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REVIEW ARTICLE

Cochlear Implants in Children with Inner Ear Malformations, A Review of Current Literature.

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ABSTRACT

Cochlear implants (CIs) are a key option of hearing rehabilitation in certain children. Scientific, surgical and technological advances in CI technology have enabled implantation in a significant number of children with sensorineural hearing loss (SNHL). There are established criteria to characterize appropriate patients, improving successful performance post operatively by standard metrics of speech perception. Clinical outcomes are, however, modified by several patient-related. An increasing body of evidence in such domains has resulted in expansion of candidacy criteria. Approximately 20% of all congenital hearing loss is associated with inner ear malformations (IEMs). Implantation of such children was associated with uncertain surgical and clinical success, and hence was limited in the early years of CI surgery. To date, the literature regarding outcomes in this special population has varied. Studies have been mixed when comparing children with IEMs to those with normal anatomy regarding success with CI. Results may be related to the type and severity of IEM. There may also be differences in children with congenital deafness versus progressive hearing loss. The current data are limited as there is not a standardized testing paradigm for evaluation in children. The variability in the data suggests further research is required to fully understand the nuances in management of these complex children. The goal of this review is to discuss surgical management and outcomes of children who meet criteria for CI in the setting of an IEM.

Introduction:

Cochlear implants (CIs) are a key option of hearing rehabilitation in certain children. Scientific, surgical and technological advances in CI technology have enabled implantation in a significant number of children with sensorineural hearing loss (SNHL). There are established criteria to characterize appropriate patients, improving successful performance post operatively by standard metrics of speech perception. Clinical outcomes are, however, modified by several patient-related factors including age at implantation[1], onset and duration of hearing loss[2], cochleovestibular anatomy[3], physiology of the auditory nerve[4], presence of neurodevelopmental disorders[5], level of psychosocial support[6] and the quality of postoperative rehabilitation efforts.[7] An increasing body of evidence in these domains has resulted in expansion of candidacy criteria. However, about 20% of all congenital hearing loss is associated with inner ear malformations (IEMs).[8] Implantation of such children was associated with uncertain surgical and clinical success, and hence was limited in the early years of CI surgery. The goal of this review is to discuss surgical management and outcomes of children who meet criteria for CI in the setting of an IEM.

Use of CI

Cochlear implantation was initially described in the 1980s with single channel devices.[9] Over time, multi-channel devices were developed with an ability to implant them in children.[10] These implants were initially exclusively offered to children with normal anatomy. It was not until recently that children with IEMs were considered candidates for a CI.[11,12]

Small case series from the 1990s initially established the safety and feasibility of CI in children with IEMs. These studies also showed that satisfactory speech perception could be achieved in most instances.[9–11] A systematic review of CI in children with abnormal anatomy found that the most common IEM are incomplete partition (IP) unspecified type (30%), enlarged vestibular aqueduct (EVA) (29%), cochlear hypoplasia (11%), common cavity (11%), IP-II (7%), and IP-I (6%).[13] These findings are in agreement with a more recent publication of 102 children undergoing CI which noted an overall IEM rate of 27%.[14] In this study, cochlear dysplasia was the most common anomaly (30%), followed by vestibular dysplasia (18%), enlarged vestibular aqueduct or endolymphatic sac (17%), and labyrinthitis ossificans (14%).[14] Less common findings included cochlear nerve hypoplasia (7%) or aplasia (6%), stenotic internal auditory canal (IAC) (3%), vestibular nerve hypoplasia (2%) and cochlear aplasia, atretic IAC, or abscess (2%).[14]

Operative modifications

Children with IEMs undergoing CI are at risk of certain complications due to the atypical anatomy which often necessitates modification of surgical techniques. Early studies discussing CI in children with IEMs noted increased rates of cerebral spinal fluid (CSF) gushers and an anomalous location of the facial nerve.[14–17] Rates of perilymphatic gushers range from approximately 5-10% of children with an IEM at time of surgery.[3,13,14,18] In addition to a CSF gusher, these children are also at risk for a low flow CSF leak at a rate of approximately 32%. Interestingly, the degree of CSF leak risk does not vary based on the degree of cochlear malformation.[13] CSF leaks and perilymphatic gushers should be addressed intraoperatively. Management strategies include use of autologous fat and fascia[19], such as a fascial donut around the electrode.[14] Other authors have reported use of temporalis muscle or temporalis fascia to seal the leak.[20] There are also specialized electrodes with a 'cork' feature which is meant to block the chochleostomy to prevent CSF leak.[19] Rarely, the middle ear needs to be obliterated to prevent persistent leak.[14,20]

The position of the facial nerve must be assessed when evaluating a patient with an IEM for CI. An anomalous facial nerve may be expected in approximately 25% of cases, with higher rates in children with severe dysplasia as compared to mild or moderate.[13] The position of the nerve may alter the ability to fully insert the electrode as well as dictate the approach for placement to limit the risk of facial nerve injury. This is critical because although rare, paralysis or paresis are possible.[12,16] Approaches which may be utilized for CI placement in these cases include the standard facial recess approach, transaditus approach, or a combination.[12]

