# **Current Issues With Pediatric Cochlear Implantation**

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# Introduction

Cochlear implants (CIs) have enabled children with severeto-profound sensorineural hearing loss (SNHL) to access sound sufficient to derive a clear functional benefit. As a result, CIs have gained wide acceptance as an ideal treatment option for deafness in the pediatric population. As with many issues related to the medical and surgical care of young children, cochlear implantation in a child has various nuances that require special attention beyond the standard considerations for evaluation and surgery in adults with hearing loss. Resulting from improvements specifically related to CIs and post-implant rehabilitation, as well as general technological advances, such as better imaging resolution and greater understanding of the significance of genetic testing, various aspects of cochlear implantation in children remain a moving target. There has been an evolution in the approach to patient evaluation, changes in candidacy guidelines and vaccine requirements as well as a lowering of the age requirement for surgery. In addition, there are special considerations to account for in children due to differences in the anatomy and physiology of infants. Current criteria and guidelines of cochlear implantation in children as well as, special surgical considerations and the outcomes of CI surgery in children will be discussed.

# **Patient Evaluation**

Comprehensive evaluation for cochlear implantation requires thorough medical, surgical, audiological, and developmental assessments performed by a multi-disciplinary care team. Initial medical evaluation includes a complete history and physical with additional focus on birth history, family history, and otologic history. Detailed birth history should focus on risk factors associated with SNHL: prematurity, low birth weight, low Apgar score, anoxia, history of intubation, aminoglycoside/loop diuretic administration, meningitis, sepsis, hyperbilirubinemia, and neonatal intensive care unit stay. Congenital SNHL has been associated with in utero exposure to teratogens including drugs of abuse, alcohol, thalidomide, and mercury or infections like toxoplasmosis, varicella, syphilis, rubella, herpes, or cytomegalovirus (CMV). Congenital CMV infection is the most common non-genetic cause of SNHL-screening for CMV infection should be considered

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in all infants that fail the newborn hearing screen [1].

It is important to discuss family history of hearing difficulties as roughly half of pediatric SNHL is genetic in origin. Close relatives with history of early onset or congenital SNHL should increase suspicion for genetic etiology. Genetic causes of hearing loss may be associated with a syndrome, or it can exist in isolation. Non-syndromic genetic hearing loss is more prevalent than syndromic. The most common defect leading to non-syndromic genetic hearing loss is mutation in the gap junction protein B2, also referred to as connexin 26 [2-6]. The mutation is inherited in an autosomal recessive pattern and results most commonly in severe-to-profound SNHL. These patients are considered excellent CI candidates with some studies demonstrating superior outcomes in comparison to patients with acquired hearing loss [7,8]. Many patients with a syndromic etiology of SNHL are also good CI candidates, including those affected by Pendred, Waardenburg, Usher, branchio-oto-renal (BOR) and Jervell Lange-Nielsen (JLN) syndromes.

Cochlear implantation in patients with CHARGE should be approached with caution as outcomes are affected by the associated developmental delay [9]. In disease states that can cause both vision and hearing loss, like Usher syndrome or Refsum disease, CI should be completed prior to severe vision loss when possible [10]. All patients with congenital SNHL without otherwise identified etiology should undergo an electrocardiogram due to the association with JLN with prolonged QT interval, arrhythmias, and syncope. If a diagnosis of JLN is confirmed, cardiology consultation and family testing should be completed as cardiac intervention may be required preoperatively and medical treatment of the cardiac electrophysiologic anomaly can reduce risk of sudden cardiac death [11].

Blood tests and other laboratory evaluations can be considered in the correct clinical setting but have low yield when performed indiscriminately. Kılıç, et al. [12] reviewed 150 children who underwent complete blood count, thyroid function study, treponemal titer, cholesterol, triglyceride, chemistries, and EKG as part of routine workup of congenital SNHL. They found 45 abnormalities in the results but none that contributed to the etiology of hearing loss. In a similar study, Preciado, et al. [13] also found routine laboratory evaluation in a simultaneous testing approach carried an extremely low diagnostic yield. However, they additionally found that 22% of patients with severe-to-profound SNHL had abnormal connexin screening. The diagnostic yield was lower in patients with less severe SNHL however there were positive screens with every degree of SNHL. In this same series, every child underwent temporal bone imaging with either computed tomography (CT), magnetic resonance imaging, or both. The diagnostic yield of CT was higher in unilateral SNHL (36.7%) than in bilateral (24.7%). Of the abnormal imaging findings, enlarged vestibular aqueduct was the most common finding (67.45%). Thus, a targeted, stepwise approach is recommended when evaluating the etiology of congenital SNHL.

