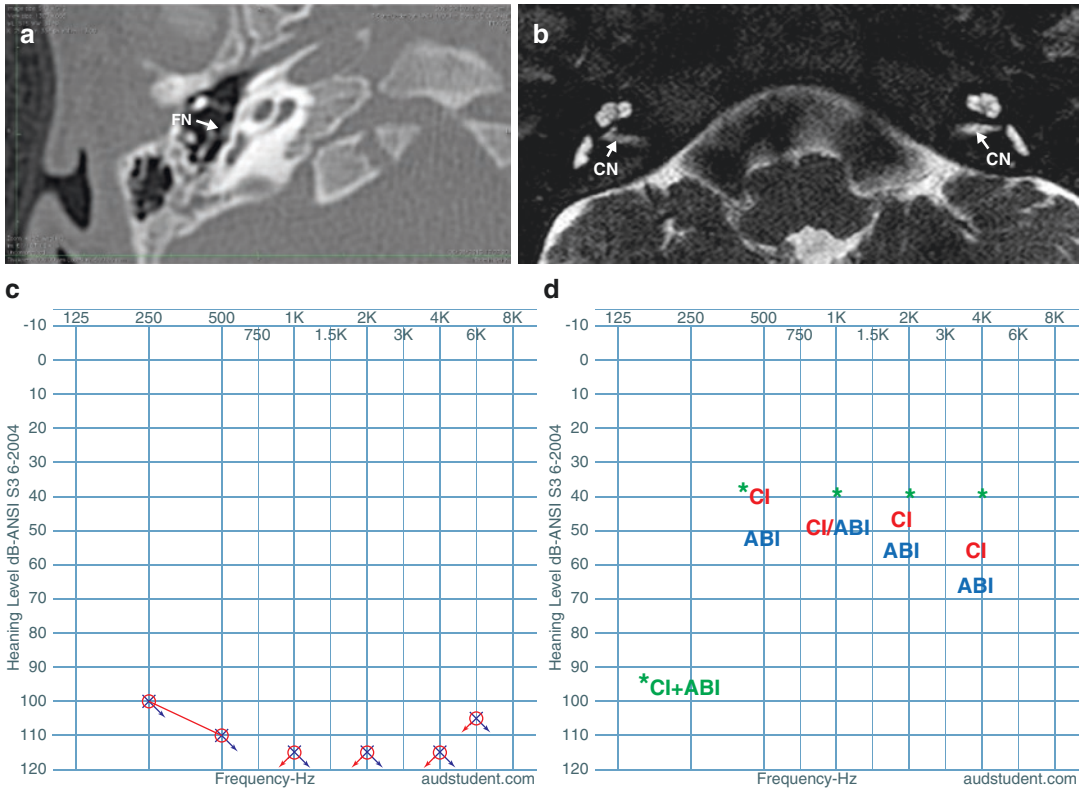


Fig. 26.12 Case 26.8. (**a**) Axial section showing CH-III and facial nerve (FN) lying on the promontory on the right side, (**b**) MRI showing extremely hypoplastic CN nerves bilaterally, (**c**) Audiogram showing response only on the right side with insert earphones, (**d**) Postoperative audiogram showing thresholds with CI and ABI. (Courtesy to Margaret Winter, House Ear Clinic, Los Angeles)

to the oval window (Fig. 26.5c). MRI was repeated two times and it revealed extremely hypoplastic CN nerves bilaterally (Fig. 26.12b). Audiologically response was present only on the right side (Fig. 26.12c). CI and ABI were planned simultaneously. Right side was operated with CI. Facial nerve was anteriorly dislocated, preventing identification of RW (Video 26.2). Facial recess was enlarged posteriorly towards the usual location of FN. Vestibule was opened. Scala vestibuli was identified and electrode was rotated 90° into scala vestibuli, obtaining full insertion. ABI was performed to the left side simultaneously. This is the first simultaneous CI and ABI surgery in the world. Initially there was no clear response to auditory stimuli in free field but she started to respond to various musical instruments. After some time she could be conditioned with VRA during CI programming. Even though there was some facial stimulation on ipsilateral side, all intracochlear electrodes were active. However, with time, she showed improvement and her current audiogram, performed 3.5 years after operation demonstrated thresholds with only CI between 40 and 55 dB, only ABI between 50 and 65 dB, and with CI + ABI around 40 dB (Fig. 26.12d).

In addition facial nerve abnormality seen on HRCT should alert the surgeon to use alternative approaches for CI surgery. If FN is anteriorly dislocated on the promontory, usual location of FN at the FR is not occupied by the FN. Therefore, recess can be extended posteriorly until the vestibule was exposed.

References

1. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
2. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
3. Melo AS, et al. Cochlear implantation in children with anomalous cochleovestibular anatomy. Auris Nasus Larynx. 2017;44(5):509–16.
4. Feng YM, et al. Cochlear implantation in a patient with severe cochlear hypoplasia. J Laryngol Otol. 2012;126(11):1172–5.
5. Puram SV, et al. Auditory brainstem implantation in a 16-month-old boy with cochlear hypoplasia. Otol Neurotol. 2015;36(4):618–24.
6. Cinar BC, et al. Audiologic and radiologic findings in cochlear hypoplasia. Auris Nasus Larynx. 2017;44(6):655–63.
7. Sennaroglu L, et al. Cochlear hypoplasia type four with anteriorly displaced facial nerve canal. Otol Neurotol. 2016;37(10):e407–9.
8. Pamuk G. Measurement of cochlear dimensions in cochlear hypoplasia using temporal bone computerized tomography and magnetic resonance imaging and its effects on cochlear implant selection. Department of Otolaryngology, Hacettepe University Medical Faculty; 2018.
9. Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. Otol Neurotol. 2002;23(1):39–42; discussion 42–3.

Enlarged Vestibular Aqueduct

27

Levent Sennaroglu, Merve Ozbal Batuk, and Burce Ozgen Mocan

Special Features

1. Enlarged vestibular aqueduct can be isolated or accompany cochlear deformities.
2. No typical audiological configuration. Hearing loss usually fluctuating and progressive throughout lifetime, with occasional sudden SNHL. Air-bone gap is usually present at the lower frequencies.
3. Usually they become a candidate for cochlear implantation during follow-up.
4. The reason for the progressive SNHL may be CSF pressure transmission into the inner ear.

27.1 Definition

Vestibular aqueduct (VA) is a bony canal transmitting the endolymphatic duct between endolymphatic sac and vestibule. Normally it is less than 1.5 mm in diameter at the midpoint and also less than the diameter of the posterior semicircular canal (Fig. 27.1a, b).

Enlarged Vestibular Aqueduct (EVA): This entity describes the presence of an enlarged vestibular aqueduct (i.e. the midpoint between posterior labyrinth and operculum is larger than 1.5 mm on axial HRCT of the temporal bone) in the presence of a normal cochlea (Fig. 27.2a, b).

EVA was discovered by Carlo Mondini [1] during temporal bone dissection for the first time. Radiologically EVA was defined by Valvassori and Clemis [2] in 1978 when its horizontal dimensions are greater than 1.5 mm at the midpoint and 2 mm at the external aperture of the vestibular aqueduct on the axial plane. It can be isolated as a separate group of inner ear malformations (IEM) in the presence of normal modiolus and cochlea (Figs. 27.2 and 27.3) or it may be accompanying incomplete partition type II (IP-II) malformation (Fig. 27.4).

In the past, the term "Large vestibular Aqueduct Syndrome" was used to describe this entity. A syndrome is defined as "a group of signs and symptoms that occur together and characterize a particular abnormality or condition." However, EVA can be present as an isolated finding or in association with cochlear abnormalities as in IP-II. Sometimes it is seen in association with congenital hypothyroidy (Pendred Syndrome). Therefore, it is more appropriate to accept this entity as "enlarged vestibular aqueduct" rather than "large vestibular aqueduct syndrome."

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

M. Ozbal Batuk (✉)
Department of Audiology, Hacettepe University, Faculty of Health Sciences, Ankara, Turkey

B. O. Mocan
Department of Radiology, Hacettepe University, Medical Faculty, Ankara, Turkey

University of Illinois at Chicago, Chicago, IL, USA

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_27

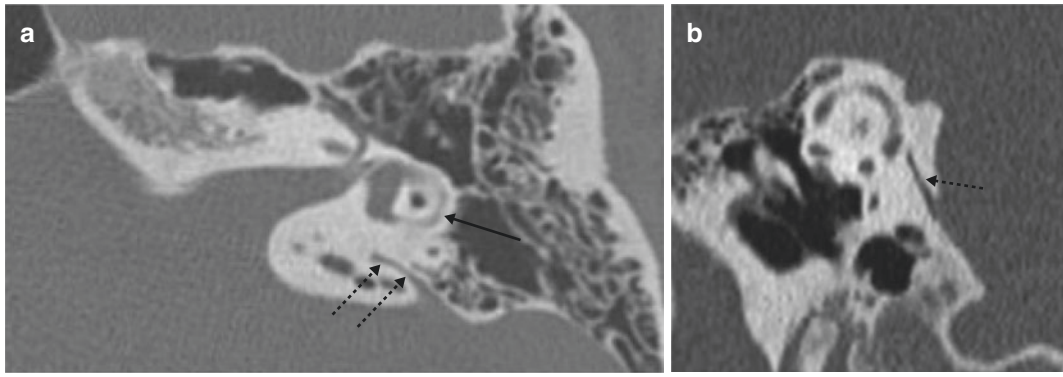


Fig. 27.1 Computerized tomography demonstrating normal vestibular aqueduct on axial (**a**) and 45° oblique reformat (Poschell view) (**b**) views (*dotted arrows* vestibular aqueduct, *straight arrow* lateral semicircular canal)

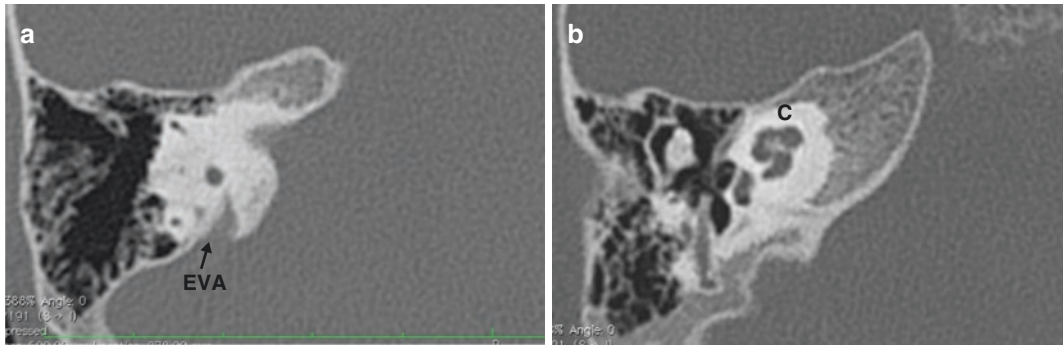


Fig. 27.2 Axial computerized tomography of enlarged vestibular aqueduct (**a**) in the presence of normal cochlea and modiolus (**b**) (*C* cochlea, *EVA* enlarged vestibular aqueduct)

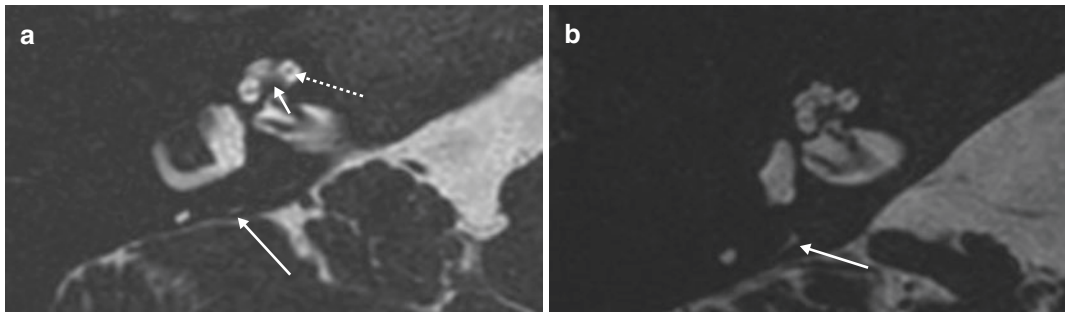


Fig. 27.3 (**a**, **b**) Magnetic resonance imaging findings showing normal modiolus (small white arrow) and osseous spiral lamina (dotted white arrow). Note that the normal sized vestibular aqueduct (straight long white arrow) is difficult to appreciate with MR imaging

According to a recent paper by Sennaroglu [3], etiology of enlarged vestibular aqueduct (EVA) appears to be the result of a genetic abnormality based on literature findings. It is usually bilateral. Rarely there may be asymmetry between the two sides. Depending on the time of the pathology and extent of CSF pressure transmission into the inner ear, other cochleovestibu-

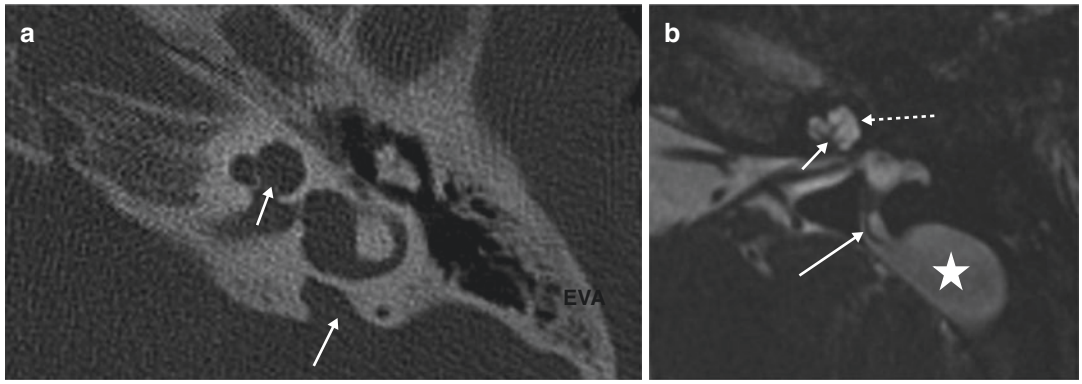


Fig. 27.4 Computerized tomography (**a**) and magnetic resonance imaging (MRI) (**b**) findings in an incomplete partition type II patient with enlarged vestibular aqueduct (white arrow), with defective modiolus (thin white arrow) and defective interscalar septa (dotted white arrow). MRI demonstrates a very large endolymphatic sac (white star)

lar findings may accompany EVA (see Chap. 24 IP-II for further details).

The difference between EVA and IP-II is the presence of additional findings in the latter. IP-II is characterized by a triad of anomalies, which were first described by Mondini [1]:

1. EVA.
2. Minimally dilated vestibule.
3. Cochlear modiolar defects resulting in cystic apex.

EVA appears to be responsible for the changes observed in IP-II. High CSF pressure transmitted into inner ear via EVA may cause different modiolar defects resulting in IP-II. Thus in the setting of an EVA, the cochlea should be evaluated carefully for additional anomalies and if the cochlea is completely normal on imaging the term EVA should be used.

In the past EVA was diagnosed more often. However, with the development of better scanners it has become possible to detect smaller cochlear changes with the present day HRCT and MRI (Fig. 27.3). MRI particularly can demonstrate even scala vestibuli dilatation [4]. Therefore, many of the pathologies may be in fact IP-II rather that EVA.

It is appropriate to accept EVA as a separate IEM group because the cochlea and vestibule are normal on imaging in spite of EVA. However, audiological findings are similar to IP-II cases.

27.2 Anatomy of Vestibular Aqueduct (VA), Endolymphatic Duct (ED), and Sac (ES)

According to Emmet [5], Valvassori and Clemis [2] described VA as an inverted "J" shaped structure. It has two segments: short descending proximal segment and a longer descending distal segment:

1. Proximal segment or isthmus, which appears as the short limb of "J"; it is approximately 1.5 mm in length, with a mean diameter of 0.3 mm.
2. Distal segment which appears as the long limb of "J"; it is triangular in shape with the apex at the isthmus and base at the opening into the posterior fossa. It increases from 0.5 mm at the isthmus towards the outer opening.

Vestibular aqueduct houses endolymphatic duct (ED) and the endolymphatic sac (ES) and the anatomy of the latter structures were described in detail by Lo et al. [6]. The ED and ES are filled with endolymph and are the non-sensory components of the membranous labyrinth. The ED forms from the confluence of the utricular and saccular ducts. It passes through the VA to the ES, which extends through the distal VA out of the external aperture of the aqueduct to terminate

in the epidural space of the posterior cranial fossa. Therefore, ED and ES consist of components both inside and outside the otic capsule connected by a narrow passageway through the capsule.

Lo et al. [6] indicated that its proximal segment, the sinus, lies in a groove on the posteromedial surface of the vestibule, while its major portion is contained within the short, slightly upwardly arched, horizontal segment of the VA. After entering the VA, the sinus tapers to its intermediate segment within the horizontal segment of the VA and then narrows at its isthmus of the VA. Endolymphatic sac begins distal to the isthmus and has a proximal, intraosseous portion, lying within the transversely widening, vertical segment of the VA, which is covered posteriorly by a thin scale of bone, the operculum. The distal, extra-osseous portion of the sac rests on a fovea on the posterior wall of the petrous bone, between layers of dura.

27.3 Histopathology and Pathophysiology

EVA appears to be responsible for the changes observed in IP-II. High CSF pressure transmitted into inner ear via EVA may cause different modiolar defects resulting in IP-II. The mildest deformity observed in the cochlea is dilatation of scala vestibuli and the most significant deformity is the complete modiolar absence. It is also possible that patency of the EVA is minimal or EVA occurs after the development of the inner ear is complete and no pathology is observed in the cochlea. If the cochlea is completely normal on imaging it is appropriate to use the term EVA. In Chap. 24 on IP-II, histopathology and pathophysiology are described in more detail.

27.3.1 Literature Findings

Pritchett et al. [7] evaluated the results of cochlear implantation in 55 individuals diagnosed with EVA. They reported that the severity of IEM affected the time of the implantation. They found that the age of CI was approximately 6 years in EVA due to the progressive nature of hearing loss. They also emphasized that individuals diagnosed with isolated EVA could benefit from hearing aids for a longer period without implantation than those diagnosed with IP-II malformation.

Ahadizadeh et al. [8] reported the audiological outcomes (word recognition scores, speech reception thresholds, and pure tone audiometry) in patients with EVA and IP-II in their retrospective longitudinal study. They mentioned that the severity of the hearing loss in patients with EVA had no correlation with the presence of the IP-II. The main reason for the severity of the hearing loss was thought to be the EVA.

In the paper by the authors [9], relationship between the size of the vestibular aqueduct and air-bone gap in patients with EVA was evaluated. It was found out that the size of the vestibular aqueduct is not associated with air-bone gap. Due to the progressive nature of the hearing loss in EVA, it is not possible to find a correlation between the size of the vestibular aqueduct and air-bone gap. While hearing changes over the time, the size of the vestibular aqueduct remains same.

Madden et al. [10] evaluated the audiometric thresholds and the configuration of the hearing loss and compared them with the size of the VA in patients with EVA. They found no correlation between the degree of the hearing loss and the size of the VA.

In contrast to these findings, a study by Ascha et al. [11] in 2017 identified a direct relationship between the size of the VA and the audiometric thresholds in the repeated measures of hearing test results. They reported an increase of 17.5 dB in speech reception threshold per each millimeter increase of the size of the VA. They emphasized that the size of the VA is directly associated with the hearing loss in patients with EVA.

Aimoni et al. [12] described the clinical and genetic characteristics of 14 adolescents with EVA depending on the pendrin gene mutations (SLC26A4) in their study. Participants were divided into three subgroups: Group 1 non-syndromic EVA; Group 2 EVA with SLC26A4 gene mutation without thyroid dysfunction; and

Group 3 EVA with Pendred syndrome with two pathological mutations of the SLC26A4 gene. They defined that degree of the hearing loss changes from mild to profound in patients with non-syndromic EVA. Conversely, severe to profound hearing loss was identified in most of the patients (%70–75) with SLC26A4 gene mutation (for more details see Chaps. 6 and 12).

In the literature many papers showed that the audiological performance of the patients with EVA are similar to the patients without any cochlear malformation [13–15]. Isaiah et al. [14] retrospectively analyzed the speech perception and radiological characteristics of the pediatric cochlear implant users with IEMs. They found out that most of the children with EVA are able to achieve open set speech perception and perform as good as others with normal anatomy. Manzoor et al. [15] also compared the performance of the patients with isolated EVA and EVA with IP-II and could not find any significant difference between two the groups.

In addition to audiological findings, vestibular symptoms in EVA were also reported in a recent study by Song et al. [16]. In their series vestibular complaints were reported in more than half of the patients with EVA and surprisingly, the incidence of benign paroxysmal positional vertigo (BPPV) was 18.2%. Although the percentage of vestibular symptoms are less than cochlear findings, it was recommended to evaluate vestibular symptoms particularly BPPV, in EVA.

27.3.2 Clinical Findings

Clinical and audiological presentation is similar to IP-II patients. Hearing may be normal at birth. Usual presentation is progressive mixed or SNHL. There may be sudden hearing loss attacks. In case of sudden hearing loss, there is a strong possibility for hearing recovery if the treatment (steroids and vasodilators) is started immediately. Usually with more attacks hearing progresses to a point with no benefit from hearing aids and they become candidates for CI surgery. Meningitis and facial nerve anomaly are extremely rare in EVA.

27.3.3 Audiological Findings

Audiological presentation and management is similar to that of IP-II. There is no typical audiological configuration for EVA. A heterogeneous hearing pattern, such as progressive sensorineural hearing loss, mixed type hearing loss, and fluctuating hearing loss, is encountered in EVA malformation. At birth they may obtain pass-on hearing screening with otoacoustic emission and automatic auditory brainstem response tests. Hearing loss is usually progressive. At the beginning they benefit from hearing aids. With progressive hearing loss or sudden hearing loss attacks they become candidates for CI. A study by Govaerts et al. [17] emphasized that initial diagnosis of the hearing loss in EVA is between the age of 3.5 and 5 years.

Hearing loss can be symmetric or asymmetric (Fig. 27.5a, b). The most characteristic audiological finding is air-bone gap (ABG) particularly present at low frequencies, but sometimes ABG can be seen in all frequencies. **These patients should not be operated for ossicular fixation**. The reason for air-bone gap is the third window phenomenon caused by the EVA which creates a bony defect in the labyrinth.

The pathological third window effect of EVA malformation causes the acoustic energy to go in a different direction from the cochlea, unlike the normal third window effect. This increases the compressive mechanism of bone conduction and results in better bone conduction hearing thresholds and lower air conduction hearing thresholds. Third window effect refers to selective stimulation of bone pathway in the presence of EVA [18].

27.3.4 Radiology

According to the IEM database of Hacettepe University Department of Otolaryngology, out of 776 patients with various IEMs, 69 of 1652 ears had EVA (4.2%).

Vestibular aqueduct enlargement is diagnosed when the midpoint between posterior labyrinth and operculum is larger than 1.5 mm in the pres-

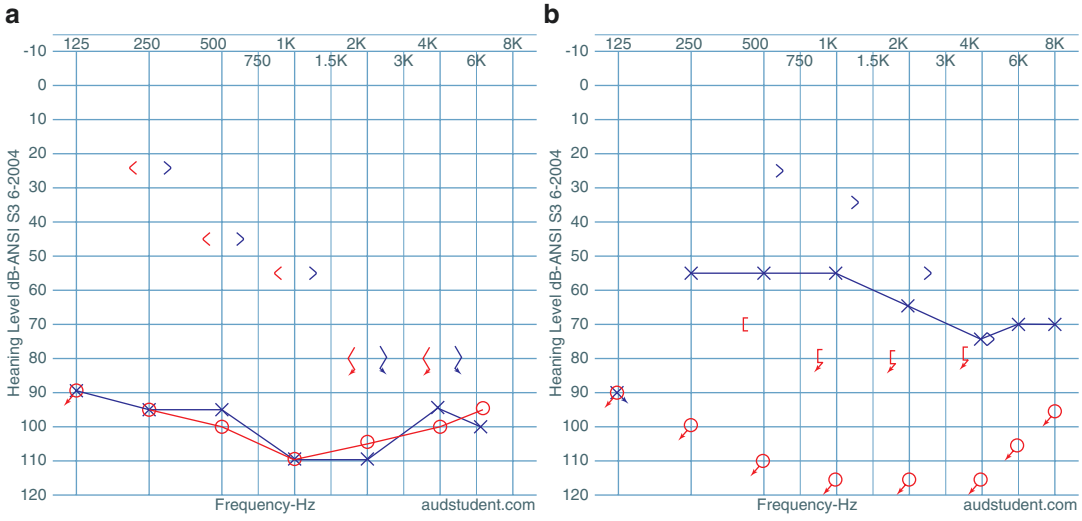


Fig. 27.5 Audiological findings in enlarged vestibular aqueduct: (**a**) Symmetrical and (**b**) Asymmetrical audiological findings

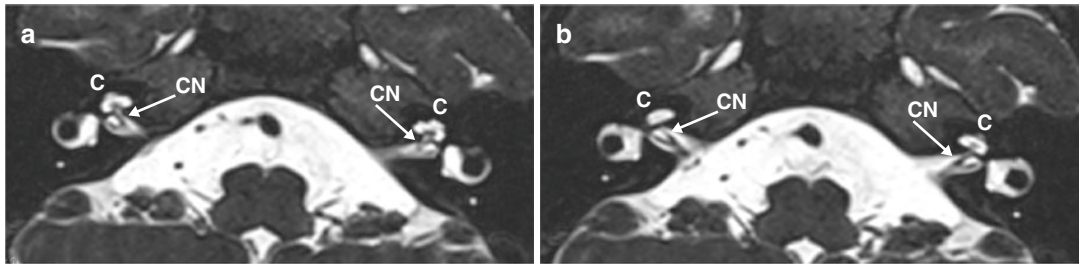


Fig. 27.6 Magnetic resonance imaging showing normally developed cochlear nerves (CN) on both sides (**a** and **b**)

ence of a normal cochlea (Fig. 27.2a, b). The external size of the cochlea as well as internal architecture should be normal. Higher field strength scanners with dedicated sequences such as three-dimensional constructive interference in steady-state (CISS), fast imaging employing steady-state acquisition (FIESTA), or driven equilibrium (DRIVE) allow higher resolution imaging of the cochlea with a better demonstration of modiolus, interscalar septa, scala tympani, and vestibule, allowing differentiation of EVA from IP-II (Figs. 27.3 and 27.4) [19–21]. In some cases of minor EVA, it may be difficult to diagnose EVA with only MRI. CT has been reported to have a higher accuracy compared to MRI in the depiction of enlargement (Fig. 27.2 and 27.3).

