



COCHLEAR IMPLANTS

From Principles to Practice

David S Haynes
René H Gifford
George B Wanna
Alejandro C Rivas

Contents

Section 1: Aspects of Hearing Loss

- 1. Etiology of Hearing Loss: Implications for Cochlear Implant Performance 3**
Timothy J Davis, Kristen L D'Onofrio

Section 2: Assessment of Cochlear Implant Candidacy

- 2. Assessment of Adult Cochlear Implant Candidacy..... 15**
Robert T Dwyer, René H Gifford
- 3. Expanding Criteria for Cochlear Implantation 23**
René H Gifford
- 4. Preoperative Imaging in Cochlear Implantation 35**
Jennifer Dang, Alex D Sweeney

Section 3: The Cochlear Implant/Surgical Procedures

- 5. Surgery for Cochlear Implantation: Standard Approach 47**
David R Friedmann, Daniel Jethanamest, J Thomas Roland Jr
- 6. Surgery for Cochlear Implantation—Bilateral Cochlear Implants 55**
David R Friedmann, Sean O McMenomey
- 7. Surgery for Cochlear Implantation—Congenital Anomalies 58**
Nicholas L Deep, Brendan P O'Connell, Mai-Lan Ho, John I Lane, Matthew L Carlson
- 8. Hearing Preservation Cochlear Implantation..... 83**
Robert J Yawn, Alejandro C Rivas
- 9. Surgery for Cochlear Implantation—Suprameatal Approach..... 90**
Eric Applebaum, Alex D Sweeney, Robert J Yawn, David S Haynes
- 10. Surgery for Cochlear Implantation—Subtotal Petrosectomy Approach 95**
Jacob B Hunter, Brandon Isaacson
- 11. Revision Cochlear Implantation 106**
Matthew G Crowson, Howard W Francis

12. Auditory Brainstem Implants	114
<i>Craig A Buchman, Nedim Durakovic</i>	
13. Electrophysiological Testing for Cochlear Implants and Auditory Brainstem Implants	123
<i>Michelle L Hughes, Shuman He</i>	

Section 4: Results and Outcomes of Cochlear Implantation

14. Outcomes for Children.....	147
<i>Emily Lund, Holly FB Teagle</i>	
15. Outcomes for Adults	165
<i>Kristen L D'Onofrio, Jourdan T Holder</i>	
16. Combined Electric and Acoustic Stimulation (EAS) and Electric Complementation (EC): Considerations for Candidacy, Fitting, and Long-Term Audiological Management	177
<i>Artur Lorens</i>	
17. Music Perception in Cochlear Implantation	191
<i>Kate Gfeller, Virginia D Driscoll</i>	

Section 5: Current Concepts and Future Technology

18. Importance of Scalar Location in Cochlear Implant Surgery	201
<i>George B Wanna, Brendan P O'Connell, Matthew L Carlson</i>	
19. Drug-Eluting Electrodes and the Next-Generation Electrodes	207
<i>Adrien A Eshraghi, Christopher A O'Toole</i>	
20. Future of Intraoperative Electrophysiologic Testing in CI Surgery	218
<i>Michael S Harris, William Jason Riggs, Oliver F Adunka</i>	
21. Cost-Effectiveness in Cochlear Implantation	228
<i>Nicolas-George Katsantonis, Jacob B Hunter, Marc L Bennett*</i>	
22. Cochlear Implant Device Reliability Reporting.....	237
<i>P Cody Buchanan, Cedric Navarro, Douglas D Backous*</i>	
Index.....	243

Expanding Criteria for Cochlear Implantation

René H Gifford

INTRODUCTION

Food and Drug Administration (FDA) labeled indications for adult conventional cochlear implant (CI) candidacy were last changed in 2005 with the expansion of Medicare's National Coverage Determination (NCD) for individuals with bilateral sensorineural hearing loss scoring up to 40% correct for open-set sentences. Pediatric CI indications were last amended in 2000 with change in minimum age for implantation from 18 to 12 months. In the United States, the most recent change in candidacy has been with the approval of the Nucleus Hybrid-L24 system in 2013 and its commercial availability in 2014. This indication specifies hybrid CI candidacy for adults with precipitously sloping high-frequency hearing loss who score up to 60% for monosyllabic word recognition in the ear to be implanted and no >80% in the contralateral ear. Though the hybrid approval significantly broadened the candidacy umbrella for cochlear implantation in the United States, Hybrid-L24 implant recipients represent just a fraction of the total CI recipients across the globe.

In order for implant manufacturers to broaden or expand their labeled indications, they must petition the national agency responsible for overseeing safety and approval of biomedical devices in each country the device is offered. In the United States, that agency is the FDA, which will typically require a clinical trial to determine the safety and efficacy of the revised indication. This is both

a time intensive and costly endeavor. Thus, although CI technology and associated clinical practice has evolved rapidly in recent years, the labeled indications have largely remained unchanged.

