

# Cochlear Implants in Children: Recent Advances

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## ABSTRACT

Cochlear implants (CIs) are the best-performing neural prostheses today. Clinical data have demonstrated that early implantation facilitates advancements in auditory, cognitive and developmental milestones, enabling children to succeed in mainstream schools. With recent improvements in engineering design, signal processing, as well as surgical and rehabilitation techniques, CIs have ushered in expanded candidacy criteria. This review aims to provide a critical evaluation of recent developments in CI strategies --specifically within the areas of implantation of malformed inner ears, outcomes following bilateral CIs, implantation for single-sided deafness and newer, adjuvant biological therapies to augment CI technology.

**Keywords:** Bilateral Implants, Children, Cochlear implants, Inner ear malformation, New therapies, Single sided deafness.

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## INTRODUCTION

Since their introduction in the 1960s, cochlear implants (CIs) have undergone many advances, positioning themselves as probably the best-performing neural prosthesis available today. According to the most recent data from the National Institutes of Health, more than 300,000 individuals have received CIs thus far in the world, and of these, about 40,000 are children in the United States. From the development of the first successful commercial single-channel device, they have evolved into multichannel devices that are part of the national health programs of several countries. From simple perception of basic speech sounds alone with early CIs, these devices currently are able to deliver intelligible speech and even some musical information to the auditory system. This is the result of numerous advances in hardware design,

surgical technique, and signal processing. Concerted efforts from several disciplines, including engineering, acoustics, neurobiology, otolaryngology, and audiology, have ensured that the continued development of CI technology has resulted in significant benefits to children with profound sensorineural hearing loss. In addition, CIs have been recognized as a useful tool for studying the potential protective effects of patterned electrical stimulation on the developing auditory system. As a result, data from animal models have changed the manner in which CIs are used clinically to deliver auditory information to the brain.

Fifty years have passed since the first patients received commercial CI devices. Implantation criteria for both pediatric and adult recipients have undergone many changes over the years, keeping abreast with recommendations that include candidates not previously considered suitable for fitting of CIs. Current Food and Drug Administration (FDA) criteria have also largely discarded the premise that profound bilateral sensorineural hearing loss is a critical and sometimes the sole criterion for the indications of CIs in children. In keeping with a number of changes that have taken place in the realm of hearing restoration, this review examines the key themes of recent development within pediatric CI and aims to provide a synopsis for reference.

## BENEFITS OF EARLY IMPLANTATION— LESSONS FROM SENSITIVE PERIOD FOR CORTICAL DEVELOPMENT

The importance of acoustic input in the development of the auditory system cannot be overstated. In order to prevent potential irreversible changes resulting from lack of input, hearing restoration should only be focused on medical and surgical factors once minimum age criteria are met. A sensitive period seems to exist for restoration of hearing in congenitally deaf children, with a mean upper limit of ~3.5 years. Hence, there is a strong case to implant children as early as possible to optimize development of speech and language.<sup>1</sup>

An important tool for assessing the maturation of cortical auditory pathways is the cortical auditory evoked potential (CAEP), which is an averaged electroencephalographic trace recorded from the scalp in response to acoustic stimuli. The P1 component of a CAEP waveform (Figs 1A to D) is thought to reflect the maturation of thalamocortical pathways. Insertion of CIs within

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the critical period leads to near-normal development of the P1 waveform, whereas CIs fitted later lead to their aberrant development.<sup>2</sup> In addition, there is also strong evidence from physiological and imaging studies that point to recruitment of nonauditory cues, such as visual cues that take over secondary auditory association areas, leading to limited abilities to harness the full benefits of a CI in such a setting. Cortical rewiring that takes over in the absence of auditory input may lead to cross-modal takeover of the cortex and potentially other parts of the auditory pathway by senses such as vision.<sup>3</sup>