There are some controversies in the literature about the dimensions of VA to be accepted as enlarged. Vijayasekaran et al. [22] reported that VAs with midpoint or opercular widths of 1.0 and 2.0 mm or greater, respectively, are enlarged. Mohammed El Badry et al. [23] supported these measurements for diagnosis of EVA.

MRI always reveals normally developed cochlear nerves (Fig. 27.6). Therefore, ABI is not indicated in EVA.

EVA was mainly diagnosed on axial sections. Ozgen et al. [24] reported that the 45° oblique reformate provides better visualization and more accurate measurement of the VA compared to the routine images in the axial plane (Fig. 27.1b). It is very important to evaluate coronal sections as well (Fig. 27.6). In a recent paper by Sennaroglu and Bajin [25], the authors added the vertical dimension as well in the diagnosis of EVA. We have made the observation that EVA is present in

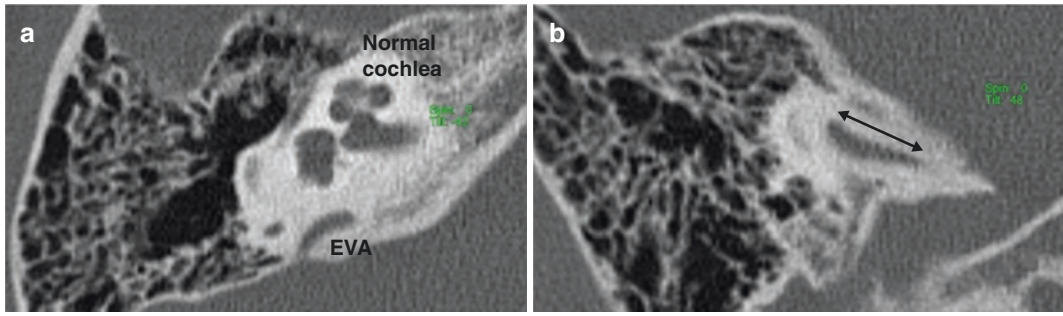


Fig. 27.7 Enlarged vestibular aqueduct diagnosed on coronal section. (**a**) Axial section showing normal vestibular aqueduct. (**b**) Coronal section showing enlarged vestibular aqueduct on oblique dimensions

a number of axial cuts (Fig. 27.7). This means that it can be enlarged in superior-inferior or oblique dimensions as well. It is advisable to use the same measurement of 1.5 mm at the midpoint on coronal sections for the diagnosis of EVA as well. Sometimes VA is enlarged in an oblique position; when axial measurements are taken into consideration, it may not be enlarged but it may be enlarged when oblique measurements are used (Fig. 27.7).

EVA is frequently bilateral and symmetric; rarely, it can be asymmetric.

27.4 Management

In patients with EVA, head trauma should be avoided. This may cause increase in CSF pressure and this may be transmitted into the inner ear resulting in sudden or progressive hearing loss. Helmets are advised during sports. It is advisable to avoid contact sports. In a recently published review study of Brodsky and Choi [26], it was reported that despite the possible association between EVA and sudden HL, head traumas are not a risk factor for overall progression of HL in EVA.

As hearing loss shows a progressive pattern, hearing aids are sufficient at the beginning. Language development in EVA is excellent.

With bilateral profound SNHL CI is indicated. There may be cases with asymmetric hearing loss with one side moderate, and other side profound SNHL. In this situation, CI is indicated on the side with profound HL.

27.5 Surgery

During CI surgery, all kinds of electrodes can be used as the modiolus and basal part of the cochlea is normal. Dimensions of cochlea and semicircular canals are normal; therefore, no facial nerve abnormality or difficulty during surgery is expected.

27.6 Hacettepe Experience

In our department between 1997 and September 2018, 2646 patients underwent CI and ABI operations. 279 of CI cases had IEMs. 30 had EVA deformity. During this period 30 patients with isolated EVA, out of 279 patients with IEMs, were implanted with CI. In this particular group of patients with EVA, minimum age of CI was 12 months and maximum age was 30 years. Mean age at CI was 9.7 years for our series of EVA. These findings suggest two important audiological characteristics of EVA malformation:

1. Patients with EVA can benefit from their hearing aids for a longer period due to progressive nature of hearing loss.
2. Patients with EVA may have sudden hearing loss at any time in their life.

Therefore, in patients with EVA, CI may be required at any time in their life.

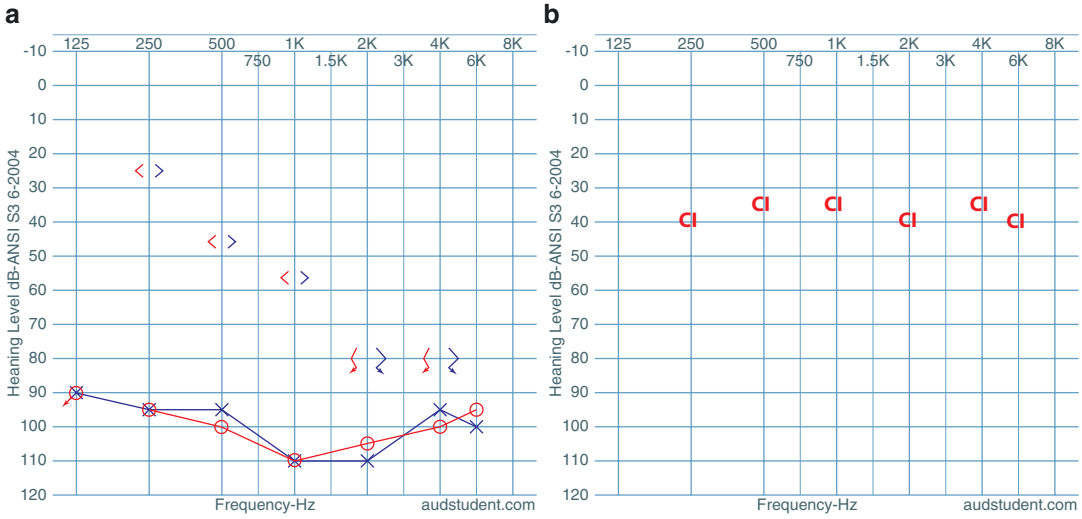


Fig. 27.8 Case 27.1: 17-year-old male patient with bilateral EVA. (**a**) Preoperative audiogram, (**b**) hearing thresholds with right cochlear implant

During surgery, a facial recess approach was successfully used in all 30 patients. While pulsation was frequently observed, none of them had gusher. Six had oozing during surgery. This was easily controlled with any kind of electrode using soft tissue around the electrode.

Cochlear nerve is always present in EVA. None of the patients with EVA received an ABI.

27.7 Cases

Case 27.1: YY, 17-Year-Old Male Patient, Operated October 2016

He had progressive hearing loss necessitating a CI (Fig. 27.8a). HRCT of the patient revealed bilateral EVA. Despite a large air-bone gap especially at the low frequencies, no middle ear surgery was planned because of EVA. He received a CI on the right side in October 2016 (Fig. 27.8b).

Case 27.2: EÇ, 9-Year-Old Male Patient, Follow-Up with Hearing Aids

He had bilateral EVA with severe mixed type hearing loss (Fig. 27.9). His follow-up was done with bilateral hearing aids. Family was informed about the possibility of progressive hearing loss and necessity of a CI during follow-up.

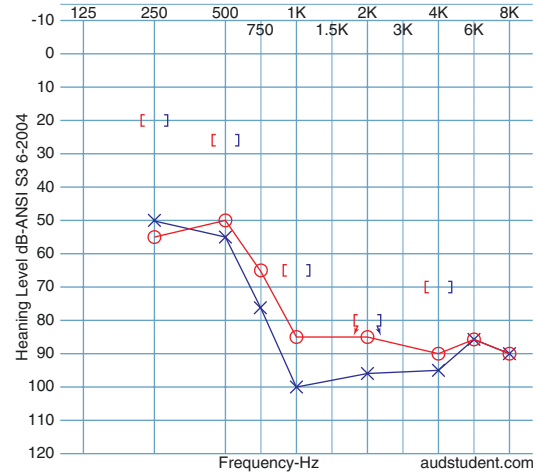


Fig. 27.9 Case 27.2: 9-year-old male patient who is followed up with bilateral hearing aids

Case 27.3: MI, 31-Year-Old Male Patient, Operated November 2017

History revealed sudden hearing loss on the right ear at the age of 4. His hearing on the left side was normal and he did not use any hearing aids on the right ear. He experienced sudden hearing loss on the left ear in February 2016 (Fig. 27.10a). Despite medical therapy, his hearing did not recover. An audiogram 10 months later showed another attack of hearing loss on the left ear (Fig. 27.10b). He started to use hearing aids on

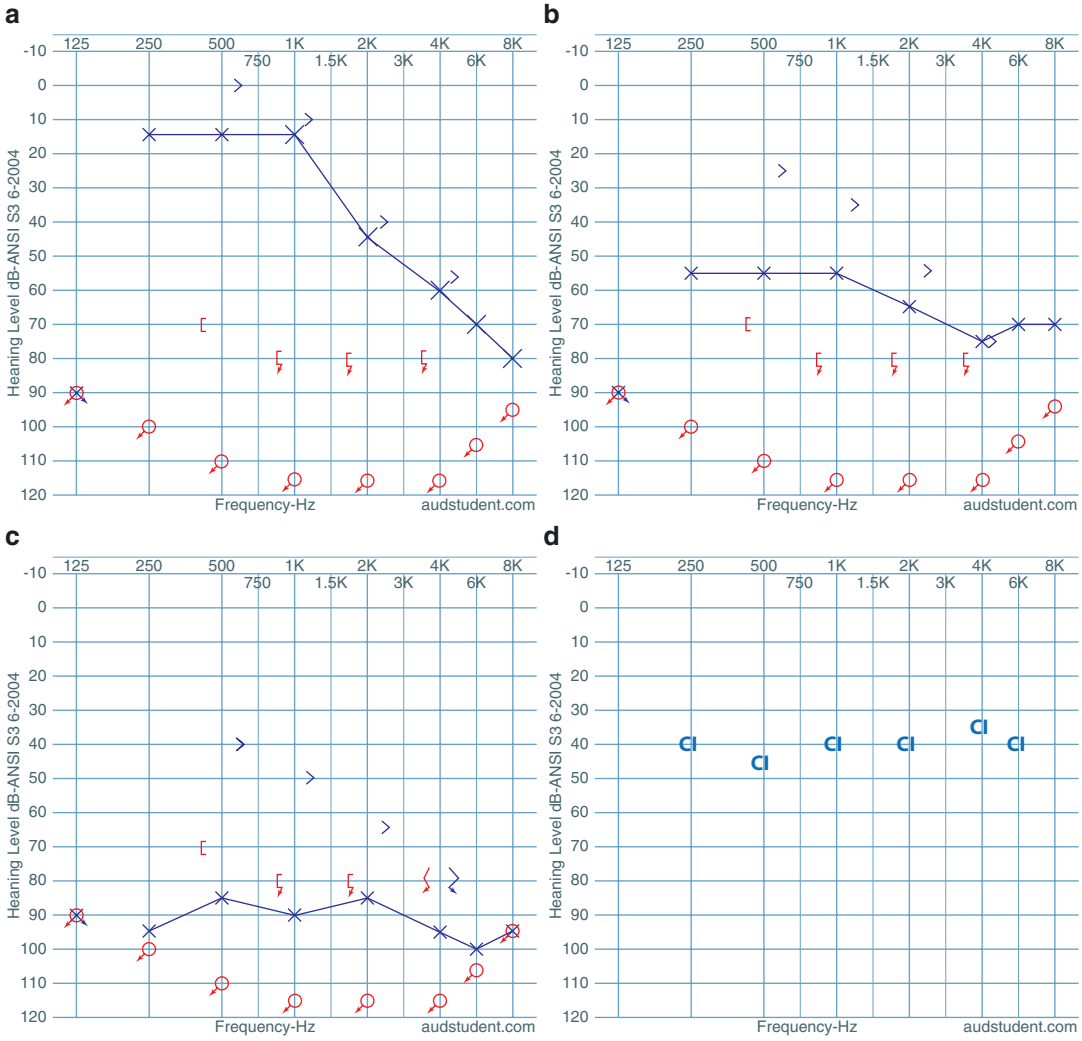


Fig. 27.10 Case 27.3: 31-year-old male patient with sudden hearing loss attack. (**a**) First sudden hearing loss attack on the left ear, (**b**) second hearing loss attack (left side), (**c**) preoperative audiogram, (**d**) hearing thresholds with left cochlear implant

both sides. After the last attack, he applied for cochlear implantation in May 2017 (Fig. 27.10c). His left ear was operated with CI (Fig. 27.10d).

Present day approach with radiological imaging techniques and cochlear implantation was not present 30 years ago when his hearing loss on the right side occurred. At that time appropriate rehabilitation option was not planned for the right ear. At present time when a hearing loss occurs unilaterally, in a child of 4 years of age with bilateral EVA, cochlear implantation should be planned if there is no recovery after medical treatment. Only in this way bilateral hearing can be restored in a patient with EVA.

Case 27.4: MS, 3-Year-Old Male Patient, Operated December 2016

He was diagnosed with profound mixed type hearing loss (Fig. 27.11) and bilateral EVA. He was implanted with CI at the age of 3.

In contrast to Case 27.3, at present time CI is provided at the time of diagnosis of profound hearing loss. Because of the nature of the EVA, hearing loss is usually progressive and is expected

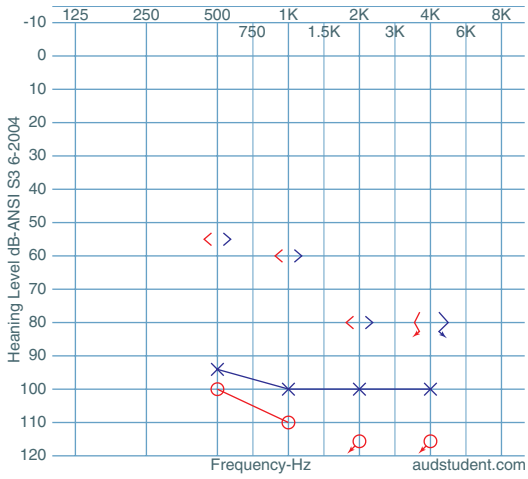


Fig. 27.11 Case 27.4: 3-year-old male patient operated with CI

on the left side as well. Therefore, in cases of EVA, CI should be performed in unilateral profound hearing loss.

References

1. Mondini C. Minor works of Carlo Mondini: the anatomical section of a boy born deaf. Am J Otol. 1997;18(3):288–93.
2. Valvassori GE, Clemis JD. The large vestibular aqueduct syndrome. Laryngoscope. 1978;88(5):723–8.
3. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
4. Sennaroglu L. Another evidence for pressure transfer mechanism in incomplete partition two anomaly via enlarged vestibular aqueduct. Cochlear Implants Int. 2018;19(6):355–7.
5. Emmett JR. The large vestibular aqueduct syndrome. Am J Otol. 1985;6(5):387–415.
6. Lo WW, Daniels DL, Chakeres DW, Linthicum FH Jr, Ulmer JL, Mark LP, et al. The endolymphatic duct and sac. AJNR Am J Neuroradiol. 1997;18(5):881–7.
7. Pritchett C, Zwolan T, Huq F, Phillips A, Parmar H, Ibrahim M, et al. Variations in the cochlear implant experience in children with enlarged vestibular aqueduct. Laryngoscope. 2015;125(9):2169–74.
8. Ahadizadeh E, Ascha M, Manzoor N, Gupta A, Semaan M, Megerian C, et al. Hearing loss in enlarged vestibular aqueduct and incomplete partition type II. Am J Otolaryngol. 2017;38(6):692–7.
9. Batuk MÖ, Çınar BÇ, Özgen B, Sennaroğlu G, Sennaroğlu L. Audiological and radiological characteristics in incomplete partition malformations. J Int Adv Otol. 2017;13(2):233.
10. Madden C, Halsted M, Benton C, Greinwald J, Choo D. Enlarged vestibular aqueduct syndrome in the pediatric population. Otol Neurotol. 2003;24(4):625–32.
11. Ascha MS, Manzoor N, Gupta A, Semaan M, Megerian C, Otteson TD. Vestibular aqueduct midpoint width and hearing loss in patients with an enlarged vestibular aqueduct. JAMA Otolaryngol Head Neck Surg. 2017;143(6):601–8.
12. Aimoni C, Ciorba A, Cerritelli L, Ceruti S, Skarżyński P, Hatzopoulos S. Enlarged vestibular aqueduct: audiological and genetical features in children and adolescents. Int J Pediatr Otorhinolaryngol. 2017;101:254–8.
13. Clarós P, Fokouo JVF, Clarós A. Cochlear implantation in patients with enlarged vestibular aqueduct. A case series with literature review. Cochlear Implants Int. 2017;18(3):125–9.
14. Isaiah A, Lee D, Lenes-Voit F, Sweeney M, Kutz W, Isaacson B, et al. Clinical outcomes following cochlear implantation in children with inner ear anomalies. Int J Pediatr Otorhinolaryngol. 2017;93:1–6.
15. Manzoor NF, Wick CC, Wahba M, Gupta A, Piper R, Murray GS, et al. Bilateral sequential cochlear implantation in patients with enlarged vestibular aqueduct (EVA) syndrome. Otol Neurotol. 2016;37(2):e96–e103.
16. Song J-J, Hong SK, Lee SY, Park SJ, Kang SI, An Y-H, et al. Vestibular manifestations in subjects with enlarged vestibular aqueduct. Otol Neurotol. 2018;39(6):e461–e7.
17. Govaerts P, Casselman J, Daemers K, De Ceulaer G, Somers T, Offeciers F. Audiological findings in large vestibular aqueduct syndrome. Int J Pediatr Otorhinolaryngol. 1999;51(3):157–64.
18. Merchant SN, Nakajima HH, Halpin C, Nadol JB Jr, Lee DJ, Innis WP, et al. Clinical investigation and mechanism of air-bone gaps in large vestibular aqueduct syndrome. Ann Otol Rhinol Laryngol. 2007;116(7):532–41.
19. Dahlen RT, Harnsberger HR, Gray SD, Shelton C, Allen R, Parkin JL, et al. Overlapping thin-section fast spin-echo MR of the large vestibular aqueduct syndrome. AJNR Am J Neuroradiol. 1997;18(1):67–75.
20. Simons JP, Mandell DL, Arjmand EM. Computed tomography and magnetic resonance imaging in pediatric unilateral and asymmetric sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 2006;132(2):186–92.
21. Kachniarz B, Chen JX, Gilani S, Shin JJ. Diagnostic yield of MRI for pediatric hearing loss: a systematic review. Otolaryngol Head Neck Surg. 2015;152(1):5–22.
22. Vijayasekaran S, Halsted MJ, Boston M, Meinzen-Derr J, Bardo DM, Greinwald J, et al. When is the vestibular aqueduct enlarged? A statistical analysis of the normative distribution of vestibular aqueduct size. AJNR Am J Neuroradiol. 2007;28(6):1133–8.
23. El-Badry MM, Osman NM, Mohamed HM, Rafaat FM. Evaluation of the radiological criteria to diagnose

large vestibular aqueduct syndrome. Int J Pediatr Otorhinolaryngol. 2016;81:84–91.
24. Ozgen B, Cunnane ME, Caruso PA, Curtin HD. Comparison of 45 degrees oblique reformats with axial reformats in CT evaluation of the vestibular aqueduct. AJNR Am J Neuroradiol. 2008;29(1):30–4.
25. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397.
26. Brodsky JR, Choi SS. Should children with an enlarged vestibular aqueduct be restricted from playing contact sports? Laryngoscope. 2018;128(10):2219–20.

Cochlear Aperture Abnormalities

28

Emel Tahir, Burce Ozgen, and Levent Sennaroglu

Special Features

1. In spite of profound SNHL, they may pass hearing screening if only OAE is done
2. ABR demonstrates profound SNHL
3. CI results usually not satisfactory
4. Possible indication for ABI

28.1 Definition

The bony canal between the internal auditory canal (IAC) and the cochlear modiolus is termed as the cochlear aperture (bony cochlear nerve canal) [1]. The cochlear aperture (CA) lies between the fundus of the internal auditory canal (IAC) and the base of the cochlea. It carries cochlear nerve fibers from the spiral ganglion to the cochlear nerve [2]. Sennaroglu et al. included cochlear aperture abnormalities into the classification of inner ear malformations (IEMs) in 2013 [3]. This is because inner ear can be normal apart from CA atresia/stenosis which changes the outcome of cochlear implantation in such a patient. Therefore, it is very important to evaluate CA abnormalities as the eighth category in the classification of IEMs.

E. Tahir (✉)
Department of Otolaryngology, Medical Faculty, Ondokuz Mayıs University, Samsun, Turkey

B. Ozgen
Department of Radiology, Medical Faculty, Hacettepe University, Ankara, Turkey

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

28.2 Histopathology and Pathophysiology

The cochlear aperture is a short anatomical bony canal with a circular cross-section. The cochlear nerve passes through this canal. In the current literature there are only a few in vivo studies regarding the cochlear aperture [1]. In a histopathological study, mean cochlear aperture (CA) diameter was found as 2.26 (± 0.25) mm at the mid-modiolar level in normal human temporal bones. Measurements were made by selecting the mid-cochlear slide in axial histopathologic sections. In that study, 110 temporal bones from 110 patients, 62 (56.4%) men and 48 (43.6%) women were investigated [1]. There were no differences with increasing age or between males and females in terms of the CA measurements [1].

In another temporal bone histopathological study which was combined with computed tomography (CT) scan, CA was measured in 117 casts from temporal bone specimens [2]. The mean diameter in the axial plane was 2.59 mm. The mean length and diameter determined from CT scans of the specimens were 1.19 and 1.98 mm, respectively [2]. The authors concluded that if the diameter of the CA is <1.4 mm, then the possibility of cochlear nerve abnormality should be considered.

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_28

Normal development of the inner ear can be affected by a number of factors in various stages of the development. The exact embryological cause of the narrow CA remains unclear. It was speculated that a developmental insult of the otic capsule may inhibit normal production of neural growth factor [4, 5]. This might result in excessive neuronal degeneration and prevent normal growth of the developing cochlear nerve. Fatterpekar et al. [6] indicated that as the majority of patients with profound SNHL are thought to have an abnormality involving the membranous labyrinth, it is possible that such a malformation may inhibit the normal trophic effects of nerve growth factor. This would result in a hypoplastic cochlear nerve and hypoplasia of the cochlear aperture [7]. Just as the developing internal acoustic meatus requires the presence of a normal cochlear nerve as a stimulus for attaining normal adult dimensions, it is likely that the cochlear aperture also requires a similar neural stimulus for normal development [7, 8]. Lack of an adequate stimulus because of a hypoplastic CN may prevent the CA reaching its normal diameter.

Labyrinthine development begins at approximately 3 weeks' gestation with the formation of the otic placode. At 7 weeks, the spiral organ of Corti develops and fibers from the spiral ganglia form the cochlear nerve [9]. At approximately 9 weeks' gestation, the mesenchyme surrounding the otic vesicle begins to transform chondrocytes which will ossify and form the otic capsule. In addition, presence and development of the nerve seems to require the release of a neural growth factor from the otic capsule. The IAC is thought to be formed by inhibition of cartilage formation and this inhibition requires the presence of the normal vestibulocochlear nerve. It might be speculated that CA is also dependent on the formation of the nerve as CA is a continuation of IAC. If CN is deficient, it is difficult to expect a proper IAC development [10, 11].

It is obvious that an aplastic IAC does not contain a cochlear nerve. Also there may be variation in the diameter of the IAC depending on the volume of vestibulocochlear nerve fibers traversing the canal and thus the volume producing an inhibitory substance to cartilage formation. That is why the deficiency of all components of the vestibulocochlear nerve causes an aplastic or hypoplastic IAC [12].

Developmental insults such as mechanical destruction, acoustic trauma, hypoxia, and ototoxic medication causing neuronal degeneration may be the cause of cochlear nerve hypoplasia. Degeneration of the cochlear neuroepithelium may cause the retrograde destruction of the spiral ganglia and neurons [13]. This variability in cochlear neuronal destruction with different types of end organ insults may explain clinical variability in patients with sensorineural hearing loss.

28.3 Literature Review

Lim et al. [14] studied IEMs in CA stenosis cases in a small cohort. They found a narrow IAC in 4.8% of the affected ears and cochleovestibular anomalies in 7.1% of the affected ears. Nakano et al. [15] reported the coexistence of CA stenosis and CN deficiency in 28 of 58 patients with unilateral hearing loss. A narrow CA together with a narrow IAC indicated CN deficiency in that study. The authors of this chapter conducted a similar study and found CN deficiency in all ears with CA atresia [16]. If the CA was stenotic, only 16% of the ears had normal CN. The mean IAC was narrower in CA atresia/stenosis cases which also supports the theory that development of these two structures is complementary. Also the distribution of IEMs was investigated in that study. Among the ears with CA atresia, 90.5% of them had cochlear hypoplasia. Among the ears with CA stenosis, 66.6% of them had cochlear hypoplasia and 17.5% of them had incomplete partition type I. The causal relationship between cochlear hypoplasia and cochlear nerve deficiency can be explained by insufficient growth factor production by a hypoplastic cochlea, which may adversely affect neural development, resulting in CN deficiency.