# **Candidacy Guidelines**

Children with bilateral SNHL or unilateral hearing loss (UHL)/single-sided deafness (SSD) are considered candidates for implantation. UHL is generally described as any amount of hearing loss in one ear with normal hearing in the contralateral ear. SSD is generally defined as profound SNHL in one ear with normal to mild SNHL in the contralateral ear although definitions vary in the literature for both conditions. CI was first approved for children 2 years and older with bilateral SNHL in 1990. Most recently in 2020, the US Food and Drug Administration (FDA) approved the Nucleus 24 Cochlear Implant System (Cochlear Americas, Sydney, Australia) for infants 9 months and older. At present, each manufacturer has unique FDA approved device labelling based on age and hearing characteristics. As a whole, candidacy for children is far more stringent than for adults. Word recognition testing thresholds exist for children who can participate.

Advanced Bionics (CA, USA) produces the HiResolution Ultra 3D implant for children 12 months and older with bilateral profound SNHL greater than 90 dB hearing level. The pediatric labelling requires the patient score  $\leq$ 20% words correct on a standard lexical neighborhood or multisyllabic test in the best aided condition.

Cochlear Americas produces the Cochlear Nucleus Cochlear Implant System for infants and children 9 months or older. Patients 9 months to 2 years of age must have profound bilateral SNHL to be candidates, whereas patients 2 and older can have severe-to-profound bilateral SNHL. The FDA approved labelling requires  $\leq$ 30% word recognition performance in best aided condition. This device has additional approval for UHL/SSD in children 5 years or older with limited benefit from appropriate amplification demonstrated by trial of contralateral routing of signals (CROS) aid or other device and monosyllabic word score  $\leq$ 5%. Additionally, they must have a duration of profound deafness of 10 years or less.

Med-El (Innsbruck, Austria) produces the OPUS 2 cochlear implant system for children 12 months and older with thresholds  $\geq$ 90 dB at 1,000 Hz and  $\leq$ 20% word recognition performance in best aided condition. The device is also approved for UHL/SSD with the same requirements as the Cochlear device.

Candidacy for cochlear implantation has gradually expand-

ed as further evidence of its safety and efficacy have emerged. However, there is general consensus that current candidacy guidelines for pediatric patients are too strict. The Joint Committee on Infant Hearing recommends appropriate hearing intervention by the third month of life [14]. Park, et al. [15] argue that current FDA guidelines cannot meet this standard for infants younger than 9 months of age with no response on auditory brainstem reflex as non-CI interventions cannot provide enough access to sound to be considered an "appropriate intervention."

Limited data exists regarding implanting children who do not otherwise meet FDA criteria. Carlson, et al. [16] described their experience with implanting children between 2–17 years old with <70 dB HL pure-tone average (PTA) in the ipsilateral ear and <2 years old with <90 dB HL PTA. In both groups, children who could participate in best aided word recognition testing performed better than 30%. In their series of 51 children with an average follow-up duration of 17.1 months, they demonstrated a statistically significant increase in speech recognition score in the ipsilateral (mean 62.8%) and binaural (mean 39.9%) conditions. Children who could not participate in speech recognition testing showed a significant increase in language development questionnaire score (mean 26.5%). The authors conclude that expanding CI candidacy for children should be considered based on their outcomes.

Lovett, et al. [17] conducted an observational study of 71 children receiving audiological care in the United Kingdom and found that an unaided four-frequency PTA of 80 dB HL or poorer in both ears carried four times odds of having a better outcome with an implant than with hearing aids. The threshold of 4:1 odds ratio had been previously used to define criteria for implant candidacy in the United Kingdom, and as a result, this study was designed to influence policymaking.

An exception to the age requirement is made for patients with post-meningitis SNHL. Delaying implant in this scenario risks cochlear fibrosis and obliteration of the cochlear lumen making attempts at future implantation difficult or impossible by standard surgical technique.