The age at onset of deafness appears to limit the extent to which individuals who are implanted in adulthood can realize the full benefits of CIs without utilizing concurrent visual information.<sup>4</sup> While these findings highlight the importance of early auditory experience, other factors, such as duration of implant use and targeted auditory training, can impact the performance of CI users. Indeed, there is more recent evidence that supports the use of specialized multisensory training to improve outcomes following cochlear implantation with behavioral and electrophysiological evidence of reversal of perverse sensory development.<sup>5</sup>

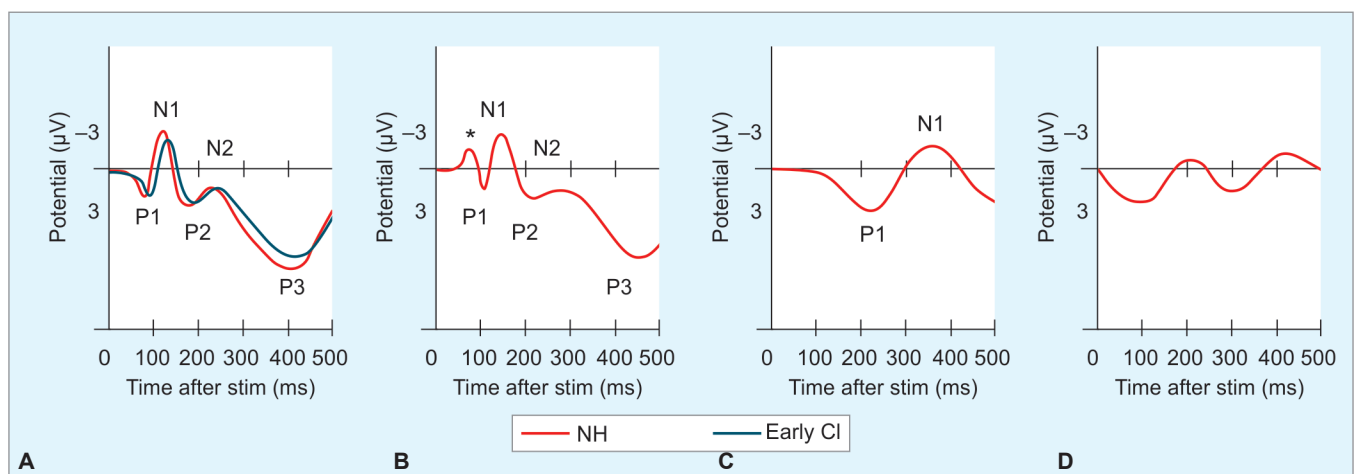
Although there is general consensus toward early implantation, guidelines from the FDA as well as pediatric anesthesiology literature<sup>6</sup> generally encourage waiting until 12 months of age prior to considering elective surgery, given the lack of a clear hearing advantage vis-à-vis perioperative anesthetic risks.

## BILATERAL COCHLEAR IMPLANTATION IN CHILDREN—STANDARD OF CARE?

A sensitive period for cochlear implantation has been recognized as a major determinant in guiding fitting

of devices in children, thereby affirming that early intervention is the standard of care in hearing restoration. Consistent with this recommendation, benefits were first seen in children with profound bilateral sensorineural hearing loss, in whom a CI was inserted in one ear and the contralateral ear with residual hearing was fitted with a hearing aid. These recipients were shown to have the ability to merge the inputs from the two different ears and derive significant binaural gain from this bimodal arrangement.<sup>7</sup> However, the question remained as to whether a hearing aid was the optimal way of providing input to the contralateral ear. While the rationale to save that ear for a more advanced intervention in the future has uncertain merit, providing a CI instead of a hearing aid to that second ear with minimal to no residual hearing seems to be well supported. Data from a recent study appear to substantiate this, with marked improvement seen in children with sequential bilateral CIs when tested for speech perception, sound localization performance, and overall quality of life.<sup>8</sup>