Since CA encases the cochlear nerve, a narrow CA likely indicates anatomic or functional deficiency in the cochlear nerve. The question of which value for the CA should be accepted as "normal" is an ongoing debate. Until now, there

has been no consensus regarding the cut-off points and normative data of the length and width of the CA. There are various reports in the literature about the measurement methods and normal values of the CA length and width. Many authors have reported that the CA length in normal hearing inner ears ranges from 0.93 to 1.17 mm and that the width ranges from 1.88 to 2.13 mm [2, 17, 18]. In 2000, Fatterpekar et al. [6] reported that the width of the BCNC was significantly smaller in patients with sensorineural hearing loss than in a normal control group. In a report by Komatsubara et al. [19], patients with a narrow CA on CT scan were diagnosed as having CN hypoplasia on MRI with 88.9% sensitivity and 88.9% specificity. Those authors stated that in ears in which BCNC was <1.5 mm on CT, CN hypoplasia could be seen on MRI. In a report by Kono, a CA diameter <1.7 mm suggested CN hypoplasia, even if no cochlear abnormality could be found on CT [17]. In the study of Miyasaka et al. all ears with CN hypoplasia had a small CA (≤1.5 mm) [20].

As the CA is defined as a bony canal between the IAC and the cochlear modiolus that carries the cochlear nerve fibers, any abnormality in CA may diminish neural transmission. CA stenosis is associated with IAC stenosis and cochlear nerve deficiency [21]. The cell bodies of the cochlear nerve reside in the spiral ganglion of the modiolus, with axonal projections passing from the base of the cochlea into the IAC. Because these nerve fibers must pass from the modiolus to the IAC, it is often hypothesized that a narrow CA is evidence of a deficient cochlear nerve. In CA and IAC stenosis it has been postulated that a small number of cochlear nerve fibers remain, and that they are able to transmit action potentials required for hearing [22]. Stenosis could reflect cochlear nerve deficiency, however, it could also be a manifestation of a broader embryologic insult to the developing inner ear.

In 2000, Fatterpekar et al. [6] reported that the width of the CA was significantly smaller in patients with SNHL than in a control group. Yi et al. [21] reported that the degree of hearing loss increased with narrower CA. Narrow CA cases showed statistically significant, severe to profound hearing loss compared to the normal CA group. In their series, most of the unilateral sensorineural hearing loss ears had CA stenosis/atresia. When the diameter of the CA was less than 1.4 mm, pure tone thresholds were more than 70 dB HL in most ears. Kono et al. [17] also showed that in unilateral SNHL group, a diameter of BCNC less than 1.7 mm indicated cochlear nerve hypoplasia.

However, the mechanism for hearing loss in these patients is unknown and no causal relationship between CA stenosis and SNHL has been proven. One possible association is that SNHL in patients with CA stenosis is due to a deficient number of cochlear nerve fibers passing through the CA [23]. If so, then we might expect a stable hearing loss and less likelihood of progression over time when compared with patients with other anomalies such as EVA. Conversely, CA stenosis may reflect a more serious developmental abnormality involving the cochlea itself. In which case, patient may be at risk of progression in ipsilateral, or even contralateral, ears [24].

Purcell et al. [24] investigated unilateral SNHL cases and found that baseline pure tone average (PTA) was 70.8 dB in children without CA stenosis and 75.6 dB in children with stenosis. Among children with CA stenosis 18% had progressed by 12 months rates of progression were lower among children without CA stenosis. They found that the children with stenosis had nearly two times greater risk estimate for progression. The study by Purcell et al. [24] demonstrated that CA stenosis is significantly associated with the impairment of speech perception in children with unilateral hearing loss.

Cinar et al. [25] reported that cases with CA aplasia mostly had severe to profound SNHL or no response to sound. However, when there was CA stenosis, the degree of SNHL showed variability. The degree of SNHL changes from mild to profound in CA stenosis cases and some cases had no response.

On contrary, Lim et al. [14] analyzed the differences in hearing level at each frequency of pure tone audiogram according to the presence or absence of CA stenosis. However, there were no significant differences in hearing threshold at each frequency between normal ears and ears with hearing loss. They also checked the correla-

tion between severity of hearing loss and the diameter of the CA to determine whether hearing level was affected by the degree of CA stenosis, but no significant correlation was found.

28.4 Radiological Evaluation

Technological improvements in imaging have led to the identification of IEMs that had not been previously identified, such as CA abnormalities. Recently, there have been more studies about the imaging of inner ear malformations. High-resolution computed tomography (HRCT) of the temporal bone provides additional information regarding temporal bone pathologies especially in bony malformations. As the CA is a bony structure it should be evaluated on HRCT scans.

Although there is no real consensus on the diagnostic criteria of the CA abnormalities, as mentioned in the previous paragraph, it is generally accepted that the CA should be considered abnormal when narrower than 1.5 mm. In literature the recommended evaluation method for the measurement of the CA width and length is the measurement at mid-modiolar section in axial CT. In order to evaluate the CA dimensions, first the fundus of the IAC should be recognized by using axial slices on a CT scan. In axial section, the length of the CA can be measured by drawing a perpendicular line from the base of the modiolus to the fundus of the internal auditory canal (Fig. 28.1a). The width of the CA can be measured at its mid-portion at mid-modiolar sections (Fig. 28.1b). The authors of this chapter accepted 1.5 mm as cut-off value and found a significant relationship between a narrow CA and CN deficiency [16]. The above mentioned papers are summarized in Table 28.1.

Magnetic resonance imaging (MRI) is able to show the vestibulocochlear nerve and it is the only tool to evaluate the status of the cochlear nerve such as aplasia or hypoplasia [20]. Generally, MRI is recommended in the evaluation of cochlear nerve integrity in patients with profound SNHL; however, estimation of CA by using TBCT may play a supportive role. A narrow CA and a narrow IAC may indicate that there are no sufficient nerve fibers to transmit sound from cochlea to brainstem. The fundamental goal of preoperative imaging is the prediction of CI outcomes. Therefore, a narrow CA may cause higher rates of cochlear nerve deficiency and poor cochlear implantation outcomes. Differentiation between hypoplasia and a normal sized cochlear nerve can also be challenging and requires the highest possible resolution [19]. There is no well-defined consensus regarding the definition of cochlear nerve hypoplasia. The cochlear nerve hypoplasia was previously defined as a cochlear nerve with a diameter smaller than that of the facial nerve, seen on the oblique sagittal images [20] or when it appeared decreased in size compared with the other nerves of the IAC [19].

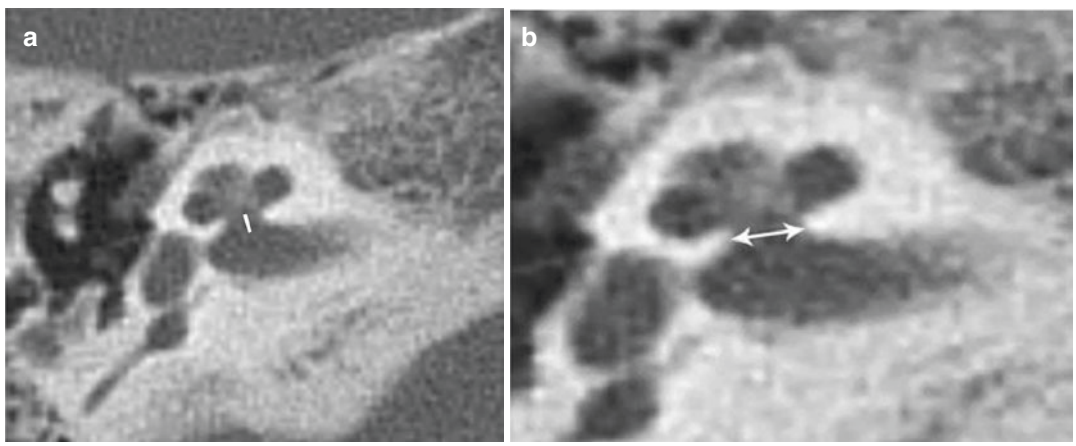


Fig. 28.1 Axial high-resolution computed tomography sections showing (**a**) the length of the bony cochlear nerve canal from the base of the modiolus to the fundus of the internal auditory canal (white line) (**b**) the width of the bony cochlear nerve canal at the mid-modiolar level (white arrow)

Table 28.1 Measurement methods and measured width of the CA in previous studies with normal hearing and sensorineural hearing loss

Author	Measurement method	Mean CA width(±SD) (mm) in control	Mean CA width(±SD) in SNHL
Stjernholm et al.	Mid-portion of inner walls	1.91 (±0.24) (*N* = 100 ears)	n.a
Fatterpekar et al.	Mid-portion of inner walls	2.13 (±0.44) (*N* = 50 ears)	1.82 (±0.24) (*N* = 33 ears)
Pagarkar et al.	Maximum width	1.9 (±0.7) (*N* = 19 ears)	1.0 ± 0.2 (*N* = 8 ears)
Komatsubara et al.	Midline between the IAC fundus/base of the modiolus	1.91 (±0.27) (*N* = 100 ears)	0.99 (±0.37) (*N* = 12 ears)
Kono	At the base of the modiolus	2.1 (±0.2) (*N* = 118 ears)	n.a
Teissier et al.	At the entry of the cochlea	2.16 (±0.24) (*N* = 174 ears)	2.12 (±0.55) (*N* = 120 ears)
Tahir et al.	Mid-portion of inner walls	1.99 (±0.36) (*N* = 72 ears)	0.96 (±0.43) (*N* = 59 ears)
Jang et al.	At the fundus of the IAC	2.39(±0.23) (*N* = 87 ears)	1.57(±0.74) (*N* = 87ears)

SD standard deviation, *N* number, *CA* cochlear aperture, *SNHL* sensorineural hearing loss, *n.a* not available

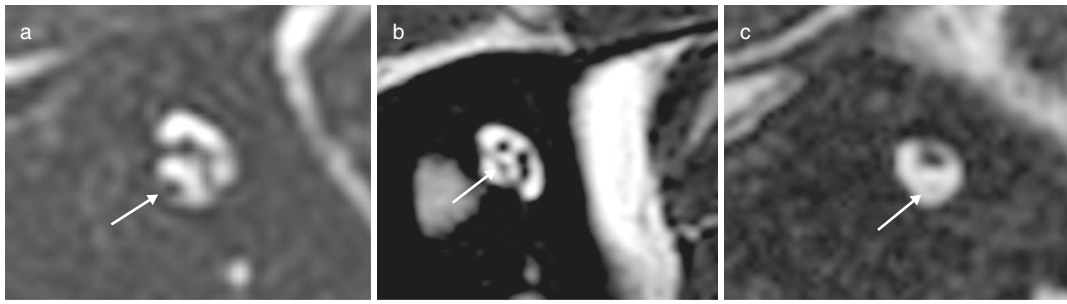


Fig. 28.2 Cochlear nerve. Cochlear nerve is evaluated on axial and sagittal oblique T2-weighted MRI images. And it is classified as normal (**a**), hypoplastic (**b**), and aplastic (**c**), according to its size relative to the ipsilateral facial nerve. CN can be defined as "*aplastic*" if it cannot be seen in the IAC and "*hypoplastic*" if smaller than the facial nerve inside the IAC

It was accepted by many authors that cochlear nerve can be evaluated on axial and sagittal oblique T2-weighted MRI images and classified as normal, hypoplastic, and aplastic, according to its size relative to the facial nerve (Fig. 28.2a–c). CN can be defined as *aplastic* if it cannot be seen in the IAC and *hypoplastic* if smaller in diameter than the facial nerve inside the IAC.

Although the axial slices can demonstrate the size of the IAC and help to evaluate the course of the vestibulocochlear nerve, the sagittal oblique images obtained perpendicular to the long axis of the IAC are best to distinguish each of the individual components of the vestibulocochlear nerve as the nerves are visualized in cross-section [26].

The normal size of the cochlear nerve on MRI measures 1.8 ± 0.2 mm at the porus acusticus and 1.2 ± 0.2 mm in the mid to distal IAC [27].

In a normal sized IAC the diagnosis of cochlear nerve aplasia/hypoplasia is relatively straightforward with dedicated sagittal oblique high-resolution images [7] (Fig. 28.2a–c).

Preoperative measurement of the CA on temporal bone CT images may help to decide whether cochlear implantation will be beneficial or not [26]. In turn, CA stenosis can be used to select children who should undergo further evaluation using MRI. These measurements have important implications for clinicians who evaluate children with SNHL [28, 29]. In recent years there has been a reversal in this protocol because of the concerns regarding radiation with CT. MRI

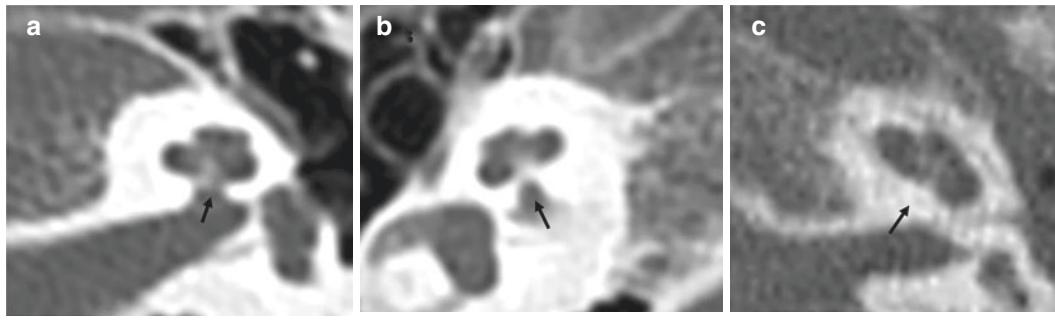


Fig. 28.3 Cochlear aperture. Axial high-resolution computed tomography images showing normal cochlear anatomy with normal cochlear aperture (**a**) isolated aperture stenosis and (**b**) isolated aperture atresia (**c**) (black arrows)

can be done as an initial investigation method and CT is reserved for cases planned to have surgery. If the audiological findings suggest a CI surgery, it is advisable to perform both modalities under general anesthesia together with ABR.

It is critical to recognize that there might be occasional discrepancy between the imaging and audiological findings regarding the presence/functionality of the cochlear nerve [29]. Several studies have shown that subsets of patients with cochlear nerve aplasia have positive audiological responses and might derive benefit from cochlear implantation [30–32]. Anatomical connections between the cochlear nerve and other branches of the vestibulocochlear complex that are below the resolution of the current MR imaging might be responsible for this radiological–audiological inconsistency.

As there is no single finding on imaging that could ascertain absence of a CN with certainty, cochlear nerve aplasia should be determined on a combination of findings from the MRI, CT, and audiological assessment. Indicator of the presence of a cochlear nerve.

28.5 Cochlear Aperture Abnormalities (Fig. 28.3a–c)

Normal CA*:* The mid-portion or mid-modiolar width of the CA is ≥1.5 mm (Fig. 28.3a). A normal CA can be seen together with other IEMs and also can be seen in otherwise normal cochlear morphology.

Stenotic CA*:* The mid-portion or mid-modiolar width of the CA is <1.5 mm (Fig. 28.3b). This abnormality can be seen together with other IEMs and also can be seen in otherwise normal cochlear morphology.

Atretic CA*:* The CA is not visible and it is occluded by bony tissue (Fig. 28.3c). In this case CA is not visualized as a continuation of the IAC. CN is aplastic in this situation. This abnormality can be seen together with other IEMs and also can be seen in otherwise normal cochlear morphology.

All of the above mentioned abnormalities may be isolated or may accompany another IEM. If these abnormalities do not accompany any other IEMs regarding the bony labyrinth they can be named as follows:

Isolated Cochlear Aperture Atresia*:* The CA is not visible and other cochlear structures are normal. Cochlea is normal in dimensions and shape. There is no other IEM such as incomplete partition or cochlear hypoplasia. Cochlear nerve is usually deficient. A narrow or atretic IAC can be determined.

Isolated Cochlear Aperture Stenosis*:* The mid-portion or mid-modiolar width of the CA is <1.5 mm. Cochlea is normal in dimensions and shape. There is no other inner ear malformation such as incomplete partition or cochlear hypoplasia. Cochlear nerve is usually deficient.

28.6 Audiological Findings

Physiologically, transient evoked otoacoustic emissions (TEOAEs) represent the response of the cochlea's external hair cells and can therefore

be identified in individuals with a normal cochlea but an abnormal cochlear nerve. During neonatal screening, these children may have otoacoustic emissions present in the ear with CA stenosis along with an absent auditory brainstem response confirming auditory neuropathy. In itself, a normal TEOAE does not guarantee normal hearing and a normal cochlear nerve so, even in healthy neonates, hearing screening should involve both TEOAE and automated ABR tests [16].

In 2006, Buchman et al. [33] screened 65 auditory neuropathy patients and noted an aplastic CN to be present in 18%. They also observed cochlear microphonics in 70% of ears with cochlear nerve aplasia, despite a negative ABR, thus proving that hair cells can function in the absence of a cochlear nerve.

28.7 Management

Papsin reviewed the results of CI in patients with cochlear and vestibular anomalies and reported poorer outcomes in those with narrow IAC and CA stenosis compared to other patients [34].

Both BCNC and IAC stenosis have been recommended as relative contraindications for cochlear implantation. However, stenosis of these structures does not always indicate cochlear nerve deficiency. MRI studies of the cochlear nerve have found that CA stenosis, as visualized on TBCT, is not necessarily an accurate indicator of nerve deficiency, and cochlear nerve deficiency can occur in ears with normal bony architecture [33]. Because of this fact, the decision between CI and auditory brainstem implantation (ABI) becomes more complicated. Sennaroglu et al. regarded CN deficiency as relative indication for an ABI [35, 36].

Bilateral complete labyrinthine aplasia, cochlear aplasia, aplastic internal auditory canal, absence of the CN, CA aplasia, or any combination of these malformations cause prelingual total hearing loss that is accepted as a definite indication for an ABI [35]. As hearing loss is prelingual in such patients, age of implantation and the decision making process for implantation surgery are critically important for a successful outcome. When a cochlear anomaly, or a stenotic BCNC and IAC, is detected via preoperative temporal bone CT, cochlear implantation should not be performed before establishing that the cochlear nerve is normal. Unfortunately, in the presence of a hypoplastic cochlear nerve, it is difficult to predict whether auditory-verbal abilities will develop sufficiently after cochlear implant surgery. If a patient does not benefit from a cochlear implant, then an ABI should be considered as a treatment option on the contralateral side. There are cases where CI and ABI can be performed together (for more detail about management of cochlear nerve deficiency please refer to Chap. 32).

As CA stenosis/atresia is seen as a part of other inner ear malformations, CI surgery might be complicated. In a recent study it was reported that facial nerve stimulation was correlated with a narrow CA and CN hypoplasia/aplasia. The underlying mechanism was thought to be the requirement of strong stimulating electric current levels compared to normal patients. Therefore, in CI for patients with a narrow CA or CN hypoplasia/aplasia, the possible occurrence of facial nerve stimulation should be considered, and the side of implantation or programing strategies should be selected carefully [35].

28.8 Cases

Case 1 SGG, 2-year-old female patient, operated January 2013.

She had bilateral cochlear hypoplasia type III with bilateral CA stenosis (Fig. 28.4a, b). On MRI there is no separate cochlear nerve on either side (Fig. 28.4c, d). She received an ABI on the right side and Fig. 28.4e demonstrates thresholds with ABI.

She had unilateral implantation in 2013. With our management strategy at present day, we would have planned CI or ABI surgery on the left side as well. This decision is done according to the presence of thresholds with insert earphones. In this particular patient, even though the nerve is absent on MRI, thresholds on the left side may indicate limited benefit from CI. CI and ABI combination

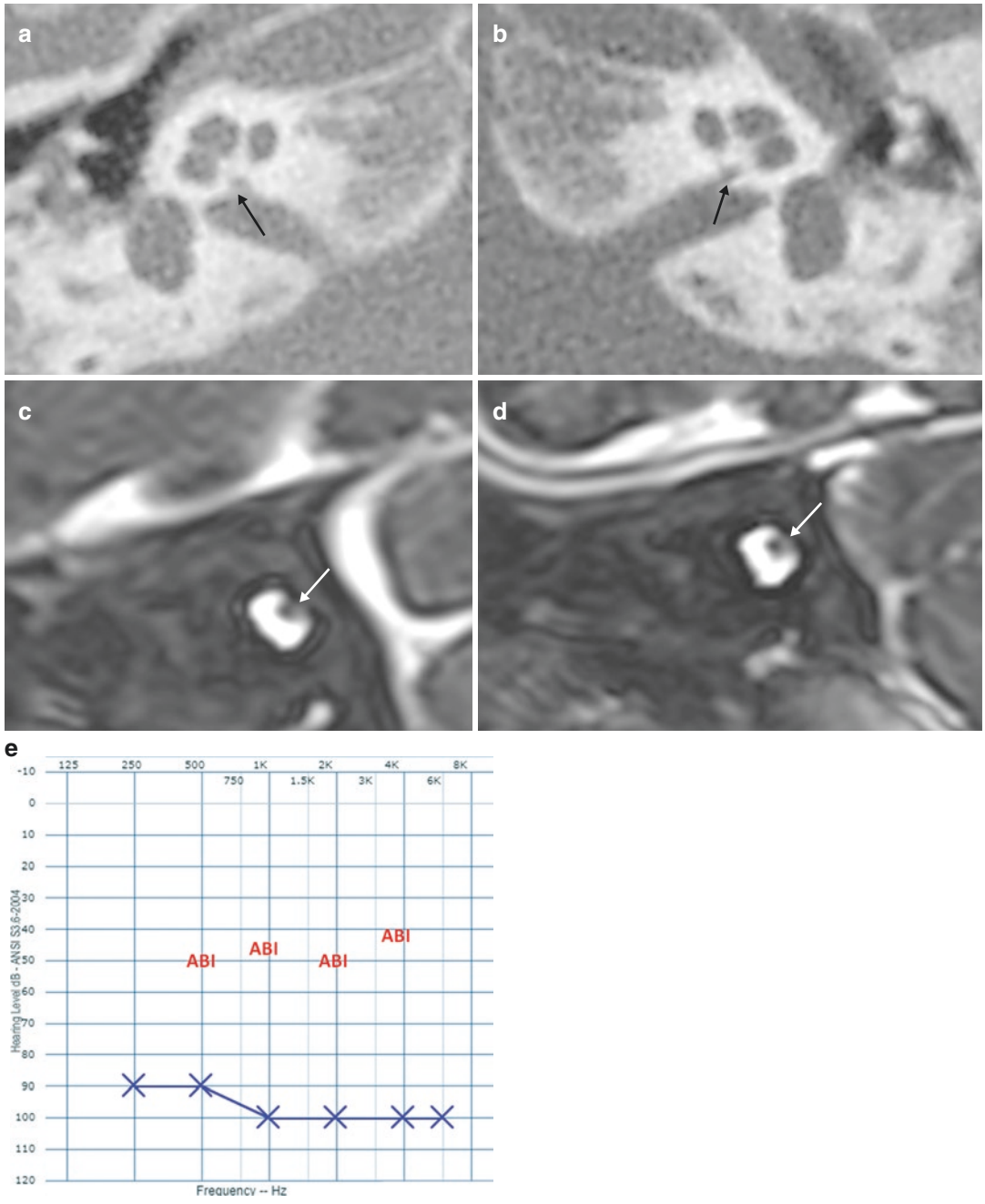


Fig. 28.4 Case 1. Temporal bone CT axial mid-modiolar section showing bilateral cochlear hypoplasia type III, bilateral CA stenosis (**a** and **b**) and sagittal oblique section perpendicular to internal auditory canal showing no separate CN on either side (**c** and **d**). Postoperative thresholds with ABI (**e**)

appears to be the best management option of these cases providing bilateral stimulation.

Case 2 MC, 15-year-old male patient, operated June 2015.

He had bilateral cochlear hypoplasia type III (CH-III), with bilateral CA stenosis (Fig. 28.5a, b); CN was present on the right side and absent on the left side (Fig. 28.5c). He received a CI on the right side. Figure 28.5d

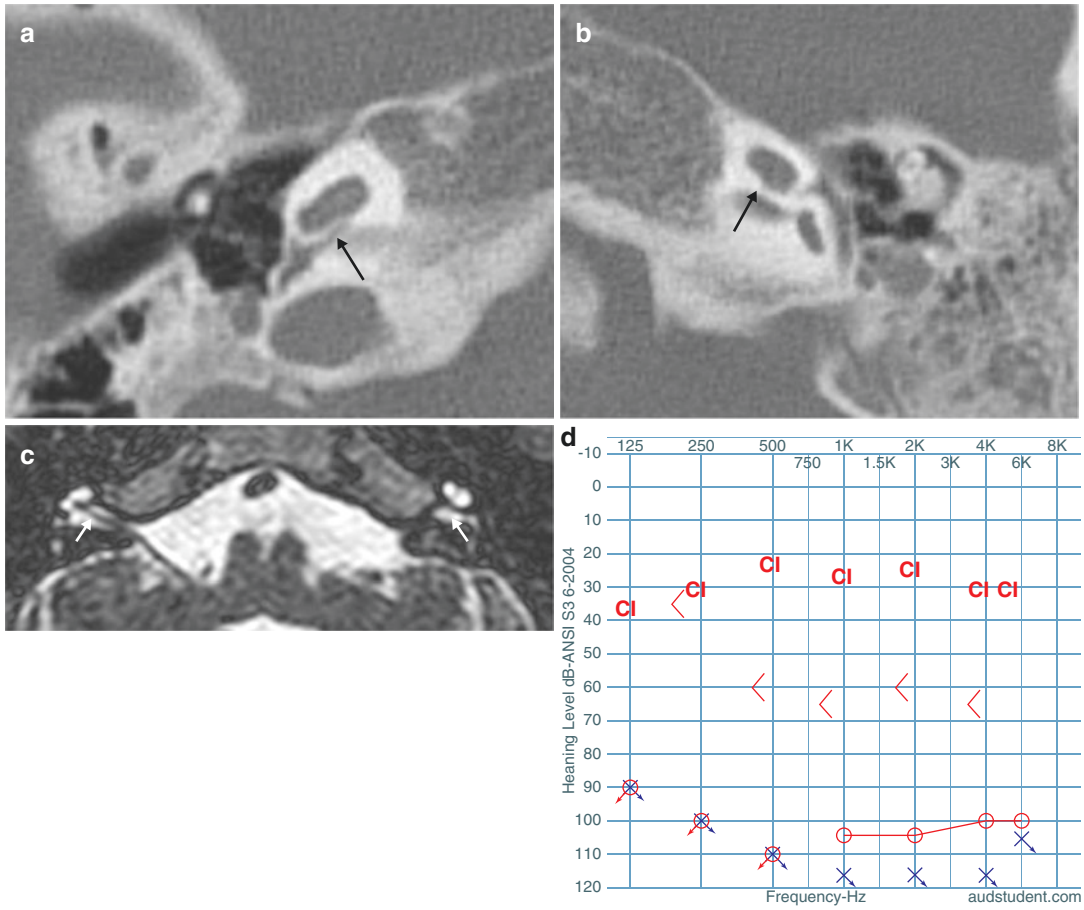


Fig. 28.5 Case 2. Temporal bone CT axial mid-modiolar section showing bilateral cochlear hypoplasia type III, bilateral CA stenosis (**a** and **b**), MRI section showing CN on the right side where left CN was aplastic (**c**). Preoperative and postoperative thresholds with CI on the right side (**d**)

demonstrates pre-op and post-op thresholds on the right side.