## Vaccine Requirements

Cochlear implantation is associated with increased risk of pneumococcal meningitis [18-20].

Reefhuis, et al. [21] described a 30-fold risk increase for pneumococcal meningitis post-implantation, with compounded risk when using an electrode positioner. The inner ear communicates with the cerebrospinal fluid (CSF) space via the cochlear aqueduct connecting the basal turn to the posterior fossa and via microscopic canaliculi connecting the basal turn to the internal auditory canal. Children with cochleovestibular malformation are at additional risk for meningitis due to increased rates of CSF gusher—31%-35%—depending on severity of dysplasia as found in a systematic review [22]. There is no evidence that CI patients have increased risk for *Hemophilus influenzae* type b (Hib) or meningococcal meningitis so the Centers for Disease Control (CDC) recommends routine vaccination based on age and other risk factors [23].

The CDC and Prevention Advisory Committee on Immunization Practices recommend children complete all recommended doses of pneumococcal vaccine at least 2 weeks prior to implantation. There are multiple formulations of the pneumococcal vaccine, the major difference is whether it is a pneumococcus conjugated vaccine (PCV) or pneumococcus polysaccharide vaccine (PPSV), as well as the number of strains covered. Children younger than 2 years old should receive PCV15 or PCV20 according to the routine childhood immunization schedule. Children 2-18 years old who have completed their childhood series require an additional dose of PCV20 or PPSV23 if they never received PCV20 in their original series. Children 5 years and younger who have not completed the childhood series require a unique catch-up series designed by their pediatrician. Children 6-18 years who have not completed their childhood series require either a dose of PCV20 or a dose of PCV15 followed by PPSV23. Children 2-18 years who completed their childhood series and do not meet the above exception do not require additional doses [24]. The vaccine series is complex due to multiple formulations, agerelated recommendations, and quantity of doses required. The CDC provides a quick reference called PneumoRecs VaxAdvisor which simplifies decision-making about individual patient's required vaccines [25]. While the CDC recommends administration at least 2 weeks prior to implantation, the Medicines and Healthcare Products Regulatory Agency of Great Britain does not recommend delaying implantation to complete the vaccination series. There are no human experimental studies that directly investigate the effect of pneumococcal vaccination on rates of meningitis in CI patients and further research in this area is needed [26].

#### Age at Implantation

As discussed in the prior section, the FDA-approved minimum age of pediatric cochlear implantation has decreased since its initial approval in 1990. These changes were supported by multiple studies highlighting the importance of early implantation and the resultant improvement in language and auditory outcomes, including comprehension and expression, educational achievement, functional performance, and quality of life [27-30]. In fact, the current American Academy of Otolaryngology Position Statement regarding pediatric cochlear implantation states that children with bilateral severeto-profound SNHL should receive CIs as soon as practicable and ideally within 6 to 12 months of age [31]. A multicenter study from Australia in 2016 reviewed 403 children with congenital bilateral severe-to-profound SNHL who underwent implantation prior to 6 years of age and examined the effect of age on open-set speech perception, language, and speech production [32]. Children implanted prior to 12 months of age had improved language standard scores and speech production scores compared to those over 12 months on standardized testing; those implanted at younger than 24 months also had improved open-set speech perception [32]. Similarly, Nicholas and Greers [33] compared children implanted at less than 12 months old to those at 12 to 18 months; they found that those implanted earlier had improved scores in receptive vocabulary, expressive and receptive language, as measured by the Peabody Picture Vocabulary Test-III and Preschool Language Scale-IV. Additional studies have demonstrated comparable findings [34-36].

### Surgical Considerations

#### Preoperative considerations

At present, a majority of pediatric cochlear implantations performed in the United States are done so in the outpatient setting. In support of this practice, a review by Alyono and Oghalai [37] in 2015 determined that pediatric otologic surgery, particularly cochlear implantation, could safely be performed without overnight admission and reduce overall hospital costs. According to several studies, the most common reason to admit patients for overnight observation is postoperative nausea and vomiting related to recovery from anesthesia, which occurred in roughly 14% to 23% of patients [38-40]. While Liu, et al. [38] found that over 90% of families were satisfied with outpatient surgery, some families would have preferred overnight observation, primarily for concerns regarding pain, nausea, and wound care questions.