Outcomes following CI in infants and children are shaped by an interplay of factors including etiology, duration of deafness, and acquisition of language prior to implantation. These findings have been incorporated into technical reports and treatment guidelines established by government agencies throughout the world. Establishment of these guidelines thus excluded certain populations from receiving auditory rehabilitation using a CI, either unilateral or bilateral. Given the growing body of evidence, it is clear that early intervention using bilateral CIs can restore, at least partially, the sensory experience necessary for maturation of the neural circuitry responsible for processing binaural cues. In light of this, and as the concept of saving an



**Figs 1A to D:** Cortical auditory evoked potential waveforms in normal hearing and deafness. Cortical auditory evoked potential waveforms are plotted as a function of time after offset of stimulus in scenarios indicating different durations of deafness: (A) Normal waveform morphology (N1, P1, N2, P2) when development of hearing is normal through the sensitive period (black), and when cochlear implant (CI) is performed early (<3.5 years) within development (gray); (B) An abnormal negative peak (asterisk) in an unstimulated central auditory system following acute implantation and; (C) significantly increased latency of P1 in a partial stimulated auditory system (CI performed after 7 years of age), and (D) Polyphasic waveforms, sometimes seen in a reorganized auditory cortex. Figure adapted from Sharma et al<sup>2</sup>

ear for future interventions has yet to be justified, bilateral CI should be considered as the primary option for restoration of hearing in children with profound sensorineural hearing loss.

### COCHLEAR IMPLANTATION FOR SINGLE-SIDED DEAFNESS

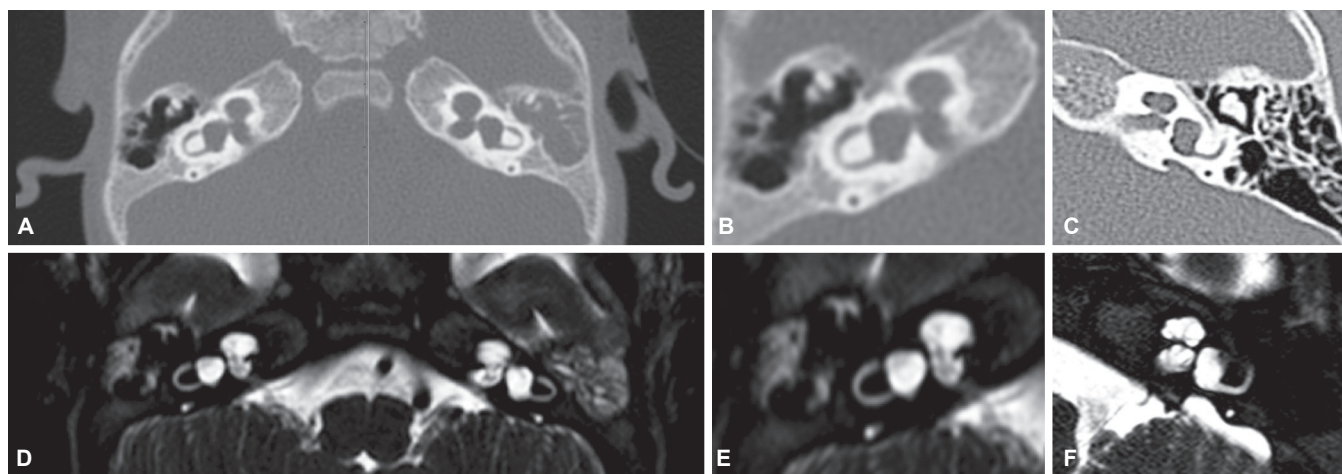
More recently, there is increased interest in CI as durable treatment for single-sided deafness, specifically when associated with incapacitating tinnitus. While the literature in this regard continues to evolve, the limited number of studies thus far has demonstrated sound localization and speech perception benefits in adults. Few studies in children also have shown rapid development of speech discrimination in the implanted ear, improvements in sound localization and speech perception in noise, and a high degree of patient satisfaction, hinting toward potential benefits in a wider population.<sup>9</sup>