There are a number of reports of clinicians routinely implanting patients who may not perfectly fit the CI candidate profile—as specified in the labeled indications. The reason that this is occurring is that the peer-reviewed literature and clinical practice has provided us with evidence supporting the expansion of CI criteria. This chapter describes the evidence, evolving clinical practices, and importance of assessing the whole patient for the determination of pediatric and adult CI candidacy.

WHAT ARE THE BARRIERS TO COCHLEAR IMPLANTATION?

As of 2012, over 529,000 registered devices had been implanted worldwide¹—a number that has been increasing most notably in the past decade. Even though the number of implant recipients has been increasing worldwide, the World Health Organization (WHO) estimates that there are 360 million people worldwide who have disabling hearing loss. The WHO defines disabling hearing loss as thresholds poorer than 40 and 30 dB HL for adults and children in the better hearing ear. The WHO estimation of individuals with disabling hearing loss represents 5% of the world's population, a number that is expected to

increase with a global prevalence toward an aging population. In the United States, specifically, approximately 37.5 million Americans (~12% of the U.S. population) have hearing loss² with up to 750,000 having severe-to-profound hearing loss.² With an estimated 96,000 CI recipients in the United States as of 2012,³ that means that as of 2012, just 12.8% of the 750,000 Americans with severe-to-profound hearing loss had actually been implanted.

Underutilization of CI technology is believed to be most applicable to the adult population as it has been reported that 50%–55% all pediatric implant candidates in the United States receive an implant.^{4,5} In contrast to the U.S. data, 95%–97% of children with severe-to-profound sensorineural hearing loss born in Europe and Australia receive an implant.⁵ In Japan, it is estimated that just 1% of individuals meeting candidacy for cochlear implantation are utilizing this technology.⁶ No published data regarding uptake of cochlear implantation in adults and children in developing countries are available, though one could reasonably assume that access is more restricted than in the United States, Europe, Japan, and Australia.

There are a number of potential factors explaining the underutilization of CI technology. One possibility is the lack of patient awareness about CIs. Given the success of CIs and an ever-growing presence of research in the peer-reviewed literature in the fields of otology, hearing science, audiology, deaf education, language, and speech, one would assume that public awareness is also widespread. Unfortunately, this is not the case. A general lack of public awareness regarding CIs—including function, candidacy, and insurance coverage—is a large part of the mission for a number of not-for-profit advocacy and awareness foundations including the American Cochlear Implant Alliance, British Cochlear Implant Group, Cochlear Implant Awareness Foundation, Alexander Graham Bell Association, Hear the World Foundation, Hearing Health Foundation, Hands and Voices, and others. Despite these efforts, we still have much to do to improve access and public awareness of this life-changing technology.

Another explanation for the underutilization of CIs is financial in nature. Stern et al.⁷ examined the zip codes of CI recipients and the associated median income in those codes, and found that more children living in higher socioeconomic areas were more likely to have CIs. This finding is concerning given that epidemiological research has shown a greater prevalence of childhood hearing loss in lower socioeconomic areas.⁸ A recent study investigating attitudes regarding hearing healthcare and

cochlear implantation revealed that African-Americans with severe-to-profound hearing loss cited a multitude of reasons for not seeking hearing health care including physician mistrust, lack of finances, and lack of awareness and education regarding CIs. Finances also present a barrier to implant technology for adult candidates.⁹

Another financial burden exists for individuals without access to universal healthcare coverage or employer-subsidized coverage, the latter of which being most common in the U.S. system. Individuals who are uninsured may choose to forego health insurance coverage completely or purchase high-deductible plans which may exclude cochlear implantation. Implant exclusionary policies may only become problematic in cases of sudden or acquired adult hearing loss leaving the policyholder without CI coverage and lacking the finances to pay for the surgery, device, and external activation kit. Moving toward a national health-care system offering coverage for CIs—similar to that in Canada, Europe, the United Kingdom, and Australia—should allow greater access to this technology, though could paradoxically limit access to individuals who are borderline candidates, candidates for electric-acoustic stimulation (EAS), or for adults pursuing a second CI.

Another barrier to CI technology is a lack of knowledge amongst referring medical providers. There is evidence that primary care physicians¹⁰ and hearing aid dispensers¹¹ may lack up-to-date knowledge about CI candidacy. There is also reason to believe that even general otolaryngologists, audiologists, and speech-language pathologists may not fully recognize the audiologic profile of today's implant candidate.^{12,13} Thus, we have much work to do to educate both the public and healthcare providers regarding appropriate referrals and the potential of this implantable technology.