Because of his late presentation no surgery was done on the left side. According to our present day management strategy, if applied early, a similar very young candidate would receive a CI on the right and an ABI on the left, if they were evaluated between 1 and 2 years of age.

Case 3 CMC, 4-year-old male, operated in March 2011.

He had bilateral CA atresia (Fig. 28.6a, b) and bilateral CN hypoplasia (Fig. 28.6c, d). Preoperative subjective and objective audiological tests were performed. The auditory brainstem response (ABR) test and otoacoustic emissions (OAE) showed no responses. However, behavioral testing with insert earphones showed some auditory response at 250 Hz, 500 Hz, 1000 Hz, and 2000 Hz (90 dB, 110 dB, 115 dB, and 115 dB) (Fig. 28.6e). In addition, there was some awareness to speech noise at 85 dB bilaterally. These auditory responses exhibited by the patient indicated that there might have been some functioning auditory nerve fibers. Because of audiological findings, he underwent CI surgery on the right side. Hearing thresholds with CI for 250–6000 Hz were 30–55 dB HL. Before CI, his Meaningful Auditory Integration Scale (MAIS) score was 3/40; the score was obtained just for using the device, but he did not produce meaningful speech. Postoperatively his MAIS score was 32/40, his Sentence Recognition Test

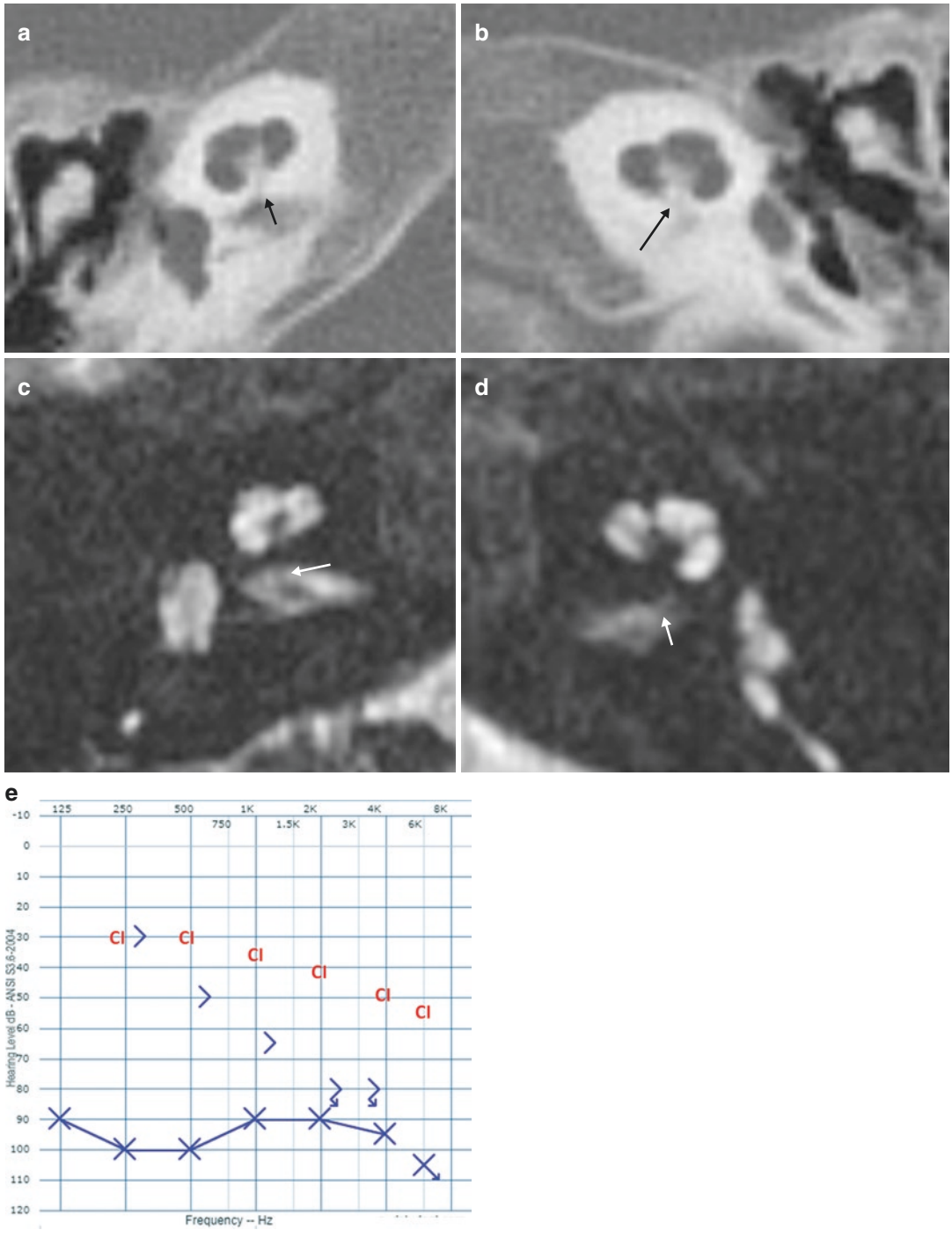


Fig. 28.6 Case 3. Bilateral CH-III with CA atresia (**a** and **b**) and bilateral CN hypoplasia (**c** and **d**). Postoperative thresholds with CI on the right side (**e**)

Auditory-Verbal Score was 97%, and the auditory only score was 85%. His CAP-II score was 8, FAPCI score was 88.6%, and PEACH score was 78.3%. The patient has been integrated into a mainstream primary school with satisfactory academic performance.

This patient shows us the importance of audiological evaluation in the decision making. Although CA was aplastic, because of thresholds with insert ear phones a decision for CI was made. This intervention resulted in satisfying speech and language development.

28.9 Auditory Rehabilitation

The outcome of children with hypoplastic CN with a cochlear implant is known to be poor, and it remains to be determined if using a combination of imaging and electrophysiological techniques can identify children with a present nerve to optimize their language outcome. It is likely that although the nerve was present, it was hypoplastic and not sufficient to provide the input required to support speech and language development.

As the cochlear nerve is less likely to be normal in the presence of CA atresia or stenosis, auditory rehabilitation and language development after cochlear implantation are possibly to fail in patients with undetermined cochlear nerve status.

Besides age at the time of CI and preoperative residual hearing, which were already known as prognostic factors for CI outcome, the anatomical factors (inner ear malformations, CA atresia/stenosis, CN deficiency) are related to post-CI outcome. It is shown that, in CI patients, a narrow CA on TBCT is strongly correlated with CN deficiency and poor CI outcomes and speech performance tests [21, 31]. These data suggest that CA width indirectly reflects the residual capability of the cochlear nerve. Hence, along with MRI of the IAC, TBCT may contribute to preoperative estimation of cochlear nerve residual functioning and it may be helpful in patient counseling.

Several case reports have included information on the speech performance of patients with CN deficiency. Recent reports of CI among children with CN deficiency have been reported with generally poor results. Buchman et al. [32] reported children with CN deficiency had higher pure tone averages and required greater charge for CI stimulation than other inner ear malformation types. In addition, open-set speech perception after CI was achieved in only 19% of CND cases and participating in mainstream education is more limited.

References

1. Henderson E, Wilkins A, Huang L, Kenna M, Gopen Q. Histopathologic investigation of the dimensions of the cochlear nerve canal in normal temporal bones. Int J Pediatr Otorhinolaryngol. 2011;75(4):464–7.
2. Jang J, et al. Implication of bony cochlear nerve canal on hearing in patients with congenital unilateral sensorineural hearing loss. Audiol Neurootol. 2012;17(5):282–9.
3. Stjernholm C, Muren C. Dimensions of the cochlear nerve canal: a radio- anatomic investigation. Acta Otolaryngol. 2002;122(1):43–8.
4. Sennaroglu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397–411.
5. Sennaroglu L. Histopathology of inner ear malformations: do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3–20.
6. Fatterpekar GM, Mukherji SK, Alley J, Lin Y, Castillo M. Hypoplasia of the bony canal for the cochlear nerve in patients with congenital sensorineural hearing loss: initial observations. Radiology. 2000;215(1):243–6.
7. Casselman JW, Offeciers EF, Govaerts PJ, et al. CT and MR imaging of congential abnormalities of the inner ear and internal auditory canal. Eur J Radiol. 2001;40(2):94–104.
8. Davidson HC. Imaging evaluation of sensorineural hearing loss. Semin Ultrasound CT MR. 2001;22(3):229–49.
9. Larsen WJ. Human embryology. 2nd ed. New York: Churchill Livingstone; 1997. p. 385–411.
10. McPhee JR, Van De Water TR. Epithelial-mesenchymal tissue interactions guiding otic capsule formation: the role of the otocyst. J Embryol Exp Morphol. 1986;97:1–24.
11. Adunka OF, Jewells V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. Otol Neurotol. 2007;28(5):597–604.
12. Shelton C, Luxford WM, Tonokawa LL, Lo WW, House WF. The narrow internal auditory canal in

children: a contraindication to cochlear implants. Otolaryngol Head Neck Surg. 1989;100:227–31.
13. Lefebvre PP, Leprince P, Weber T, Rigo JM, Delree P, Moonen G. Neuronotrophic effect of developing otic vesicle on cochleo-vestibular neurons: evidence for nerve growth factor involvement. Brain Res. 1990;507:254–60.
14. Lim CH, Lim JH, Kim D, Choi HS, Lee DH, Kim DK. Bony cochlear nerve canal stenosis in pediatric unilateral sensorineural hearing loss. Int J Pediatr Otorhinolaryngol. 2018;106:72–4.
15. Nakano A, Arimoto Y, Matsunaga T. Cochlear nerve deficiency and associated clinical features in patients with bilateral and unilateral hearing loss. Otol Neurotol. 2013;34(3):554–8.
16. Tahir E, Bajin MD, Atay G, Mocan BÖ, Sennaroğlu L. Bony cochlear nerve canal and internal auditory canal measures predict cochlear nerve status. J Laryngol Otol. 2017;131(8):676–83.
17. Kono T. Computed tomographic features of the bony canal of the cochlear nerve in pediatric patients with unilateral sensorineural hearing loss. Radiat Med. 2008;26(3):115–9.
18. Nelson EG, Hinojosa R. Aplasia of the cochlear nerve: a temporal bone study. Otol Neurotol. 2001;22(6):790–5.
19. Komatsubara S, Haruta A, Nagano Y, Kodama T. Evaluation of cochlear nerve imaging in severe congenital sensorineural hearing loss. ORL J Otorhinolaryngol Relat Spec. 2007;69(3):198–202.
20. Miyasaka M, Nosaka S, Morimoto N, Taiji H, Masaki H. CT and MR imaging for pediatric cochlear implantation: emphasis on the relationship between the cochlear nerve canal and the cochlear nerve. Pediatr Radiol. 2010;40(9):1509–16.
21. Yi JS, Lim HW, Kang PC, Park SY, Park HJ, Lee KS. Proportion of bony cochlear nerve canal anomalies in unilateral sensorineural hearing loss in children. Int J Pediatr Otorhinolaryngol. 2013;77(4):530–3.
22. Valero J, Blaser S, Papsin BC, James AL, Gordon KA. Electro-physiologic and behavioral outcomes of cochlear implantation in children with auditory nerve hypoplasia. Ear Hear. 2012;33(1):3–18.
23. Clemmens CS, Guidi J, Caroff A, et al. Unilateral cochlear nerve deficiency in children. Otolaryngol Head Neck Surg. 2013;149(2):318–25.
24. Purcell PL, Iwata AJ, Phillips GS, Paladin AM, Sie KC, Horn DL. Bony cochlear nerve canal stenosis and speech discrimination in pediatric unilateral hearing loss. Laryngoscope. 2015;125(7):1691–6.
25. Cinar BC, Batuk MO, Tahir E, Sennaroglu G, Sennaroglu L. Audiologic and radiologic findings in cochlear hypoplasia. Auris Nasus Larynx. 2017;44(6):655–63.
26. Noij KS, Remenschneider AK, Kozin ED, et al. Direct parasagittal magnetic resonance imaging of the internal auditory canal to determine cochlear or auditory brainstem implant candidacy in children. Laryngoscope. 2015;125(10):2382–5.
27. Nadol JB Jr, Xu WZ. Diameter of the cochlear nerve in deaf humans: implications for cochlear implantation. Ann Otol Rhinol Laryngol. 1992;101(12):988–93.
28. Chung J, Jang JH, Chang SO. Does the width of the bony cochlear nerve canal predict the outcomes of cochlear implantation? Biomed Res Int. 2018;21:5675848.
29. Teissier N, Van Den Abbeele T, Sebag G, Elmaleh-Berges M. Computed tomography measurements of the normal and the pathologic cochlea in children. Pediatr Radiol. 2010;40(3):275–83.
30. Peng KA, Kuan EC, Hagan S, Wilkinson EP, Miller ME. Cochlear nerve aplasia and hypoplasia: predictors of cochlear implant success. Otolaryngol Head Neck Surg. 2017;157(3):392–400.
31. Young NM, Kim FM, Ryan ME, Tournis E, Yaras S. Pediatric cochlear implantation of children with eighth nerve deficiency. Int J Pediatr Otorhinolaryngol. 2012;76(10):1442–8.
32. Acker T, Mathur NN, Savy L, Graham JM. Is there a functioning vestibulocochlear nerve? Cochlear implantation in a child with symmetrical auditory findings but asymmetric imaging. Int J Pediatr Otorhinolaryngol. 2001;57(2):171–17631.
33. Buchman CA, Roush PA, Teagle HF, Brown CJ, Zdanski CJ. Auditory neuropathy characteristics in children with cochlear nerve deficiency. Ear Hear. 2006;27(4):399–408.
34. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(1 Pt 2 Suppl 106):1–26.
35. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, Ramsden R, Sarac S, Freeman S, Andersen HR, Zarowski A, Ziyal I, Sollmann WP, Kaminsky J, Bejarano B, Atas A, Sennaroglu G, Yucel E, Sevinc S, Colletti L, Huarte A, Henderson L, Wesarg T, Konradsson K. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32:187–91.
36. Rah, et al. Facial nerve stimulation in the narrow BCNC. Laryngoscope. 2016;126(6):1433–9.

29 Current Indications and Long-Term Results of Auditory Brainstem Implantations in Children with Inner Ear and Cochlear Nerve Malformations

A. Zarowski, E. Offeciers, and Levent Sennaroglu

29.1 Introduction

Stimulation of the auditory pathways at the level of the cochlear nuclei in the brainstem is feasible and is able to provide auditory sensations allowing for open-set speech discrimination in selected patients. This was demonstrated by different publications reporting results of application of the auditory brainstem implant (ABI) in postlingually deafened (adult) patients with bilateral destruction of the auditory nerves due to bilateral tumors of the cerebellopontine angle, trauma, etc. [1–3]. Also patients with bilateral destruction of the labyrinth leading to total cochlear ossification excluding successful application of cochlear implants could benefit from ABIs.

Pioneering work of Colletti and Sennaroglu [4–6] demonstrated that ABIs are also able to provide auditory sensations, stimulate the development of the auditory pathways, and result in some degree of speech understanding in children born with severe malformations of the inner ear structures and/or the auditory nerves.

Application of ABI in small children remains however controversial since it requires major intracranial surgery for an elective and only functional indication. The last multi-center consensus meeting was organized in Kyrenia in 2013 in order to evaluate the functional benefits against the potential risks and to provide state-of-the art advice for treatment of this group of patients. This chapter summarizes the conclusions of this meeting.

29.2 Indications

Indications for ABIs in children are all conditions leading to bilateral profound hearing loss of peripheral type where application of cochlear implants (CIs) is impossible. The indications comprise two groups of pathologies. The first group are cochlear aplasias, dysplasias, or ossifications offering no possibility for placement of the stimulating electrode in the vicinity of the neuronal structures. The second comprises the cochlear nerve aplasias, hypoplasias, and cochlear nerve damages resulting in the absence of functional neuronal connection between the inner ear and the auditory nuclei in the brainstem.

All cases with total aplasia of the labyrinth, the cochlea, the cochlear aperture, and/or the cochlear nerve are definitive indications for ABI. Dysplastic cochlea's and/or hypoplastic cochlear nerves are potential indications that must be considered on individual basis. In these patients state-of-the-art audiological and radiological assessment (see further) is necessary, but

A. Zarowski (✉) · E. Offeciers
European Institute for Otorhinolaryngology, Antwerp, Belgium

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_29

will not always allow for a clear-cut decision. In these cases a trial application of a CI may be the optimal approach. Insufficient results obtained with CIs after 6–12 months of observation period would justify a switch to ABI.

29.3 Contraindications

Decision to exclude patients from the ABI option is complex and is based on pragmatic considerations. Children with multiple handicaps and especially with pronounced mental retardation will show significant problems during revalidation and their chances to achieve significant benefit from ABI are very low. In patients with minor disabilities, the surgical option should be favored in order to optimize the communication skills of the child. In patients with intermediate grade of handicap the indications should be considered on individual basis and preferably in a multidisciplinary setting involving specialists in ENT, neurology, neurosurgery, pediatrics, psychology, genetics, and speech and language therapy.

29.4 Timing of the Intervention

Development of the auditory pathways necessitates adequate sound and speech input that is present directly after birth and possibly even before. Delaying of this input beyond the critical period of development of the auditory pathways significantly decreases the chances for acquiring normal speech and language capabilities. Therefore, in children with bilateral profound deafness (independent of the causative factors), the age of adequate intervention is the major factor influencing the expected results of speech discrimination. From the developmental point of view it is obvious that the earlier the intervention the better, however the earliest age of intervention is also determined by the surgical and anesthesiologic risks as well as the timing, accuracy, and certainty of audiological evaluation, etc.

As of today it is considered that the optimal age for cochlear implantation (CI) in bilaterally deaf children is between 9 and 18 months with the upper limit estimated for 3 years. There is recent data showing that even earlier implantations from the age of 6 months [7] can provide additional benefit, but the limiting factor remains the accuracy of audiological diagnostics at this very young age. Children implanted after the age of 3 show significantly lower average long-term results of speech and language development than children implanted at earlier age.

The same rules apply for ABIs. However, due to an increased risks of intracranial surgery and more complicated diagnostic work-up, the age of intervention is usually later than in the case of CIs. Also in some cases application of ABI is preceded by a trial with a CI. In these cases, a CI should be implanted as early as possible (around the age of 9–12 months) in order to allow around 6–12 months' time for observation and evaluation of the results. In the case of insufficient benefit from CI the final decision for application of the ABI should be made as soon as possible and definitely not later than at the age of 3 years. Implantation at older age will result in unfavorable outcomes and the risk for discrediting this method of treatment.

29.5 Audiological and Electrophysiological Assessment

Audiological and electrophysiological assessment plays a very important role in defining the indications for ABI, in intraoperative confirmation of correct positioning of the electrode array and in optimization of the ABI fitting process.

1. Preoperative audiological and electrophysiological evaluation of the potential ABI candidates is, together with radiological assessment (see further), the basis for confirmation of the candidacy for ABI. The goal is to accurately measure the degree of the hearing loss and to define the site of lesion. In the preoperative setting both behavioral and objective tests are being used. Usually the diagnostics starts after referral from the universal newborn hearing screening. At first the normal status of

the middle ears has to be confirmed by otoscopy and tympanometry. Then the next tests performed are the objective acoustically evoked auditory brainstem responses (ABR) and transient or distortion-product otoacoustic emissions (OAE). Some centers perform also additional auditory steady state response audiometry (ASSR).

In deaf patients with hypoplastic auditory nerves demonstrated by imaging the question remains if intracochlear electrical stimulation with CI would be able to elicit adequate auditory sensations. In order to evaluate this, electrically evoked auditory brainstem responses (EABR) are being measured under general anesthesia. In the past a transtympanic promontorium needle was used as the stimulating electrode, currently round window ball or "golf club" electrodes are being used. In order to position such an electrode in the round window niche surgical access to the middle ear has to be created by elevation of the tympanomeatal flap. Some of the cochlear implant manufacturers deliver intracochlear stimulators with customized test electrodes allowing for performing the preoperative EABR evaluation. Improved quality of eABR registration can be achieved by using muscle relaxants during general anesthesia in order to decrease the influence of the myogenic potentials. If there is a positive eABR response, CI can be performed during the same operative session.

It has however to be remembered that objective measures have also their limitations related to the delivered stimulus intensity and may be misleading in cases of dysmaturation of the auditory pathways or in the radiologically diagnosed hypoplastic nerves. In these patients testing of the behavioral responses to acoustic stimuli (without and with hearing aids) is indispensable. Depending on the age of the child behavioral observation audiometry (BOA), conditioned orientation reflex (COR) or visual reinforcement audiometry (VRA) is performed. It can confirm functionality of the auditory pathways even if the objective measures show no responses. In these cases a trial with a CI might be a good option before taking the decision for ABI.

In adult patients it is also possible to register behavioral responses during a trial electrical stimulation at the round window under local anesthesia. Evaluation of the dynamic range of the auditory perceptions, the rate pitch discrimination, and the tone decay parameters allows for evaluation of the functional status of the auditory nerve and offers an alternative to EABR registration under general anesthesia.

2. Intraoperative electrophysiological testing is necessary to confirm correct positioning of the electrode array in the lateral foramen of the fourth ventricle. EABR is measured in monopolar or bi-polar configuration during stimulation with the ABI electrode. In this way each of the electrode contacts can be defined as giving purely auditive sensations or eliciting side effects coming from other cranial nerves (vestibular, facial, trigeminus, or mixed nerves). This evaluation allows for direct intraoperative adjustment of the electrode position and its contact with the cochlear nuclei. Optimization of electrode position is very important since the number of active electrode contacts generating auditory sensations correlates with the postoperative results. More than six active electrodes showing auditory stimulation are prognostic for a better outcome [2].
3. Postoperatively it is important to re-define the electrodes eliciting only acoustic sensations and the electrodes giving mixed percepts or only side effects. This is performed during the fitting sessions and the thresholds and comfort level stimulation amplitudes are adjusted accordingly. Electrodes giving predominantly side effects from other cranial nerves should be switched off. Due to safety reasons the first 1–2 fitting sessions should be performed with cardiac monitoring in a setting where immediate cardio-pulmonary resuscitation is possible (operation theater, intensive care unit).

The fitting parameters are also adjusted based on behavioral testing of the audiometric

thresholds and the results of speech discrimination (in older children).

Postoperative performance testing is especially important in children who first received a CI for a trial period. In these children accurate evaluation of the auditory performance is crucial for the decision to switch to an ABI. In order to test the efficiency of cochlear stimulation an auditory phoneme discrimination test may be extremely helpful. This test uses the COR paradigm and is feasible already from the age of 7–8 months. It is generally independent of the mother language and the cognitive strength of a child. Good phonemic discrimination with CI denominates good functioning of the CI and strongly correlates with good results on speech identification tests.

Among objective tests electrically evoked cortical responses (ECR) may be helpful for evaluation if the signal delivered by CI is sufficient to promote auditory maturation. Presence of a P1 waveform with decreasing latency at consecutive testing will indicate that adequate activation at cortical level is achieved by the CI [8, 9].

29.6 Radiological Evaluation

Precise radiological assessment of the status of the labyrinths and the cochlear nerves on both sides is crucial for defining indications for ABIs. In pediatric population the radiological examinations have to be performed under general anesthesia and therefore are advocated no earlier than at the age of 6 months.

The standard diagnostic tests comprise cone-beam CT scan (CBCT) and magnetic resonance imaging (MRI).

Current clinical CBCT devices present excellent resolution due to a voxel size of 3 × 75 μm. This allows for very precise evaluation of the type of labyrinthine malformations, the size of the cochlear aperture, and the diameter of the internal auditory meatus.

MRI is the best modality for evaluation of the cochlear nerves. Differentiation between aplasia, hypoplasia, and a normal size of the cochlear branch can be however very difficult and requires the highest possible resolution. When abnormality is seen or suspected on axial images, then the image acquisition should be performed in the direct parasagittal planes perpendicular on the nerves in the internal auditory canal and cerebellopontine angle. These direct images have a better resolution and are sharper than the images reconstructed from the standard axial acquisition.