Of note, a retrospective study of 579 pediatric CIs by Sivam, et al. [41] found a statistically significant, almost two-fold increased odds (odds ratio 1.95, 95% confidence interval 1.18– 3.28) of postoperative nausea/vomiting in patients undergoing bilateral cochlear implantation, primarily associated with increased operative times. Based on this, these patients should be considered for overnight observation.

The role of preoperative and postoperative antibiotics

The current literature does not draw any definitive conclu-

sions regarding the role of perioperative antibiotics, and no double-blinded randomized controlled trials have been performed. Anne, et al. [42] performed a systematic review of the current literature; the included articles were all low-quality evidence with heterogeneous outcomes measures, and thus, no definitive conclusions could be made. The majority of surgeons will administer one dose of an antistaphylococcal antibiotic intravenously within 20 minutes prior to incision. Postoperatively, a retrospective review by Farinetti, et al. [43] found that acute otitis media was the most common complication among pediatric CI patients, affecting 14 of 235 children (6.0%). These patients were all effectively treated with a course of oral antibiotics of amoxicillin-clavulanic acid 80 mg/kg/day or amoxicillin 70 to 100 mg/kg/day.

### Surgical procedure

Most commonly, a postauricular skin incision is made measuring 3–4 cm in length and roughly 0.5 cm behind the postauricular crease. After the skin incision, a single or double layer flap may be elevated. In the single layer flap, the incision is carried down through periosteum to bone and the soft tissue elevated with the periosteum. In the double layer flap, the superficial layer is elevated first, and a separate periosteal incision is made. The periosteum is elevated as an anteriorly based Palva flap and retracted anteriorly to expose the mastoid cortex.

A mastoidectomy is then performed. It is encouraged that the mastoidectomy cavity not be saucerized in order to aid in retention of the electrode array beneath overhangs within the mastoid cavity. A facial recess is then widely drilled once the horizontal semicircular canal and incus are identified. This is done until the round window niche is visualized. In pediatric patients, as is often described, the facial nerve may be located in a more lateral position than expected in adult patients. The width of the facial recess, however, was believed to be relatively constant from children to adults [44]. However, a recent study by Wojciechowski, et al. [45], using cone beam CT scans of 130 adults and 140 children, demonstrated 0.6-mm difference which was statistically significant (3.99±0.69 mm and 3.39±0.98 mm in children and adults, respectively).

In the case of anomalous facial nerve anatomy, the nerve is usually displaced anteriorly and medially. In a 2012 review by Pakdaman, et al. [22] of cochleovestibular anomalies, anomalous facial nerve anatomy was found in 25% of cases, including 54% of cases of cochlear hypoplasia. In cases of severe dysplasia, anomalies were encountered in 23 of 45 patients (51%), compared to 11% of cases of mild/moderate dysplasia. In these cases, the usual posterior border of the facial recess (i.e., the facial nerve) will not be reliable, and caution should be taken. The nerve often will instead turn directly into the hypotympanum and run inferior to the round window area.

Once the facial recess is opened, the round window niche may be identified, and a small 1 mm diamond burr can be used to remove the bony overhang. There is often a "false membrane" or layer of middle ear mucosa which can be removed to visualize the round window membrane. This may also be confirmed with gentle palpation of the ossicular chain through the facial recess to visualize the round window reflex. Once visualized, the scala tympani may be accessed via one of three approaches-the round window, extended round window, or cochleostomy. Many surgeons prefer the round window approach, whereby the membrane is opened with a sharp fine instrument. In the cochleostomy approach, a small drill is used to drill a cochleostomy anteroinferior to the round window membrane; in the extended round window approach, the bony round window overhang is removed with a drill and the window enlarged at its anteroinferior portion. This approach may be necessary in cases of unfavorable round window angles. The results of investigation suggest decreased rates of intracochlear fibrosis postoperatively via the round window approach. However, other findings regarding differences in audiologic outcomes are equivocal [46].