Yet another potential barrier to implantation is related to a lack of CI referrals from hearing aid dispensers and private practice audiologists for fear of losing hearing aid patients. Though no data exist suggesting that hearing aid dispensers and/or private practice audiologists are reluctant to refer for CI evaluation for financial reasons, it would not be an unreasonable supposition. For professionals whose income relies either solely or predominantly upon hearing aid sales, a CI referral may be perceived as a lost patient; however, this is absolutely not the case. Individuals making use of a CI and a contralateral hearing aid (also referred to as *bimodal* hearing) integrate the electric and acoustic signals to derive significant communicative benefit for speech recognition in quiet and in

noise¹⁴⁻¹⁸ as well as for the recognition of music.¹⁸⁻²⁰ The reality is that the bimodal listener can continue to be a unilateral hearing aid patient for years to come, and grateful patients will most certainly provide high recommendations for the referring audiologist or hearing aid dispenser.

EVIDENCE FOR THE EXPANSION OF COCHLEAR IMPLANT CANDIDACY

Adults

There are several peer-reviewed studies reporting significant benefit for speech understanding for adult implant recipients who had not met all FDA labeled indications for cochlear implantation. Adunka et al.²¹ reported significant postoperative benefit for 21 subjects with substantial residual hearing who had preoperative speech recognition scores of 72% for City University of New York (CUNY) sentences and 18% for consonant-nucleus-consonant (CNC) words. In addition, they demonstrated that these 21 non-traditional implant candidates had demonstrated similar benefit to conventional implant recipients—all of whom had met FDA labeled indications for implantation. Furthermore, speech recognition improved for all participants in the study—that is, no one exhibited a decline in performance following cochlear implantation.

In a similar study, Tremblay et al.²² demonstrated significant benefit for 17 adult implant users who had preimplant sentence scores ranging from 40% to 77% correct. They reported a mean postoperative improvement of 38% points for sentence recognition and 31% points for word recognition. Speech understanding improved for all study participants.

Gifford et al.¹⁶ reported significant benefit for 22 adult CI users who had achieved preoperative, bilaterally aided speech recognition scores of 47% for AzBio sentences and 41% for CNC words. The mean postoperative improvement for CNC word recognition was 27% and 41% points in the implant only and best aided bimodal condition, respectively. Furthermore, all recipients demonstrated improvement in speech recognition following implantation.

In a similar study, Amoodi et al.²³ reported significant postoperative improvement on measures of speech recognition for 27 adult implant recipients who had preoperatively scored 60% or greater for hearing in noise test (HINT) sentence recognition. Mean improvement for their recipients was 37% points for CNC words and 27% points for HINT sentences—though HINT sentence recognition benefit was limited postoperatively by ceiling effects.²⁴ As

with all studies mentioned here, speech understanding improved for all study participants.

Finally, Sladen et al.²⁵ reported the outcomes of a multicenter clinical trial investigating a revised indication for adult cochlear implantation for which the 21 adult recipients scored up to 40% correct for CNC word recognition in the ear to be implanted and up to 50% in the bilaterally aided condition. In the ear to be implanted, mean preoperative CNC word recognition was 23.6% correct and mean postoperative CNC word recognition was 65.1% correct, for a mean improvement of 41.5% points. In the best aided condition, mean preoperative CNC word recognition was 38.7% correct and mean postoperative CNC word recognition was 73.6% correct, for a mean improvement of 34.9% points. In addition to significant improvement for speech recognition, the 21 adult recipients also demonstrated significant subjective benefit based on the Abbreviated Profile of Hearing Aid Benefit (APHAB²⁶) and the Health Utilities Index (HUI²⁷).

Figure 3.1 summarizes the findings of the above-cited studies comparing mean pre- and postimplant outcomes for tasks of sentence and word recognition in the best aided condition, or the condition in which the listener is most often communicating. The degree of benefit averaged across all four studies was 33% points for sentence recognition and 40% points for CNC word recognition; however, postoperative sentence recognition for stimuli presented in quiet was limited by ceiling effects for many of the participants. Thus, to date, the research on non-traditional adult CI recipients has been unequivocal in demonstrating statistically significant improvement for various measures of speech perception. In fact, *the point of diminishing returns has yet to be identified.*

Children

CI programs affiliated with progressive medical centers have been routinely implanting children with bilateral sensorineural hearing loss who may not meet all labeled indications for pediatric cochlear implantation. This is considered common practice given the increasing amount of evidence in favor of expanding implant criteria for children with significant hearing loss who are not making expected progress for auditory, speech, and language skills with appropriately fitted hearing aids, expected hearing aid wear time (verified via data logging), and the recommended intervention schedule. There have been a number of older studies advocating for the expansion of pediatric CI criteria.²⁸⁻³³ There are also reports of children

