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

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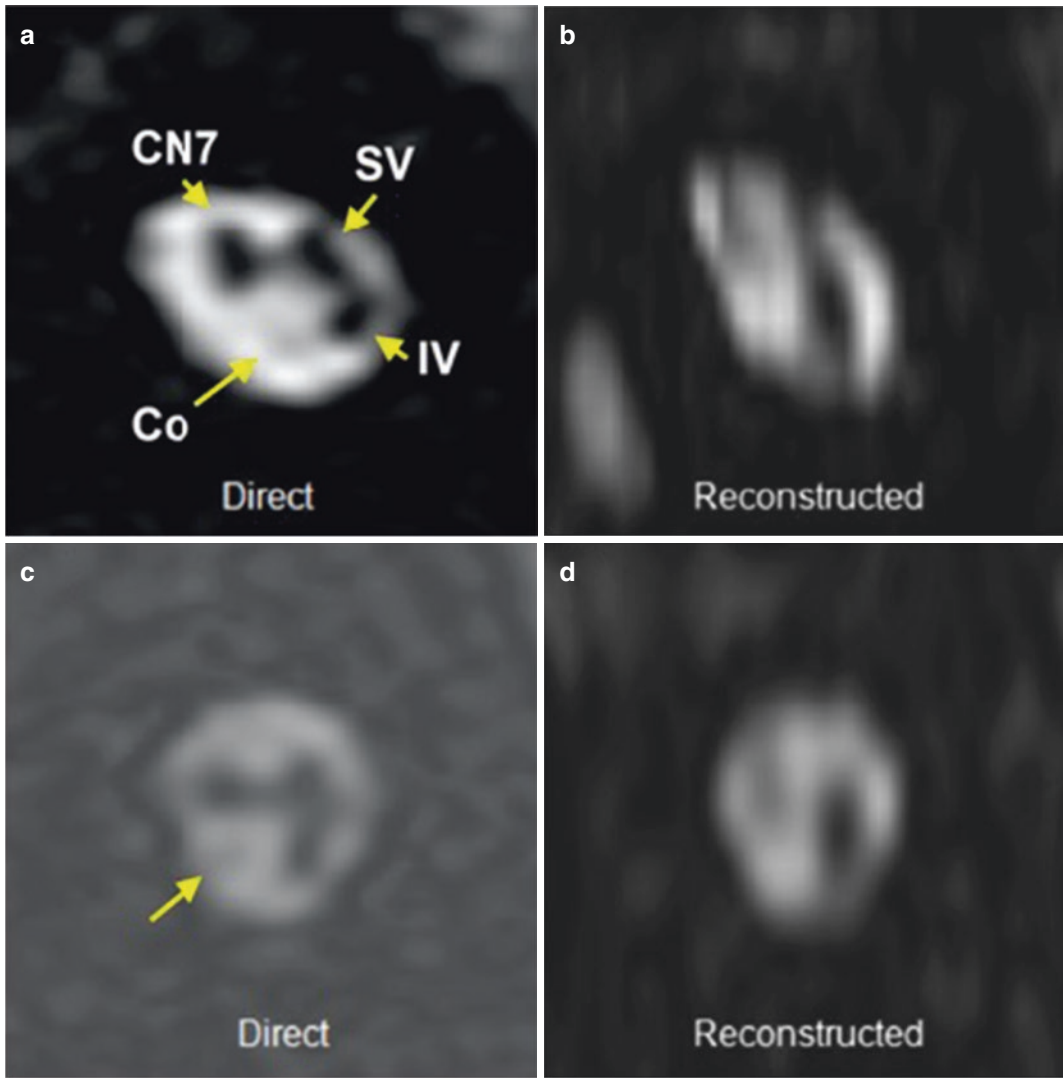

Fig. 17.1 Direct and reconstructed parasagittal T2-weighted MRI sequences illustrate internal auditory canal (IAC) morphology in two pediatric patients with congenital profound hearing loss. Reformatted images perpendicular to the IAC (image **b**) suggest cochlear nerve aplasia but provide poor resolution, 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

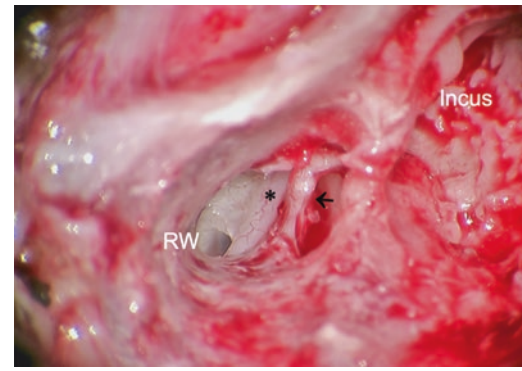

Fig. 17.2 Intraoperative transmastoid view of the left ear through a posterior tympanotomy demonstrates an aberrant facial nerve in cochlear hypoplasia type I (CH-I) in CHARGE association. Absent horizontal semicircular canal with an aberrant facial nerve (*) is identified inferior to the location of 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