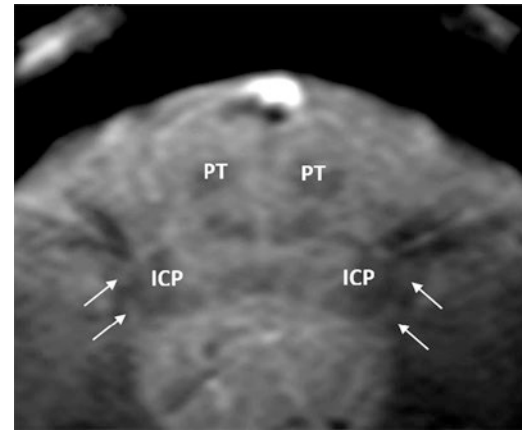


Fig. 29.1 MRI Multi-Echo Fast Field Echo (mFFE) sequences at the level of the cochlear nuclei in the brainstem. Arrows indicate the normal position of the ventral and dorsal cochlear nuclei in the brainstem. The nuclei are the gray oval structures projecting on the black background of myelinated fibers of the inferior cerebellar peduncles (ICP). *PT* pyramidal tracts

The condition for successful ABI application is the presence and normal localization of the ventral and dorsal cochlear nuclei in the brainstem. Nowadays it is possible to demonstrate presence of these nuclei by magnetic resonance imaging (MRI) using the Multi-Echo Fast Field Echo (mFFE) sequences (Fig. 29.1).

29.7 Surgical Technique

In majority of pediatric ABI cases the retrosigmoid approach is used. It is advisable to place the electrode fully into the lateral recess taking care that sufficient excess lead is left between the implant bed and the lateral recess. According to

Sennaroglu trimming the mesh around the electrode down to 1 mm around the electrode appears to be sufficient. Leaving the entire mesh around the electrode intact can make potential revision surgery impossible due to excessive fibrosis and total integration of the plate electrode into the surrounding neural tissue.

29.8 Expected Results

Based on the results from different centers it is possible to obtain a pure tone average (PTA) with an ABI between 30 and 60 dBHL in most of the implanted patients.

It has been observed that the majority of children obtain CAP scores between 4 and 5, but occasionally certain ABI users obtained scores of up to CAP 8 (Categories of Auditory Performance (CAP)-II, NEAP—Nottingham Early Assessment Package. The Ear Foundation 2009).

In a series of 35 children of Sennaroglu [10] majority of implanted children (80%) achieved scores above 50% with closed set pattern discrimination test. 30% of the children scored above 50% with open set sentence recognition test.

Conversely, audiometric thresholds between 30 and 60 dBHL are suboptimal and necessitate higher attention and concentration levels in order to hear and to recognize soft sounds. This can significantly decrease the chances for incidental learning in children with ABIs and be one of the factors resulting in longer time ABI users need to spontaneously respond to sounds in their environment comparing to the hearing aid or CI users.

ABI supports the development of language, but even after long-term ABI use, expressive and receptive language development lags behind the results observed in peers with normal hearing or with CIs. Therefore ABI users need intensive revalidation and it is advisable to stimulate the use of lip reading and cued speech in children with ABIs. If this is insufficient for adequate communication, use of sign language should also be considered.

In spite of the fact that the results achieved by pediatric patients with ABIs are on the average worse than the results of children with CIs, the majority of the patients use their device daily, with less than 10% device nonuser ratio. This suggests that these children achieve significant benefit from their ABIs.

It has also been agreed that important modifying factors influencing the obtained results are the cognitive development of the child, presence of multiple handicaps, the number of the electrode contacts eliciting auditory sensations, and the quality of ABI fitting and postoperative revalidation.

Success with an ABI decreases considerably in children with additional handicaps such as intellectual or cognitive deficits and visual impairment. These children are however even in greater need of hearing in order to establish communication with their environment. As already mentioned before in these cases indications should be defined on individual basis and best in a multidisciplinary setting. If the pathology is so severe that it would compromise the revalidation process of the child after ABI implantation, decisions should be made with great caution and consider the expected risk–benefit ratio.

29.9 Complications

ABI surgery should be performed in centers experienced in CI and neurotological surgeries. In these settings, the potential intraoperative complications are very infrequent in children. Intraoperative cerebellar swelling or postoperative leaks of cerebrospinal fluid are possible but rarely observed.

More frequent are the postoperative complications related to electrode malpositioning or migration that can be caused by a number of factors: insufficient securing of the electrode in the lateral recess of the fourth ventricle, torqued electrode, too tense or too short electrode lead causing dislocation out of the recess when the child grows. In these situations, ABI stimulation would no longer produce auditive sensations, but will result in side effects due to the stimulation of the nearby cranial nerves V, VII, IX, and X.

Electrode dislocation or device failure may require revision surgery in limited number of children. In certain patients the electrode can be easily tracked, dissected, and removed completely. In other instances, the electrode may be completely fixed to the brainstem and impossible to remove and replace.

29.10 Postoperative Follow-Up

Development of speech and language capabilities in children with ABIs is slower than in their peers with CIs. Therefore, well-designed and long-term follow-up is necessary in ABI children.

First of all, proper functioning of the device and the number of stimulating electrodes have to be regularly controlled in order to use the maximum electrode contacts providing different pitch sensations and to avoid occurrence of side effects. Regular measurement of the behavioral audiometric thresholds, phoneme discrimination and, at later age, speech discrimination should aid the revalidation program. Also data concerning the time of device use should be recorded.

Performance should be monitored using a standardized age-related set of tests and reported in a standardized way using the CAP scale allowing for comparison with the normal listening and CI peers. Other useful performance measures are standardized questionnaires allowing for collection of information about the child's auditory functioning from the family members and the teachers, e.g. Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), LittlEARS Auditory Questionnaire, etc.

29.11 Conclusions

ABIs are able to provide adequate auditory input in children with severe inner ear malformations and/or without any neural connection between the inner ear and the auditory nuclei in the brainstem. ABIs can help these children to develop speech and language, but the long-term average results of speech and language development are worse than in the case of CIs. Therefore, performing cerebellopontine angle surgery in small children for only elective functional indication remains controversial and requires careful weighing of the expected benefits against the surgical risks. Decision to apply ABI should be based on thorough audiological and radiological evaluation. In order to be able to obtain optimal results, application of ABI should be performed as soon as possible and not later than at the age of 3 years. In some doubtful cases a trial period with less invasive CI is advocated. In experienced centers the rate of complications is very low. Revision surgeries are possible, but sometimes very difficult and it might be impossible to remove and replace the ABI device.

Acknowledgement This chapter is based on the conclusions from the "Second Consensus Meeting on Management of Complex Inner Ear Malformations: Long-Term Results of ABI in Children and Decision Making Between CI and ABI" that took place on 5–6 April 2013 in Kyrenia, Northern Cyprus with the participation of 20 centers from 11 countries.

References

1. Portillo F, Nelson RA, Brackmann DE, Hitselberger WE, Shannon RV, Waring MD, Moore JK. Auditory brain stem implant: electrical stimulation of the human cochlear nucleus. Adv Otorhinolaryngol. 1993;48:248–52.
2. Nevison B, Laszig R, Sollmann WP, Lenarz T, Sterkers O, Ramsden R, Fraysse B, Manrique M, Rask-Andersen H, Garcia-Ibanez E, Colletti V, von Wallenberg E. Results from a European clinical investigation of the nucleus multichannel auditory brainstem implant. Ear Hear. 2002;23(3):170–83.
3. Colletti V, Carner M, Miorelli V, Colletti L, Guida M, Fiorino F. Auditory brainstem implant in post-traumatic cochlear nerve avulsion. Audiol Neurootol. 2004;9(4):247–55.
4. Colletti V, Fiorino F, Sacchetto L, Miorelli V, Carner M. Hearing habilitation with auditory brainstem implantation in two children with cochlear nerve aplasia. Int J Pediatr Otorhinolaryngol. 2001;60(2):99–111.
5. Sennaroglu L, Ziyal I, Atas A, Sennaroglu G, Yucel E, Sevinc S, Ekin MC, Sarac S, Atay G, Ozgen B, Ozcan OE, Belgin E, Colletti V, Turan E. Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. Otol Neurotol. 2009;30(6):708–15.

6. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, Ramsden R, Sarac S, Freeman S, Andersen HR, Zarowski A, Ziyal I, Sollmann WP, Kaminsky J, Bejarano B, Atas A, Sennaroglu G, Yucel E, Sevinc S, Colletti L, Huarte A, Henderson L, Wesarg T, Konradsson K. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.
7. Ching TYC, Dillon H, Button L, Seeto M, Van Buynder P, Marnane V, Cupples L, Leigh G. Age at intervention for permanent hearing loss and 5-year language outcomes. Pediatrics. 2017;140(3):e20164274. https://doi.org/10.1542/peds.2016-4274. Epub 2017 Aug 3.
8. Sharma A, Martin K, Roland P, Bauer P, Sweeney MH, Gilley P, Dorman M. P1 latency as a biomarker for central auditory development in children with hearing impairment. J Am Acad Audiol. 2005;16(8):564–73.
9. Gordon KA, Papsin BC, Harrison RV. Effects of cochlear implant use on the electrically evoked middle latency response in children. Hear Res. 2005;204(1–2):78–89.
10. Sennaroğlu L, Sennaroğlu G, Yücel E, Bilginer B, Atay G, Bajin MD, Mocan BÖ, Yaral M, Aslan F, Çnar BÇ, Özkan B, Batuk MÖ, Kirazl ÇE, Karakaya J, Ataş A, Saraç S, Ziyal İ. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.

30 Audiological Outcome with Cochlear Implantation

Hilal Burcu Ozkan, Betul Cicek Cinar, Gonca Sennaroglu, Esra Yücel, and Levent Sennaroglu

30.1 Introduction

Surgical and technological developments in CI technology have made a significant difference in the rehabilitation of hearing loss in individuals with inner ear malformations (IEMs). It is not surprising that there has been an increase in studies on the results of cochlear implantation in IEMs over the last decade. IEMs represent approximately 20–35% of the etiology of congenital sensorineural hearing loss (SNHL) cases based on radiology [1–4]. Incidence of IEMs was reported as 20% in our clinic [5].

There are certain challenges in the management of IEMs such as cerebrospinal fluid gusher, which is a risk for meningitis, facial nerve anomalies, decision making for the surgical approach and the type of electrode, choosing the correct implantation method; CI versus auditory brainstem implantation (ABI) and timing of surgery. It is very important to know about these possible risks for better rehabilitative counseling after surgery. Sennaroglu's classification correlates the surgical issues related to specific IEMs [5].

Classification of IEMs is based on differences in cochlear anatomy in various malformations. In addition to duration of deafness and preoperative auditory perception, IEMs have to be considered as an important limiting factor for successful CI outcomes. Children who qualify for and undergo CI surgery participate in follow-up testing at regular intervals for a period of 3 years.

30.2 Literature Review

Tucci et al. [6] reported CI outcomes in five children and one adult with IEMs. IEMs included common cavity (CC) deformity ($n = 1$), cochlear hypoplasia (CH) ($n = 2$), and incomplete partition (IP) ($n = 3$) anomalies. According to their results all patients showed improved performance after implantation. Four patients obtained open-set speech perception. Two remaining patients, whose poor language skills precluded administration of standard tests, showed increased awareness of environmental sounds and increased vocalization after implantation.

Luntz et al. [7] evaluated 10 CI users with IEMs: 3 CC deformity, 4 IP anomalies, 2 membranous deformity, and 1 enlarged vestibular aqueduct (EVA). Their study indicated that all 22 electrodes were inserted in 9 of 10 children. Each of the patients demonstrated speech awareness at 25 dB HL or better. Data was available after 30 months of experience in 4 of the 10 patients, and 3 (75%) of the 4 showed some degree of open-set word recognition. Six patients demon-

H. B. Ozkan (✉) · B. C. Cinar · G. Sennaroglu
E. Yücel
Audiology Department, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey

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

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_30

strated only speech detection and/or closed-set recognition scores.

Eisenman et al. [8] showed GASP-W scores of the 17 children with IEMs 12 months after implantation. Their results were worse, with slower rate of improvement than those of children with normal cochlea; however, by 24 months there was no significant difference between the two groups.

Buchman et al. [9] analyzed 28 pediatric CI users with the constellation of an IP, EVA, and dilatation of vestibule. Those with an isolated EVA or partial semicircular canal aplasia have relatively good levels of speech perception. Users with total semicircular canal aplasia, isolated IP, cochlear hypoplasia, or common cavity demonstrated lower levels of performance.

Papsin et al. [10] reported that children with IP obtained higher average speech perception outcomes because they were more likely to have a progressive hearing loss and, as a group, had superior linguistic skills before implantation. Children with CC deformity and CH demonstrated a tendency toward poorer performance despite inclusion in the data set of speech perception scores from children who were older and had significant language before implantation. Even the poorest performers with CH and CC showed speech perception gains with increased implant use.

Sainz et al. [11] reported word perception scores of CI users with common cavity and cochlear hypoplasia. They reported that these subjects demonstrated poor word perception and were unable to discriminate more than 50% of words and mostly relied on visual cues.

Isaiah et al. [12] illustrated that speech perception scores following cochlear implantation in children with IEMs were overall below than CI users with normal anatomy.

30.3 Results of Hacettepe University

Between November 1997 and September 2018, 2639 patients underwent CI and ABI in our department. Out of 2639 cases, 278 children have different IEMs, such as common cavity (CC), enlarged vestibular aqueduct (EVA), incomplete partition of the cochlea (IP-I, IP-II, IP-III), cochlear hypoplasia (CH), and dilatation of vestibule were implanted with CI. A retrospective study on auditory performance and language development of CI children with different IEMs was conducted in our clinic and these results were recently submitted for publication. In this chapter, auditory performance and language development of CI children with different IEMs were reported based on the results of this study. One hundred thirty-seven of 278 CI users were younger than 18 years old and had at least 1 year of cochlear implant experience.

All IEMs, aged between 12 months to 18 years, were matched with their peers in control group according to their chronological and implantation age (±8 months). All children are using their CI in daily basis. The distribution according to the number of IEMs and participants is shown in Table 30.1. These numbers were similar to those reported by a study in 2021 [13].

Depending on the wound healing, pediatric audiologists perform the initial activation of the electrodes between 3 days and 4 weeks after surgery. All children underwent hearing thresholds verification by audiometric testing after each CI programming session. Free field thresholds with CI were obtained at 0.25, 0.5, 1, 2, and 4 kHz using warble-tone or narrow-band stimuli and speech detection test was done through live voice using /ba/, /ss/, and /sh/ phonemes. Although some children get thresholds at 25–35 dB at 0.25, 0.5, 1, 2, and 4 kHz, others with limited benefit from CI get thresholds at 35–55 dB. Characteristic hearing thresholds of various IEMs and their free field tests are presented in Chapters 21 and 23–27.

As mentioned in Chap. 8, auditory perception skills were evaluated with a comprehensive test battery called as “Children’s Auditory Perception Skills Test in Turkish (CIAT)” [14]. All participants were evaluated before and after CI with Ling’s sound detection test, the Meaningful Auditory Integration Scale (MAIS) or Infant-

Table 30.1 Chronological age and duration of CI use of children with IEMs

	Common cavity		IP-I		IP-II		IP-III		EVA		Cochlear hypoplasia		Dilatation of vestibule	
	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)
Mean	96.3	32.8	111.8	42	115.6	45.3	95.6	32.8	140.5	60	103.6	48.3	101.1	30
Min.	40	16	42	18	31	12	39	12	60	12	87	42	24	12
Max.	177	60	216	88	216	143	215	93	182	144	123	55	216	106

Toddler Meaningful Auditory Integration Scale (IT-MAIS), and CIAT's subtests such as closed-set Pattern Perception Test and open-set Daily Turkish Sentence Recognition Test. All testing was done at 1–6 months after CI activation and follow-ups were done during 1–3 years with different intervals.

Language development skills were assessed using the Test of Early Language Development-Third Edition (TELD 3). This test provides us with receptive and expressive language performances of children [15]. However, this test was applied only to the group with IEMs and the results were not compared with the group of children with NC.

The speech perception and language development outcomes are presented according to the classification of IEMs as follows:

30.3.1 Common Cavity (CC)

We evaluated eight children with CC in terms of auditory perception performances and language skills (Figs. 30.1 and 30.8). In the Ling's Sound Test and MAIS evaluation, preoperative performance of children with CC had lower scores compared to children with NC and statistically significant differences were found ($p < 0.05$). After cochlear implantation, for auditory perception Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was a statistically significant difference between children with CC and NC in open-set and closed-set tests except Ling's Sound Test ($p < 0.05$). One to three years after CI, children with CC had a score of 25.38 points, while children with NC had a score of 40 full points in MAIS test. Similarly, closed-set Pattern Perception Test results were obtained 35.38% in children with CC and 82.63% in children with NC. The lowest score belongs to the Daily Turkish Sentence Recognition Test, which is an open-set 0% for CC and 46.13% for NC. Language skills assessments conducted with TELD-3 showed that the receptive language age of the children with CC is average 56.25 months, while the expressive language age is average 42.5 months. Children with CC obtained the lowest scores in terms of auditory perception performance among IEMs.

30.3.2 Cochlear Hypoplasia (CH)

We evaluated 26 children with CH in terms of auditory perception performances and language skills (Figs. 30.2 and 30.8). Before CI in the Ling's Sound Test, children with CH had lower scores than children with NC and statistically significant differences were found ($p < 0.05$). There was no statistically significant difference in MAIS test. One to three years after cochlear implantation, Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed for auditory perception. There was a statistically significant difference between children with CH and NC in open-set and closed-set tests except Ling's Sound Test ($p < 0.05$). In the MAIS test the children with CH had a score of 31.08 points, while the children with NC had a score of 38.73 points. Pattern Perception as closed-set test results showed that children with CH had lower scores (47.65%) than children with NC (85.12%). Similar to the results of CC, the lowest score belongs to the Daily Turkish Sentence Recognition Test (CH 5.96% and NC 45.15%). According to language skills assessments conducted with TELD-3, the receptive language age of the children with CH is average 58.34 months, while the expressive language age is average 41.53 months. Children with CH constituted the group of children with the second lowest scores in terms of auditory perception performance among IEMs.

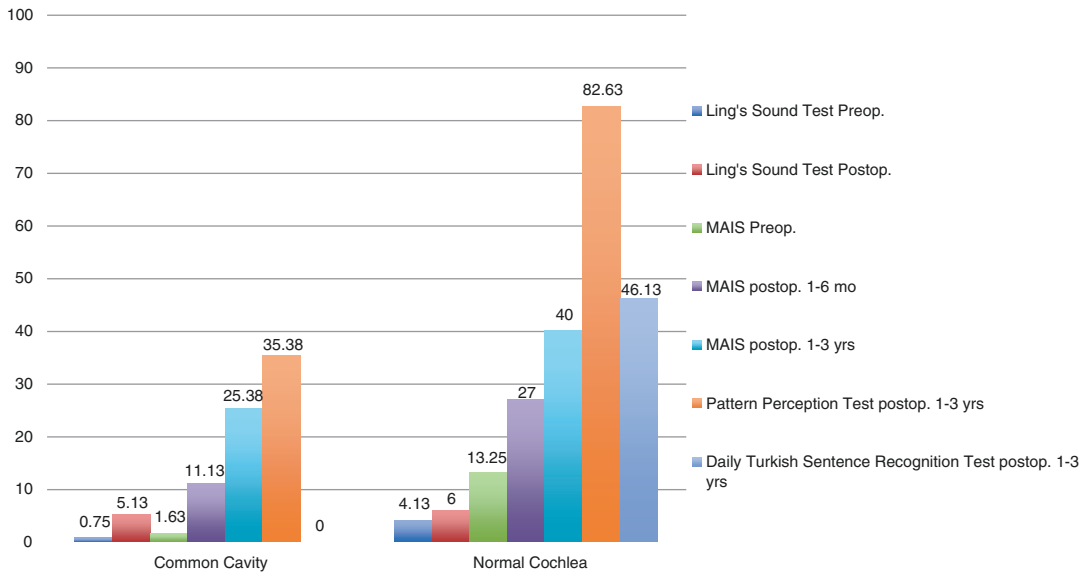


Fig. 30.1 Comparison of children with CC and NC in terms of auditory perception performance (CC = common cavity, NC = Normal cochlea)

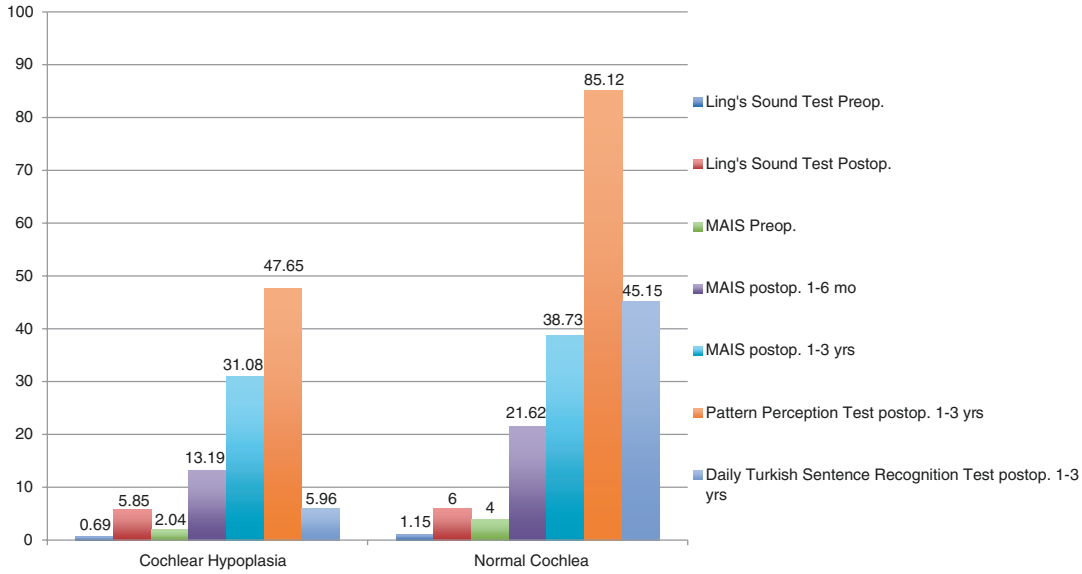


Fig. 30.2 Comparison of children with CH and NC in terms of auditory perception performance (CH = Cochlear hypoplasia, NC = Normal cochlea)

30.3.3 Incomplete Partition Anomalies of the Cochlea

30.3.3.1 Incomplete Partition Type I (IP-I)

We evaluated 36 children with IP-I in terms of auditory perception performances and language skills (Figs. 30.3 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-I had lower scores than children with NC and statistically significant differences were found ($p < 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were per-

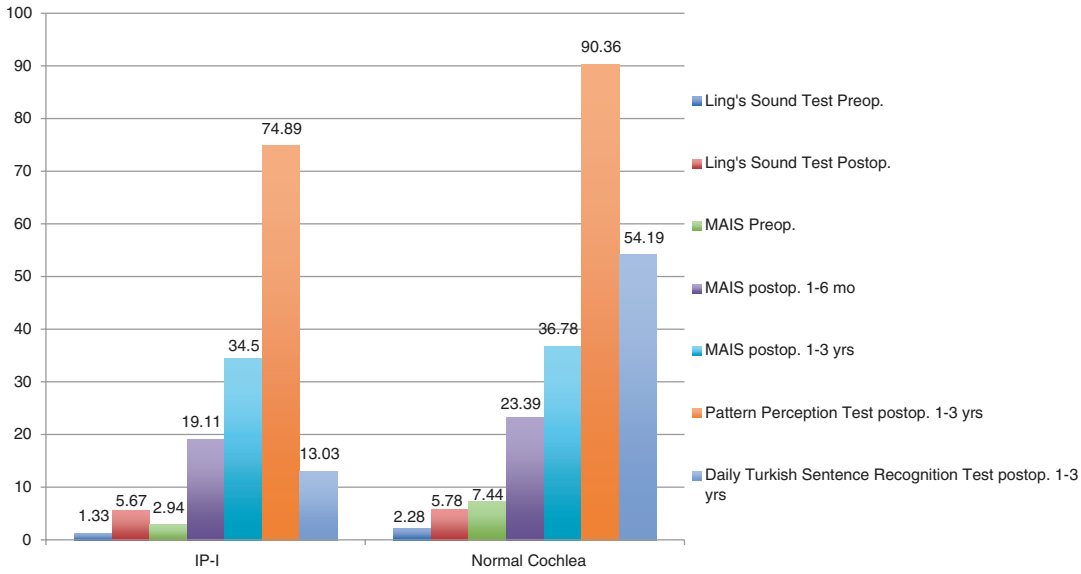


Fig. 30.3 Comparison of children with IP-I and NC in terms of auditory perception performance

formed. There was a statistically significant difference between children with IP-I and NC in Pattern Perception Test and Daily Turkish Sentence Recognition Test ($p < 0.05$). In the MAIS test between 1 and 3 years after CI, children with IP-I had a score of 34.5 points while the children with NC had a score of 36.78 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 90.36% and IP-I children had 74.89%. The lowest score belongs to the Daily Turkish Sentence Recognition Test (IP-I 13.03% and NC 54.19%). In addition to auditory perception evaluation, language skills assessments conducted with TELD-3, the receptive language age of the children with IP-I is average 67.61 months, and while the expressive language age is average 50 months. Children with IP-I constituted the group of children with the third lowest scores in terms of auditory perception performance among IEMs.

30.3.3.2 Incomplete Partition Type II (IP-II)

We evaluated 40 children with IP-II in terms of auditory perception performances and language skills (Figs. 30.4 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-II obtained similar scores to children with NC. After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with IP-II and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, children with IP-II had a score of 37.98 points while the children with NC had a score of 38.28 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 88.5% and children with IP-II had 87.75%. The Daily Turkish Sentence Recognition Test showed close performance between the two groups (IP-II 48.48% and NC 50.05%). In addition to language skills assessments conducted with TELD-3, the receptive language age of the children with IP-II is 83 months, while the expressive language age is average 78 months. Children with IP-II completed all test items as the receptive language age. Children with IP-II constituted the group of children with the second highest scores in terms of auditory perception performance among IEMs.