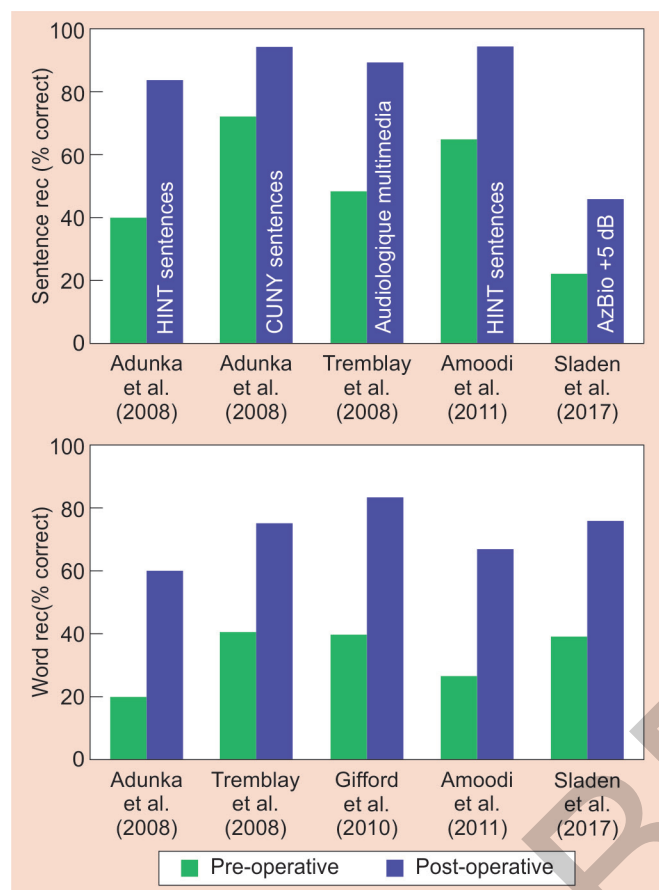


Fig. 3.1: Comparison of pre- and postimplant sentence and word recognition across studies specifically investigating the efficacy of cochlear implantation for adult candidates with significant residual hearing and/or above criterion preoperative speech recognition.

with residual hearing successfully combining EAS across ears for bimodal hearing.^{14,34-42} There has also been an increasing number of peer-reviewed papers dealing *specifically* with the issue of expanded criteria for pediatric cochlear implantation.

Dettman et al.⁴³ evaluated pre- and postimplant performance for open-set speech recognition in a group of 16 children who had achieved preoperative sentence recognition scores above 30% correct. All but one of the study participants demonstrated statistically significant improvement in word and sentence recognition performance. Important to note here, however, was that the children in this study would have met FDA labeled indications relative to degree of hearing loss as the mean pure tone average (PTA) was 109 and 96 dB HL for the implanted and non-implanted ears, respectively.

Yoshinago-Itano et al.⁴⁴ studied repeated measures of auditory-oral language growth for 87 children with

severe-to-profound hearing loss who had either been fitted with bilateral acoustic amplification ($n = 38$) or at least one CI ($n = 49$). The children were followed over time and trajectory of language growth was tracked up to 7 years of age. They showed that children with CIs exhibited a more rapid growth of receptive and expressive language than the children with hearing aids. They also reported that children with CIs closed the language gap relative to age equivalent peers with normal hearing. Children with hearing aids, however, closed the gap at a much slower pace for receptive language and indeed, *did not close the gap for expressive language*. Thus these data could be interpreted as further evidence for expanding criteria to include at least severe hearing losses for the youngest candidates.

Leigh et al.⁴⁵ evaluated speech recognition performance for 142 children with hearing loss who were either fitted with bilateral acoustic amplification ($n = 62$) or at least one CI ($n = 80$). They reported that for children with a PTA >60 dB HL, there is a 75% chance of improvement with a CI for word recognition performance. Using sentence recognition performance as a guide, then a PTA > 72 dB HL would have a 75% chance of improvement with an implant. Thus they conservatively concluded that children with a PTA of 75 dB HL or greater should be recommended for cochlear implantation—at least unilaterally. Indeed one could interpret their findings to promote cochlear implantation for children with unaided PTA above 60 dB HL—provided that the child is not demonstrating *at least* year-for-year progress on speech, language, and auditory skills (see also Chapter 14).

