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Cochlear Implantation in Unique Pediatric Populations

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Abstract

Purpose of review—Over the last decade, the selection criteria for cochlear implantation have expanded to include children with special auditory, otologic, and medical problems. Included within this expanded group of candidates are those children with auditory neuropathy spectrum disorder, cochleovestibular malformations, cochlear nerve deficiency, associated syndromes, as well as multiple medical and developmental disorders. Definitive indications for cochlear implantation in these unique pediatric populations are in evolution. This review will provide an overview of managing and habilitating hearing loss within these populations with specific focus on cochlear implantation as a treatment option.

Recent findings—Cochlear implants have been successfully implanted in children within unique populations with variable results. Evaluation for cochlear implant candidacy includes the core components of a full medical, audiologic, and speech and language evaluations. When considering candidacy in these children, additional aspects to consider include disorder specific surgical considerations and child/care-giver counseling regarding reasonable post-implantation outcome expectations.

Summary—Cochlear implantations are accepted as the standard of care for improving hearing and speech development in children with severe to profound hearing loss. However, children with sensorineural hearing loss who meet established audiologic criteria for cochlear implantation may have unique audiologic, medical, and anatomic characteristics that necessitate special consideration regarding cochlear implantation candidacy and outcome. Individualized pre-operative candidacy and counseling, surgical evaluation, and reasonable post-operative outcome expectations should be taken into account in the management of these children.

Keywords

auditory neuropathy spectrum disorder; cochlear implant; cochlear nerve deficiency; pediatric; cochleovestibular malformations; congenital hearing loss; syndromic hearing loss; unique population

Introduction

Since its introduction more than 40 years ago, cochlear implantation (CI) has grown to become an accepted, well-recognized treatment for pediatric patients with severe to profound sensorineural hearing loss (SNHL). Over the last decade, the selection criteria for CI have expanded to include children with special auditory, otologic, and medical problems.

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Conflicts of Interest

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Included within this expanded group of candidates and discussed in this chapter are those children with auditory neuropathy spectrum disorder (ANS), cochleo-vestibular malformations, cochlear nerve deficiency (CND), associated syndromes, as well as multiple medical and developmental disorders. Definitive indications for implantation in these unique pediatric populations currently are still in evolution. This review will provide an overview of managing and rehabilitating hearing loss within these populations with specific focus on CI as a treatment option

Pediatric CI candidacy evaluation begins with a thorough audiologic assessment. Electrophysiologic testing such as auditory brainstem response (ABR) and oto-acoustic emissions (OAE) testing are implemented in newborn hearing screening, but are also particularly useful in patients who cannot participate in behavioral testing due to young age or developmental issues. They are also critical in the diagnosis of ANS. When possible and practical, pre-operative pure tone audiometry, speech perception, and best aided performance should be assessed(1). Speech and language evaluation can help identify children with cognitive and motor delays that might hinder post-implant auditory development so that modified rehabilitation strategies can then be implemented early. Experienced pediatric audiology and cochlear implant program personnel are critical to the proper candidate selection, pre-operative counseling, and post-operative outcomes for children receiving CIs.

The medical evaluation includes a comprehensive history, physical examination, and adjunct studies with emphasis on identifying both the cause of hearing loss and significant co-morbidities. Approximately 15–40% of congenital SNHL can be attributed to acquired causes such as exposure to intrauterine infections and teratogens, prematurity, perinatal anoxia, hyperbilirubinemia, sepsis, or the use of ototoxic drugs (1). Hereditary causes account for 40–50% of congenital SNHL in most reports and can be further divided into nonsyndromic or syndromic SNHL(1). Syndromes associated with hearing loss include Usher syndrome, Pendred syndrome, Jarvill Lange-Nielsen syndrome, Waardenburg syndrome, and CHARGE syndrome. Early recognition of associated syndromes through history, physical examination, and adjunct studies (such as an electrocardiogram, genetic or infectious disease testing, and ophthalmologic findings) may significantly impact the management of hearing loss as well as identify potentially significant co-morbidities.