Other potential complications of CI insertion in the setting of an IEM include difficult insertion[12,14], kinked electrodes, needing a drillout[14], and incomplete insertion leading to misplaced electrodes outside of the cochlea.[21,22] The electrode may also extrude from an abnormal

cochlea and cause facial nerve stimulation in the setting of an aberrant nerve location.[11]

Outcomes

An important consideration when discussing CI placement is the expected outcome regarding speech and language. In children, these factors are impacted by multiple variables, complicating decision making and counseling. It has been established that age at time of implantation is significant[1] with a younger age at time of placement being important to promote development of the auditory system.[23] This is independent of the amount of their residual hearing at time of implantation.[24] Medical comorbidities[23] and neurodevelopmental disorders[4] play a key role in a child's ability to perform after placement. Psychosocial support[6] and post operative rehabilitation[23] are critical in influencing a child's results. A recent article noted children with IEMs require more frequent audiology evaluation for mapping adjustments compared to those with normal inner ears to have the best outcomes.[25]

The literature regarding auditory performance and speech production has been evolving. In children with normal inner ear anatomy, the expected success after cochlear implant regarding speech perception ranges from 57% to 85%.[7,26–28] Children with IEMs typically have less success after CI. Eisenman et. al established that speech perception performance in children with IEMs is delayed when compared to normal counter parts.[29] Isaiah et. al also noted poor performance in all children with IEM, aside from those with enlarged vestibular aqueducts or endolymphatic sacs.[14] This finding was described in several other studies. Buchman et. al found that children with IEMs were slower to develop speech perception than those without malformations.[11] This study also identified only 46% of children were able to develop some open-set speech perception.[11] In addition, they noted differences in performance based on the type of malformation. Children with EVA, IP, and dilated vestibule (Mondini malformations), isolated EVA, and partial semicircular canal aplasia did well with open set speech perception, while those with more severe malformations, such as cochlear hypoplasia and common cavity, performed at lower levels.[11] A systematic review of CI with IEMs noted 45% of implanted children had speech perception data.[13] Patients with mild or moderate IEMs were able to perform significantly better (84%) on open word testing compared to those with severe anomalies (54%) using PBK (phonetically balanced kindergarten) and GASP (Glendonald auditory screening procedure) tests.[13] Loundon et. al studied CI in children with IEMs using the test d'evaluation des perceptions (TEPP), which is similar to PBK, and Lafon children's list for speech perception and linguistic testing.[30] This study found that the majority of children had an improvement in perception and language after implantation as early as the first year, but there was a significant difference between children with congenital (including those with IEMs) versus progressive deafness.[30] The authors concluded that outcomes in children with IEMs are less predictable.[30] Rachovitsas evaluated CI in children with and without IEMs and evaluated post operative hearing outcomes using the listening progress profile (LiP), capacity auditory performance (CAP) and speech intelligibility rating (SIR) tests.[31] In their cohort, children with normal anatomy had significantly better scores on each test compared to those with IEMs.[31] However, those with IEMs did show improvement compared to pre-operative testing.[31]

In contrast, Melo et. al noted similar outcomes between children with IEMs and normal anatomy as assessed using the European Portuguese word discrimination tests, CAP and SIR tests.[32] Celik et. al did not find a significant difference in outcomes on the listening progress profile test (LPPT), monosyllabic trochee polysyllabic test (MTP) and evaluation of auditory responses to speech (EARS) tests for children with IEMs compared to children with normal inner ear anatomy.[33] Outcomes regarding speech and language may vary based on the type of anomaly[11], with some studies noting more severe malformations having poorer outcomes.[11,13,34,35] Other studies have not found a difference based on anatomy.[36–38] However, it is important to note that most children will have some benefit after implantation.[31,39]

The current data are limited as there is not a standardized testing paradigm for evaluation in children. The mentioned studies used multiple testing methods, making comparison challenging. Additionally, there may be some subtleties in how inner ear malformations were characterized or diagnosed at different institutions, with differences in imaging protocols for example. Follow up times and interval evaluations are not consistent, which makes outcome assessments difficult. As criteria for CI

change and decisions are made with improving technology, patients included in some of these series may be managed differently in the modern era, which may also affect results. The variability in the data suggests further research is required to fully understand the nuances in management of these complex children.

Conclusion:

In the setting of congenital profound SNHL, the prevalence of inner ear anomalies is about 20% in children who are candidates for cochlear implantation. Heightened awareness is necessary to ensure anticipation and preparation prior to surgery due to potential intraoperative surgical challenges such as perilymphatic gusher, which may impact outcomes following surgery. Modifications to surgical technique such as use of fascial packing and specialized electrode arrays may be beneficial in reducing morbidity. Pre-operative assessment of facial nerve position is also critical to avoid injury with paralysis or paresis. Related to this, while the authors of this review use primarily MRI for imaging analysis of children with SNHL, when severe anomalies are identified on MRI, we recommend also obtaining a temporal bone CT to assess the course of the facial nerve. The literature regarding speech and language outcomes following cochlear implantation in children with inner ear anomalies varies, indicating a knowledge gap where future studies are required. However, the benefit of sound awareness even in situations where speech recognition may not occur may support offering a CI to these patients. Counseling families prior to surgery on realistic expectations and risks of the procedure is necessary for patient safety and caregiver satisfaction.

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