In a follow-up study, Dettman et al.⁴³ describe a longitudinal study of 403 children with bilateral sensorineural hearing loss who had either been fitted with acoustic amplification or received at least one CI prior to 2.5 years of age. Based on phoneme recognition scores obtained at 5 years of age, they reported that children with a PTA of 65 dB HL or higher (i.e., poorer) were 75% more likely to achieve greater outcomes with a CI as compared to bilateral hearing aids. Thus these outcomes corroborated their earlier findings reported by Leigh et al.⁴⁵

Carlson et al.⁴⁶ reported outcomes for 51 pediatric CI recipients who had been implanted off-label defined as having a PTA lower (i.e., better) than 90 dB HL for children under 2 years of age, PTA lower (i.e., better) than 70 dB HL for children over 2 years of age, and/or word recognition >30% correct in the bilaterally aided condition. They reported significant improvement in auditory skills development and/or speech recognition performance at the

group level with mean speech recognition benefit of 63% points in the implanted ear and 40% points in the best aided condition. Furthermore, they reported that every child exhibited better postoperative performance. That is, no child exhibited a decrement in performance for speech understanding, auditory skills development, or language development—consistent with Leigh et al.⁴⁵ Thus, the point of diminishing returns has not yet been identified for either adult or pediatric listeners with bilateral moderate sloping to profound sensorineural loss.

Infants Under 12 Months of Age

Current U.S. labeled indications for pediatric CI candidacy specify children 12 months of age and older. This age limitation should not be interpreted as infants under 12 months would not benefit from cochlear implantation. Rather, there is a growing body of evidence demonstrating higher levels of word and language acquisition,⁴⁷⁻⁵¹ speech perception,⁵² speech production intelligibility,⁵³ and vocabulary development^{29,47,54} for children implanted under 12 months of age—even when compared to children implanted in the second year of life.

One of the primary concerns regarding implantation under 12 months of age is the issue of specificity—or the risk of implanting a child *without* permanent sensorineural hearing loss. With a number of audiologic tests at our disposal including otoacoustic emissions, electrophysiologic measures, and advanced behavioral audiometric techniques for use in children as young as 5-6 months, initial diagnostic and then confirmatory diagnostic testing is certainly possible.

Cosetti and Roland⁵⁵ explained that the majority of concerns and complications reported in the literature have been confounded by emergency surgery in infancy. These surgeries are complicated by a lack of fasting and hence a greater risk of aspiration, as well as the risks of surgery in the young and possibly medically fragile infant. As related to CI surgery, there are a number of studies demonstrating no greater anesthetic risk for infants under 12 months.^{43,56-63} Cosetti and Roland⁵⁵ provided a thorough description of the surgical issues unique to the infant population including intraoperative blood loss, facial nerve anatomy, skull thickness (<1 mm), fixation of the receiver/stimulator package, thin scalp flap, and device migration with skull growth. They reported, however, that these are known variables which may be relieved with a highly skilled surgical team having extensive pediatric implantation experience.

In addition to the growing evidence demonstrating significant auditory and speech/language benefit for implantation in the first year of life, another critical consideration is that infants with severe-to-profound sensorineural hearing losses are missing critical language-learning opportunities in that first year. This is true even for infants with appropriately fitted hearing aids given that *audibility will not be sufficient to allow for consistent access to spoken language at average conversational levels*. Language-related developmental changes occurring rapidly in the first year of life include word segmentation, auditory memory, and phonological/lexical/semantic representation. Word segmentation is the process of dividing connected discourse into meaningful units, such as individual words. Research has shown that word segmentation develops rapidly between 7.5 and 10.5 months of age.^{64,65} By 8 months of age, infants have the capacity for auditory memory and long-term storage of new words—both of which are important prerequisites for auditory-based language learning.^{66,67} Development of phonological, lexical, and semantic representation also rapidly emerges during the first year of life.⁶⁸⁻⁷¹ Given the developmental changes occurring rapidly during the first year of life for the typically developing child, an infant with severe-to-profound sensorineural hearing loss with limited aided audibility will miss out on these auditory-based, language-learning opportunities. It is actually possible that language-learning opportunities begin before birth as research has shown that neural reorganization in response to speech is likely initiated in the womb. Newborns have been shown to respond differentially to familiar sounds that they were exposed to in utero⁷²⁻⁷⁵ with greater brain activity observed in response to familiar sounds.⁷⁶ Thus, cochlear implantation under 12 months of age will likely be included in the labeled indications in the near future.