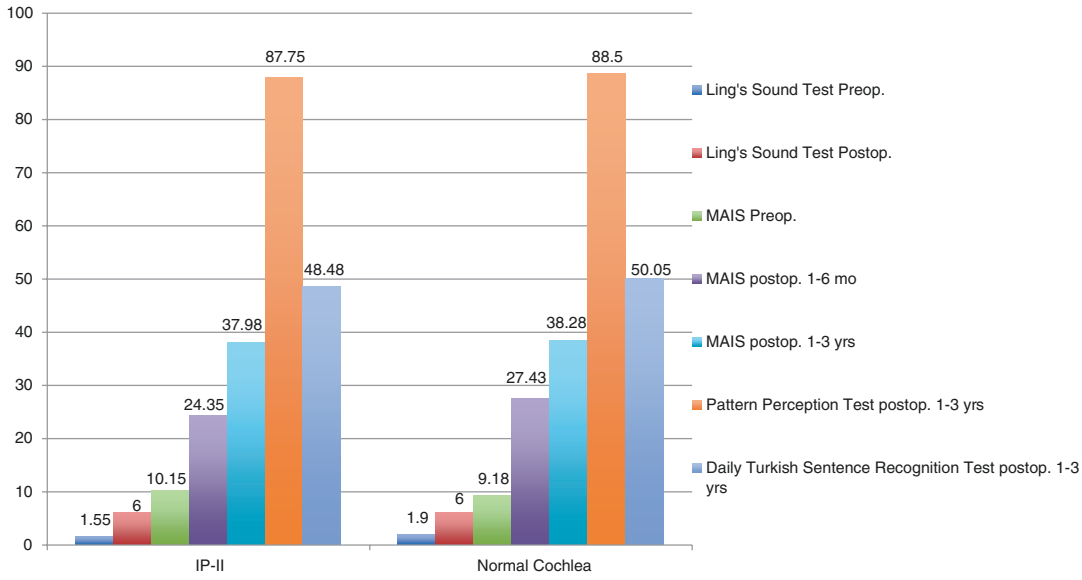


Fig. 30.4 Comparison of children with IP-II and NC in terms of auditory perception performance (IP-II = incomplete partition Type II, NC = Normal cochlea)

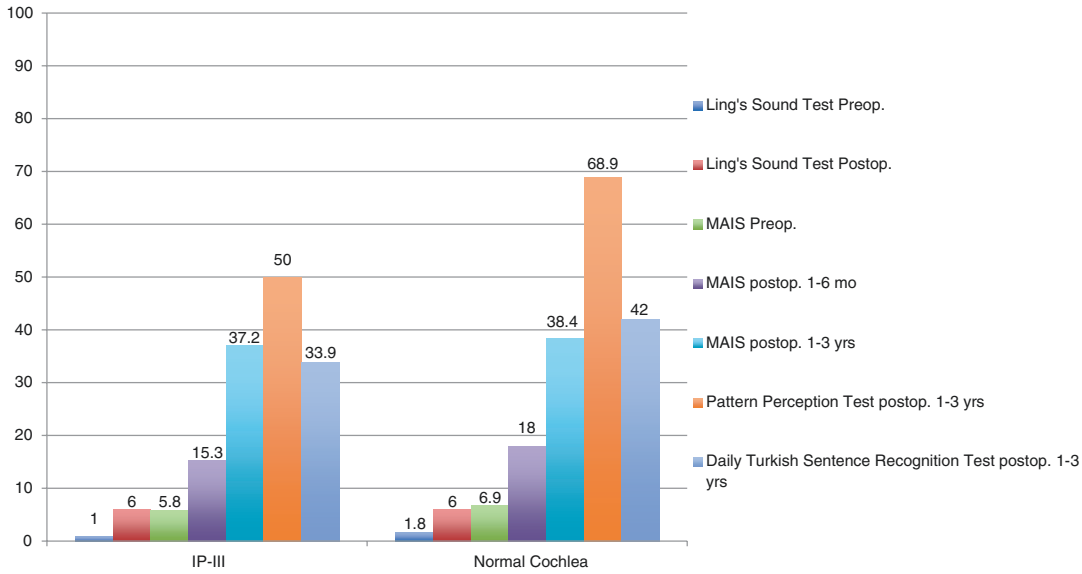


Fig. 30.5 Comparison of children with IP-III and NC in terms of auditory perception performance (IP-III = Incomplete Partition type III, NC = Normal cochlea)

30.3.3.3 Incomplete Partition Type III (IP-III)

We evaluated ten children with IP-III in terms of auditory perception performance and language skills (Figs. 30.5 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-III had scores close to children with NC and there was no statistically significant difference ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with IP-III and NC in open-set and closed-

set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with IP-III had a score of 37.2 points, while the children with NC had a score of 38.4 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 68.9% and children with IP-III had 50%. The Daily Turkish Sentence Recognition Test showed close performances between the two groups (IP-III 33.9% and NC 46.13%). Additionally, language skills assessments conducted with TELD-3, the receptive language age of the children with IP-III is average 68.9 months, while the expressive language age is average 54 months. Children with IP-III constituted the group of children with the fourth lowest scores in terms of auditory perception performance among IEMs.

30.3.4 Dilatation of Vestibule

We evaluated three children with dilatation of vestibule in terms of auditory perception performance and language skills (Figs. 30.6 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with dilatation of vestibule had the scores close to children with NC and statistically significant differences were not found ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with dilatation of vestibule and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with dilatation of vestibule had a score of 39.33 points, while the children with NC had a score of 40 full points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 93.33% and children with dilatation of vestibule had 56.67%. The Daily Turkish Sentence Recognition Test showed no statistically significant difference between the two groups (dilatation of vestibule 13.33% and NC 57%) but we found that children with dilatation of vestibule had difficulty in this open-set test. According to language skills assessments conducted with TELD-3, the receptive language age of the children with dilatation of vestibule is average 71.66 months, while the expressive language age is average 58 months. Children with dilatation of vestibule constituted the group of children with the third highest scores in terms of auditory perception performance among IEMs.

30.3.5 Enlarged Vestibular Aqueduct (EVA)

We evaluated 14 children with EVA in terms of auditory perception performance and language skills (Figs. 30.7 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with EVA obtained scores very close to children with NC and statistically significant differences were not found ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with EVA and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with EVA had a score of 35 points while the children with NC had a score of 38.57 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 82.64% and children with EVA had 89.71%. The Daily Turkish Sentence Recognition Test showed close performances between the two groups (EVA 79.64% and NC 87.29%). According to language skills assessments conducted with TELD-3, the receptive language age of the children with EVA is 83 months, while the expressive language age is average 83 months. Children with EVA completed all test items as the receptive and expressive language age. Children with EVA obtained the highest scores in terms of auditory perception performance among IEMs.

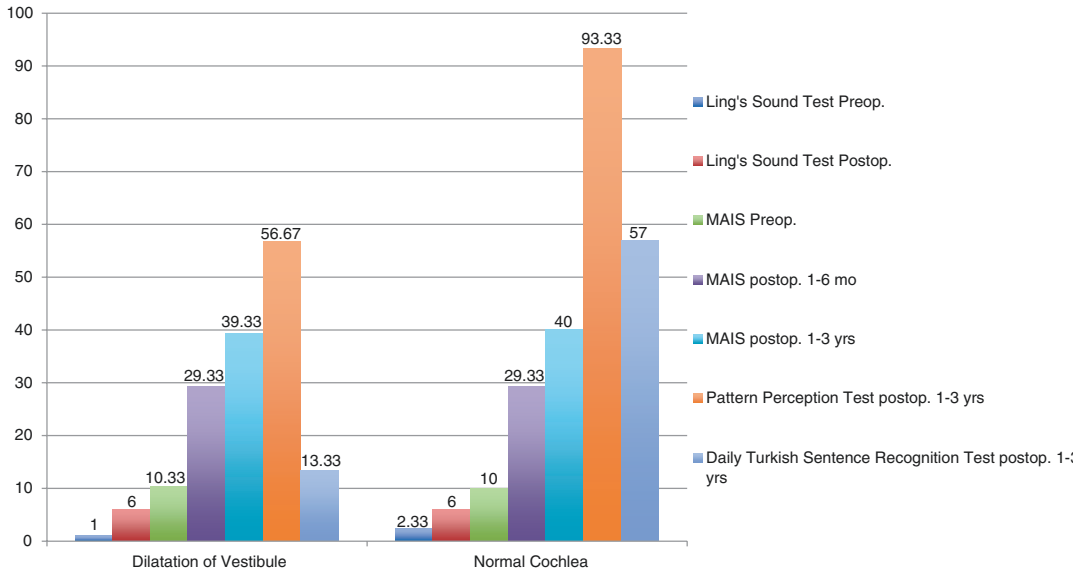


Fig. 30.6 Comparison of children with dilatation of vestibule and NC in terms of auditory perception performance (NC = Normal cochlea)

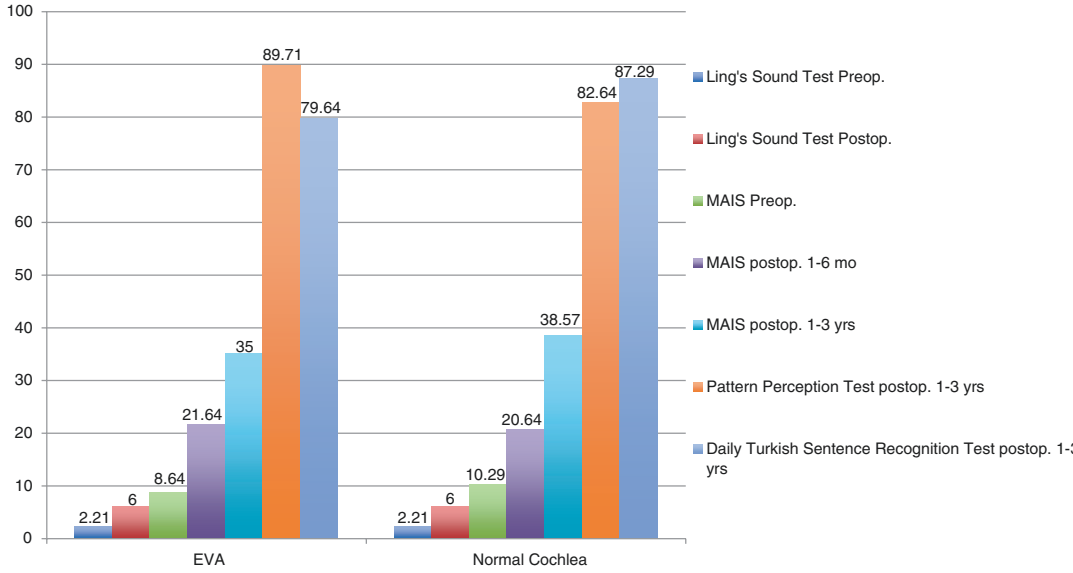


Fig. 30.7 Comparison of children with EVA and NC in terms of auditory perception performance (EVA = Enlarged vestibular aqueduct, NC = Normal cochlea)

Our data showed that children with IEMs might receive considerable benefit from CI. Taking into consideration variation in children with IEMs using CI, it is very difficult to establish outcomes. Nevertheless, with other studies, our results demonstrated that CI is a successful treatment modality in deaf children with and without IEMs. Depending on the subgroup of IEMs, their outcomes also showed variability. Additionally, this variation can also be caused by several other factors, such as age of implantation, preoperative residual and functional hearing, cognitive skills, parental and environmental support.

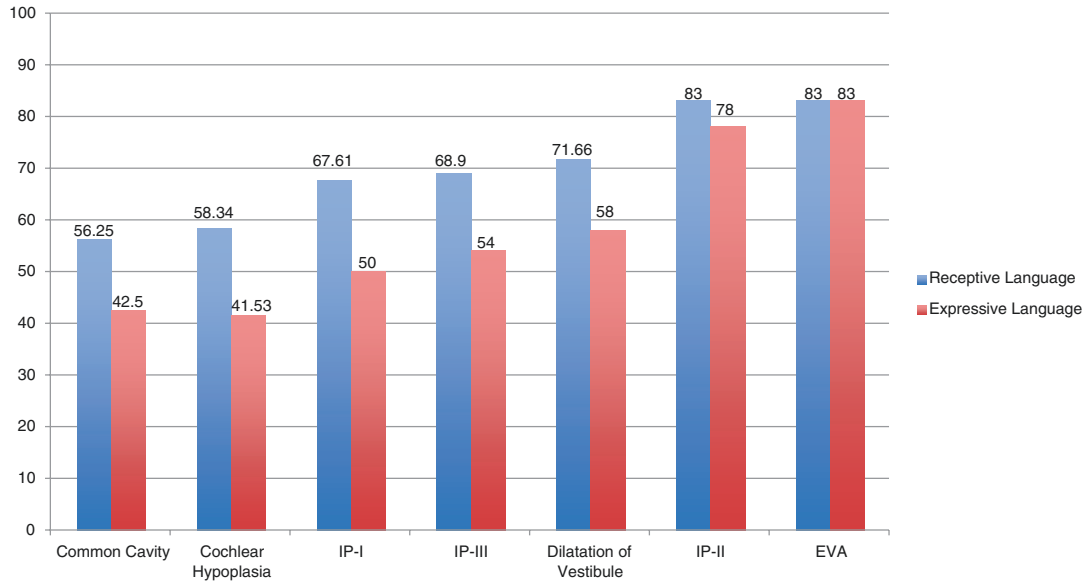


Fig. 30.8 Results of TELD-3 in children with all IEMs

30.4 Hacettepe Experience of Cochlear Implantation in Children with IEMs

Auditory perception performance in speech sounds and closed-set pattern perception assessed with CIAT test battery. All children with or without IEMs detected Ling 6 sounds in various frequencies. However, speech sounds, which are, used in this task only represent frequencies in wide ranges in speech frequency bands. Considering the results of speech perception tasks, children with all kinds of IEMs showed varying degrees of auditory benefit by the end of 3 years of CI experience.

Speech perception and language development performance variations between children with IEMs are summarized below:

30.4.1 Common Cavity (CC)

Children with CC had the lowest scores in terms of auditory perception performance among IEMs. Although they used their CIs regularly in daily basis, it was found that they develop identification and comprehension of environmental sounds 1 year after but not developed this proficiency for speech sounds.

One child with CC who could not perform any speech perception skills had only four active electrodes and had facial stimulation as side effect. He underwent ABI for his contralateral ear after 1 year of cochlear implant experience. He performed significantly better with his contralateral ABI and rejected to use CI.

Functionality of auditory perception performances was mostly evaluated with MAIS in clinical studies. The results indicated an obvious delay in children with CC; however, their performance changes with time. Unfortunately, children with CC are unable to reach the full MAIS score even 1–3 years. It is necessary to follow children with CC more closely and to study in this direction in terms of the functionality of auditory perception performances.

Children with CC showed better performance with regard to a closed-set situation, but their vocabulary was weak; they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain

conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [16]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Unfortunately, open-set sentence recognition is not improved in children with CC between 1 and 3 years.

When the language skills of children with CC were analyzed, these children have a language development of about 4.5 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be more difficult [17].

30.4.2 Cochlear Hypoplasia (CH)

Children with CH constituted the group of children with the second lowest scores in terms of auditory perception performance among IEMs.

Children with CH used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

Functionality of auditory perception performances was mostly evaluated with MAIS in researches. The results indicated an obvious delay with CH children; however, their performance changes with time. Unfortunately, children with CH are unable to reach the full MAIS score even 1–3 years, but it has been found that children with CH can use auditory perception more functionally than children with CC. Also, similar results show it is necessary to follow children with CH more closely and to study in this direction in terms of the functionality of auditory perception performances.

Children with CH showed better performance with regard to a closed-set situation, but their vocabulary was weak; they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development. It was also found that children with CH could use closed-set pattern perception skills better than children with CC.

Open-set sentence recognition is improved in children with CH between 1 and 3 years. This is a better result compared to children with CC, even though only some children with CH have already open-set sentences recognition.

Analysis of the language skills of children with CH showed that they have a language development of about 4.8 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI use is more significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.3 Incomplete Partition Anomalies of The Cochlea

30.4.3.1 Incomplete Partition Type I (IP-I)

Children with IP-I constituted the third lowest scores in terms of auditory perception performance among IEMs. They used their CI regularly on a daily basis, and they were found to

develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

The results of MAIS indicated obvious delay children with IP-I; however, their performance changes with time. Unfortunately, children with IP-I are unable to reach the full MAIS score even 1–3 years, but it has been found that children with IP-I can use auditory perception more functionally than children with CC and CH. Also, similar results show it is necessary to follow children with IP-I more intensely and to study in this direction in terms of the functionality of auditory perception performances.

Children with IP-I obtained better performance with regard to a closed-set situation, also their vocabulary was not weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development. Also it has been found that IP-I children can use closed-set pattern perception skills better than children with CC and CH.

Open-set sentence recognition is improved in children with IP-I between 1 and 3 years. This is a better result compared to children with CH, even though only some children with IP-I have already open-set sentences recognition.

When analyzed at the language skills of children with IP-I that these children have a language development of about 5.5 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.3.2 Incomplete Partition Type II (IP-II)

Children with IP-II obtained the second highest scores in terms of auditory perception performance among IEMs.

Children with IP-II used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1–3 months.

MAIS results indicated better performance children with IP-II; however, their performance changes with time. Children with IP-II are close to reach the full MAIS score 1–3 years, and it has been found that children with IP-II can use auditory perception more functionally than children with IP-I and IP-III.

Children with IP-II demonstrated better performance with regard to a closed-set situation, also their vocabulary was strong.

Open-set sentence recognition showed improvement in children with IP-II between 1 and 3 years. Most of the children with IP-II can repeat sentences correctly.

When the language skills of children with IP-II were analyzed, their receptive language was found to be same as their chronological age.

30.4.3.3 Incomplete Partition Type III (IP-III)

Children with IP-III constituted the fourth lowest scores in terms of auditory perception performance among IEMs.

Children with IP-III used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

MAIS results indicated obvious delay children with IP-III; however, their performance changed with time. Children with IP-III are close to reach the full MAIS score 1–3 years, and it has been found that children with IP-III can use auditory perception more functionally than children with IP-I.

Children with IP-III did not show a good performance with regard to a closed-set situation. In addition, their vocabulary was weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Open-set sentence recognition has improved in children with IP-III between 1 and 3 years. This is a better result compared to children with IP-I; more children with IP-III have already open-set sentence recognition.

Children with IP-III have a language development of about 5.7 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is more significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.4 Dilatation of Vestibule

Children with dilatation of vestibule constituted the third highest scores in terms of auditory perception performance among IEMs.

Children with dilatation of vestibule used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 3–6 months.

MAIS results indicated better performance children with dilatation of vestibule; however, their performance changes with time. Children with dilatation of vestibule are close to reach the full MAIS score 1–3 years.

Their performance with regard to a closed-set situation was not good. In addition their vocabulary was weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Open-set sentence recognition is improved in children with dilatation of vestibule between 1 and 3 years.

Children with dilatation of vestibule have a language development of about 6 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.5 Enlarged Vestibular Aqueduct (EVA)

EVA is the only group that Pattern Perception Test and Turkish Daily Sentence test results were better than NC group. SNHL since birth EVA is a congenital anomaly with progressive SNHL. At the beginning their hearing may be normal. The process is progressive. Final outcomes are better than children with NC.

EVA–IP-II difference is modiolar defect in IP-II. In general outcome of IP-II is almost similar to NC. EVA is better than IP-II because there

is no modiolar defect in EVA, which is the reason to make the outcome slight worse.

Children with EVA had a higher score than children with NC in the closed-set test. In these children, we can say that the children with EVA perform better than children with NC. It is thought that the reason for this is that some of the children with EVA may have been suffering from hearing loss in the peri- or post-lingual period rather than the prelingual period.

Children with IP-II and EVA scores reached implanted children with NC but children with CC, CH, and IP-I improved slower. Preoperative counseling for the parents is advised in order to explain the possible impact of the diagnosed disabilities on performance and habilitation. Nevertheless, factors influencing the success of implantation are multiple, including a thorough preoperative radiological examination, a well-performed surgery, and an individually tailored postoperative rehabilitation program.

30.5 Summary

Our major goal is to provide meaningful sound information through CI in children with IEMs who have severe to profound hearing loss. Because of the complexity of different subgroups, it is not possible to explain the performance of children with IEMs under a single group. The critical point is that the degree of malformation should be taken into consideration while evaluating functional hearing with implantation. The readiness of the children in auditory perception, language skills cognitive, psychosocial, and similar areas should be evaluated comprehensively.

According to our results, in terms of auditory perception and language skills, the children with IEMs can be arranged from poor to good performance as **CC, CH, IP-I, IP-III, Vestibular Dilatation, IP-II, and EVA**.

Children with CC obtained the lowest scores among children with IEMs. All children in this group make benefit from CI almost after 3 years CI usage for auditory perception. By evaluating these low scores, we have determined that children with CC cannot bring their auditory perception and language skills to the level of children with NC at the end of the third year. Therefore, if we do not see an acceptable progress in these children, our team proposes an ABI on their contralateral ears.

Children with CH and IP-I are the second and third groups that obtained the least benefit from CI, respectively. However, these children showed heterogeneous scores in terms of auditory perception and language development. Although several children with CH and IP-I achieved closer scores to CI children with NC, some others were able to show improved performance after 3 years of CI use. Children with CH and IP-I definitely need bilateral implantation. As we know, children with CH and IP-I can be a candidate for CI or ABI. When CI was applied to one ear, according to their benefit from CI in that ear and cochlear nerve status on the contralateral ear (demonstrated with MRI), these children could also be an ABI candidate on the contralateral side. If there is well-developed cochlear nerve, bilateral CI should be done. In case of CN deficiency, contralateral ABI is advisable. In case of bilateral CH-I, which is the least developed type of CH, contralateral ABI can be proposed.

Children with IP-III and dilatation of vestibule showed poor performance in open-set scores. Consequently, in the earliest period, bilateral CI should be recommended to this group. The risk of gusher in IP-III is 100%; therefore, bilateral CI should be staged in IP-III. In vestibular dilatation bilateral CI can be done simultaneously or staged. IP-III cases do not need an ABI as they have well-developed cochlear nerves.

It was found that the open-set, closed-set, and language scores of children with IP-II and EVA were similar to children with NC. Therefore, children with IP-II and EVA were able to obtain good results with unilateral CI. Bilateral CI is also proposed to these groups to obtain the advantages of bilateral hearing.

Children with IEMs should be involved in intensive rehabilitation and follow-up at regular intervals. The open-set, closed-set, language, and speech production of children in this group should be examined one by one. The alternative

surgical method should be considered when there is a failure in these skills.

References

1. Chadha NK, James AL, Gordon KA, Blaser S, Papsin BC. Bilateral cochlear implantation in children with anomalous cochleovestibular anatomy. Arch Otolaryngol Head Neck Surg. 2009;135:903–9.
2. Ha JF, Wood B, Krishnaswamy J, Rajan GP. Incomplete cochlear partition type II variants as an indicator of congenital partial deafness: a first report. Otol Neurotol. 2012;33:957–62.
3. McClay JE, Tandy R, Grundfast K, Choi S, Vezina G, Zalzal G, et al. Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 2002;128:664–71.
4. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11:4–41.
5. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397.
6. Tucci DT, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations. Arch Otolaryngol Head Neck Surg. 1995;121:833–8.
7. Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformation. Arch Otolaryngol Head Neck Surg. 1997;123:974–7.
8. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. Otol Neurotol. 2001;22(6):834–41.
9. Buchman CA, Copeland BJ, Yu KK, Brown CJ, Carrasco VN, Pillsbury HC III. Cochlear implantation in children with congenital inner ear malformations. Laryngoscope. 2004;114(2):309–16.
10. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(S106):1–26.
11. Sainz M, Garcia-Valdecasas J, Fernandez E, Pascual MT, Roda O. Auditory maturity and hearing performance in inner ear malformations: a histological and electrical stimulation approach. Eur Arch Otorhinolaryngol. 2012;269(6):1583–7.
12. Isaiah A, Lee D, Lenes-Voit F, Sweeney M, Kutz W, Isaacson B, Lee KH. Clinical outcomes following cochlear implantation in children with inner ear anomalies. Int J Pediatr Otorhinolaryngol. 2017;93:1–6.
13. Ozkan HB, Cicek Cinar B, Yucel E, Sennaroglu G, Sennaroglu L. Audiological performance in children with inner ear malformations before and after cochlear implantation: a cohort study of 274 patients. Clin Otolaryngol. 2021;46(1):154–60.
14. Yücel E, Sennaroglu G. Çocuklar İçin İşitsel Algı Testi. Advanced Bionics. 2011.
15. Topbaş S, Güven OS. Reliability and validity results of the adaptation of TELD-3 for Turkish speaking children: implications for language impairments. In: Oral presentation, 12th Congress of the International Clinical Phonetics and Linguistics Association, Istanbul, Turkey;2008.
16. Yücel E, Aslan F, Özkan HB, Sennaroglu L. Recent rehabilitation experience with pediatric ABI users. J Int Adv Otol. 2015;11(2):110.
17. Merkus P, Di Lella F, Di Trapani G, Pasanisi E, Beltrame MA, Zanetti D, et al. Indications and contraindications of auditory brainstem implants: systematic review and illustrative cases. Eur Arch Otorhinolaryngol. 2014;271:3–13.

Audiological Outcome with ABI

31

Filiz Aslan, Esra Yücel, Gonca Sennaroglu, Mehmet Yaralı, and Levent Sennaroglu

31.1 Introduction

Cochlear implants (CI) have been widely used to provide auditory sensation in the profound sensorineural hearing loss. Despite their known effect on auditory perception, speech, and language development; in some cases (such as severe inner ear malformations (IEMs) and cochlear nerve deficiencies) CI has minimal or no effect on speech and language perception. At that point, auditory brainstem implants (ABI) can be considered for hearing restoration. In 2001, Colletti et al. reported the results of pediatric ABI patients for the first time, which showed environmental sound awareness and speech detection skills [1]. In the early 2000s, studies demonstrated that the ABI could be an option to improve speech and language development in these children [2–4]. Recently, ABI has started to be used with increasing numbers in many centers around the world with successful results on auditory perception skills in children. This chapter addresses the audiological outcomes of children with the ABI including auditory perception, language development, and speech intelligibility.

