

TITLE: Pediatric Bilateral Cochlear Implantation

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According to the Food and Drug Administration's (FDA's) 2005 data, nearly 100,000 people worldwide have received cochlear implants. In the United States, nearly 15,000 children have received them. Cochlear implants, coupled with intensive postimplantation therapy, can help young children to acquire speech, language, and social skills. Most children who receive implants are between two and six years old. Early implantation provides exposure to sounds that can be helpful during the critical period when children learn speech and language skills. In 2000, the FDA lowered the age of eligibility to 12 months. However, fewer than 2% of patients receiving cochlear implants are implanted bilaterally. This report will delve into the benefits of bilateral implantation, in children, that is currently coming to light with current research. First, a brief history of cochlear implants will be presented, followed by an orientation to the hardware that makes up a cochlear implant. Next, indications for implanting children will be discussed. Next, the surgical procedure will be outlined. Then, the well-known benefits of binaural hearing will be discussed, followed by a review of the benefits of bimodal listening (one ear with a cochlear implant and the other with a powered hearing aid). Finally, a review of the literature will be presented, highlighting the benefits of bilateral cochlear implant usage in children.

Cochlear implants as we know them now are the result of intensive research over the last four decades. However, there is a long history of attempts to provide hearing by the electrical stimulation of the auditory system. The centuries old interest in the biologic application of electricity was the basis for the development of cochlear implants. In the late 18th century, Alessandro Volta discovered the electrolytic cell. Volta was the first to stimulate the auditory system electrically, by connecting a battery to two metal rods that were inserted into his ears. When the circuits were completed, he received the sensation of 'une recousse dans la tate' ("a boom within the head"), followed by a sound similar to that of boiling of thick soup. The work of Wever and Bray (1930) demonstrated that the electrical response recorded from the vicinity of the auditory nerve of a cat was similar in frequency and amplitude to the sounds to which the ear had been exposed. Meanwhile, the Russian investigators Gersuni and Volokhov in 1936 examined the effects of an alternating electrical stimulus on hearing. They also found that hearing could persist following the surgical removal of the tympanic membrane and ossicles, and thus hypothesized that the cochlea was the site of stimulation. In 1950, Lundberg performed one

of the first recorded attempts to stimulate the auditory nerve with a sinusoidal current during a neurosurgical operation. His patient could only hear noise.

A more detailed study followed in 1957 by Djourno and Eyries. They provided the first detailed description of the effects of directly stimulating the auditory nerve in deafness. They placed a wire on the auditory nerves that were exposed during an operation for cholesteatoma. When the current was applied to the wire, the patient described generally high-frequency sounds that resembled a "roulette wheel" or a "cricket." The signal generator provided up to 1,000-Hz and the patient gradually developed limited recognition of common words and improved lip-reading capabilities. Simmons, in 1966, provided a more extensive study in which electrodes were placed through the promontory and vestibule directly into the modiolar segment of the auditory nerves. The nerve fibers representing different frequencies could be stimulated. The subject demonstrated that in addition to being able to discern the length of signal duration, some degree of tonality could be achieved. Dr. William House first heard of the research of Djourno and Eyries from one of their patients. He had previously observed the percepts of his patients when small electric currents were introduced to the promontory during middle ear procedures under local anesthesia. House envisioned an implantable device that could stimulate the auditory nerve. During the early sixties, he implanted several devices in patients that were rejected due to lack of biocompatibility. The devices worked for a short time, though, providing optimism. Between 1965 and 1970, Dr. House teamed up with Jack Urban, an innovative engineer, to ultimately make cochlear implants a clinical reality. The new devices consisted of a single electrode and benefited from microcircuit fabrication derived from space exploration and computer development. In 1972, a speech processor was developed to interface with the single-electrode implant and it was the first to be commercially marketed as the House/ 3M cochlear implant. More than 1,000 of these devices were implanted between 1972 to the mid 1980s. In 1980, the age criteria for use of this device were lowered from 18 to 2 years and several hundred children were subsequently implanted. During the late 70s, work was also being done in Australia, where Clark and colleagues were developing a multi-channel cochlear implant later to be known as the Cochlear Nucleus Freedom. Multiple channel devices were introduced in 1984, and enhanced the spectral perception and speech recognition capabilities compared to House's single-channel device.