### IMPLANTATION OF THE ANATOMICALLY MALFORMED COCHLEA

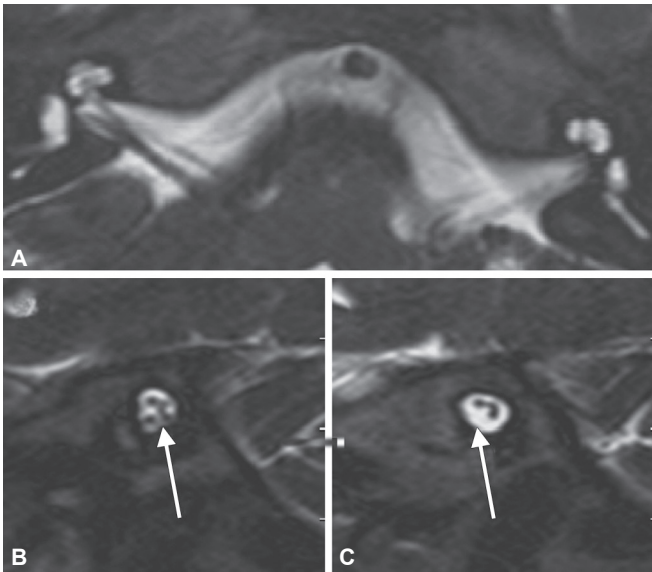
During the initial experience, implantation of the malformed cochlea represented challenges within three avenues—(i) the lack of resolution in preoperative imaging that precluded clear surgical planning, (ii) poor insertion geometry enforced by use of the standard CI array, and (iii) lack of data due to the relatively small number of patients with anatomic malformations. Since the first prominent report that described implantation in a small cohort of children with malformed cochlea, CI manufacturers have also introduced devices appropriate for a range of anatomic defects with the primary purpose of avoiding implantation trauma. Most initial studies hinted toward near-normal performance in children with specific coch-

lear abnormalities when compared with those without them. Improvements in slice acquisition time and heavily T2 weighted sequences have facilitated accurate identification of preoperative anatomic defects, thus allowing preselection of ideal arrays, e.g., those used in common cavity malformations. In light of this and other factors, at our center, we use magnetic resonance imaging (MRI) as the primary imaging modality to evaluate for CI candidacy. With current MRI protocols, detailed inner ear anatomy can be visualized with resolution often greater than that seen with computed tomography (CT) to identify various inner ear anomalies and assist with preoperative planning (Figs 2A to F). In addition, MRI allows visualization of and thus the confirmation of the presence of cochlear nerves in the internal auditory canal on both axial and sagittal images (Figs 3A to C). Given the recent concerns of ionizing radiation with CT scans in children, we reserve the use of temporal bone CT scans in patients with grossly malformed inner ears identified on MRI. The course of the facial nerve can be aberrant in these patients. Visualizing the bony anatomy with CT allows tracing the path of the Fallopian canal, which is helpful in surgical planning to minimize risk of facial nerve injury in these difficult cases.

One of the key challenges following successful implantation in the setting of severe anomalies such as common cavity malformation is the undesirable spread of monopolar current which elicits both facial nerve activity and reduction in dynamic range. Even with complete insertions, programming can be a challenge and may require selectively switching off stimulation channels as well as frequent audiologic programming sessions.<sup>10</sup> In summary, children with incompletely partitioned cochlea and enlarged vestibular aqueduct achieve near-normal performance levels.<sup>11</sup> Patients with total semicircular canal aplasia, cochlear nerve hypoplasia, cochlear nerve



**Figs 2A to F:** Comparison of CT and MRI for demonstration of inner ear anatomy: (A) Axial slices illustrating the lack of development of the cochlea and the vestibule; (B) the cochlear and vestibular dysplasia on the right; (C) an example of an enlarged vestibular aqueduct and a dilated endolymphatic sac on the left; and (D–F) heavily weighted steady-state T2-weighted axial magnetic resonance slices reveal anatomic features of the membranous labyrinth corresponding to (A–C) in greater detail