Radiologic assessment of the temporal bones is considered standard of care for CI candidacy selection. Up to one third of children with SNHL have cochleo-vestibular anomalies which can be easily identified on preoperative imaging(2). Computed tomography (CT) is particularly useful for bony malformations, aberrant facial nerve course, temporal bone aeration, and aberrant vasculature (i.e., dehiscent jugular bulb), while magnetic resonance imaging (MRI) can identify soft tissue anomalies of the inner ear, specifically the presence of cochlear nerves. In the past, the diagnosis of CND was a contraindication for CI, however there is evidence that these patients may still benefit from CI despite imaging findings(2). Management strategies of children with inner ear malformations and CND will be further discussed.

Patient and family counseling is essential for all CI candidates. Parental motivation, expectations, and participation in auditory rehabilitation significantly impact successful CI use. Children with unique considerations may require additional caregiver involvement due to the special nature of their disorder or associated co-morbidities. Early identification in the candidacy process facilitates counseling and management which may differ significantly from traditional CI candidates with isolated SNHL.

Auditory Neuropathy Spectrum Disorder

Although initially thought to be a rare disorder, recent reviews indicate that the prevalence of ANSD may be as high as 8–15% of newly diagnosed cases of pediatric hearing loss(3). The diagnosis is typically determined using electrophysiologic measures, such as otoacoustic emissions (OAEs) and ABR testing, to establish normal function of outer hair cells and absent or abnormal auditory nerve function. The presence of a cochlear microphonic (CM) with absent neural responses on electrocochleography or ABR recordings define ANSD(4) (Figure 1). The presence of OAE's in the setting of absent neural waveforms on ABR also points to ANSD, however, the absence of OAEs does not necessarily exclude ANSD. Up to 30% of ANSD ears may not have OAEs, which have been documented to disappear over time(4, 5, 6). In addition, concurrent middle ear disease can result in the absence of OAE's. It should be noted that up to 20% of cases of ANSD, neural waveforms on ABRs are not absent but may show a distorted and/or delayed wave V(5, 6). Clinically, children with ANSD often have speech perception difficulties that are disproportionate to their hearing levels, especially when hearing in noise(3).

The causes of ANSD are multifactorial, with variable sites of lesion along the auditory pathway, anywhere from the inner hair cells to the cerebral cortex(7–9). Neonatal risk factors associated with ANSD include prematurity, hyperbilirubinemia, hypoxia, CNS immaturity, low-birth weight, NICU stay, and use of ototoxic drugs(6). Due to the heterogeneity of the disorder, there is a broad range of pure-tone thresholds without clear correlation to speech performance with hearing aid (HA) or CI use(3). Many current management paradigms consists of a 3–6 month trial of conventional amplification prior to recommending CI if there is lack of benefit(3, 6, 8, 10). The main disadvantage of the stepwise approach is the potential delay of cochlear implantation. An appropriate amplification trial may be further delayed by the fact that electrophysiologic testing cannot estimate behavioral thresholds (as the ABR waveforms are absent) and children with ANSD may have associated sensorimotor, developmental, and cognitive impairments that make behavioral audiometry challenging if not impossible(4, 7). Success rates utilizing HAs alone have been reported between 30–50%(3). In some cases, hearing thresholds may actually spontaneously improve, especially in children with a history of hyperbilirubinemia(3, 6).

Proponents of universal CI for children ANSD argue that acoustic amplification only offers louder but still distorted auditory signals thereby limiting the benefit of HAs, whereas CIs can induce neural synchrony at the level of the auditory nerve(3, 5). However, post-implant speech perception performance also varies greatly, with many achieving similar outcomes with matched non-ANSD SNHL peers while others demonstrate a continued delay in speech and language development(4, 8, 11, 12). It is important to identify CND as the cause of ANSD as these patients will have poorer performance than non-CND children regardless of whether HAs or CIs are implemented(3, 8). Currently there are no accepted objective measures to consistently predict outcomes prior to CI. Post-implantation, robust electrically evoked compound action potential (ECAP) measurements correlate with development of open-set speech perception(4, 8).