Currently, there are two major companies manufacturing multi-channel cochlear implants for use in the United States: The Cochlear Corporation and Advanced Bionics. They produce the Nucleus Freedom implant, and the Hi-Res 90K implant, respectively. Both devices consist of the same basic arrangement of components. The external components consist of a microphone, a speech processor, and a signal transmitter. The microphone sits in the external acoustic meatus and picks up sound from the environment. The speech processor selectively filters sound to prioritize audible speech and sends the electrical sound signals through a thin cable to the transmitter. The transmitter, which is a coil held in position by a magnet placed behind the external ear, transmits the processed sound signals to the internal device by electromagnetic induction. The internal components are a receiver/ stimulator and an electrode array. The receiver/ stimulator is secured in bone beneath the skin, and converts the signals into electric impulses and sends them through an internal cable to electrodes. An array of up to 22 electrodes wound through the cochlea, sends the impulses to the nerves in the scala tympani and then directly to the brain through the auditory nerve system.

Cochlear implants are indicated for children with severe to profound sensorineural hearing loss who do not benefit from conventional hearing aids. For children who can respond reliably, standard pure-tone and speech audiometry tests are used to screen likely candidates. Otherwise, ABR and OAEs can be used to detect very young children with severe-to-profound hearing loss. For children aged 12-23 months, the pure-tone average (PTA) for both ears should equal or exceed 90 dB. For individuals older than 24 months, the PTA for both ears should equal or exceed 70 dB. Older children are then evaluated with speech-recognition tests with best-fit hearing aids in place in a sound field of 55-dB. One of the most common speech-recognition tests is the hearing in noise test (HINT), which tests speech recognition in the context of sentences (open set sentences). Current guidelines permit implantation in children whose recognition is <60%. 12 months is the current age limit the FDA has established for implantation. However, a child with deafness due to meningitis may develop *labyrinthitis ossificans*, filling the labyrinth with bone. In these cases, special techniques may be needed for implantation and suboptimal outcome may result. Using serial imaging, implant teams may monitor patients with new deafness due to meningitis and perform implantation at the first sign of replacement of the scala tympani with fibrous tissue or bone. Otherwise, implantation in patients with postmeningitic deafness is usually recommended after 6 months to allow for possible recovery of hearing. Preoperative CT scan should always be performed, to detect cochlear abnormalities or absence of CN VIII. Cochlear malformations, though, do not necessarily preclude implantation. In pediatric patients with *progressive* hearing loss, neurofibromatosis II and acoustic neuromas should be excluded by performing MRI.

Cochlear implantation is usually performed as an outpatient surgery. General anesthesia is used and the ear and postauricular area are shaved and prepped in the usual sterile fashion. The future site of the implant receiver is marked with methylene blue in a hypodermic needle. This site at least 4 cm posterosuperior to the EAC, leaving room for a behind-the-ear controller. Next, a postauricular incision is made and carried down to the level of the temporalis fascia superiorly and to the level of the mastoid periosteum inferiorly. Anterior and posterior supraperiosteal flaps are then developed in this plane. Next, an anteriorly based periosteal flap, including temporalis fascia is raised, until the spine of Henle is identified. Next, a superior subperiosteal pocket is undermined to accept the implant transducer. Using a mock-up of the transducer, the size of the subperiosteal superior pocket is checked. Next, using a 6 mm cutting burr, a cortical mastoidectomy is drilled. It is not necessary to completely blue-line the sinodural angle, and doing so may interfere with proper placement of the implant transducer. Using a mock-up of the transducer for sizing, a well is drilled into the outer cortex of the parietal bone to accept the transducer magnet housing. Small holes are drilled at the periphery of the well to allow stay sutures to pass through. These suture will be used to secure down the implant. Stay sutures are then passed through the holes. Using the incus to judge depth, the facial recess is then drilled out. Through the facial recess, the round window niche should be visualized. Using a 1 mm diamond burr, a cochleostomy is made just anterior to the round window niche. The transducer is then laid into the well and secured with the stay sutures. The electrode array is then inserted into the cochleostomy and the accompanying guidewire is removed. Small pieces of harvested periosteum are packed in the cochleostomy around the electrode array, sealing the hole. Fibrin glue is then used to help secure the electrode array in place. The wound is then closed in layered fashion and a standard mastoid dressing is applied.