With the scala tympani opened, the electrode is then inserted in a methodical and controlled fashion to minimize insertional trauma. In general, the electrode should be directed inferiorly along the lateral wall of the basal turn in the case of lateral wall electrode insertions. Monopolar cautery should be avoided once the implant is opened and on the field, given the risk of damage to its electronic components. The round window or cochleostomy is then packed with periosteum, muscle, or fascia to seal the inner ear from the middle ear.

The incision is then closed in a multilayered fashion. Great care is taken to ensure that the Palva flap covers the mastoid cavity and ideally covers the area where the CI electrode exits the receiver-stimulator. Many surgeons will apply a pressure dressing for a 24-hour period.

#### Subperiosteal pocket and receiver-stimulator well

At present, many surgeons do not drill a "well" for the receive-stimulator when performing a cochlear implantation. Historically, surgeons would drill a depression in the skull in order to accommodate the receiver-stimulator, which had a much larger thickness/profile than at present. This may expose dura in the case of children with thin skulls as well as increase operative times during cochlear implantation [47,48]. Instead, surgeons at present have made several different modifications. One such modification is the direct subperiosteal pocket technique, in which a posterosuperior subperiosteal pocket is made with a periosteal elevator just larger than the size of the receiver-stimulator, allowing for appropriate positioning without the need for drilling or fixation. Sweeney, et al. [47] performed a retrospective chart review of 193 patients undergoing implantation with creation of this tight subperiosteal pocket and found an 18.9% decrease in operative time, as well as no evidence of receiver-stimulator migration [47-49]. Cohen, et al. [50] also reported on their outcomes with the subperiosteal pocket on six children  $\leq 1$  year old; the authors found no evidence of device migration in this patient population. However, concerns still remain regarding device migration necessitating revision surgery [51].

Another option employed by the senior author is the utilization of tie-down sutures secured using small plating screws placed in the calvarium. In this case, 3 mm self-drilling screws are inserted on either side of the receiver-stimulator, and a non-absorbable suture is secured to each screw. The sutures are tied together over the top of the receiver-stimulator. Similarly, tie-down sutures can be secured by drilling holes into the mastoid cavity on either side of the electrode, or a miniplate or other material may be secured over the device.

### Labyrinthitis ossificans

Ossification of the cochlea necessitates different surgical approaches depending on the degree of cochlear ossification. In cases of profound SNHL after meningitis, cochlear ossification may be rapid, and it is critical to expedite these patients to the operating room for cochlear implantation before ossification can occur [52]. Smullen and Balkany [53] detail 3 stages or degrees of ossification: I, round window niche only; II, inferior segment of basal turn up to 180 degrees; and III, more than 180 degrees of the basal turn. In the case of round window obliteration, the new bone can be picked away, drilled, or removed with a laser until patent scala tympani is visualized. In stage II, a drill-through procedure is performed, drilling in the area of the round window anteriorly up to 8 mm along the basal turn until the lumen is opened. In these cases, one must be aware of the carotid artery along the anterior wall of the basal turn of the cochlea, and small amount of bleeding may indicate proximity to the vasa vasorum of the carotid artery. Finally, in stage III, a scala vestibuli insertion can be performed, as it can be patent in some cases of scala tympani ossification. As another option, a double array electrode can be used; a basal turn drill-out procedure is performed, and second middle turn cochleostomy is drilled. The incus and stapes superstructure are removed for access anterior to the oval window, and a 1 mm diamond burr is used to drill anterior to the oval window but below the cochleariform process to limit risk to the facial nerve. One array is placed in the basal turn cochleostomy, and the second array is inserted through the middle turn cochleostomy either in a retrograde or anterograde fashion [54].

# **Speech Perception Outcomes**

In general, pediatric CI candidates demonstrate acceptable language development and speech perception outcomes and self-reported significant benefit to CI recipients [55]. This is evidenced by multiple long-term follow-up studies reporting exceptional usage rates of CIs at least 10 years post-implant, ranging from 88% to 96% in usage during waking hours [27, 56,57]. Uziel, et al. [56], in 2007 published a 10-year follow-up study of 82 prelingually deafened children undergoing implantation in France. In the 10-year follow-up, 79/82 (96%) reported always wearing their device due to significant benefit, with the remaining three children all deafened post-meningitis with either significant delay to implantation or total ossification of their cochlea intraoperatively. Mean scores for the Phonetically Balanced Kindergarten word test and word recognition in noise were 72% and 44%, respectively, and 66% developed intelligible connected speech [56]. In another 10year follow-up study, Beadle, et al. [57] found that 87% of the patients understood a conversation without need for lipreading, and 66% could use a telephone. Of the 30 recipients, 77% could use intelligible speech to the average listener. These studies also showed that the majority achieved university-level education and employment and could participate normally in society, though others have shown that employment rates may be decreased relative to normal hearing peers [27,58].