At this point, stepwise management of children ANSD with a trial of amplification and CI offered to those who fail to benefit appears to be the most accepted standard of care so as to avoid the risks of surgery in children who may potentially benefit from amplification alone. In cases where when traditional audiometry cannot reliably estimate detection thresholds or assess speech perception, unilateral CI has been suggested as a conservative approach(4). A trial of aiding the contralateral non-CI ear, though somewhat controversial, may increase the benefit of CI use alone and is a low risk intervention(10).

Cochlear Nerve Deficiency

The diagnosis of CND is based solely on radiologic findings. It is defined as anatomically small or absent auditory nerve and is found in up to 18% of patients with SNHL(9, 12–14). It represents an anatomic deficiency which by definition is neuropathic (an absent nerve) and therefore can present with audiometric and electrophysiologic results which are indistinguishable from ANSD. Therefore, appropriate imaging is essential in the CI candidacy selection process for children with SNHL, but in particular those with ANSD or no response ABR.

The diagnosis is based on both high resolution CT (HRCT) and MRI findings. On HRCT, a bony cochlear nerve canal (BCNC) of <1.3mm or internal auditory canal (IAC) of <3mm is suggestive of CND. A closed BCNC confirms the diagnosis. However, in one study, 38% of CND cases on MRI were actually found to have a normal sized IAC and BCNC on HRCT(9) (Figure 2). In order to avoid a missed diagnosis of CND, MRI has been suggested as the first line imaging modality over HRCT(9). The pitfall of MRI may be in the case of a narrow IAC where resolution may be insufficient to identifying separate nerves within the canal(9). The assumption in this case is that the radiologically visible nerve represents the facial nerve however intact auditory nerve fibers may be present but not bundled as a separate nerve, making detection by MRI difficult(14).

The pathogenesis of CND involves failure in development of the nerve either completely (aplasia) or partially (hypoplasia), or as a result of post-developmental degeneration(13). Although the underlying mechanisms remain unclear, proposed theories include vascular insult, uncontrolled apoptotic nerve remodeling, neurotrophic infections, and global metabolic and neurologic disorders(13). Children with cochlear nerve hypoplasia who have unilateral residual hearing on the affected side require long-term audiometric follow-up as the hearing threshold may deteriorate in as neural degeneration continues.

Cochlear implantation in children with CND continues to be a controversial topic. While most children with CND with CI rarely achieve open-set speech perception, many do benefit in terms of auditory awareness(15). Complete absence of cochlear nerves bilaterally is a clear predictor of poor CI candidacy(13), however in the setting of cochlear nerve hypoplasia, stimulation of the even a small number of nerve fibers may provide benefit given the plasticity of the auditory cortex in young children(14). Parents should be counseled extensively prior to proceeding with cochlear implantation regarding the expected outcomes and be prepared to implement supplemental, non-verbal communication modes early in the rehabilitation process.

Cochleovestibular Anomalies

Cochlear implantation of cochleo-vestibular anomalies presents several challenges. In 1987, Jackler et al. reported that approximately 20% of congenital SNHL is related to anomalous inner ear anatomy evident on radiography(16). In early era of cochlear implantation, cochleovestibular malformations were considered a contraindication to implantation due to concerns about proper electrode insertion, array stability, absent or dysfunctional neurons which might preclude significant auditory perception, and the increased risk of complications such as facial nerve injury and cerebrospinal fluid leak(17). A better understanding of cochleo-vestibular malformations, in combination with improved cochlear devices and surgical techniques, have resulted in successful implantation in this patient population.

Cochleo-vestibular anomalies are typically classified into 7 categories: complete cochlear and labyrinthine aplasia (Michel deformity), cochlear aplasia, common cavity of the cochlea

and vestibule, hypoplastic cochlea, Incomplete Partition Type I (IP-I, cystic cochleovestibular malformation or <1.5 basal turns), Incomplete Partition Type II (IP-II Mondini deformity or 1.5–2.75 basal turns), and enlarged vestibular aqueduct (EVA)(16–18) (Figure 3). Thin-section HRCT facilitates the identification of the type of malformation and T2-weighted MRI can help determine cochlear patency and partitions.