Hearing Preservation Cochlear Implantation

Considerable research and clinical attention has been placed on preservation of acoustic hearing with minimally traumatic surgical techniques and thin, atraumatic electrode arrays. Functional hearing preservation is possible both with short electrodes and associated shallow insertion depth^{16,30,77-81} as well as for longer electrodes with deeper insertion.⁸²⁻⁸⁸ By adding ipsilateral acoustic hearing to electrical stimulation, CI users can experience nonlinear, additive gains in speech understanding and

basic auditory function.^{15-16,89-100} Further, the benefit from acoustic stimulation can be added via the non-implanted ear, the implanted ear in cases of hearing preservation, or both ears allowing for *binaural acoustic stimulation*. Current implant technology combined with hearing aids in the implanted and/or non-implanted ear yields significant benefit for the vast majority of recipients. Mean benefit obtained via acoustic hearing in the implanted ear ranges from 10% to 15% points or 2–3-dB improvement in the signal-to-noise ratio (SNR).^{81,89,91-95,101,102} This benefit of 10%–15% points (or 2–3 dB) is beyond that obtained in the bimodal condition (CI and contralateral hearing aid (HA)) for which the acoustic hearing in the implanted ear is occluded.

The majority of the peer-reviewed literature focusing on hearing preservation and combined EAS has focused on adult CI recipients. There is, however, a growing population of pediatric CI recipients with preserved acoustic hearing¹⁰³⁻¹⁰⁷ who have demonstrated similar significant EAS benefits in speech understanding. Further, a recent study of 153 ears has shown that younger age at implantation was positive prognostic variable for acoustic hearing preservation with cochlear implantation.¹⁰⁸ Thus, we may expect to see an increase in children presenting for “hybrid/EAS” CI candidacy—particularly if high-frequency audibility is not attainable with conventional amplification or with frequency lowering technology. It is important to note that U.S. approved indications for hybrid implant technology are limited to individuals aged 18 years and older. This is not to imply, however, that atraumatic electrodes should not be used for children with low-frequency acoustic hearing that could be preserved and possibly aided for combined EAS. We can expect that future FDA approved indications for hybrid/EAS hearing systems may include children younger than 18 years.

UNILATERAL AND ASYMMETRIC HEARING LOSS

CI candidacy has historically been based on speech recognition in the best aided condition. To date only one of the implant manufacturers has even made reference to considering preoperative performance in the ear to be implanted in their labeled indications (Cochlear Americas physicians package insert) for both the conventional implant criteria as well as indications for the Nucleus Hybrid-L device. The prevalence of asymmetric hearing

loss amongst adults with sensorineural losses is estimated at 50%–57%.^{109,110} For children, the prevalence of unilateral sensorineural hearing loss is estimated from 1 to >3 per 1,000 births.¹¹¹⁻¹¹⁴ Hence it is reasonable to suppose that the best aided condition may overestimate hearing and speech understanding abilities—particularly for realistic listening environments in which the target stimulus may not always be directed toward the front or the better hearing ear. Further the literature has repeatedly demonstrated that children with minimal hearing loss—including unilateral sensorineural hearing loss—experience greater fatigue, stress, and increased academic risk as compared to their peers with normal hearing.¹¹⁵⁻¹¹⁸

Cochlear implantation in cases of unilateral hearing loss, also commonly referred to as single-sided deafness (SSD), has been a hot topic for many years. There is a growing literature base including adults and children with highly asymmetric hearing losses as well as individuals with SSD receiving CIs in the poorer ear. Firszt et al.¹¹⁹ described the outcomes for 10 adult recipients with asymmetric hearing loss for which implant candidacy was determined on the basis of the poorer hearing ear. In the non-implanted ear, 4 of the 10 subjects had low-frequency audiometric thresholds in the normal to near-normal range. Even with the highly asymmetric nature of the hearing losses in their population, they showed significant improvement in speech recognition performance in quiet and noise as well as localization abilities. These findings suggest that we should consider implant candidacy on an individual ear basis even when the better hearing ear exceeds traditional candidacy criteria. They noted, however, that adults with pre- or perilingual onset of deafness in the poorer hearing ear achieved modest benefit and thus may require more extensive counseling regarding realistic expectations.

Unilateral hearing loss, or SSD, is just a special case of asymmetric hearing loss as one ear has *completely normal hearing*. Research has shown that a single hearing ear is sufficient for a child to develop speech and language within the age normative range; however, as documented previously here, children with SSD have been shown to exhibit greater fatigue, stress, and increased academic risk as compared to their normal-hearing peers.¹¹⁵⁻¹²⁰

In a study of 20 children with SSD who had received a CI,¹²¹ assessed speech understanding in colocated and spatially separated noise conditions with and without the CI in place. They also obtained subjective reports of

speech, spatial, and qualitative reports prior to and following CI surgery using the speech, spatial, and qualities [SSQ¹²²] questionnaire administered to the parents. They documented significant improvements in speech understanding with the use of the CI for conditions in which the noise was presented to the normal-hearing ear. They further reported significantly improved qualitative reports on all subscales of the SSQ following cochlear implantation. Arndt et al. also mentioned that the speech and qualitative outcomes were better for children with postlingual onset of sensory hearing loss as well as an overall shorter duration of SSD as compared to the children with congenital sensory hearing loss and longer durations of SSD.