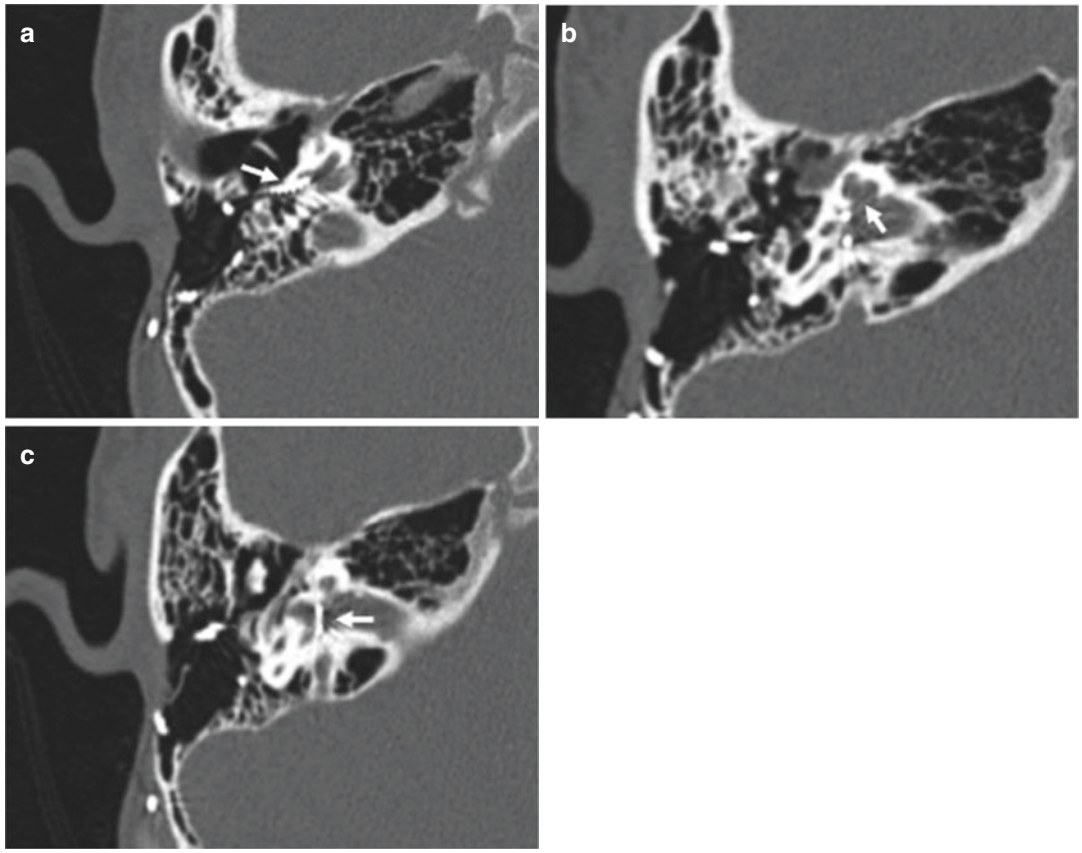

Fig. 17.3 Non-contrast axial CT scan images demonstrate CI placement into the IAC in a case of type III incomplete partitioning. Although the CI is placed through a round window approach (Image **a**), an underdeveloped modiolus and widened cochlear nerve canal (Image **b**) result in displacement of 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.

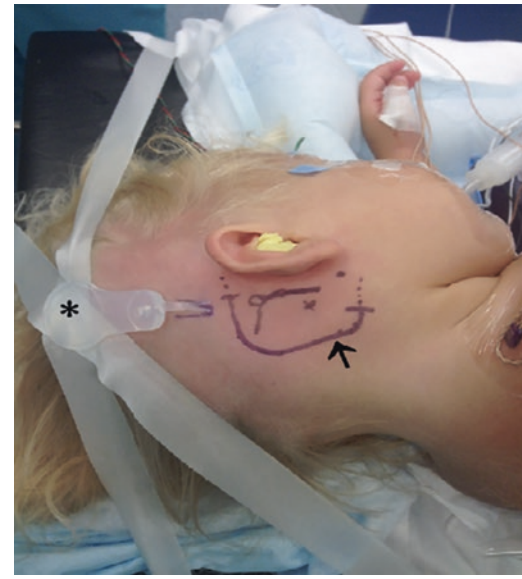

Fig. 17.4 Surgical site incision planning for a right ear retrosigmoid craniotomy with placement of the ABI receiver-stimulator (*) relative to a retrosigmoid craniotomy incision (arrow)

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.

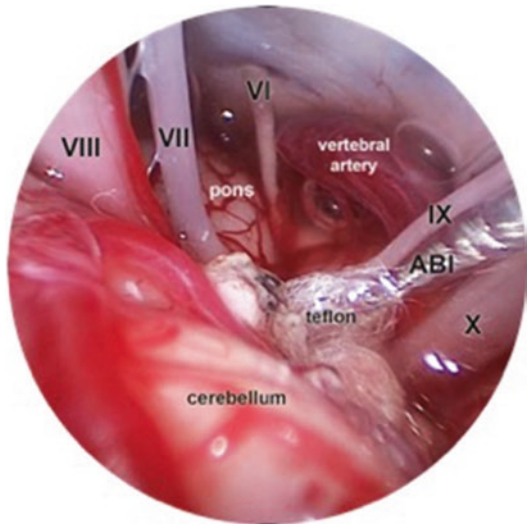

Fig. 17.5 Intraoperative 30° endoscopic view of the right cerebellopontine angle following retrosigmoid craniotomy for a 2-year-old male with congenital deafness and cochlear aplasia illustrates placement of the auditory brainstem implant (ABI) array. The Teflon felt is used to secure the array in the 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.

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