Cochlear implantation can be successfully achieved in nearly all cochleovestibular malformations, the exceptions being complete labyrinthine and cochlear aplasia. The standard transmastoid posterior tympanostomy (facial recess) approach to the middle ear can be used to place cochlear implants in patients with most inner ear malformations. The exception is common cavity malformation, which may be accessed directly via a transmastoid labyrinthotomy. However, special care must be used when implanting common cavity malformations due to the lack of a central modiolus and the inability to predict the location of the cochlear nerve ganglion cells. Modiolar conforming electrode arrays should be avoided in such cases(15).

Additional intraoperative challenges include cerebrospinal fluid (CSF) leaks, electrode malposition, incomplete electrode insertion, and aberrant facial nerve course. The risk of CSF leak is correlated with the severity of malformation(17). CSF gushers (a brisk flow of perilymph/CSF from the cochleostomy) can be well controlled with tight packing of the cochleostomy around the electrode with connective tissue(15). Preoperative pneumococcal vaccination is required for all CI patients but is especially important in patients with malformations in preventing postoperative meningitis.

Several anatomic factors may make electrode insertion difficult or result in electrode malposition. The absence of the cochlear lamina cribrosa may lead to inadvertent implantation in the IAC, potentially resulting in worsened hearing, vertigo, stimulation of the facial nerve, and CSF leakage. The electrode array should be inserted until resistance is met and no further. The depth of insertion can be predicted on preoperative imaging. If proper insertion is uncertain, intraoperative fluoroscopy, computed tomography, or transorbital x-ray can be used to confirm placement(15, 19). Some authors advocate specific electrode characteristics for each anatomic variant (i.e. non-conforming vs conforming electrode array for IP-I and IP-II, compressed arrays for extremely short cochleae and cystic cochlear variants), however a thorough pre-operative preparation with a variety of electrode arrays immediately available in the operating room is advisable.

Aberrant facial nerve course is associated with up to 14% of cochleovestibular malformations. Given this anatomic variability, special care is required to identify the facial nerve course intraoperatively(20). Facial nerve monitoring and intra-operative stimulation may be extremely helpful in nerve identification and protection and is highly advisable.

Although many children with malformations benefit significantly from CI, the severity of malformation is inversely correlated with speech perception performance. Children with EVA, IP-I, and IP-II tend to perform very well while those with common cavity or hypoplastic anomalies tend to have poorer speech performance due to reduced neural stimulation(15, 17, 20, 21). Electrode selection which is individualized to patient specific anatomic features may be desirable in patients with these anomalies. Of note, the presence of an intraoperative perilymph gusher does not significantly impact speech perception performance post-CI(21, 22).

Associated Syndromes

There are more than 400 genetic syndromes that affect hearing and up to 30% of pre-lingual deafness is due to syndromic causes(23). Each syndrome is associated with unique co-

morbidities that may impact CI candidacy. In general, significant improvement in speech comprehension is achieved when children receive CIs before the age of 2, therefore early identification of associated syndromes and appropriate management of associated comorbidities is paramount to achieving safe and effective outcomes in this population.

Usher syndrome is the most common autosomal recessive (AR) syndromic cause of hearing loss, and is associated with vestibular dysfunction and progressive visual impairment due to retinitis pigmentosa. Usher type 1 patients benefit little from amplification, and in these patients CI should be considered before age of 3 to promote successful speech and language development prior to severe visual loss(24). Patients with Usher types 1 and 3 who receive CIs demonstrate improvement in sound recognition and speech detection as well as subjective improvement in quality of life(25, 26).

Pendred syndrome is the second most common AR syndromic cause of hearing loss and inner ear dysplasias. It is associated with a euthyroid goiter. The progression of hearing loss is typically stepwise and can be associated with minor head trauma. All patients with Pendreds have EVA, but this typically does not affect electrode insertion(27, 28). The most common intraoperative complication is CSF gusher which most frequently can be controlled at the time of CI; post-operative speech performance post-CI appears to be unaffected(21, 22) (Figure 4).