Two recent studies of pediatric implant recipients with SSD documented that children wore their CI sound processors for the majority of waking hours, similar to rates of children with bilateral sensorineural hearing loss.^{123,124} Thus it may ultimately be the case that cochlear implantation is recommended for cases of SSD with short durations of deafness and/or postlingual onset. For adults with acquired SSD and children with SSD whose parents have expressed interest and motivation for obtaining a CI in the poorer hearing ear, cochlear implantation is a viable intervention capable of significantly improving auditory neural development, speech understanding in the poorer ear, speech understanding with noise presented to the better hearing ear, spatial hearing abilities, and quality of life. As of July 2019, we now have an FDA-approved indication for SSD cochlear implantation in the US. MED-EL received approval for individuals aged 5 years and older with profound hearing loss in one ear.

Cochlear Implantation for Tinnitus Suppression

Other cases of cochlear implantation with unilateral or highly asymmetric hearing losses have focused on tinnitus suppression rather than improving speech recognition. There are multiple published reports of the benefits of cochlear implantation for patients with unilateral hearing loss for tinnitus suppression.^{23,125-132} The majority of reports describing cochlear implantation for tinnitus relief in cases of unilateral hearing loss have resulted in positive outcomes. In some cases, there were even reports of improved speech recognition in noise and subjective reports of hearing benefit.^{125,129} Despite the increased interest in this topic in recent years, it remains unclear whether cochlear implantation for tinnitus suppression

in cases of unilateral hearing loss is a cost efficient and viable treatment option and whether this is a treatment that would be covered by insurance as *medical necessity*.

COCHLEAR IMPLANTATION FOR CHILDREN WITH SPECIAL NEEDS: GLOBAL DEVELOPMENTAL DELAY

Up to 40% of children with sensorineural hearing loss also have other medical and/or developmental comorbidities including cognitive, visual, motor, behavioral, and learning.¹³³⁻¹³⁶ Despite the prevalence of comorbidities in childhood hearing loss, there is no professional consensus regarding cochlear implantation as a viable, successful treatment option for children with special needs—particularly for those children with compromised cognition and/or severe global developmental delay.

An obvious, non-trivial consideration in determining CI candidacy for children with hearing loss and special needs is the difficulty in obtaining reliable behavioral estimates of hearing. In some cases, we may not be able to obtain reliable behavioral audiometric thresholds. However, given the various objective measures of auditory function, confirmation of the nature and degree of hearing loss is possible for even a non-responsive child. Further, sedated MRI can confirm the presence and structural status of the auditory nerve.

Expectations management is a vital component of pre-operative counseling for families. Hearing loss certainly complicates these diagnoses by restricting effective communication. Indeed, significant hearing loss confounds the interpretation of neurodevelopmental assessment and thus it is *possible* to observe significant improvement in a child's behavior and overall responsiveness following cochlear implantation. In most cases, however, hearing loss will not be the underlying cause of the developmental delay. This is particularly true for diagnoses including, but not limited to, agenesis of the corpus callosum, autism, cerebral palsy, Down syndrome, Fragile X syndrome, and Rett syndrome. Given this, we must have the counseling skills needed to be empathetic as well as honest and realistic about what CIs can and cannot do.

We will generally expect more modest outcomes with respect to the development of auditory skills and auditory/oral speech and language—particularly for those children exhibiting significant global developmental delay. In many reports, children with developmental delay demonstrated

significant postoperative benefit in auditory perceptual skills¹³⁷⁻¹⁴¹ and overall quality of life for the child and family. Although our definition of what constitutes a successful outcome will be individually determined for each child and family, the presence of global developmental delay should not automatically preclude cochlear implantation.

SUMMARY

CI indications have evolved over the past several decades such that CIs are no longer only for individuals with profound sensorineural deafness. A number of studies have demonstrated the efficacy of CIs for individuals falling outside current labeled indications with respect to audiometric thresholds (severity, configuration, and symmetry), aided speech recognition, age, and developmental abilities. What is of critical importance is that each implant candidate be assessed on an individual basis considering the whole patient so that we are implementing personalized medicine within the fields of audiology and otology. Though indications are in place to provide guidance for clinicians and patients, there will never be a substitute for the professional clinical judgment of the interdisciplinary CI team.

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COCHLEAR IMPLANTS

From Principles to Practice

This book is a multidisciplinary and comprehensive effort on the topic of cochlear implantation in children and adults with sensorineural hearing loss. It is essential reading for all disciplines involved in the care of these individuals and seek the best outcomes that would improve auditory perception, communication, and overall quality of life. The authors' list is a veritable *Who's Who* in the field. Their contributions to this book represent a lifetime of experiences that the reader will now have at his/her fingertips. The multidisciplinary nature of the authors' list reflects what is necessary in this field—a team approach to achieve the best outcome for our patients. The text is a must-read for all of us who have the honor to work in this field and care for individuals with hearing loss.