Audiologists are well aware of the benefits of bilateral conventional hearing aids for patients with bilateral hearing loss. Bilaterally hearing-impaired people who wear hearing aids in both ears can clearly understand speech better, especially in noise. Ricketts (2001) documented the advantages of bilateral hearing aids across a broad variety of conditions. It seems reasonable, then, that children with 2 ears that meet criteria should receive bilateral implantation. The potential benefits of bilateral implants are threefold. Firstly, it ensures that the ear with the best postoperative performance is implanted. Second, it may allow preservation of some of the benefits of binaural hearing: head shadow effect, binaural summation and redundancy, binaural squelch, and sound localization. Third, it may avoid the effects of auditory deprivation on the unimplanted ear. When speech and noise come from different directions, there is always a more favorable signal-to-noise ratio (SNR) at one ear. The head shadow effect is about 7dB difference in the speech frequency range, but up to 20 dB at the highest frequencies. With binaural hearing, the ear with the most favorable SNR is always available. Sounds that are presented to 2 ears simultaneously are perceived as louder due to summation. Thresholds are known to improve by 3 dB with binaural listening, resulting in doubling of perceptual loudness and improved sensitivity to fine differences in intensity. The auditory nervous system is wired to help in noisy situations. Binaural squelch is the result of brainstem nuclei processing timing, amplitude, and spectral differences between the ears to provide a clearer separation of speech and noise signals. The effect takes advantage of the spatial separation of the signal and noise source and the differences in timing and intensity that these create at each ear. Interaural timing is important for directionality of low-frequency hearing. For high frequency hearing, the head shadow effect is more important. Head and pinna shadow effects, pinna filtering effects, and torso absorption contribute to spectral differences that can help determine elevation of a sound source. Work with conventional hearing aids has demonstrated the effects of auditory deprivation; If only 1 ear is aided, when there is hearing loss in both ears, speech recognition in the unaided ear deteriorates over time. This effect has been shown in children with moderate and severe hearing impairments (Gelfand and Silman 1993).

Bimodal listeners use a cochlear implant on 1 ear and a conventional hearing aid on the opposite ear. Results of studies with bimodal devices paved the way for bilateral cochlear implantation. One of the earliest studies of bimodal devices, Waltzman et al (1992) demonstrated that eight adults with a unilateral Nucleus cochlear implant perceived speech better, on average, when listening bimodally than with one ear alone. This early study, though, failed to demonstrate the same advantage in children. Ching (2001) examined the efficacy of bimodal input with a hearing aid and a cochlear implant. 16 congenitally hearing impaired children aged 6 to 18 were studied. These children wore a powered hearing aid in the non-implant ear. After adjustment of the powered hearing aid, speech recognition scores were significantly better in both quiet and noise. Objective localization scores were also better in the bimodal condition. In 2005, Luntz evaluated 12 patients, 9 of who were pre-lingually impaired adults and older children (aged 7 to 16) who used hearing aids on the unimplanted ear. They were tested for speech recognition at 1-6 months post-op and then at 7-12 months post-op with speech and noise presented at 55 dB from a frontal speaker (SNR +10dB). Ching (2006) reviewed a series of their own experiments and data collected on children using bimodal devices. They reported that the children as a whole performed better with bimodal stimulation than with the cochlear implant alone on horizontal localization tasks and could take advantage of head shadow and binaural redundancy effects.

The earliest published report of bilateral cochlear implants was 1988. The primary reason for bilateral implantation in the early days was either there was a need for a technology upgrade or the device in 1 ear produced inadequate performance. In the late 1990s, bilateral implants began to be done solely with hope and intention of providing binaural benefits. Recently, there has been a trend toward simultaneous implantation, rather than sequential implantation. Bilateral implantation is becoming more common, but is still relatively rare. Laszig reported in 2004 that although over 50,000 people had been implanted with the Nucleus CI worldwide, fewer than 1% had been bilaterally implanted. Of primary interest has been determining whether or not bilateral implantation will produce improvements in understanding speech, particularly in background noise, relative to unilateral implantation. For most cochlear implant users, speech understanding in noise is relatively poor and they require higher SNR than do normal-hearing children. Kuhn-Inacker et al (2004) reported bilateral implantation on 39 German children. Age at 1st implant ranged from 8 months to 16 years. Age at 2nd implant ranged from 1 year to 16 years. Time lag between implants was 0 to 4 years. All children had pre or perilingual deafening. Speech discrimination in noise tests were done on 18 of the children. Speech was delivered at 15 dB SNR through an array of loudspeakers designed to minimize head shadow effects and thus look only at true binaural processing effects. The interval between 2nd implant and testing ranged from 6 to 24 months. All children did better with bilateral implants than with unilateral implant. Mean difference between bilateral and unilateral speech discrimination scores was 18.4%. Analysis showed neither age at 1st implant, nor interval between implants significantly influenced performance. However, there was a trend toward faster, better performance with the 2nd implant when lag time was shorter. Litovsky, in 2004, tested 3 children 3 months after activation of bilateral implants. These children had sequential procedures 3-8 years apart. Children underwent testing of speech intelligibility, with competing noise, with the first CI alone, and bilaterally. On the speech tasks, 1 child did not benefit from bilateral hearing. Two children showed consistent improvement with bilateral hearing when the noise was near the side that underwent implantation first. The authors suggested that some children might require a more prolonged period of adjustment and learning with 2 implants.