Jervell and Lange-Nielsen syndrome (JLNS) is the third most common AR syndromic cause of hearing loss. It is associated with a prolonged QT interval on electrocardiogram which can cause syncopal episodes or sudden death. Hearing loss results from mutation in the potassium channels that regulate endolymph ion concentrations in the stria vascularis(29). Cardiac assessment is imperative in the setting of borderline or abnormal QTc, positive family history, or history of unexplained falls, syncope, or seizures. With careful cardiac precautions during anesthesia, CI can be successfully performed in children with JLNS with good post-implant auditory performance results(24).

Waardenburg syndrome (WS) is the most common autosomal dominant syndromic cause of hearing loss. Patients have variable expressivity of SNHL and pigmentary abnormalities including white forelock and heterochromic irides. Hearing loss results from failure of neural crest cells migration to form the stria vascularis(24). Because WS children typically have normal intelligence and their hearing loss is cochlear in nature, their post-implant results have generally been excellent. Many are able to achieve speech perception skills comparable to patients with nonsyndromic SNHL(30, 31).

CHARGE syndrome is a rare congenital disorder with multi-organ involvement initially described in 1981. Since then, the diagnostic criteria have evolved to include partial and atypical forms of the disease. The three major diagnostic criteria are coloboma, choanal atresia, and hypoplastic semicircular canals (SCCs)(32). More than 90% of children with CHARGE have hearing loss, many of which are of mixed type. Nearly all CHARGE patients have SCC aplasia and vestibular dysplasia; CND and incomplete cochlear partitioning are common as well(28) (Figure 5). Preoperative imaging is necessary for appropriate candidate selection as well as for surgical planning. The surgeon should be aware of absence of typical anatomic landmarks, particularly the absence of the semicircular canals and aberrant facial nerve course, and abnormal cochlear anatomy which could necessitate an alternate cochleostomy site. Despite the many other medical issues associated with CHARGE syndrome during the perinatal period, it is crucial to identify hearing loss and start appropriate rehabilitation early. Cochlear implantation may be recommended earlier in CHARGE children due to presence of visual impairment which further challenges effective communication(33). Post-implantation results for CHARGE children vary

depending heavily on the extent of additional co-morbidities. In most cases, open-set speech is not achieved; however, measurable auditory benefits may be apparent in the form of sound awareness and improved quality of life(34).

Children with multiple medical problems and developmental disorders

Up to 30–40% of children with SNHL are affected by additional disabilities including cognitive, visual or motor dysfunction, and behavior, or development disorders; most of these children were considered unsuitable for CI until recently(35). With the expanding CI selection criteria, a substantial number of children with complex medical problems are now receiving CIs and have been reported to have measurable progress in speech intelligibility, auditory skill development, and language skills(36). Although children with developmental delays consistently scored lower in receptive and expressive language skills even at 3 years post-implantation when compared to the typically developing children with SNHL, many still benefit considerably from implantation(37). The definition of “benefit” can vary from achieving open-set speech perception to simply environmental sound awareness, improved quality of life, or satisfaction of the child and family. The decision for CI in a child with multiple medical or developmental issues needs to be tailored for each individual patient and family and requires special consideration. Realistic expectations for post-CI performance are integral to appropriate counseling and decision making.

Congenital cytomegalovirus (CMV) infection is the most frequent infectious cause of pediatric hearing loss. The definitive diagnosis may be easily missed as more than 90% of those affected do not show typical clinical symptoms. Hearing loss is variable and may be cochlear or central in nature(38). Variability in associated psycho-neurological disorder such as autism, learning disabilities, or cognitive delays can make CI performance unpredictable(39). Nevertheless, CI can successfully address CMV-related hearing loss when the goal is to improve overall communication.