David S Haynes MD MMHC FACS is a Professor, Department of Otolaryngology, Neurosurgery and Hearing and Speech Sciences, Vanderbilt University Medical Center, The Otology Group of Vanderbilt, Nashville, Tennessee, USA. He is the Vice Chair, Department of Otolaryngology and is hold the Directorship of Relationship Development within the department. He is the Service Chief, Division of Neurotology and The Otology Group of Vanderbilt within the department of Otolaryngology and is Co-Director of the multidisciplinary Vanderbilt Skull-Base Center. In addition, he directs the internationally recognized Neurotology Fellowship Training Program and is the Medical Director of the Vanderbilt Cochlear Implant Program. He has hosted many international conferences including the XIV International Pediatric Cochlear Implant Conference (2014) in Nashville, TN. He also serves on the Executive Board of the Hearing Health Foundation, New York, The Acoustic Neuroma Association, Atlanta, GA, and the American Cochlear Implant Alliance, Washington DC. His research interests include optimizing care delivery models in cochlear implantation and skull-base surgery and in leadership development in medical education.

René H Gifford PhD is Professor, Hearing and Speech Sciences, Vanderbilt University Medical Center, Nashville, Tennessee, USA. She is also the Director of Cochlear Implant Program, Vanderbilt University Medical Center, USA. She began her professional career as a Clinical Audiologist and have maintained an active clinical practice over the years in the Cochlear Implant (CI) Audiology Clinic at the Vanderbilt Bill Wilkerson Center. Her interest of research focuses on the study of basic auditory function, spatial hearing, and speech perception for individuals utilizing electric and acoustic stimulation with cochlear implants and hearing aids. As both a scientist and a practicing audiologist working in an academic medical environment, she has first-hand knowledge of the clinical problems associated with hearing loss, cochlear implants, and the need for evidence-based recommendations for intervention.

George B Wanna MD FACS is a Professor of Otolaryngology-Head and Neck Surgery and Professor, Neurological Surgery, Icahn School of Medicine, Mount Sinai, and the Chief of the Division of Otology-Neurotology for the Mount Sinai Health System. He has served as the Chair of Department of Otolaryngology—Head and Neck Surgery, Mount Sinai Downtown since January 2017. He is Co-Author and Editor of several other books including: *Contemporary Management of Jugular Paraganglioma* (2018), *Otolaryngology Clinics of North America, Tumors of the Ear and Lateral Skull Base, Parts 1 and 2* (2015) as well as many book chapters in medical journals and publications. He is the Creator and Course Director of the Annual Endoscopic Middle Ear Dissection Course at NYEE, the Editor-in-Chief of Otolaryngology Case Reports and a Senior Board Examiner for the American Board of Otolaryngology—Head and Neck Surgery.

Alejandro C Rivas MD is an Associate Professor, Departments of Otolaryngology—Head and Neck Surgery and Neurological Surgery at Vanderbilt University Medical Center, Nashville, Tennessee, USA. There he serves as the Associate Service Chief, Department of Otology and Neurotology. He is a Fellow of the American Neurotology Society, American Academy of Otolaryngology—Head and Neck Surgery, and Triological Society. He is board certified in Otolaryngology—Head and Neck Surgery, as well as Otology and Neurotology. He earned his medical degree from the Universidad Militar Nueva Granada, School of Medicine in his hometown of Bogotá, Colombia. He completed a research fellowship and his residency in Otolaryngology—Head and Neck Surgery, John Hopkins University in Maryland, and subsequently finished an ACGME-accredited Neurotology Fellowship, Vanderbilt University Medical Center in Tennessee. His interests include endoscopic ear surgery, cochlear implantation, single-sided deafness, congenital ear canal atresia, bone conduction implantation, acoustic neuromas and other skull-base surgeries. As an educator, he had the opportunity to host and be the Co-Director of the 2014 ACIA meeting in Nashville, TN. He has performed and taught endoscopic ear surgery and cochlear implant surgical approaches to other doctors across the globe, including countries such as Colombia, Italy, Argentina, Chile, Mexico, Brazil, Uruguay, Japan, Australia, Peru, France, Spain, and Guatemala. He has presented at numerous international, national, and regional meetings. On this subject, he has contributed more than 100 peer-reviewed publications and book chapters. Most recently, he received the Honor Award in 2018 for distinguished service from the American Academy of Otolaryngology—Head and Neck Surgery Foundation.



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