In comparing implanted children to their age-matched peers with normal hearing, Fitzpatrick, et al. [59] compared 22 children with bilateral moderately severe-to-severe SNHL using hearing aids to 21 children using CIs. They found no difference in open-set speech perception or in speech production, though they did find significant differences in domains of receptive vocabulary, language, phonological memory, and reading comprehension. In contrast, Wu, et al. [60] published a long-term follow-up study of 39 prelingually deafened children in Taiwan and found that receptive vocabulary and receptive language, expressive language, and total language measurements in Mandarin Chinese were in the low-average range of normal after implantation. Others have also demonstrated language development after implantation comparable to that of normal hearing children [55,61-63].

The current literature has also demonstrated audiologic benefits with bilateral simultaneous relative to sequential or unilateral implantation. Santa Maria and Oghalai [64] in their 2013 best practices review concluded that bilateral cochlear implantation was safe and should be performed when feasible, given improved outcomes and symmetry in central auditory pathway development. In fact, Wu, et al. [65] in 2023 performed electrically evoked auditory brainstem response testing on 58 children (33 implanted sequentially, 25 simultaneously); the authors found that latencies of waves III and V were significantly shorter between the first and second implanted sides in the sequential group, but were similar in the bilateral group. Language development is certainly improved in cases of bilateral implantation relative to unilateral implantation [66], but recent clinical data has also suggested that simultaneous implantation has improved outcomes over sequential implantation and that the interval between implants matters [67]. A retrospective study of 240 children with sequential implants by Kocdor, et al. [68] showed that a sequential implant should ideally be performed within 3-4 years of the first implant, and that very little speech recognition is achieved when the sequential implant is performed beyond 7-8 years apart. A prospective study including children with sequential implants, simultaneous implants, and normal hearing controls tested audiologic outcomes in spatial unmasking, whereby target speech and background noise are spatially separated. They found that patients with simultaneous implants scored significantly better than their sequential counterparts, with their performance almost approaching that of the normal hearing control group [69].

As previously discussed, current literature has shown that age is a significant driver in audiologic and performance outcomes in pediatric CI recipients. Age is an objective, easily measured factor. However, audiologic and performance outcomes after pediatric cochlear implantation are affected by a multitude of other variables. These factors include the etiology of hearing loss as well as the language environment in which a child is raised. Other socioeconomic and psychosocial factors play an impactful role in the ultimate outcome in these patients. For example, Quittner, et al. [70] evaluated parentchild interactions in CI patients in a prospective multicenter study and found that maternal sensitivity and cognitive stimulation predicted significant increases in oral language development, to the same degree that age of implantation did. In the same vein, maternal education has been identified as another primary driver in performance outcomes in children after CI, with one study showing increased implant usage with higher maternal education [71,72]. This is attributed to the richness of the language environment in which a CI recipient was raised, as characteristics of maternal language input such as mean length of utterance and expansions have been independently associated with rates of language development [71,73]. In addition, multiple studies using large administrative databases such as the Healthcare Cost and Utilization Project State Ambulatory Surgery Databases have shown an association between age of implantation and socioeconomic factors such as race/ethnicity and insurance, highlighting issues with access to implantation and the multitude of factors influencing outcomes [74-77].

#### **Conflicts of Interest**

The authors have no financial conflicts of interest.

#### **Author Contributions**

Conceptualization: Kenneth H. Lee. Investigation: all authors. Project administration: Kenneth H. Lee. Supervision: Kenneth H. Lee. Visualization: Donald Tan. Writing—original draft: Donald Tan, Rance J.T. Fujiwara. Writing—review & editing: Donald Tan, Kenneth H. Lee. Approval of final manuscript: all authors.

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