F. Aslan (✉) · E. Yücel · G. Sennaroglu · M. Yaralı
Department of Audiology, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey

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

31.2 Audiological Outcomes

Free field aided testing gives valuable information about the hearing level with ABI device. This test is routinely done in every follow-up after the initial device fitting in our implant group. The results from this test guide audiologists in changing the MAP parameters such as charge levels. Our pediatric ABI users with IEMs, who used their devices for a period of 1 month to 1 year after the initial fitting, have an average threshold of (average of 500, 1000, 2000, and 4000 Hz) 61 dB, whereas using the device for a period of 1–2 years brings average aided thresholds to 47 dB. Patients who used their ABI for a longer period (between 3 and 5 years) had better average aided thresholds of 44 dB. Finally, the group who used their ABI for more than 5 years had average aided threshold of 35 dB.

These are the mean average thresholds from all ABI users, but individual differences exist. Examples of individual differences in this sense were given in previous chapters with aided threshold examples of different ABI users. In fact, a user with high adherence to rehabilitation program and fitting sessions may reach the above mentioned aided threshold levels in a shorter time, and just the reverse may be observed for the ones who do not comply with our program. Existence of comorbid disorders has potential to worsen this even further.

© Springer Nature Switzerland AG 2022

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_31

The number of active electrodes is another issue to be discussed in terms of outcomes. In a previous study of our group it was found that the number of active electrodes was not found to be related with closed and open-set speech recognition. In fact, active electrode percentage is 76% in our series, and this is not correlated with better aided thresholds or length of implant use. Better aided thresholds only have significant correlation with longer implant use period ($r = -0.571$, $p = 0.001$).

31.3 Auditory Perception Outcomes

From surgery to the first fitting, the families are anxious and confused during the waiting period. Therefore, it is necessary to inform families about the waiting period and the first fitting experience during preoperative period. After the first fitting, families often hope that their children will recognize the sounds immediately. To cope with frustration and hopelessness, it is necessary to explain the families that "hearing" is not "the transmission of voice through ears"; the brain plays a crucial role to recognize the meaning of sounds.

Although the child reacts to the sound during the ABI fitting, some children may not respond to speech sounds in everyday life. This is different from the cochlear implant (CI), because sound awareness abilities of CI users improve usually after the first fitting, and their listening behaviors are relatively clearer than ABI users. For this reason, after the initial tune-up, the family should be informed about how to monitor the children's responses to sounds and how to introduce the environmental sounds. Families often expect clear and typical listening behaviors. However, the first reactions of children with ABI include vague behaviors to the sound, and frequently there are unique behaviors in each child. For example, there may be relatively more clear responses with sounds, such as becoming quiet, slowing/accelerating movements, or less clear responses such as using gestures, shrugging their shoulders, and looking around. Moreover, parents should be informed that auditory perception process includes various development areas such as attention and memory skills. In this period, according to the information received from the family, auditory perception developments in daily life can be followed. Auditory perception skills of ABI users have been investigated with family reports, checklists, and assessment tools. The information about everyday listening behaviors can be obtained with questionnaires such as IT-MAIS [5], MAIS [6], FAPCI [7], and Little EARS [8]. These tools help parents to track their child's sound awareness behaviors. For further information, please refer to Chap. 8.

In the early 2000s, the promising results on auditory perception began to be published. Nevison et al. [9] shared their experience with 26 out of 27 adult patients who received auditory sensation during initial tune-up. According to follow-up outcomes, adult patients discriminated the basic temporal and spectral features of speech patterns. In one of the first studies, Colletti et al. [2] reported that environmental sound awareness and speech detection abilities improved after 1 year of experience. In 2009, Sennaroglu et al. published their preliminary results of 11 children with several cochlear malformations. All children were enrolled in the auditory-verbal therapy sessions and their parents chose aural communication. In their study, the auditory perception assessments were performed at 3 months' interval from first month to 1 year. Six children gained basic auditory perception skills such as identifying environmental sounds, recognizing and discriminating speech sounds after 6 months of experience with ABI. There was a regular increase in the MAIS scores of all children (Fig. 31.1). Same six children began to identify the Ling's Six Sounds at the end of a year. Only two children, who were diagnosed with attention deficit hyperactivity disorder (ADHD), scored inconsistently in Ling's Sounds Test. Sennaroglu et al. reported that children with additional handicaps such as ADHD, mental retardation, and developmental delay performed worse on

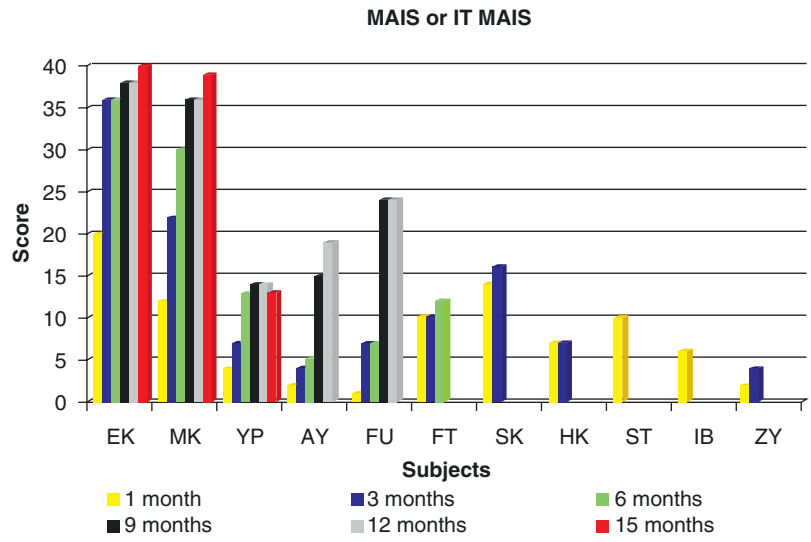


Fig. 31.1 IT/MAIS scores of ABI users in 15 months follow-up. (With permission from Otology & Neurotology)

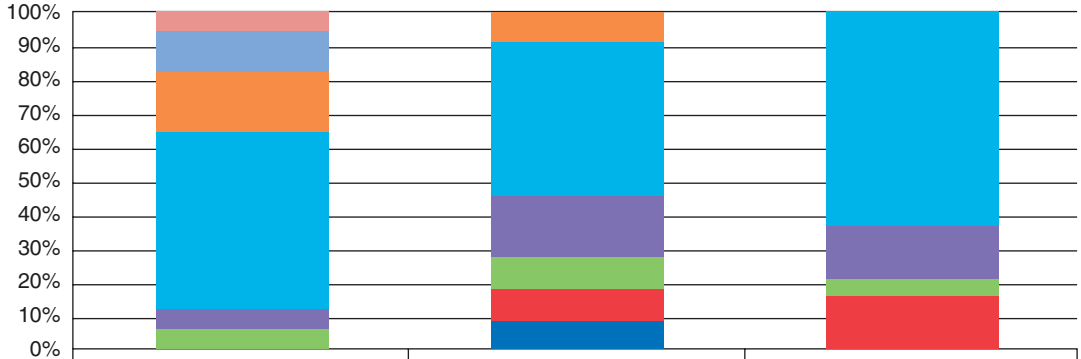
auditory perception tasks when compared to children with no additional handicap.

Findings by Choi et al. supported the developmental delay on auditory perception in children with additional handicaps such as mental retardation and blindness [10]. They believed that the reason for the delay could be the higher cognitive function, not associated with auditory sensation. This is because all eight children showed improvement in their auditory perception. In 2013, Hacettepe University pediatric ABI cohort's auditory perception outcomes were published and it included 39 patients with severe IEMs [11]. In their study, they reported the results of 29 children with ABI experience for 1.5 years. Most important finding was that children used their device regularly on daily basis, whether an additional handicap was present or not. In addition, majority of the children gained basic auditory perception skills. Eighty-six percent of children detected all six sounds in Ling's Sound Test; 75% of them recognized all sounds in the test, and 64% of them had MAIS scores between 30 and 40.

In 2016, Sennaroglu et al. reported their long-term results of 60 pediatric ABI users with complex IEMs [12]. Among 60 patients, 35 of them were followed more than 1 year. The rest of the patients that used ABI for less than 1 year were excluded from the study to eliminate the effect of inadequate experience with ABI. Auditory perception performance of the participants was assessed with CAP and they were divided into three groups according to their free field hearing thresholds. As demonstrated in Fig. 31.2, CAP scores were better with a better hearing threshold. Majority of the patients accumulated in category 5 which implies that they can understand common phrases without lip reading (Fig. 31.3).

Similarly, they compared the functional communication performance of the participants according to age at ABI surgery. Majority of the FAPCI scores were in the lowest tenth percentile, which reveals that children with ABI are worse performers compared to average CI users. There was no difference between the groups according to the age (Fig. 31.4). Moreover, they presented data that additional handicaps such as mental retardation, ADHD, and visual impairment had a negative impact on auditory perception performances of ABI users.

Wilkinson et al. reported that out of 9 children, who were enrolled in their ABI program, 4 of them completed the 1-year follow-up process. These children scored between 8 and 31 out of 40 points in IT-MAIS/MAIS, respectively [13]. They concluded that these scores demonstrated individual variability and slow progression as the children begin developing fundamental auditory skills. Moreover, they asserted that ABI outcomes



	25-40 dB	41-50 dB	>50 dB
8	5.9	0	0
7	11.8	0	0
6	17.6	9.1	0
5	52.9	45.5	57.1
4	5.9	18.2	14.3
3	5.9	9.1	4.3
2	0	9.1	14.3
1	0	9.1	0

Fig. 31.2 Distributions of CAP scores due to hearing thresholds of children with ABI. (With permission from Otology & Neurotology)

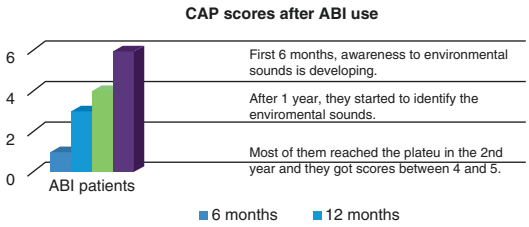


Fig. 31.3 CAP Scores of pediatric ABI users in 1 year period

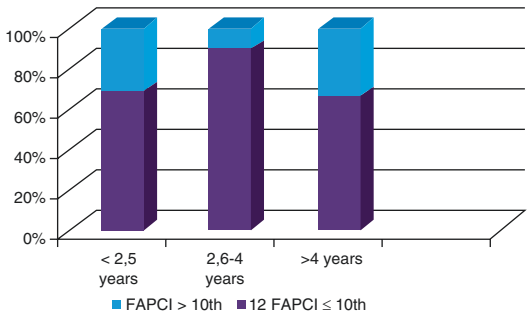


Fig. 31.4 Functional communication performances of children with ABI due to their age at surgery. (With permission from International Journal of Advanced Otology)

will occur along the mid-to-lower percentiles of CI patients due to longitudinal researches.

According to the literature [14, 15], it is evident that auditory perception of children with ABI showed individual variations. Despite the heterogeneity of the outcomes, all children used their device regularly and their auditory sensation was restored in different degrees. Conversely, children with additional handicaps showed slower development within the group. More intensive rehabilitation and integrative therapies should be considered in this latter group.

31.4 Speech Perception Outcomes

Speech perception development of pediatric ABI users was mostly assessed with closed-set pattern discrimination, word identification, and open-set sentence recognition tests. In cochlear implant (CI) technology, it is clear that speech perception

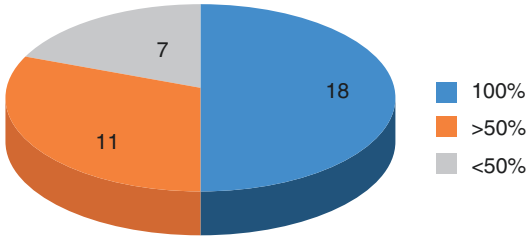


Fig. 31.5 Pattern perception performance. (With permission from International Journal of Advanced Otology)

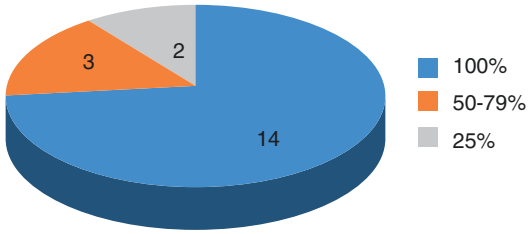


Fig. 31.6 Word identification performance. (With permission from International Journal of Advanced Otology)

ability of children with CI will improve steadily. In ABI technology, it is still unclear which factors are crucial for speech perception development. In this section, some of the factors that have impact on the speech perception outcome will be discussed.

One of the early articles on speech perception of ABI users investigated a child who was deafened in the perilingual period. Sanna et al. [16] shared their results of a 12-year-old female patient who underwent ABI after meningitis. The patient scored 100% sound identification, 90% recognition of bisyllable words, and 100% sentences recognition after 8 months. Later, Colletti et al. presented auditory perception performances of 14 ABI users [17]. Their initial findings indicated that three children began to recognize bisyllabic words and understand simple commands. While all children reached some degree of auditory sensation, none of them achieved open-set speech recognition yet. Case study by Eisenberg et al. provided information about speech perception development of 3-year-old boy who received ABI at the age of two [18]. After 1 year, he began to develop word identification in closed-set condition. Sennaroglu et al. reported 11 children's speech perception performances comprehensively [4]. Their findings showed that 4 of 11 children began to discriminate speech pattern, 2 of them had consistent word identification score of 24/24 at 9 months postoperatively (Figs. 31.5 and 31.6). Also, these children had improved their speech perception skills more rapidly than others. While four children continued to be struggling, only two children achieved open-set sentence recognition level after 15 months. They even began to use the telephone with familiar people. In the same study, children with no additional handicaps progressed steadily on speech perception tasks and showed that ABI provides a valuable opportunity for these children. In 2013, Sennaroglu et al. reported speech perception outcomes of 34 pediatric ABI patients after one and a half year [11]. The patients' sentence recognition performances improved and 10 of 29 patients scored 60–100 in auditory-verbal condition and 8 of them scored 20–100 in only auditory condition.

The first consensus meeting was organized on auditory brainstem implantation (ABI) in children and in non-neurofibromatosis type 2 (NF2) cases by Hacettepe University Implant Group in 2009. Health care professionals and scientists from ten centers who worked with pediatric ABI attended the meeting. At the end of the meeting, a consensus statement was published. One of the essential points on this statement was "from the results of different centers, it can be understood that it is possible to restore hearing perception in children with prelingual deafness with severe IEMs and cochlear nerve anomalies. In some selected cases, it also was possible to develop an open-set speech understanding. However, the family should be warned of different outcomes from this intervention so that their expectations should not be high. When compared with CI surgery, programming and rehabilitation of prelingually deafened children with ABI are much more labor intensive, and the results do not reach the level of CI users. On this basis, the candidacy assessment is much more detailed than in CI patients and requires more experienced staff. ABI

is a viable option for children including prelingually deafened patients with IEMs and cochlear nerve hypoplasia/aplasia. ABI provides auditory perception in most patients. The potential for speech and language acquisition in the longer term depends on the age of implantation, the presence or absence of additional disabilities, and the other established factors seen in CI. It was concluded that open-set speech discrimination is possible in selected cases" [19].

In 2013, "Second Consensus Meeting on Management of Complex Inner Ear Malformations: Long-Term Results of ABI in Children and Decision Making Between CI and ABI" brought professionals from 20 centers in 11 countries. According to results from all centers, the pediatric ABI users could develop speech discrimination in closed-set to open-set condition [20]. At the time of consensus meeting, the largest case series had 35 children with at least 1 year of follow-up. In the cohort, 80% of children achieved scores above 50% with closed-set pattern discrimination task, while approximately 30% reached maximum scores. Additionally, 30% of patients scored above 50% with open-set sentence recognition test. The consensus report suggested that visual information should be used as a complementary element in auditory rehabilitation programs in pediatric ABI users. Follow-ups should be determined at shorter intervals than cochlear implantation and should be intensive.

31.5 Language Development Outcomes

While evaluating language development skills after auditory brainstem implantation, verbal language skills, as well as sign language and prevocalic development (gestures, mimics) skills, are equally important. For this reason, feedback from the family and the notes of the family and the experts working with the child are valuable for the evaluation of communication skills in daily life. In order to monitor the development, it is appropriate to request video and audio recording from families in different occasions. In this way, speech and communication analysis of the patient can be done more correctly.

At the end of the first 6 months, there is a significant increase in the vocalization of children. Their production has similar characteristics to babbling. In line with the development of pattern discrimination skills in auditory perception, they can produce sounds that are compatible with the patterns of words they try to imitate. The most common problem is the continuous repetition of the sounds produced by the child and the reduction of their intelligibility. For example, instead of "mama," they repeat "mamamama." In this period, children like to listen to their own voices, even the children who do not produce similar words like joy or anger are seen to make the sound plays.

In this period, experts should also consider to support the concept knowledge (such as colors, numbers) which is an important part of the language development. Additionally, another developmental area which is fine motor skill should not be ignored by focusing on the development of language and auditory perception. There is an individual difference in language performance in pediatric ABI patients due to chronological age, duration of ABI use, additional handicaps, and cognitive development. Sennaroglu et al. suggested that the main reasons could be the associated comorbidity due to additional handicaps such as attention deficit hyperactivity, slight mental retardation, and visual problems on language development delay [11]. They claimed that majority of the patients with limited improvement in performance have additional handicaps. Many studies have reported that supporting cognitive and fine motor skills can help support language development in children [21]. Eisenberg et al. [18] reported a 3-year-old boy whose language development reached the level of 2-year-old children. Although he had a normal intelligence, he had difficulties in completing structured tasks due to attention deficits.

According to the studies, language development skills of children with ABI are not

compatible with their typically developing peers, but they are more prone to show language development compatible with the time they begin to hear [22]. Depending on the individual differences of children, the time, when the child is ready for standardized language development tests, shows variation. When language skills are examined in two subtests (receptive and expressive language skills), receptive language is slightly better than the expressive language [4]. The long-term follow-up outcomes of language development showed that the language skills of the patients have entered into a stagnation period [12]. The language development in the first 2 years of ABI surgery is beyond the duration of ABI use. In 2009, the language scores of 15 patients were above the duration of ABI use in means of language parameters as age equivalent scores (Figs. 31.7 and 31.8). However, when this same group of patients is examined almost 5 years later, only 4 of 20 are above the line (Figs. 31.9 and 31.10). It is assumed that the rate of language development was much faster in the first 2 years, and although it still continues, the pace declines afterward.

Manual communication options (such as sign language, coded language, cued speech) should be recommended to parents' of children with ABI to support their communication skills. It is observed that communication skills increase with the support of sign language, behavioral problems decrease, provide children to follow the conversation with more than one person and in noisy environments [14, 22]. These findings were supported by the first consensus statement as "Auditory-verbal therapy in these children, where only auditory stimulation is conveyed, may not be as efficient as in children who are using CI. Total communication and speech reading also should be encouraged to convey more linguistic and language information to these children. In this method, speech reading assumes considerable importance as a source of information, whereas tactile and motor kinesthetic stimulation provides supportive avenues for spoken language acquisition. In addition, the involvement in speech reading training programs has a positive effect on postoperative perceptive and expressive linguistic skills" [19].

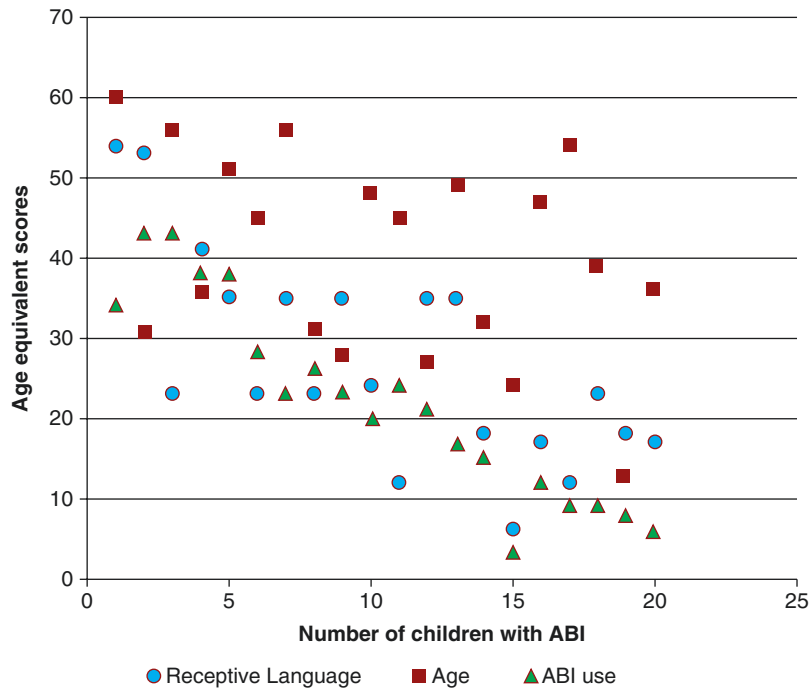


Fig. 31.7 Receptive language performance in 2009 study. (With permission from Otology & Neurotology)

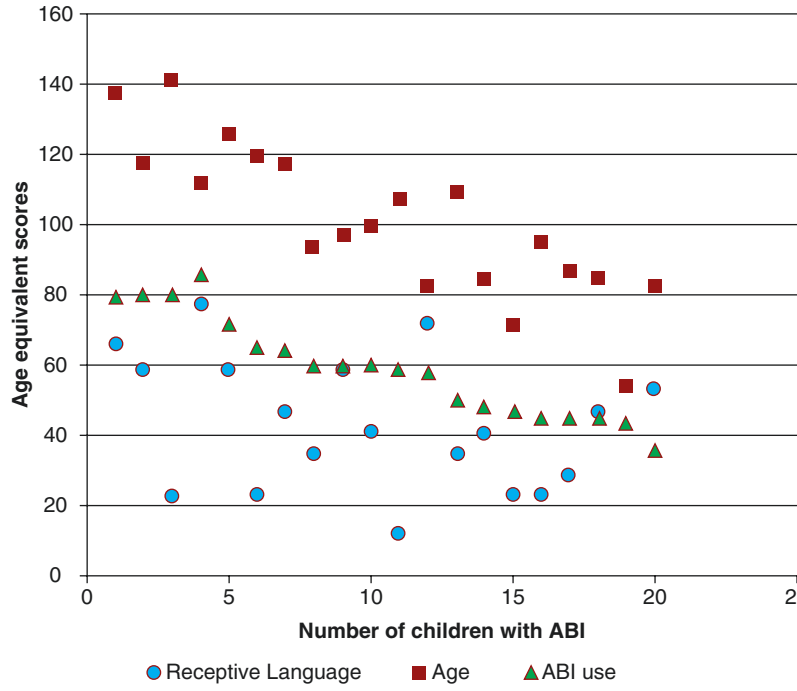


Fig. 31.8 Receptive language performance of the same patients in 2013 study. (With permission from Otology & Neurotology)

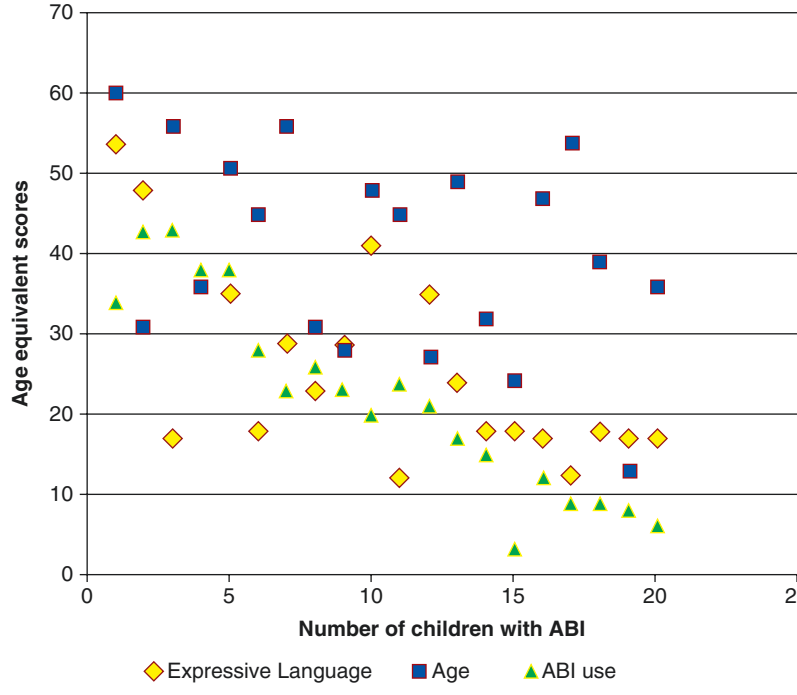


Fig. 31.9 Expressive language performance in 2009 study

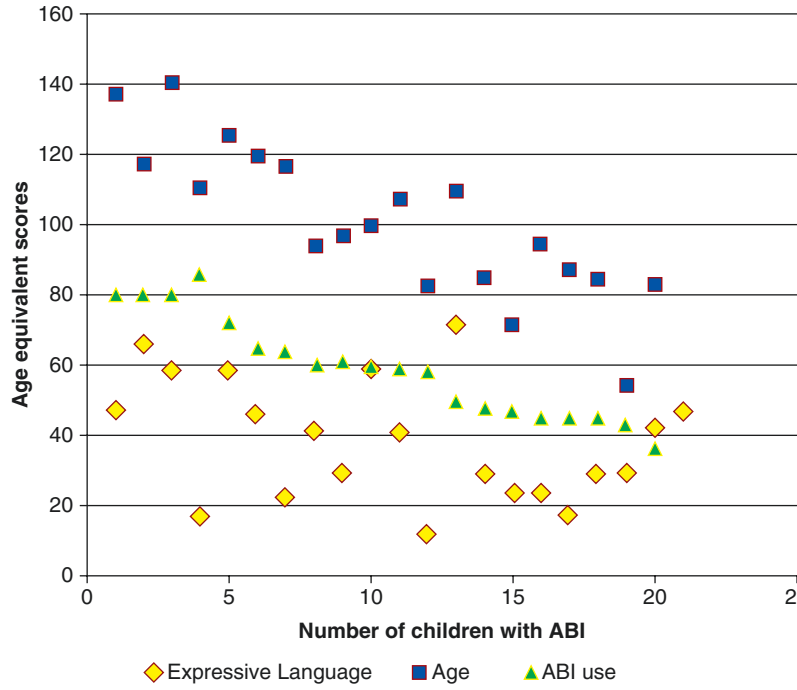


Fig. 31.10 Expressive language performance in 2013 study

31.6 Speech Intelligibility Outcome

In addition to expressive language skills, there are difficulties in developing speech intelligibility which is low in pediatric ABI patients [12]. Speech intelligibility is reported to be one of the most challenging skills in daily life in the assessments and information obtained from the family [22]. According to Eisenberg et al. parents reported that their children's speech remains unclear, especially for unfamiliar listeners [14]. Therefore, they may need additional cues such as sign language for communication. Further studies are needed in the literature on the difficulties of speech intelligibility in this population.