Special considerations in CI in children with Down Syndrome include developmental issues, higher prevalence of otitis media, under- or non-aerated mastoid cavities, and hypoplastic cochleae(40). Recurrent otitis media can be a challenge to control, but can be managed with ventilation tubes so as to not delay the CI process and to reduce the risk of meningitis. Caregivers should be aware that although it is possible to achieve improvement in auditory performance, the rehabilitation process may be prolonged.

Hearing loss has been reported to affect 12% of children with cerebral palsy (CP), but many are diagnosed late due to the severity of other co-existing medical issues and the difficulties with assessing audiometric thresholds(41). Not surprisingly, those with mild or no cognitive impairments and early implantation tend to perform well with CIs. However, even with severe impairments, speech perception can still be achieved with implantation over time(41). When considering CI for a child with CP, appropriate selection of receiver position should take into account the natural head posture in relation to wheelchair head supports. Improper positioning can lead to partial or non-use of the device due to difficulties with compliance or retention.

Conclusion

CIs are accepted as the standard of care for restoration of hearing and improved speech development in children with severe to profound hearing loss. Some children with SNHL who are appropriate candidates for CI may have unique audiologic, medical, and anatomic characteristics. Special consideration is required when undertaking hearing rehabilitation and cochlear implantation in these unique populations.

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Key Summary Points

- Children with ANSD should be offered CI if a trial of amplification fails to show benefit, however post-implant speech perception outcomes can be variable due to heterogeneous underlying etiologies of the disorder.
- Most children with CND with CI rarely achieve open-set speech perception, but many can benefit in terms of auditory awareness.
- Most cochleo-vestibular malformations are amenable to CI, and many children show post-implant benefit with speech perception performance correlating with the severity of malformation.
- Congenital syndromes with associated hearing loss present with unique co-morbidities that should be identified early for appropriate and timely CI candidacy evaluation.
- The decision for CI in children with multiple medical or developmental issues needs to be tailored for each individual patient and family with appropriate counseling regarding realistic expectations for post-CI performance.

Figure 1.

Auditory brainstem response testing diagnostic for auditory neuropathy spectrum disorder. It shows no response or flat auditory brainstem response with alternating clicks, mirror image cochlear microphonics with positive and negative polarity clicks, and a no response auditory brainstem with a no sound run.

Figure 2.

Cochlear nerve deficiency. (a) Axial temporal bone computed tomography suspicious for bilateral cochlear nerve deficiency as evidenced by narrow bony cochlear nerve canals. (b) Axial MRI, constructive interference in the steady state (CISS) sequence demonstrating bilateral cochlear nerve deficiency. (c) Sagittal MRI, CISS sequence of the internal auditory canal demonstrating four nerves within the internal auditory canal (left; normal) versus one nerve within the internal auditory canal (right; cochlear nerve deficiency).

Figure 3.

Radiographic features and classification scheme and Michel aplasia. (a) Radiographic feature and classification scheme. The upper row (A, B, C) shows examples of inner ear malformations from the incomplete partitioning spectrum (I–III). Incomplete partitioning refers to normal cochlear dimensions with decreased or absent partitioning (type I); apical fusion with present interscalar septum (type II), or X-linked stapes gusher syndrome (type III). The middle row (D, E, F) demonstrates anomalies from the hypoplastic spectrum, in which the cochleae have smaller overall dimensions (subclassification as type I: bud like; type II: cystic hypoplasia; type III: cochleae with less than 2 turns). G shows a cystic cochleovestibular anomaly, and H shows a common cavity malformation, both from the

cystic spectrum. Reproduced from [19]. (b) Michel aplasia, courtesy of Dr. Craig Buchman, University of North Carolina at Chapel Hill, Chapel Hill, NC.

Figure 4. Axial computed tomography and axial constructive interference in the steady state (CISS) sequence MRI. (a) Axial computed tomography demonstrating enlarged vestibular aqueduct. (b) Axial CISS sequence MRI demonstrating enlarged endolymphatic ducts and sacs.

Figure 5.
Axial computed tomography demonstrating absent semicircular canals in child with CHARGE association.