**Figs 3A to C:** Magnetic resonance imaging of cochlear nerve anatomy. High-resolution T2-weighted slices of the internal acoustic canal (IAC) in the axial plane (A), when compared with the right ear (B), further demonstrating lack of a developed cochlear nerve in a sagittal section through the IAC in the left ear (C)

aplasia, or common cavity demonstrated lower levels of performance.<sup>10</sup> In addition, poor performance may be a function of poor functional capacity related to other sensory deficits such as visual and motor. Despite anatomic factors that complicate implantation in this cohort of children, satisfactory clinical outcomes may be achieved with careful planning and modification of surgical technique.<sup>12</sup>

### ADJUVANT THERAPIES TO ENHANCE INTRACOCHLEAR STIMULATION

Spiral ganglion cells (SGCs) represent the first site of activation within the afferent auditory pathway when stimulated by a CI. In human CI users, there appears to be clear evidence that their performance, especially in psychophysical tests, may be related to the number of surviving SGCs.<sup>13</sup> A number of authors have now advocated relatively novel techniques for a further increase in trophic support to surviving SGCs. At the apex is genetic therapy that involves transfection with adenoviral vectors that can satisfactorily release neurotrophic factors. Challenges remain with their implementation, as the safety of viral vectors for use in human therapeutics is yet to be demonstrated. Others have tried modifications to the implant design in order to incorporate release of these factors to increase the benefits normally expected with chronic intracochlear electrical stimulation, either through special polypyrrole coatings or by use of nanoparticles.<sup>14</sup> In the latter, a novel chitosan-based nanoparticle delivery system is described, which could release both drugs and biomaterials to the inner

ear, including cochlear fluids, in a sustained fashion. This technology shows promise in noninvasive delivery of therapeutics to the neural elements of the inner ear without adversely affecting their structures. There is scope for adding this technology to CIs as an efficient combination for enhancing neurotrophic support to SGCs without increasing the size or creating significant alterations to current designs of CIs.

### REPLACING CIs

Novel replacements for CIs are now being suggested in the form of either biological or optical (laser) techniques. At the forefront of biological treatment options are the concept of hair cell regeneration and the pursuit of genetic factors that determine their fate. Advances in the area can be summarized in the form of three strategies—gene therapy, stem cell therapy, and molecular therapy. Efforts to improve functional recovery at system and behavioral levels have reached a difficult horizon, although there is considerable promise in overcoming the limitations of modern day CIs, centered on the delivery of energy to the inner ear. The most promising of these biologic therapies combines optical technology with viral expression of light-sensitive cation channels at different sites in the auditory pathway.<sup>15</sup> As other authors have also shown, pulsed delivery of light in optogenetics can potentially stimulate viral-vector-induced channelrhodopsin-2 and similar light-sensitive proteins within the cochlea in a tonotopic fashion. This technology has highlighted improved frequency selectivity by limiting the spatial spread of excitation, which has been the main issue associated with electrical CIs, even with the most modern signal processing strategies.

From an optical technology standpoint, several groups around the world are examining the role of laser and other forms of optically coherent radiation as a means to provide focused stimulation of intracochlear locations, reducing the possibility of spread of excitation within the cochlea. Even with pulsed infrared stimulation, tissue heats up quickly, and the parameters for stimulation within the context of useful delivery of stimulation have not been described. Recent advances in optical stimulation within the cochlea have fueled the development of micro-light-emitting diode arrays that showed promise in a small animal model. The intrinsic limitation of using this technology is the issue with safety of long-term delivery of ionizing radiation to the inner ear, and to date, the data have not been entirely satisfactory. Improvements in CI technology and methods for maximizing the survival of the SGCs will have to go hand in hand with a better understanding of the plasticity of the developing brain if we have to realize the full benefits of this area of research.

In summary, there is clear evidence to conclude that early intervention in the form of cochlear implantation, and where possible, bilateral implantation, has the potential to provide sensory experience necessary for optimal development of the deafened auditory system in children, with enormous benefits to the hearing impaired, and society at large. While CIs have come a long way since its first introduction as a therapeutic technology, current research efforts may lead to further advances and improved function to patients in the near future.

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