31.7 Recent Results of Pediatric ABI Patients

Between June 2006 and September 2018, 124 children with complex IEMs received an ABI by Hacettepe University Hospital Implant Team. Five of these are revision due to device failure. The results of 84 primary pediatric ABI patients who have been using their ABI for more than a year were analyzed and presented in 15th The International Conferences on Cochlear Implants and Other Implantable Auditory Technologies in Belgium. Inclusion criteria for this study include: (1) age younger than 18 years, (2) monolingual children, (3) only unilateral ABI users, (4) at least 1-year ABI experience. Children with additional difficulties were not excluded from this study, only one child with autism who was not enrolled in the sessions regularly was excluded.

Fifty-nine percent of the patients were female. Mean age at ABI surgery was 34.54 months old (range: 12–96 months old, SD = 18.52). Of all patients, 26% had additional difficulties such as CHARGE syndrome, Goldenhar Syndrome, ADHD. Seventy-six percent of these children are using auditory-verbal communication mode and the rest have chosen total communication.

In this study, average MAIS score was 26.50 (range: 5–40, SD = 10.83). There is a significant relationship between MAIS scores and age of

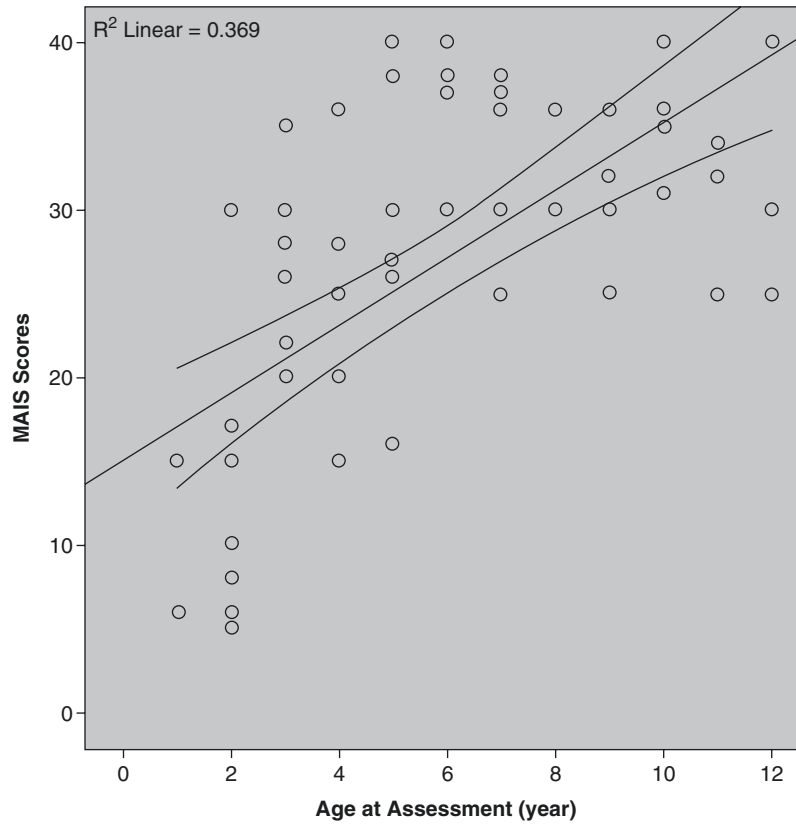


Fig. 31.11 MAIS performances due to age at assessment

evaluation (Fig. 31.11). Although there was a linear development due to chronological age in MAIS performances, no significant relationship due to age at ABI surgery was found.

Out of 84 children who met the inclusion criteria, we determined that approximately half of the participants completed the ability to distinguish the closed-set pattern. We found that 17% of the children could do more than half of the test and 36% of them continued to improve this skill. While the difficulty in "word discrimination" testing increased, we wanted them to find the word that has the same number of syllables; 22% completed this skill, 31% completed more than half of the test, and 46% still continue to improve this skill. The next stage is the open-ended sentence recognition phase where clues are eliminated. At this stage, 16% of children recognized the sentences in daily life, 24% of them are just starting to gain this skill, and 63% of them have not yet reached this level in auditory perception.

When we evaluated long-term language development results of children with ABI, the gap between chronological age and language age persists. In contrast to the difference between chronological age and language age, the gap between the age equivalence of language performance and the duration of he/she begins to hear (hearing age) decreases. We found that language development of children who started to use ABI in the early period was similar when they started to hear. There was no statistically significant difference between the language development of the participants who had started using ABI before and after the age of 3 years. Therefore, the following long-term results are important for determining whether the gap would be opened or closed (Figs. 31.12 and 31.13).

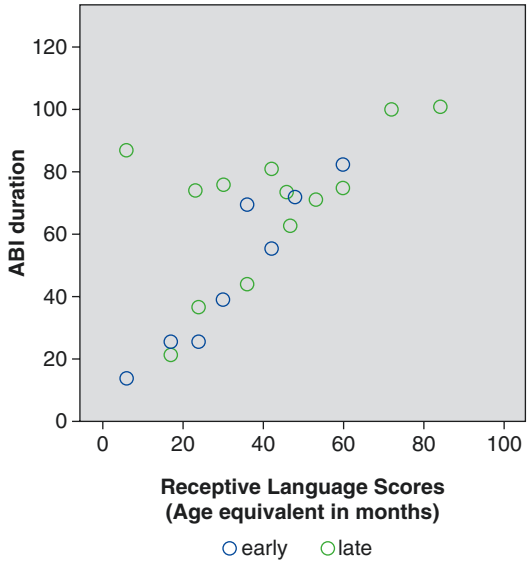


Fig. 31.12 Receptive language scores of ABI users due to duration of ABI use

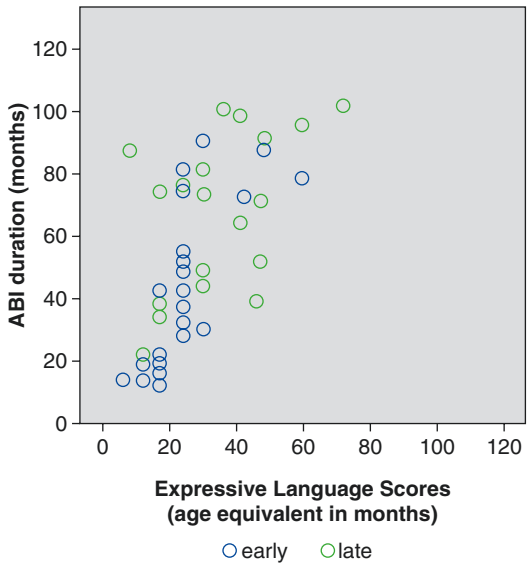


Fig. 31.13 Expressive language scores of ABI users due to duration of ABI use

31.8 Educational Settings

Children who are being considered for ABI candidacy should be evaluated in terms of cognitive skills and learning behavior. Their participation in educational tasks is decisive for postoperative progress and building of a therapy program. The assessment process involves their communication behaviors such as eye contact, speech reading, turn-taking, initiating, and sustaining joint attention, following directions, etc. [20]. These nonverbal forms of communication reflect the child's ability to structure a particular communication method [20]. In 2018, 47 of 75 children with ABI at school age, only 9% of them attended the school of Deaf. All children were enrolled in Individualized Education Programs in local centers. Majority of the children used total or verbal communication. Further studies need to be done to determine which factors have a significant impact on the choice of school settings (Figs. 31.14 and 31.15).

Factors that can influence auditory perception and language outcomes

- ✓ Length of auditory deprivation
- ✓ Age at surgery
- ✓ Etiology of deafness
- ✓ Additional handicaps
- ✓ Surgical issues
- ✓ Mode of communication
- ✓ Educational settings
- ✓ Motivstion and participation of parents
- ✓ Post-operative hearing thresholds
- ✓ Training options and frequency

Fig. 31.14 Factors that can influence auditory perception and language outcome in pediatric ABI users

Standard protocol includes in pediatric ABI users

- ✓ Reports about child's listening behaviors and use of ABI
- ✓ Comment of the therapist
- ✓ Behavioral audiometry
- ✓ Check of device function
- ✓ Measurement of electrodes
- ✓ NRT recording

Fig. 31.15 The parameters should be included in standard assessment and follow-up protocol in ABI

31.9 Conclusion

An auditory brainstem implant is an auditory prosthesis that provides acceptable and effective treatment option for pediatric population with complex IEMs. Auditory brainstem implants have enabled children, who show no or inadequate benefit from cochlear implants, to develop speech perception and language skills. It is evident that the majority of the children gain auditory sensation with ABI. Therefore, the speech perception and language outcomes varied widely.

Although children with ABI demonstrated improved speech perception ability with auditory-verbal approach, visual cues and sign language should be offered to enhance their communication skills. Even children, who achieved open-set sentence recognition, need visual cues in difficult listening environment.

It is recommended that the family and the professionals working with the child are informed carefully about the (re)habilitation objectives. More realistic targets keep therapists and parents on the track to support children with ABI. The motivation and participation of the family and the child in the rehabilitation process are very important.

References

1. Colletti V, Fiorino F, Sacchetto L, Miorelli V, Carner M. Hearing habilitation with auditory brainstem implantation in two children with cochlear nerve aplasia. Int J Pediatr Otorhinolaryngol. 2001;60(2):99–111.
2. Colletti V, Carner M, Fiorino F, Sacchetto L, Miorelli V, Orsi A, et al. Hearing restoration with auditory brainstem implant in three children with cochlear nerve aplasia. Otol Neurotol. 2002;23(5):682–93.
3. Colletti V, Carner M, Miorelli V, Guida M, Colletti L, Fiorino F. Auditory brainstem implant (ABI): new frontiers in adults and children. Otolaryngol Head Neck Surg. 2005;133(1):126–38.
4. Sennaroglu L, Ziyal I, Atas A, Sennaroglu G, Yucel E, Sevinc S, et al. Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. Otol Neurotol. 2009;30(6):708–15.
5. Zimmerman-Phillips S, Osberger M, Robbins A. Infant-toddler meaningful auditory integration scale. Sylmar, CA: Advanced Bionics Co.; 2001.
6. Robbins AM, Renshaw JJ, Berry SW. Evaluating meaningful auditory integration in profoundly hearing-impaired children. Am J Otol. 1991;12:144–50.
7. Lin FR, Ceh K, Bervinchak D, Riley A, Miech R, Niparko JK. Development of a communicative performance scale for pediatric cochlear implantation. Ear Hear. 2007;28(5):703–12.
8. Coninx F, Weichbold V, Tsiakpini L, Autrique E, Bescond G, Tamas L, et al. Validation of the LittlEARS® Auditory Questionnaire in children with normal hearing. Int J Pediatr Otorhinolaryngol. 2009;73(12):1761–8.
9. Nevison B, Laszig R, Sollmann W-P, Lenarz T, Sterkers O, Ramsden R, et al. Results from a European clinical investigation of the Nucleus® multichannel auditory brainstem implant. Ear Hear. 2002;23(3):170–83.
10. Choi JY, Song MH, Jeon JH, Lee WS, Chang JW. Early surgical results of auditory brainstem implantation in nontumor patients. Laryngoscope. 2011;121(12):2610–8.
11. Sennaroglu L, Sennaroglu G, Atay G. Auditory brainstem implantation in children. Curr Otorhinolaryngol Rep. 2013;1(2):80–91.
12. Sennaroglu L, Sennaroglu G, Yücel E, Bilginer B, Atay G, Bajin MD, et al. Long-term results of ABI in children with severe inner ear malformations. Otol Neurotol. 2016;37(7):865–72.
13. Wilkinson EP, Eisenberg LS, Krieger MD, Schwartz MS, Winter M, Glater JL, et al. Initial results of a safety and feasibility study of auditory brainstem implantation in congenitally deaf children. Otol Neurotol. 2017;38(2):212.
14. Eisenberg LS, Hammes Ganguly D, Martinez AS, Fisher LM, Winter ME, Glater JL, et al. Early communication development of children with auditory brainstem implants. J Deaf Stud Deaf Educ. 2018;23(3):249–60.
15. Noij KS, Kozin ED, Sethi R, Shah PV, Kaplan AB, Herrmann B, et al. Systematic review of nontumor pediatric auditory brainstem implant outcomes. Otolaryngol Head Neck Surg. 2015;153(5):739–50.
16. Sanna M, Khrais T, Guida M, Falcioni M. Auditory brainstem implant in a child with severely ossified cochlea. Laryngoscope. 2006;116(9):1700–3.
17. Colletti L. Beneficial auditory and cognitive effects of auditory brainstem implantation in children. Acta Otolaryngol. 2007;127(9):943–6.
18. Eisenberg LS, Johnson KC, Martinez AS, DesJardin JL, Stika CJ, Dzubak D, et al. Comprehensive evaluation of a child with an auditory brainstem implant. Otol Neurotol. 2008;29(2):251–7.
19. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol. 2011;32(2):187–91.

20. Sennaroğlu L, Colletti V, Lenarz T, Manrique M, Laszig R, Rask-Andersen H, et al. 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.
21. Diamond A. Close interrelation of motor development and cognitive development and of the cerebellum and prefrontal cortex. Child Dev. 2000;71(1):44–56.
22. Yücel E, Aslan F, Özkan HB, Sennaroglu L. Recent rehabilitation experience with pediatric ABI users. J Int Adv Otol. 2015;11(2):110.

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

© Springer Nature Switzerland AG 2022

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

	CAP		SIR	
CASE	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,

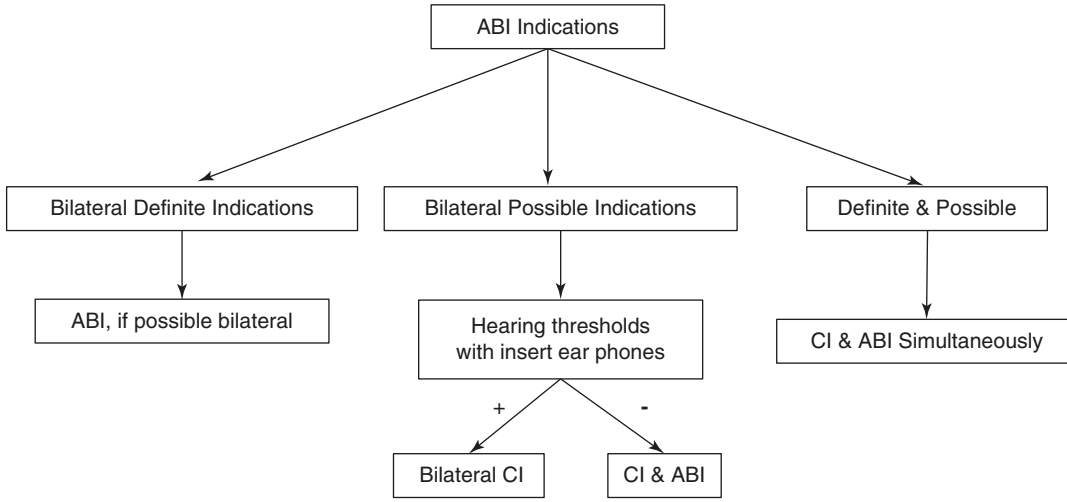


Fig. 32.1 Current management strategy in severe inner ear malformations with cochlear nerve deficiency

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

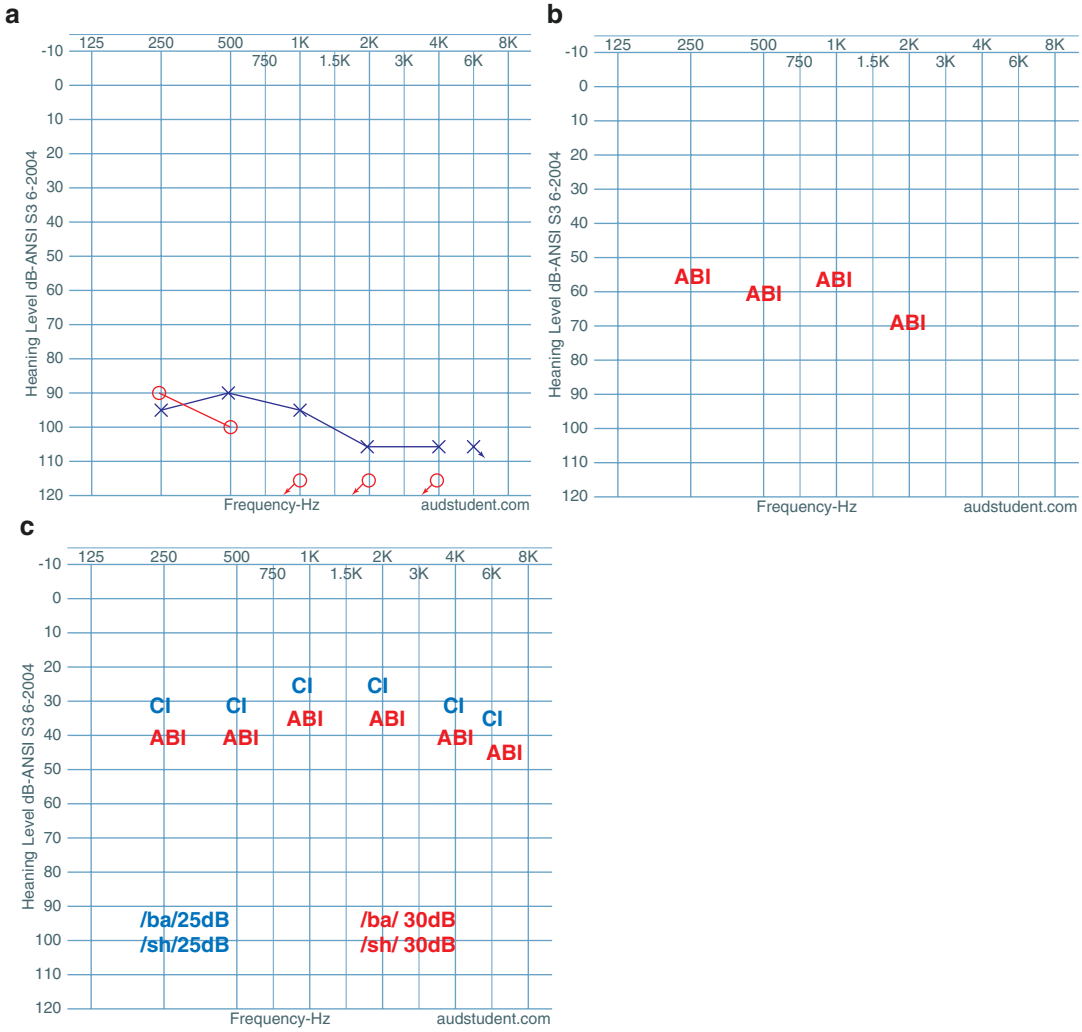


Fig. 32.2 Case 1. (**a**) Preoperative audiogram with insert ear phones showing a response on the left side, (**b**) first audiogram with ABI, (**c**) postoperative thresholds with CI and ABI after 5 years

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

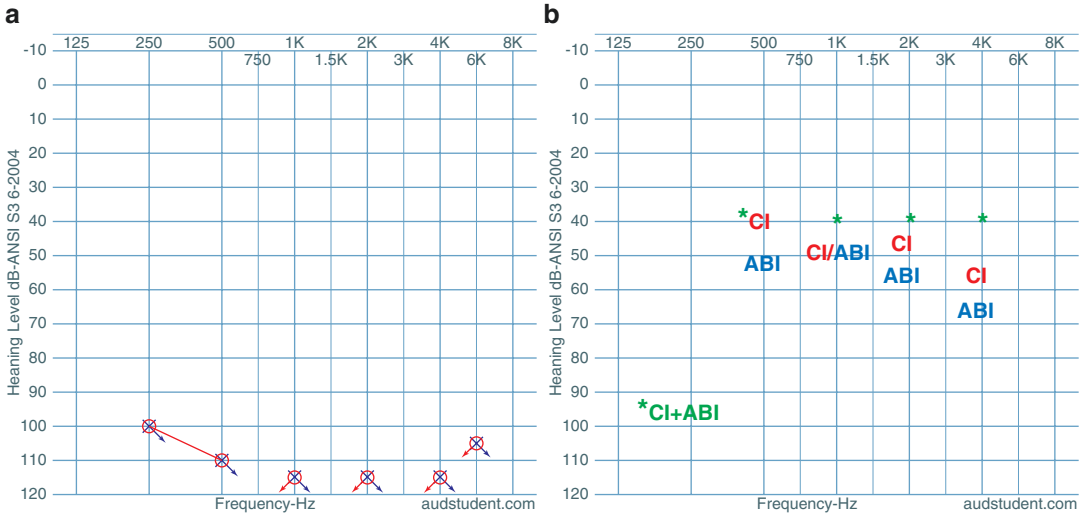


Fig. 32.3 Case 2. (**a**) Preoperative audiogram showing auditory responses on the right side where there was a common cavity with a well-developed cochleovestibular nerve, (**b**) Postoperative audiogram after 2 years

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

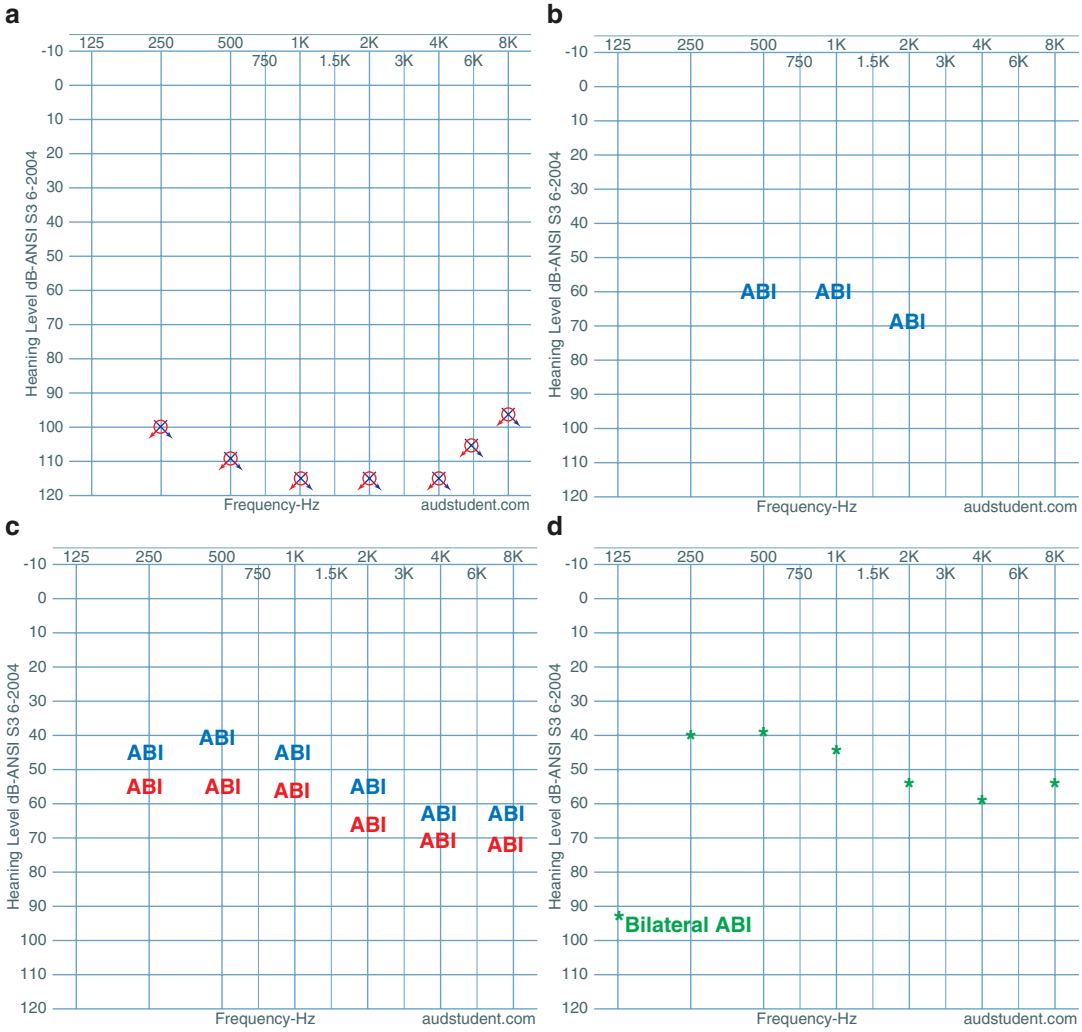


Fig. 32.4 Case 3. (**a**) Preoperative audiogram showing no auditory response, (**b**) auditory responses with left ABI, (**c**) thresholds with both ABIs, (**d**) better auditory response in the bilateral condition

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.