Litovsky continued to investigate bilateral implants and in 2006 evaluated children ages 4-14, 10 using two CIs (sequentially implanted) and 10 using one CI and one HA. Speech intelligibility was measured in quiet, and in the presence of 2-talker competing speech using the CRISP forced-choice test. Results indicated clear and significant improvements in speech results with two-eared versus one-eared listening for the CI group across all conditions. The results were somewhat less compelling for the bimodal users. Peters et al in May 2007 published reports of children aged 3 to 13 years who were recipients of 2 cochlear implants, received in sequential operations, a minimum of 6 months apart. All children received their first implant before 5 years of age and had acquired speech perception capabilities with the first device. They were divided into 3 age groups on the basis of age at time of second ear implantation: Group I, 3 to 5 years, Group II, 5.1 to 8 years, Group III, 8.1 to 13 years. Results for speech perception in quiet show that children implanted sequentially acquire open-set speech perception in the second ear relatively quickly (within 6 mo). However, children younger than 8 years do so more rapidly and to a higher level of speech perception ability at 12 months than older children. Speech intelligibility for spondees in noise was significantly better under bilateral conditions than with either ear alone when all children were analyzed. The bilateral benefit in noise increased with time from 3 to 9 months after activation of the second implant. This bilateral advantage is greatest when noise is directed toward the first implanted ear, indicating that the head shadow

effect is the most effective binaural mechanism. Wolfe et al, August 2007, evaluated speech recognition in quiet and in noise for a group of 12 children, all of whom underwent sequential bilateral cochlear implantation at various ages. The primary outcome measure for speech recognition in noise assessment was the signal-to-noise ratio needed for 50% performance. The results of these assessments were contrasted between children receiving their second cochlear implant before 4 years of age versus after 4 years of age. Speech recognition scores were significantly worse in quiet for the later implanted ear when the 2nd implant was received after age 4 demonstrating auditory deprivation effects. There was not a significant difference in speech scores in quiet between individual ears when the 2nd implant was received before age 4. Both groups of children possessed better speech recognition scores in noise in the bilateral condition relative to either unilateral condition. However, there was not a statistically significant relationship between speech recognition performance in noise and the duration of deafness of the later implanted ear.

Scherf (2007) published a report on 33 children who underwent a second, sequential cochlear implant. Assessments took place pre-second implant and at several time intervals post-fitting on pure tone audiometry and speech recognition in quiet and noise. Speech perception in noise testing was performed at 18 months post-op. Speech recognition scores in quiet were for all children superior in the bilateral condition. In the noisy condition, only significant bilateral better results were obtained in the group of younger children. The data appear to show a beneficial performance for those children who received their second implant before the age of 6, especially in the more challenging conditions. In a study by Galvin, 2007, a second cochlear implant was received by 11 children. The principal selection criteria were being age 4 to 15 yr with a bilateral profound hearing loss and being a consistent user of a first implant. The children were tested for speech recognition in noise at 9 months post-op. When noise was presented ipsilateral to the first implant, 8 of 10 subjects showed a benefit in the bilateral condition. None of the nine subjects tested showed a benefit when noise was contralateral to the first implant. Generally, there was no benefit to localization in the bilateral condition.

Modern cochlear implants, being the result of decades of research and development, are an excellent therapeutic modality for the treatment of pediatric hearing loss. Most children who use bilateral cochlear implants have better speech recognition in noise and better sound localization than children who use a unilateral implant. Some evidence points toward benefits of earlier bilateral implantation. More studies need to be done to elicit the effects of age at time of implants (1st and 2nd) and the effects of sequential versus simultaneous implantation.

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